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Ocular diseases in SOAP format

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Abstract
The purpose of this thesis project was to develop a comprehensive reference source of various ocular diseases and disorders. The format used was based on the SOAP format which is very common amongst disease profiles. An attempt was made to obtain the most recent accepted treatment plans including therapeutic drug regimens where applicable. It is advised that this compilation of SOAP's be used as a quick summary resource and if detailed knowledge be required, appropriate research be done on the desired topic. It is also believed that this manuscript may be used as a valuable study aid while preparing for disease and treatment sections on optometry board examinations.

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Thesis

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Author
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OCULAR DISEASES

IN SOAP FORMAT

By

DUANE A. CHANG
DAVID Y. FOK
BRIAN T. KUBO
IRIS Y. KUWABARA
STANLEY K. SATO
JUDY Y. TSUJIUCHI

A thesis submitted to the faculty of the
College of Optometry
Pacific University
Forest Grove, Oregon
for the degree of
Doctor of Optometry
May, 1988

Advisor:

Al Furie, O.D.
ABSTRACT

The purpose of this thesis project was to develop a comprehensive reference source of various ocular diseases and disorders. The format used was based on the SOAP format which is very common amongst disease profiles. An attempt was made to obtain the most recent accepted treatment plans including therapeutic drug regimens where applicable. It is advised that this compilation of SOAP's be used as a quick summary resource and if detailed knowledge be required, appropriate research be done on the desired topic. It is also believed that this manuscript may be used as a valuable study aid while preparing for disease and treatment sections on optometry board examinations.

Subjective, Objective, Assessment, Plan
David Y. Fok

Attended San Francisco State University. Received a B.S. degree in Visual Science in 1985 at Pacific University. He is a member of the Beta Sigma Kappa Society, the College of Optometrists in Vision Development, the American Optometric Association, and the California Optometric Association. Areas of optometric interest include: Contact lenses, Pediatrics, Vision therapy, and Sports vision.

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Attended the University of California, Los Angeles. Received B.S. in Visual Science from Pacific University in 1985. While at Pacific University he was a teaching assistant for Contact Lens II, and Procedures I and II. He is a member of Beta Sigma Kappa, a student member of the American Optometric Association, a member of the College of Optometrists in Vision Development, and a member of the California Optometric Association. Areas of optometric interest include: Contact Lenses, Pediatric Optometry, and Surgical Co-Management.

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Attended the University of Washington and University of Hawaii. Received B.S. in Visual Science with honors from Pacific University in 1985. While at Pacific she was a teaching assistant for a semester of Ocular Anatomy Laboratory. She is a member of Beta Sigma Kappa, a student member of the American Optometric Association, and the College of Optometrist in Vision Development. Areas of optometric interest include: Vision Therapy, Contact Lenses, Environmental Vision, and Low Vision.

Judy Y. Tsuiuchi

Attended the University of Hawaii and received a B.A. in pre-optometry under the liberal studies program in 1984. While at the University of Hawaii she received a service certificate from the Ho'opono School for the Blind where she did some volunteer work. She is also a member of Phi Eta Sigma freshman honor society. At Pacific University she was a student member of the American Optometric Association and the College of Optometrists in Vision Development. Areas of optometric interest include: Vision Therapy, and Contact Lenses.
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Cone dystrophy
Rubella
Acute posterior multifocal placoid pigment epitheliopathy
Acute retinal pigment epithelitis
Senile macular holes
Idiopathic preretinal macular fibrosa
Retinoblastomas

VITREOUS:
Posterior vitreous detachment
Asteroid hyalosis
Cholesterolosis bulbi
Syneresis
Muscae volitantes
Hemorrhage
Persistant hyperplastic primary vitreous
Vitreous prolapse
Retinitis proliferans

UVEAL TRACT:
Granulomatous uveitis
Non-granulomatous uveitis
Anterior uveitis
Acute posterior uveitis
Chronic uveitis
Panuveitis
Syphilitic uveitis
Herpes simplex iridocyclitis
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Adult cytomegalio inclusion disease
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- Reiter's syndrome
- Relapsing Polychondritis
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- Systemic lupus erythematosus
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SCLERA AND EPISCLERA:

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Episcleritis
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Scleromalacia perforans
Posterior scleritis
Sclerokeratitis
Staphyloma
Ectasia
Scleral thinning
Senile hyaline plaque

ADNEXA:
Sebaceous cysts
Suderiferous cysts
Serous cysts
Squamous cell papilloma
Nevi
Verruca vulgaris
Xanthelasma
Ciliary hemangiomas of the eyelid
Epidermoid and Dermoid cysts
Actinic keratosis
Basal cell carcinoma
Squamous cell carcinoma
Meibomian gland carcinoma
Malignant melanoma
Neurofibroma
External hordeolum
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Staphylococcal blepharitis
Entropion
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Blepharospasm
Keratoacanthoma
Meibomianitis
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Orbital cellulitis
Epicanthus
Lid coloboma
Molluscum contagiosum
Contact dermatitis
Poliosis
Blepharochalasis
Dermatochalasis

CORNEA:
Terrien's degeneration
Moore's degeneration
Salzmann's degeneration
Wessely ring
B-fibrillosis
Corneal edema
Bullous keratopathy
Band keratopathy
Neurotrophic keratopathy
Punctate epithelial erosions
Epithelial microcyst
Punctate Epithelial keratitis
Subepithelial infiltrates
Bacterial corneal ulcers
Corneal dellen
Dendritic ulcers
Stromal Herpes (Disciform keratitis)
Trophic ulcers
Stromal keratitis
Interstitial keratitis
Corneal deposits (calcium, silver, etc.)
Anterior megalophthalmos
Posterior keratoconus
Anterior Embryotoxin (arcus juvenilis)
Corneal keloid
Peter's anomaly
Axenfeld's anomaly
Sclerocornea
Spherical (keratinoid) degeneration
Coat's white ring
Lipid degeneration
Keratoconjunctivitis sicca
Xerophthalmia and keratomalacia
Posterior crocodile--shagreen
Meesman's epithelial dystrophy (Stocker-Holt)
Dot fingerprint map (Cogan's microcystic)
Reis-Buckler's
Fleischer (Vortex) dystrophy
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Idiopathic band keratopathy
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General Name: Morning Glory syndrome
Specific Name: Coloboma of the optic disc
ICD 9-CM#: 743.57
Location: Optic disc
Associated Conditions: Congenital forebrain anomalies; Basal encephalocele.

S: Decreased visual acuity; visual field defects.

O: Decreased visual function and pupillary defects. Enlarged, excavated disc with white, fibroglial tissue located at its center; an elevated subretinal peripapillary annulus of chorioretinal pigment changes encircles the disc.

A: Incomplete closure of the retinal fissure. The optic nerve is deeply excavated. It occurs equally frequent unilaterally and bilaterally and it is transmitted as an autosomal dominant hereditary defect. Patients are particularly prone to develop central serous choroidopathy that suggests a communication between the subretinal space and the subarachnoid space. Nonrhegmatogenous retinal detachment is seen in greater than 1/3 of the cases. These are connected to the disc and usually are confined to the posterior pole. Etiology is unknown although the association of the Morning Glory disc with hyperplastic primary vitreous has been noted.

P: No treatment is available. Just treat the symptoms.
General Name: Congenital optic disc anomaly

Specific Name: Pseudopapilledema

ICD 9-CM#: 377.24

Location: Optic Nerve

Associated Conditions: Papilledema, pseudoneuritis

S: Patient is asymptomatic. Vision is not impaired. It is most often seen in severely hyperopic eyes (>5 diopters).

O: Ophthalmoscopic examination reveals an elevation of the disc similar in appearance to papilledema. The disc margins are blurred due to the heaped up nerve fibers and drusen. Drusen may enlarge the nerve head, obliterate the physiologic cup and give the edge of the disc a crenated appearance. Illumination along the disc margin frequently causes drusen to glow and red-free light is often helpful. The color of the disc is gray-yellow and unlike hyperemia of papilledema, drusen when pronounced, show auto-fluorescence. Ultrasound and fundus fluorescein angiography is helpful in identifying buried drusen. Visual acuity is not impaired and the retinal blood vessels are normal. Spontaneous hemorrhage occurring with disc drusen may present as peri-papillary flameshaped, intravitreous, and/or subretinal hemorrhage around the disc that may extend under the macula. In this case, there will be VA and central field loss.

A: Pseudopapilledema, pseudoptic neuritis, or pseudophthalmits describes a bilateral asymmetric congenital anomalous elevation of the optic nerve head that is associated with high hyperopia, hyaloid remnants, myelinated nerve fibers, hyaline borders, drusen, or other unexplainable, congenital elevations. Accurate diagnosis depends upon eliciting the concurrent history and physical signs. Any patient with suspected optic disc swelling needs a careful case history, ophthalmoscopy, and examination of visual acuity, refraction, color vision, pupillary response, ocular movements, and visual fields. There are three patterns of field defects: enlargement of blind spot; arcuate or nerve fiber bundle defects with sector cuts, ring, paracentral, and Bjerrum scotomas; or irregular peripheral conditions. Narrowing of the lower nasal field is the most characteristic defect. Typically defects progress very slowly. Differential diagnosis from other similar clinical conditions of papilledema and papillitis is indicated.

P: The condition is not progressive and requires no treatment. However, in cases when there is difficulty in fundus diagnosis, patient should be observed even though he may be symptom-free and healthy. Follow up examination should consist of re-examination of eyes, re-inquiry of neurologic symptoms of diplopia, headache, nausea, vomiting, or drowsiness, and arrange for a serial fundus photographs.
General Name  Papilledema
Specific Name  Papilledema
ICD  9-CM#  377.0
Location  Optic Disc

Associated Conditions
Intracerebral or Subarachnoid Hemorrhage, Tumors of the Spinal Cord, Inflammatory Polyneuritis, Infectious Disease, Toxic Metabolic Diseases, Trauma, Benign Intracranial Hypertension, Therapeutic Doses of Lithium Carbonate, Congenital Brain Malformations, Any Intracranial Space-Occupying Lesion

S:  Transient visual obscurations lasting 10-30 seconds. May also have diplopia. Visual acuity is unaffected except in cases of chronic papilledema with optic atrophy. Headaches which may be acute or chronic, lateralized or generalized, mild, moderate or severe. Often it is worse on waking in the morning and aggravated by coughing, sneezing, and straining. When accompanied by nausea and vomiting the condition is particularly serious.

O:  Optic disc edema is the first sign observable of papilledema. Striations of the nerve fibers first appear in the inferior, then superior, then nasal aspect of the disc with the temporal aspect least susceptible to swelling. A reliable and important first sign is obscuration of the wall of a vessel crossing the disc margin. Following signs include blurring of the disc margins, hyperemia of the disc and capillary dilation. Late papilledema includes elevation of the disc, obliteration of the physiologic cup and hemorrhage on or near to the disc. A general but not absolute rule is absence of spontaneous venus pulsation. Visual field defects includes an enlarged blind spot and constriction of the peripheral field. Long term changes include gliosis surrounding the disc, sheathing of vessels, and progressive optic atrophy.

A:  Papilledema is optic disc swelling due to elevation of the intracranial cerebrospinal fluid pressure. Papilledema must be differentiated from pseudo papilledema which includes high hyperopia, hyaline bodies, myelinated nerve fibers and other unexplainable congenital elevations. When difficulty in fundus diagnosis persists and the patient is otherwise symptom free and healthy, observe the patient. Re-examine the eyes, inquire again for neurological symptoms of diplopia, headaches, nausea, vomiting or drowsiness, and arrange for serial fundus photographs.

P:  True papilledema is a major ophthalmic and neurologic emergency. The patient requires immediate admission to a hospital.
General Name Anterior Ischemic Optic Neuropathy (AION)
Specific Name Arteriosclerotic-Hypertensive AION
ICD 9-CM# 377.41
Location Optic Nerve

Associated Conditions
Hypertension, Optic atrophy, Marcus Gunn pupil

S: Patients complain of loss of vision over a 24 hour period.

O: Visual acuity is decreased and ranges from mild to severe.
- Marcus Gunn pupil
- Field losses, either inferonasal, altitudinal defect or central scotoma
- Optic disk edema and hyperemia with splinter hemorrhages
- Progressive optic atrophy

A: Most cases tend to occur in men 45-65 years of age. Very often there is a family history of diabetes or hypertension. In addition the patients themselves often have a history of vascular disease. Based on the presence of hypertension, decreased visual acuity, and optic disk changes the diagnosis is Arteriosclerotic-Hypertensive AION. Optic neuropathy occurs due to the compromise of the short posterior ciliary arteries. Generally, it is caused by an acute imbalance of the perfusion pressure at the disk. Any drop in perfusion pressure including the type caused by an antihypertensive drug can cause an acute ischemic attack.

P: The condition is treated by controlling the underlying problem. The perfusion ratios must be rebalanced by either lowering IOP or bringing the hypertensive swings under better control. Steroids are sometimes used to decrease the swelling of the nerve head and permeability of the capillaries, but the treatment is not universally accepted.
General Name: Anterior Ischemic Optic Neuropathy (AION)
Specific Name: Temporal Arteritic AION
ICD 9-CM#: 377.41
Location: Optic nerve

Associated Conditions:
Elevated sedimentation rate, Optic atrophy

S: Patients may complain of: incredible headaches in the temporal area, hazy vision, spots of vision loss, flashing lights and anorexia due to pain associated with chewing.

O: Signs include:
- Lowered VA's ranging from 20/60 to light perception.
- Pale edematous disk with splinter hemorrhages sometimes found around the disk.
- Temporal arteries are visible and are hard and knobby upon palpation.

A: Based on headaches, optic disk involvement and hard tortuous temporal arteries, the patient should be referred to an internist for a sedimentation rate and a possible temporal artery biopsy. In cases of temporal arteritis the sed rate will be elevated as high as 80-100; normal being 0-10 in males and 0-20 in females. The white cell count will also be elevated. Giant cells in the walls of the temporal arteries constitute the definitive diagnosis for temporal arteritis.

P: Temporal arteritic AION is an ocular emergency because the fellow eye will present with the condition in 3 days to 3 weeks after the initial eye in 100% of the cases. Prednisone given orally in massive doses reduces inflammation and capillary permeability and stops the infiltration. Doses are then tapered to a maintenance level. Prognosis is poor for the involved eye and total optic atrophy usually occurs. Involvement of the fellow eye is guaranteed unless the patient is put on aggressive therapy immediately.
General Name Anterior Ischemic Optic Neuropathy (AION)
Specific Name Diabetic Papillopathy
ICD 9-CM# 377.41
Location Optic nerve

Associated Conditions
Juvenile onset diabetes

S: Patients complain of marked vision loss.

O: Signs include:
  - Optic disk edema which is frequently bilateral
  - Normal pupil responses
  - Visual field defects vary from an enlarged blind spot to isolated scotomas

A: The condition is usually seen in juvenile onset diabetes. The presence of a history of diabetes and optic nerve involvement indicate diabetic papillopathy. The condition is neither a true optic neuropathy or papilledema and signs of both conditions are present. The cause of the condition is pericyte death. It is important to rule out the causes of papilledema (conditions which create an increase in intracranial pressure) and other causes of ischemic optic neuropathy such as hypertension.

P: Referral to an internist is indicated if patient is not under medical care. The treatment of the condition is to control the diabetes. Once the diabetes is under control the condition will resolve without specific therapy.
General Name: Optic Disc Infiltration

Specific Name: Optic Disc Infiltration

ICD 9-CM#: 377.49

Location: Optic Nerve

Associated Conditions: Sarcoid, Behcet's disease, Systemic lupus erythematosus (SLE), Multiple sclerosis (MS), Optic nerve glioma

S: A general term used when the optic disc becomes infiltrated by a lymphoma, metastasis, or granuloma. Visual acuity and field loss can vary.

O: The usual field loss pattern is a central scotoma with poor acuity.

A: It is important to search for a vitreous cellular infiltration or peripheral vasculitic lesions in the retina when disc swelling is present.

P: Find and treat, if possible, the (systemic) cause.
General Name: Leber’s Optic Neuropathy
Specific Name: Acute Leber’s Optic Neuropathy
ICD 9-CM#: 377.16
Location: Optic Nerve

Associated Conditions:
Optic Neuritis

S: A simultaneous acute or subacute bilateral vision loss. However a 1-6 month interval between involvement of the two eyes may occur. The patient will also complain of headaches during the disease active stage.

O: 1) Preacute signs: Atrophy of the retinal nerve fibers, altered Farnsworth-Munsell 100 hue test, a decreased VER amplitude.
   2) Acute Signs: A mild optic disc edema with early circumpapillary telangiectatic microangiopathy, central scotoma, visual acuities in the range of 20/200-20/400.

A: The etiology of the disease is unknown although some authors believe it to be hereditary. Men are more often affected with the age of onset for men between 18-30 years and for women between 10-40 years. The optic atrophy is a sequela to an acute or subacute optic neuritis.

P: The prognosis of those affected with the disease is poor. The disease will usually proceed into a permanent optic atrophy that affects the temporal sector of the disc more severely. There is no known medical intervention once the disease has become symptomatic. However vitamin B-12 and B-12A have been used with limited success in 1 mg dosages parenterally daily for 7 days for those suspected of the disease.
General Name: Glioma
Specific Name: Optic Nerve Glioma
ICD 9-CM#: 225.1
Location: Optic Nerve
Associated Conditions: Neurofibromatosis, Von Recklinghausen's disease, Cafe au lait spots

S: Symptoms include:
- proptosis
- loss of vision

O: Observation of the fundus shows an unilateral optic disc swelling. It is a well-differentiated tumor. There is also an associated visual field defect.

A: Optic nerve gliomas are benign tumor of the optic nerve. Approximately one-quarter of optic nerve gliomas are found among patients with Neurofibromatosis (Von Recklinghausen's Disease). Therefore, a child with cafe au lait spots, exophthalmos, and abnormal optic nerve head has a high probability of having optic nerve glioma. These findings, together with an enlarged optic canal on plain-film and tomographic x-ray, are virtually pathognomonic of this tumor. The glioma may occur anywhere along the length of the nerve including the optic chiasm. If it is in the intraorbital region of the optic nerve it may produce vision loss, ptosis, and unilateral swelling of the optic disc. If it is in the optic chiasm it may produce optic atrophy and visual field defects.

P: No treatment is required. However, it may be wise to advocate simple observation. Furthermore, if the optic nerve glioma is causing severe proptosis and endangering the cornea, excision may be indicated.
General Name Meningioma
Specific Name Meningioma
ICD 9-CM# 192.0
Location Intraorbital
Associated Conditions Neurofibromatosis

S: Very slowly progressive axial proptosis and/or loss of vision. There may be transient obscurations of vision.

O: Ophthalmoscopically there is the appearance of retinochoroidal striae. At first the disc may appear normal but optic atrophy or chronic disc swelling ensues. The presence of optociliary venous shunts on the disc when accompanied by disc pallor and visual loss is highly suggestive of indolent nerve sheath meningiomas. In addition there may be proptosis and increased hyperopia.

A: Meningiomas arise from meningothelial cells of the arachnoid, at multiple intracranial sites and from the intradural tissue that invests the optic nerves in the orbit. There is a distinct predilection for meningiomas and the intracranial variety occurs predominantly in adults. However, perioplic meningiomas occur at an earlier age. Meningiomas presenting in youth may be associated with neurofibrosis.

P: In adults, delay in diagnosis is the rule and any surgical procedure is ineffective with regard to preservation to vision. With indolent tumors there may be no advantage to any form of surgical intervention. Radiotherapy is of no value. Since many of these tumors grow very slowly, unless radical operative procedures are curative they cannot be justified on the basis of short-term evaluation. In cases of proliferating orbital meningiomas, complete surgical excision with posterior optic nerve transection is the treatment of choice.
General Name  Orbital Pseudotumor

Specific Name  Idiopathic Inflammation Syndrome

ICD 9-CM# 376.11

Location  Orbit

Associated Conditions

Proptosis.

S: Patient complains of pain that comes on suddenly; discomfort, double vision, burning and foreign body sensation. At times painless, quiet, and slowly progressive proptosis. Usually unilateral and in all age-groups.

O: Biomicroscopy will reveal conjunctival injection and chemosis, periocular and lid edema and erythema. There might be corneal exposure causing corneal and conjunctival dessication. Variable degrees of ophthalmoplegia (and pain), but visual loss is infrequent. No distinct swelling is found and no systemic or identifiable local cause can be found. More frequent in middle-age.

A: Idiopathic inflammation syndrome should be separated clinically form Graves' Disease by appropriate thyroid tests. In Graves' Disease the onset of inflammation signs is less explosive, and pain, extraocular motility disturbances, and visual decrease are observed later in the clinical course. Rule out Graves' disease, Orbital cellulitis, Benign lymphoid hyperplasia, Malignant lymphoma.

P: Usually a benign disease; therefore, conservative therapy should be considered. Patients can be given a trial of 80 mg. of prednisone daily for 2 weeks; this may provide rapid symptomatic improvement and validation of the diagnosis. CAT scan also helps, before and after trial therapy. When there are atypical features of the clinical course or when there is failure to respond to steroids or recurrence of the disease, a biopsy must be considered. For therapy, high doses of prednisone should be continued for a total of 3 weeks and slowly tapered to prevent a rebound. In this syndrome, there can be recurrences and even bilaterality; in adults the latter may signal the presence of a systemic disease, but in children it does not. A vasculitis diagnosis needs to be assessed in the presence of a bilateral pseudotumor. Recurrences are common. Optometric supportive care should be directed at the diplopia, blurred vision and the resultant dessication of the cornea due to the severe proptosis. Artificial tears and wet chamber concepts should be applied. The diplopia might require occluder lens to eliminate the symptom and allow the patient to function on a monocular basis, until the condition is resolved.
General Name: Toxic Amblyopia

Specific Name: Nutritional and Toxic Optic Atrophy

ICD 9-CM#: 377.33, 377.34

Location: Optic Nerve

Associated Conditions:
Malnutrition, Liver problems

S: A gradual, bilateral, painless reduction of visual acuity with eventual centrocecal scotomas.

O: The patient often has a sickly appearance. The optic disc may look edematous with isolated splinter hemorrhages in the early stages. The nerve head will eventually become atrophic in the later stages with the vision limited to detecting hand motion.

A: The differential diagnosis can be performed primarily from the visual fields and case history. The centrocecal scotomas can usually be better defined using a red isopter. The densest portion of the scotoma typically corresponds to the papillomacular bundle. The scotoma also rarely ever crosses the vertical meridian. An acquired dyschromatopsia is also associated with the nerve loss. As far as case history, the patient is typically poor and undernutritioned in nutritional optic atrophy. In toxic optic atrophy, typical drugs or substances which can induce the atrophy are:

1. alcohol
2. barbituates
3. chloramphenicol
4. chlorquine
5. corticosteroids
6. digitals
7. iodoquinol
8. disulfiram
9. ethambutol
10. ethchlorvynol
11. heavy metals
12. vitamin D
13. hydroxychloriquine
14. iodide compounds
15. isoniazide
16. phenylthiazines
17. streptomycin
18. tobacco

P: The condition is reversible with favorable prognosis if the toxic agent or nutritional deficiency is detected and removed in the early stages. Vitamin B12 therapy using 300 mg oral thiamin and 1000 micrograms of intramuscular hydroxycobalamin each week for 10 weeks has been shown to be effective in tobacco and alcohol amblyopia.
**General Name**  Embryonic Remnant  
**Specific Name**  Bergmeister's papilla  
**ICD 9-CM#**  743.51  
**Location**  Optic nerve  
**Associated Conditions**  Hyaloid membrane  

**S:** Asymptomatic

**O:** Ophthalmoscopy shows a hyaloid vasculature which extends from the optic nerve.

**A:** During embryologic life, the hyaloid vasculature extends from the optic nerve through the vitreous and nourishes the developing lens. It normally regresses completely in later development but in some individuals remnants remain at the surface of the optic disc as Bergmeister's papilla.

**P:** Bergmeister's papilla do not interfere with vision. Treatment is not required.
General Name: Morning Glory Syndrome
Specific Name: Morning Glory Syndrome
ICD 9-CM#: 377.23
Location: Optic Disc

Associated Conditions:
Persistent Hyperplastic Primary Vitreous, Basal Encephalocele

S: Visual acuity falling between the 20/100 to hand motion range.

O: Ophthalmoscopically it appears as an enlarged excavated disc with white, fibroglial-appearing tissue located at its center; an elevated subretinal peripapillary annulus of chorioretinal pigmentary changes encircles the disc. The retinal vessels enter and leave the nerve head at its margins.

A: MGS is a congenital optic disc malformation with the majority of cases being unilateral. No hereditary tendency is evident and no associated intrauterine insults have been identified. The etiology of the anomaly is uncertain, although the association of a morning glory optic disc with persistent hyperplastic primary vitreous has been noted. Optic nerve dysplasia is the term that has been applied to the morning glory disc anomaly when the center is elevated rather than excavated.

P: No available treatment.
General Name: Optic Neuritis
Specific Name: Papillitis
ICD 9-CM#: 377.31
Location: Optic Nerve
Associated Conditions: Syphilis.

S: Patient complains of sudden, moderate to severe loss of vision.

O: Papillitis is disc swelling caused by a local inflammatory process of the nerve head. It may be thought of as an intraocular form of optic neuritis although the etiology is not the same. In children, papillitis is the common form of optic neuritis. Also, simultaneous bilateral neuritis is far more common in children than adults. Unilateral retrobulbar neuritis is the common form in adults.

A: Papillitis is distinguished from papilledema by: 1) rapid loss of vision, central or paracentral scotoma and enlarged blind spot (good acuity may be retained on occasion) 2) pain 3) afferent pupil defect (Marcus Gunn) 4) cells in the vitreous especially just anterior to the disc (therefore, vitreous haze and margins blurred) 5) no trace of a central cup, but venous pulsation present or easily provoked 6) deep retinal exudates or macular star. Differentiate from the somewhat related papillary vasculitis. Rule out Papilledema, Papillary vasculitis.

P: Visual prognosis is surprisingly good, even in the presence of massive disc edema and flame-shaped hemorrhages or with initial severe visual loss. However, progressive atrophy may result regardless of therapeutic intervention (get secondary optic atrophy when swelling goes away and disc turns white and have nerve fiber drop-out). Therefore good visual outcome should not be guaranteed. Oral or sub-bulbar corticosteroids are used.
General Name: Optic Neuritis
Specific Name: Optic Neuritis
ICD 9-CM#: 377.30
Location: Optic Nerve
Associated Conditions: Syphilis.

S: Inflammation of the optic nerve, including that accompanying demyelinating disease or contiguous spread of inflammation from meninges, orbital tissues, or paranasal sinuses. Secondary inflammation uses the more general term, optic neuropathy. Patient complains of acute impairment of vision progressing rapidly for hours or days. Typically unilateral in adults and bilateral in children. Globe tenderness and deep orbital or brow pain especially with eye movements.

O: Field defects usually involve the central field, therefore diminished acuity also usually observed. Diminished light reaction on side of neuritis; desaturation of colored objects, especially red; apparent reduction of light intensities (light appears dimmer through affected eye in light comparison test); impairment of depth perception especially with moving objects (large VER latency in affected eye) (Pulfrich phenomenon); and increase in visual deficit with exercise or other elevations of body temperature (Uhthoff's sign). All of these visual symptoms may persist after return of acuity to normal levels. Some degree of disc swelling is usually present.

A: Optic neuritis is a syndrome and not a primary disorder. Most are monosymptomatic events without identifiable underlying cause. Rule out Papilledema, Papillary vasculitis.

P: For most cases, visual function spontaneously begins to improve by the third week with near-normal vision by the fifth week. Vision slowly but steadily improves over several months. There is no form of therapy to date which significantly alters the course of optic neuritis, but ACTH and corticosteroid therapy have been used.
General Name: Optic Neuritis
Specific Name: Retrobulbar Neuritis
ICD 9-CM#: 377.32
Location: Optic Nerve
Associated Conditions: Syphilis.

S: An inflammatory process of the optic nerve posterior to the globe. The subjective symptoms are the same as for optic neuritis, except that the pain is much more intense.

O: The objective symptoms are the same as for optic neuritis. However, the doctor does not see any abnormal signs with ophthalmoscopy—the eye appears normal. The classic "patient sees nothing, doctor sees nothing" is present. Marcus Gunn pupil always present as well as variable central or centrocecal scotomas.

A: The etiology of this condition remains obscure. Rule out Papilledema, Papillary vasculitis.

P: The treatment plan is the same as for optic neuritis.
General Name Papillophlebitis
Specific Name Papillophlebitis
ICD 9-CM# 362.18
Location Retina
Associated Conditions
Central Retinal Vein Occlusion.

S: Vision is nearly normal in a healthy young adult, age 20 to 40 years old.

O: Unilateral optic disc edema due to central retinal vein (CRV) occlusion. Highly engorged veins and widespread, mainly peripheral hemorrhages and microaneurysms.

A: Differentiate from papilloedema and optic neuritis. This occlusion is thought to be initiated by phlebitis.

P: Resolves in 12 to 18 months without treatment and without residue except optociliary shunt vessels on the disc (CRV branches stopping abruptly at the disc margin) (this last characteristic is often associated with optic atrophy and blindness).
General Name: Pits of the Optic Disc
Specific Name: Pits of the Optic Disc
ICD 9-CM#: 377.22
Location: Optic Nerve
Associated Conditions:
Serous Retinal Detachment

S: Variable from normal vision to complaints of severe vision loss.

O: The pit usually presents itself as a grey depression in the inferior temporal region of the optic disc. The depth may vary from very shallow to 8mm. The pits size varies from 1/8 to 1/3 disc diameters.

A: The pits are believed to be incomplete colobomas of the nerve head. Optic pits as a single entity create few problems. However pits located on the papillomacular bundle are frequently associated with a serous detachment of the macula. A definitive diagnosis can be performed using fluorescein angiography. It gives the appearance of leakage at the pit and masking of background fluorescence in the region of the serous detachment. Visual field defects are often common with this congenital defect. An arcuate bundle scotoma or central scotoma are present in about 60% of cases.

P: The prognosis for non-complicated optic pits is good. The patient generally remains asymptomatic with good vision and stable scotomas. However in those cases with associated serous macular detachment, prognosis is poor. Treatment with photocoagulation has been relatively ineffective.
General Name: Hypoplastic disc, Double ring sign
Specific Name: Optic nerve hypoplasia
ICD 9-CM#: 743.9
Location: Optic disc
Associated Conditions:
Septo-optic dysplasia (De Mosier's syndrome), Microphthalmos, Hydrocephalus

S: Slight to severe depression of visual acuity.

O: Small atrophic disc which can be very pale in color if the hypoplasia is pronounced. It is partially or totally surrounded by a yellow white ring that may be variably pigmented (causing the second ring).

A: By clinical presentation. Hypoplastic disks usually come about due to the failure of the ganglion cells to reach the disk. This atrophic disk is often associated with decreased size of the optic foramen. It occurs in otherwise normal eyes, in microphthalmus and in hydrocephalus. In 1/3 of the cases, major concomitant central nervous system anomalies have been recognized. One of them, Septo-optic dysplasia (DeMosier's syndrome) is characterized by the clinical triad of shortness of stature, nystagmus and optic nerve hypoplasia. Neurologically there is an absence of the septum pellucidum. This forebrain dysplasia is also accompanied by a deficiency of growth hormone and even by diabetes insipidus. Early recognition permits correction of the hormonal imbalance and a chance for the child to achieve normal growth and stature.

P: If the condition is a result of a deficiency of growth hormone, early correction of the hormone imbalance can decrease the effects.
**General Name** Drusen of the Optic Nerve  
**Specific Name** Drusen of the Optic Nerve  
**ICD 9-CM#** 377.21

**Location** Optic nerve

**Associated Conditions**

**S:** The majority of cases of disc drusen are harmless and remain asymptomatic. There is no associated refractive error. As a rule central vision is intact.

**O:** Ophthalmoscopic appearance of disc drusen in the child is atypical. When the drusen is embedded beneath the surface of the disc, it may produce a fullness of the papilla, mild elevation, and blurring of the disc. There may also be an abnormal number of vessels on the disc. Ophthalmoscopic appearance in the adult is usually diagnostic. Both discs are involved in 73% of the cases but asymmetry in appearance is common. Drusen presents as a small mulberry-like mass or as a waxy tumor composed of a conglomerate of smaller masses. They may enlarge the nerve head, obliterate the physiologic cup, and give the edge of the disc a crenated appearance. Illumination along the disc margin frequently causes drusen to glow. Drusen when pronounced show autofluorescence. The color of the disc is gray-yellow. Three patterns of field defects are found: 1) enlargement of the blind spot 2) arcuate or nerve fiber bundle defects 3) irregular peripheral contraction. Narrowing of the lower nasal field is the most characteristic defect. Occasionally there is spontaneous hemorrhage occurring with disc drusen presenting as peripapillary flame shaped, intravitreous, or subretinal.

**A:** Drusen of the disc are thought to be a congenital excess of immature neuroglia that undergoes degeneration. Disc drusen are inherited as an autosomal irregular dominant trait. It is not visible at birth and rarely observed in children under the age of 11 years. Drusen erupt on the disc surface in the second or third decade. With increasing age they become more easily visible and recognizable. Overt disc drusen may be seen ophthalmoscopically in parents of children with anomalously elevated discs without visible drusen. Examination of family members is mandatory if the distinction between true papilledema and pseudopapilledema in the child is in doubt.

**P:** No available treatment.
General Name: Medullated Nerve Fibers
Specific Name: Medullated Nerve Fibers
ICD 9-CM#: 743.41
Location: Optic Nerve
Associated Conditions: None

S: The patient reports no symptoms.

O: Ophthalmoscopy reveals a white opaque area with a glistening appearance and soft feathered edges continuous with the optic disk. Visual fields will reveal an enlarged blindspot or sometimes a partial scotoma.

A: The myelinated fibers involve a small sector of retina near the disk. The absence of pigment proliferation, feathered edges and relatively normal visual field differentiates the condition from chorioretinitis.

P: No treatment is indicated.
**General Name** Tilted Disc  
**Specific Name** Tilted Disc  
**ICD 9-CM#** 377.2  
**Location** Optic Nerve  

**Associated Conditions**  
Field Defects

**S:** The patient is usually asymptomatic except for the possibility of blurry vision.

**O:** Ophthalmoscopic evaluation shows an optic nerve head of tilted appearance. Accompanying scleral crescents are usually seen especially in the inferior margins. The fundus in the sector adjacent to the crescent is usually hypopigmented and takes on a tigroid appearance. A decreased foveal reflex is also common.

**A:** Assessment is made primarily from ophthalmoscopic evaluation. Visual fields can be taken usually revealing superior temporal field defects which do not respect the horizontal midline. This condition is usually bilateral and can most commonly found with myopia and oblique astigmatism.

**P:** The tilted disc is a stable condition with good prognosis. It is a congenital condition which requires no treatment other than patient education.
General Name: Multiple sclerosis (MS)
Specific Name: Multiple sclerosis
ICD 9-CM#: 340
Location: All over the body
Associated Conditions:

S: Optic neuritis is the initial episode of the disease in 15% of the patients. In 25% of the patients, muscle palsy occurs (diplopia, nystagmus). Of those patients hospitalized for MS, 70% have or have had optic neuritis. The fully developed disease is characterized by the Charcot triad of nystagmus, scanning speech and optic atrophy. Sheathing of peripheral retinal veins occurs in 55% of the patients. Diplopia occurs because of an internuclear ophthalmoplegia or because of palsies of individual muscles.

O: If the optic neuritis is retrobulbar, no signs are seen. For signs of anterior optic neuritis, see Optic Neuritis.

A: MS is very difficult to diagnose. The best method appears to be the VER in which the latency period of nerve impulse transmission increases with MS. Even in periods of remission of the disease, the latency period will still be high because remyelination of the optic nerve doesn't occur. The optic nerve is the first one affected because it is short, therefore the percentage of myelin lost is the greatest. MS is an autoimmune demyelinating disease of the CNS. It affects mainly the population between 15 & 30 yrs. of age. Prevalence is high in the northern U.S. and in Canada and low in the southern U.S.. The average duration is 27 yrs. and in many patients there are remissions without permanent neurologic residue.

P: No effective treatment known. Just treat the symptoms and prevent more damage. Salicylates can be used as anti-inflammatories. Steroids may also be used for the optic neuritis.
General Name: Pupillodilator dysfunction
Specific Name: Horner's syndrome
ICD 9-CM#: 337.9
Location: Pupil
Associated Conditions:
Mediastinal tumors, Hodgkin's disease, Metastatic tumors, Adenomas, Neurofibromatosis, Surgical and accidental traumas to the neck, Occlusion of the posterior inferior cerebellar artery, Multiple sclerosis, Syringomyelia, and Tumors of the cervical chord.

S: Symptoms include:
- Miosis (seen in 98% of the patients)
- Ptosis (seen in 88% of the patients)
- Anhydrosis (seen in 0.4% of the patients)
- Heterochromia (seen in 0.3% of the patients)
- Some patients may also experience headaches.

O: Ocular examination reveals:
1. Miotic pupil which reacts equally to light and near. The miosis is not marked and is often missed especially in lighted conditions.
2. Ptosis is due to the loss of sympathetic nerve supply to the Muller's muscle. The eyelid usually droops 1-2 mm.
3. Less pigmented iris (heterochromia iridis) may be observed in the affected eye of a patient with Horner's syndrome.

Anhydrosis is caused by the loss of the sympathetic nerve fibers to the neck and face. The triad of Horner's syndrome consists of: miosis, ptosis, and anhydrosis.

A: Horner's syndrome is due to an interruption of the oculosympathetic pathway which can be divided into 3 parts: central (1st order neuron: hypothalamus to C8-T2); Pre-ganglionic (2nd order neuron: C8-T2 to the superior cervical ganglion at the bifurcation of the internal and external carotid; Postganglionic (3rd order neuron: superior cervical ganglion to dilator muscle and the muller's muscle. There are two tests which are helpful in the diagnosis of Horner's syndrome: 1) Cocaine Test: 5%-10% cocaine will normally dilate eyes. If it does not, it confirms the presence of Horner's syndrome. Dilation occurs in normal eyes because cocaine dilates the pupil by preventing reuptake of norepinephrine at the postganglionic nerve terminal. However, in Horner's, because there is a decreasing norepinephrine, instillation of cocaine has little effect. 2)Paradine Test: 1% Hydroxyamphetamine (paradine) is used to distinguish between the 1st and 2nd from the 3rd order neurons. If the lesion is at the 1st & 2nd neuron, the drug causes dilation. If at the 3rd neuron, there's no dilation.

P: Appropriate patient management depends on accurate localization of the lesion. The following referrals are indicated:
- Central: Neurologist
- Preganglionic: Neurologist, Internist, Thoracic Surgeon
- Post ganglionic with typical headache: Internist
- Post ganglionic with atypical headache or without headache: Neurologist

Most patients with preganglionic lesions should receive further diagnostic evaluations with a neurologist to confirm or deny the presence of benign or malignant tumor. Malignant tumors are a frequent cause of Horner's syndrome and are almost exclusively located in the 1st or 2nd order neurons (77%), as compared to 3rd order neuron (16%).
General Name Tonic Pupil
Specific Name Adie's Tonic Pupil
ICD 9-CM# 379.46
Location Pupil
Associated Conditions

S: Blurred vision at near due to unilateral defective accommodation. The patient may also notice a difference in pupil size. Bright lights frequently bothers these patients.

O: Relative mydriasis in bright illumination. Poor to absent light reaction. Slow contraction to prolonged near effort. Slow redilation after near effort. Segmental vermiform movements of iris border. Defective accommodation. Pupil constricts with mecholy 2.5%, pilocarpine 0.125%. Associated with diminished deep tendon reflexes.

A: Tonic pupil is a benign condition which in the absence of other signs and symptoms heralds no neurologic or systemic disease state. Although the pupil rarely recovers much light function there is reinnervation of the ciliary body and recovery of accommodative function. After a tonic pupil has been present for years, it becomes smaller and may even be the smaller of the two pupils. However, bilateral pupillotonia has been associated with other autonomic nervous system dysfunction.

P: Tonic pupil is a benign condition which in the absence of other signs and symptoms heralds no neurologic or systemic disease state. Although the pupil rarely recovers much light function, there is reinnervation of the ciliary body and recovery of accommodative function. After a tonic pupil has been present for years, it becomes smaller and may even be the smaller of the two pupils.
General Name: Pupillary Light-Near Dissociation
Specific Name: Argyri Robertson Pupil
ICD 9-CM#: 094.89
Location: Pupil
Associated Conditions: Syphilis

S: Patient may report small irregular unequal pupils and may have systemic symptoms of syphilis.

O: Observations include:
   Pupils that fail to constrict with light but retain constriction with convergence. The syndrome includes miotic irregular and unequal pupils, presence of some vision in each eye, failure of pupils to dilate after scopolamine instillation and miosis after eserine instillation.

A: When all aspects of the syndrome listed above are present the diagnosis is of tabe dorsalis of central nervous system syphilis. Lesion is thought to be in the pretectal region where the afferent fibers synapse. In cases of tumour and hemorrhage involving the pretectal region there may be associated failure of pupils to react to light and retain reaction to convergence, but pupils won't be miotic, unequal and irregular.

P: Patient should be referred to an internist after vision needs have been taken care of.
General Name: Unilateral Fixed and Dilated Pupil

Specific Name: Unilateral Fixed and Dilated Pupil

ICD 9-CM#: 379.40

Location: Parasympathetic Nervous system and/or Sphinctor

Associated Conditions:
Stroke, 3rd Nerve Palsey, Tumors, Diabetes

S: The patient complains of uneven pupil size.

O: The patient presents an anisocoria with one dilated pupil. This pupil is fixed and unreactive to direct light or accommodative stimulus.

A: There are many potential causes of this disorder but most can be narrowed to: 1) Adie's pupil; 2) defect in the 3rd intracranial nerve; 3) anticholinergic mydriasis. Instillation of .125% pilocarpine bilaterally can be used to access the degree of sphincter or innervational damage. Damage to the 3rd cranial nerve will reveal all or some of the following: ptosis, exotropia, hypotropia, and a dilated pupil all on the same side. Typically a tonic pupil due to the third nerve damage will only constrict to .5 or 1% pilocarpine. Anticholinergic mydriasis is due to inoculation of the eye by an anticholinergic substance. This may be either through drug contact or exposure to such plants as Jimson weed. These pupils will not contract with 1% pilocarpine. * See Adies Syndrome for diagnosis of Adies pupil.

P: The prognosis for anticholinergic mydriasis is excellent. Normal pupil appearance and function will resume once the effects wear off, usually between a few days to a few weeks. The prognosis for 3rd nerve damage is poor since actual nerve damage has occurred. Many serious systemic diseases such as diabetes or tumor can cause a 3rd nerve palsey and the patient should be referred for a neurologic consultation. Adies pupil tends to slowly return to normal over a period of a couple of years.
General Name: Marcus Gunn (MG) Pupil; Afferent pupillary defect
Specific Name: Marcus Gunn Pupil
ICD 9-CM#: 379.40
Location: Pupil

Associated Conditions:
Optic neuritis, optic atrophy, direct pressure on any part of the optic nerve, retinal detachment, central retinal artery occlusion.

S: Decreased light sensitivity on the affected eye. Pupils are equal in size.

O: Diminished amplitude of pupillary light reaction, a lengthened latent period and pupillary dilation (escape) with continuous light stimulus.

A: The swinging flashlight test is used for the detection of MG pupil. There is dilation of both pupils when light is moved from the unaffected eye to the affected eye and a constriction of both pupils when the light is moved back to the unaffected eye. This phenomenon occurs due to an afferent pupillary defect of the visual pathway anterior to the optic chiasm. It is most conspicuous in optic neuritis and distinguishes the reduction of visual acuity caused by optic neuritis from that caused by cystoid macular edema or central serous retinopathy. MG pupils are seen with conditions such as papillitis and anterior ischemic optic neuropathies. The dilation seen in MG pupils is rapid and must be distinguished from the slow dilation of the pupil due to retinal adaption. Patients with strabismus and related types of amblyopia have nearly normal pupillary reflexes; patients with conversion reactions or simulated loss of vision have normal pupillary reflexes.

P: Refer patient for secondary neurological testing or treat the etiology.
General Name: Central serous retinochoroidopathy
Specific Name: Central serous retinochoroidopathy
ICD 9-CM#: 362.41
Location: Posterior pole
Associated Conditions:

S: Decreased visual acuity (if macula is elevated).

O: One or more circumscribed elevations of the pigment epithelium or sensory retina at the posterior pole. The lesions remain unchanged for long periods of time and resolve spontaneously. There may be active proliferation of the pigment epithelium, producing elevated pigmented lesions that suggest chorioretinitis. The lesions tend to disappear leaving a residue of whitish-yellowish deposits deep to a sensory retina or areas of hypopigmentation and hyperpigmentation.

A: Fluorescein angiography indicates one or more areas of leakage in the choriocapillaris. The macular photostress test decreases visual acuity one line or more whereas eyes with optic nerve disease recover within 30 seconds. Pupillary response to light is prompt with choroidopathy whereas in optic nerve inflammation, an afferent pupillary lesion occurs and there is a slow or absent constriction (Marcus Gunn pupil). There is no obvious cause to central serous choroidopathy. Men are affected more than women, especially those with excessively anxious personalities. The peak incidence occurs at about 55 yrs. of age. Both eyes may become involved but usually only one at a time.

P: Photocoagulation of the area gives prompt resolution but most cases resolve spontaneously. Usually only recurrent attacks are photocoagulated.
General Name: Macular Degeneration (Serous detachment of the macula)

Specific Name: Disciform degeneration of the macula

ICD 9-CM#: 362.52

Location: Posterior Pole

Associated Conditions:
Senile macular degeneration, Maculopathy of presumed histoplasmosis, High myopia, Angioid streaks, Traumatic choroidal ruptures, Drusen of the optic nerve, and Retinal dystrophies.

S: The most common cause of poor vision in the elderly (Kuhnt-Junius disease). Both sexes are affected equally. It manifests itself most often in the seventh and eighth decade.

O: Ophthalmoscopic evaluation reveals drusen appearing as small, bright, and sharply defined circular points lying beneath the retinal vessels confined mostly in the posterior pole. These bright yellow to white masses may coalesce to form larger rounded masses, often with some pigmentation around their edges.

A: Typically, disciform degeneration of the macula is seen in elderly patient in good general health. This condition results when the retina and the retinal pigment epithelium are detached from the underlying structures by serous fluid, and the subpigment epithelial space becomes occupied with choroidal neovascular membrane (CNM). These CNM may cause subpigment epithelial hemorrhages and the subsequent organization of these hemorrhages forms the typical subretinal fibrovascular disciform scar around the macular region. Foveal vision is severely impaired, but extrafoveal vision is spared. After approximately one year, the lesion becomes a fibrous mass and vision is reduced to a level of 3/200. The cause of CNM are senile macular degeneration (often associated with drusen), maculopathy of presumed histoplasmosis, high myopia, angioid streaks, traumatic choroidal ruptures, drusen of the optic nerve, and retinal dystrophies.

P: Treatment should follow immediately after identification of CNM with the use of fluorescein angiography. Treatment consists of obliteration of CNM by photocoagulation. However, photocoagulation is effective only if the lesion is one-quarter of a disc diameter away from the fovea. After the initial treatment, patient should be followed at close intervals with repeated fluorescein angiography and, if necessary, retreat if some residual CNM is present. Low vision aids may be useful.
General Name: Pigment Epithelial Detachments
Specific Name: Pigment Epithelial Detachments
ICD 9-CM#: 362.42
Location: Retina

Associated Conditions

S: Decreased visual acuity, distortion, generalized darkening of the visual field.

O: Increased hypermetropia. Decreased recovery from glare as seen with a macular photostress test. Ophthalmoscopy shows a local detachment of the RPE occurring in the macula region. The margins are sharply demarcated and may involve a much greater area than seen in central serous retinochoroidopathy. Flourescein angiography shows that the entire dome of the pigment epithelial detachment lights up early in the angiogram.

A: This condition usually occurs in young men and women 20 - 40 years old. In older patients these detachments occur with drusen and there may be a disciform lesion in the fellow eye. The etiology is unknown.

P: Spontaneous resolution can occur but there may be minor disturbances in visual function associated with disturbance of the RPE. Guidelines for the use of photocoagulation in this condition are not well established, although it has been shown that these detachments can be flattened by a grid of photocoagulation marks covering the entire lesion but avoiding the fovea.
General Name: Macular Degeneration
Specific Name: Dry Senile Macular Degeneration
ICD 9-CM#: 362.51
Location: Posterior Pole
Associated Conditions: Drusen

S: An older patient (over 50) usually complains of not being able to see well.

O: Signs include:
- Decreased VA
- Drusen and pigmentary disturbances at the macula
- Varying stages of retinochoroidal atrophy may be seen, with minor atrophy of the RPE to large punched out areas of complete loss of the choroid, RPE, and sensory retina.

A: It is important to differentiate the condition from wet macular degeneration in which retinal edema is present. The Amsler grid will be normal in dry macular degeneration but metamorphopsia is present in wet macular degeneration.

P: Dry atrophic macular degeneration is an acceptable ageing change in patients over 50. Treatment is symptomatic and consists of rehabilitation with low vision aids. It is also important to monitor precataract VA and be aware of vision potential in case a cataract should become a problem in the future.
General Name: Toxic Maculopathy
Specific Name: Toxic Maculopathy
ICD 9-CM#: 362.55 (use add'l 'E' code, if desired, to identify drug, if drug induced)
Location: Macula
Associated Conditions:

S: Insidious and slowly progressive bilateral loss of function in the central fields with resultant diminished acuity and central scotomas. Chloroquine is an antimalarial agent causing adverse ocular reactions such as corneal epithelial opacity, decreased vision, macular or retinal pigmentary degeneration, and visual field defects. Chlorpromazine (thorazine) is an antipsychotic agent (a phenothiazine) causing corneal endothelial opacity, decreased vision, dyschromatopsia, lacrimation, anterior lens pigmentary deposits, myopia, oculogyric crises, paralysis of accommodation, photosensitivity, pigmentary retinopathy and punctate keratitis.

O: Chloroquine is concentrated in the eye and has a particular affinity for the pigmented structures. Chloroquine keratopathy (greyish in color) is observed primarily in patients receiving dosages greater than 250 mg daily for rheumatoid arthritis or systemic lupus. Chlorpromazine produces a brownish haze over vision and macula area (speckled pigmentation). Also pigment deposits between the equator and the posterior pole, with subsequent retinal edema, loss of night vision and decreased vision is seen 4 to 5 weeks later. Unfortunately, once ocular toxicity has developed, it usually does not regress even if the drug is withdrawn. Also, loss of foveal reflex and increased pigmentation in the macula. Eventually, a ring of depigmentation develops, resembling a “bull’s-eye” target. Decreased visual acuity. Central, paracentral, and peripheral scotomas may be present. Color vision may be abnormal, especially red. Possible enlarged A wave or a reduced B wave in ERP, and the EOG depressed. Dark adaptation is usually normal even in late stages. Diplopia due to paresis of extraocular muscles has been reported.

A: The most important difference is the phenothiazine toxicity occurs after a relatively short time and chloroquine toxicity occurs after a long period. Furthermore, chloroquine toxicity is much less reversible than that due to phenothiazine. Rule out Nutritional/Tobacco/Alcohol Amblyopia, Stargardt's disease, Progressive cone dystrophy.

P: Treatment consists mainly of discontinuing the medication.
General Name: Toxic Maculopathy

Specific Name: Nutritional/Tobacco/Alcohol Amblyopia

ICD 9-CM#: 362.55

Location: Macula

Associated Conditions:

S: Regarding the etiology, the great weight of clinical evidence overwhelmingly favors a dietary deficiency of B-complex vitamins (predominantly thiamine) rather than the direct toxic effects of tobacco or chronic alcoholism. The fundus appears perfectly normal.

O: Bilateral, relatively symmetric centrocecal scotomas, with preservation of the peripheral field, is the characteristic field defect encountered in nutritional-toxic neuropathy. The defects are characterized by "soft" margins which are difficult to define for white stimuli but are larger and easier to plot for colored targets, especially red. A dense "nucleus" may at times be found between the blind spot and the fixational area, but nerve fiber bundle defects do not occur. Visual acuity is usually reduced to 20/200 levels but may be surprisingly good despite a symptomatic central defect, in which case the scotoma may be demonstrable with red targets only. Diplopia due to paresis of extraocular muscles has been reported.

A: Nutritional neuropathies differ from chiasmal interference in that visual acuity is diminished, the defects extend across the vertical meridian (especially demonstrable with red targets), there is no peripheral hemianopic depression, and the defects appear more cecocentral and less hemianopic as the defects progress. Rule out Chloroquine & Chlorpromazine Toxic Maculopathy, Stargardt's disease, Progressive cone dystrophy.

P: Prognosis for recovery is excellent for all but the most chronic cases. Treatment consists of a well-balanced diet and B-complex (e.g. B12) vitamin supplement. Among the least expensive preparations is yeast, either in powder form or tablets (500 mg, 20 tablets/day). Intramuscular thiamine may also be used.
General Name: Best's Disease
Specific Name: Vitelliform Macular Dystrophy
ICD 9-CM#: 362.76
Location: Macula
Associated Conditions: None

S: Vision is normal in early stages progressing toward complaints of loss of central vision.

O: In the early stages, the ophthalmoscopic view presents a yellowish accumulation of material underneath the macula. It has often been described to have an egg yolk appearance. The second stage is a result of a rupture of the lesion and resembles what is called "scrambled eggs" along with mottling of the macula.

A: Vision is normal during the egg yolk stage. However the EOG is decreased and a mild disturbance in the patient's dark adaptation is observed. Between the ages of 7 and 15, the material is dispersed into the scrambled egg. Scarring and retinal pigment epithelial dispersion occurs at this time. The definitive test shows a decreased EOG as mentioned previously with a normal ERG.

P: The prognosis for this dystrophy is poor. Best's disease is an autosomal dominant disorder. In most cases it will progress into the scrambled egg appearance with loss of central vision. However some cases have shown only decreased EOG without any gross lesions. Explanation of the disease's course and genetic counseling should be given.
General Name: Stargardt's disease  
Specific Name: Fundus Flavimaculatus  
ICD 9-CM#: 362.75  
Location: Fundus  
Associated Conditions:

S: Decreased visual acuity and color vision.

O: Ophthalmoscopy reveals multiple round and linear, pisciform, yellow or yellow-white lesions, usually involving the posterior fundus. These flecks vary in size, shape, outline, density, and apparent depth. Flecks may also appear and can cause a decrease in vision due to deposition beneath the macula causing secondary changes to the overlying cones. The deposits usually first appear in the perimacular area and are isolated by fairly sharp borders and variable shapes, many of them linear and fishlike. Size, shape, and amount of the flecks vary a lot as new ones are formed and old ones fade.

A: The condition is transmitted primarily autosomal recessively but occasionally dominantly. Fluorescein angiography is only useful on older lesions since new ones do not fluoresce. ERG may be normal. EOG is usually subnormal but not as bad as Vitelliform disease. The disease usually comes on before 20 or 30 yrs. of age. Differential diagnosis includes fundus albipunctatus, drusen, and Kandori's fleck retina.

P: There is no therapy for this disease.
General Name  Primary Retinal Dystrophy
Specific Name  Cone Dystrophy
ICD 9-CM#  362.75
Location  Posterior Pole
Associated Conditions
Retinitis pigmentosa, Bull's eye macula, Decreased color vision

S: Symptoms include:
- poor vision
- poor color vision
- photophobia
- bilateral condition

O: Ophthalmoscopy may show a bull's eye macula with a ring-like depigmentation around the macula. The end stage of cone dystrophy has the same appearance as retinitis pigmentosa.

A: Retinal dystrophies are genetically transmitted diseases of the retina that lead to premature cell changes and cell death. In cone dystrophy, the cone system is initially affected, with changes in the rod system later in the course of the disease. Electrophysiologic tests ERG and EOG are important in diagnosing these disorders. Differential diagnosis from Retinitis Pigmentosa is indicated.

P: There is no effective treatment.
**General Name** Rubella  
**Specific Name** Congenital Rubella Syndrome  
**ICD 9-CM#** 055  
**Location** Generalized  
**Associated Conditions**  
Nuclear Cataract, Microphthalmia, Infantile Glaucoma, Corneal Opacities, Nystagmus, Strabismus

**S:** Widespread fetal ocular and systemic defects.

**O:** Ocular findings include nuclear cataract with surrounding zone of clear cortex with gradual progression to become entirely pearly white, microphthalmia, retinal hyper and hypopigmentation, infantile glaucoma, corneal opacities, nystagmus and strabismus. Nonocular findings include neurosensory deafness (congenital and acquired in early childhood), congenital heart disease, thrombocytopenia and consequent purpura, bone lesions, hepatitis, hemolytic anemia, CNS involvement, generalized rash with seborrheic features, interstitial pneumonitis, lymphocytic infiltration to the pancreas and panencephalitis.

**A:** Rubella infection during pregnancy may cause many malformations at birth. The severity of the complications varies with different strains of the virus and is more severe the earlier in the pregnancy infection occurs. Approximately 50% of mothers with clinical evidence of infection during the first month of gestation will bear offspring with congenital defects.

**P:** Ophthalmologic care of these children starts with attention to the cataracts and glaucoma in infancy. Since the response of infantile glaucoma to medical therapy is nearly always poor, surgical treatment is usually necessary. When cataracts are present they should be aspirated in the first few months of life. All lens cortex should be removed at the initial operation to prevent an endophthalmitis. Once a patient with congenital rubella has had surgery for glaucoma or cataracts it is essential that he be followed regularly throughout life to watch for any onset of intraocular inflammation and/or glaucoma.
S: Main patient complaint is headaches. If the central retina is involved decreased vision is also reported.

O: Signs include:
- Acute onset of multiple flat or slightly elevated yellow-white lesions of the posterior pole at the level of the pigment epithelium. The lesions resolve spontaneously leaving extensive pigment epithelial degeneration, with minimal alteration of the choroid or sensory retina.
- Possible associated mild uveitis with keratic precipitates and cells and flare in the aqueous and vitreous.

A: The disease particularly affects young women who have had an upper respiratory tract infection in the past 1-2 weeks. When this particular history is present with the fundus findings the patient should be referred to an ophthalmologist for a fluorescein angiography. In early stage the inflammation blocks the choroidal fluorescence. In the late stage the lesions gradually pick up the dye and fluoresce. After healing takes place transmission defects remain in the PE without late staining or leakage.

P: The condition resolves spontaneously, but the damage to the PE is permanent. If the central retina was involved the visual acuity shows improvement over several weeks as the condition resolves.
General Name: Acute Retinal Pigment Epithelitis
Specific Name: Acute Retinal Pigment Epithelitis
ICD 9-CM#: 362.50
Location: Retina
Associated Conditions: None

S: A rapid onset decrease in central vision.

O: An ophthalmoscopic examination reveals single to multiple lesion of the pigment epithelium, most commonly at the posterior pole. These lesions appear as deep, fine, dark spots in the acute stage and in later stages are surrounded by yellow rings.

A: The disease affects primarily males at the age of approximately 45. Bilateral involvement is seen in 38% of cases. Fluorescein angiography shows transmission defects in the RPE which do not leak or change in shape.

P: This is a self limited benign condition which resolves somewhat in 6 to 12 weeks. Visual prognosis is from blurred vision to little effect on vision. Treatment is supportive and depends on the final resolution results.
General Name: Senile macular hole
Specific Name: Macular hole
ICD 9-CM#: 362.54
Location: Macula
Associated Conditions:
Possibly, conditions where vitreous traction on the macula exist. With macular cysts, there is a 40% chance that in 2 years, a hole will form.

S: Decreased visual acuity. Interruption of a slit beam focused on the hole during contact lens biomicroscopy.

O: Visual acuity worse than 20/100. A surrounding rim of subretinal fluid around the macula. There are dots of tissue at the base of the hole, and the patient will report an interruption of a slit beam focused on the hole during contact lens biomicroscopy. Fixation of a point beam by the patient during contact lens biomicroscopy is usually wandering and eccentric.

A: With longstanding holes, the pigment epithelium is usually atrophic and there is a transmission defect on fluorescein angiography. Lamellar macular holes can appear similar to full-thickness holes. However, vision is usually better and occasionally 20/20. With lamellar holes, since many of them are actually circular dehiscences in epiretinal membrane, in these cases, a preretinal membrane may be seen surrounding the apparent hole (on slit lamp biomicroscopy). Although macular holes are unquestionably associated with trauma, the majority probably occur as an end stage of macular degeneration. Some ophthalmologists believe that vitreous traction plays a role in the formation of macular holes; such traction has been demonstrated histologically. However, these changes are seldom seen on clinical examination.

P: Even full-thickness macular holes don't lead to progressive retinal detachment. The only exception to this is in very myopic eyes and in an occasional eye with a large macular hole caused by trauma. The vision is seldom worse than 20/400 and usually does not deteriorate below that level.
No treatment is indicated. There is no medical or surgical treatment for macular holes except the use of visual aids.
General Name  Vitreoretinal macular diseases
Specific Name  Idiopathic preretinal macular fibrosa
ICD 9-CM#  362.50
Location  Posterior Pole
Associated Conditions
Vitreous detachment, Secondary to local vascular or inflammatory changes, Post photocoagulation, Post retinal detachment surgery.

S: Symptoms include:
- Blurred vision
- Distorted vision (metamorphopsia).

O: Ophthalmoscopy reveals:
- Glinting reflex
- Traction lines
- Mild, gray preretinal fibrosis in the macula.

A: Disturbances at the vitreoretinal interface are common causes of reduced central vision in elderly patients. The condition may arise from the following conditions:
- Spontaneously
- Vitreous detachment
- Secondary to local vascular or inflammatory changes.
- Post photocoagulation.
- Post retinal detachment surgery

P: There is no treatment for this condition.
**General Name:** Retinoblastoma  
**Specific Name:** Retinoblastoma  
**ICD 9-CM#** 190.5  
**Location:** Retina  
**Associated Conditions**

**S:** Parents or pediatrician notices a white reflex in the pupil.

**O:** The most common presenting sign is a white reflex in the pupil. Strabismus is the second most common mode of presentation. Apparent intraocular inflammation and glaucoma secondary either to the tumor pushing the diaphragm forward or to the tumourcells clogging the trabecular meshwork are also relatively often seen. Less common are proptosis, pseudohypopyon of tumor cells, and evidence of distal metastases. These all indicate a poor prognosis for life. Ophthalmoscopically the tumor presents as a single or more commonly, multifocal, smooth, pinkish, rounded mass growing in, on top of, or below the retina.

**A:** Retinoblastoma is the most frequent ocular tumor of childhood. The incidence is about 1 in every 20,000 live births. It has one of the highest cure rates of any malignant tumor. Untreated, it is almost invariably fatal. Retinoblastoma is inherited as an autosomal dominant with greater than 90% penetrance. Of all cases of retinoblastoma, 60% are nonhereditary and 40% are hereditary. If untreated, the tumors most commonly metastasizes to the bone marrow or extends back through the optic nerve into the subarachnoid space and spreads via the cerebrospinal fluid throughout the CNS.

**P:** If it is thought that the eye can be saved with useful vision, then radiotherapy is the treatment of choice. If the tumor is unfavorable to radiation therapy, then enucleation is the treatment of choice. The role of chemotherapy in retinoblastoma is uncertain. Genetic counseling should be provided to all patients with a family history of retinoblastoma.
General Name  Vitreous Detachment
Specific Name  Posterior Vitreous detachment
ICD 9-CM#  379.21
Location  Vitreous
Associated Conditions
Retinal tears, Aphakia, Vitreal retraction, Periretinal Proliferation

S: Patient complains of seeing floaters and flashing lights which are more noticeable when the eyes are closed or when in a dim room. Others may notice a shower of floaters or blurred vision in the affected eye.

O: With ophthalmoscopy the original site of attachment of the vitreous to the margin of the optic disk may be seen as a ring shaped opacity on the posterior hyaloid. Examination with the 90D Volk or Hruby lens may reveal strands extending from the anterior hyaloid to about 1/3 into the vitreous. With the slit lamp using an optic section one may see the floater in front of an optically empty space next to the posterior retina.

A: Posterior vitreous detachment most commonly occurs in middle aged individuals. Diabetes is a less common cause of PVD. Retinal tears are present in 10-15% of patients with acute symptomatic PVD. ~5% have vitreous hemorrhage without retinal breaks. PVD is a serious problem as it can lead to retinal detachment via retinal breaks. There is higher incidence of PVD in aphakic eyes (80%) vs phakic eyes (31%) in individuals over 65. It is likely to occur earlier in axial myopes than in emmetropes.

P: It is best to do careful exam with a BIO and scleral depression. Also inspect closely for vitreous hemorrhage with the Hruby lens. If there is no hemorrhage and symptoms have been present for 3 months or more see patient back in 3 months or sooner if symptoms are more acute. The most severe condition that can follow a PVD is a massive vitreal retraction or massive periretinal proliferation. The condition leads to fixed folds in the retina and proliferation of pigment epithelium anterior and posterior to the sensory retina and irreversible blindness is the outcome. Careful monitoring of the patient must be undertaken to prevent such an occurrence. The patient must also be instructed to RTC if they notice an increase in floaters or if they perceive a curtain or shadow in the peripheral field of vision which is indicative of a retinal detachment.
General Name: Asteroid Hyalitis
Specific Name: Benson's Disease
ICD 9-CM#: 379.22
Location: Vitreous
Associated Conditions

S: The presence of asteroid hyalitis does not often give rise to symptoms and if the patient does have symptoms it is reported as "floaters".

O: When observed with the biomicroscope they appear as spherical and stellate opacities and sparkle like Christmas ornaments in the slit illumination. They are best described as "snowball" opacities. When the patient is requested to move the eye the opacities move but always return to the original position once the eye stops moving.

A: The condition of asteroid hyalosis is first observed most commonly in males between the ages of 60 and 65. It is unilateral in 75% of patients. The observed oval white bodies of varying sizes are adherent to the framework of the normal vitreous gel and may be scattered throughout the entire vitreous cavity or may be accumulated in only one part of it. The opacities consist chiefly of calcium soaps and do not appear to disturb visual function. At one time it was thought that this condition had relation to diabetes and hypercholesterolemia. Studies seem to indicate there is little connection between asteroid hyalitis and these diseases. Rule out Scintillating Scintillans, Muscae Volitantes, Cholesterolosis Bulbi.

P: There is no treatment for this condition.
General Name: Cholesterosis Bulbi
Specific Name: Synchysis Scintillans
ICD 9-CM#: 379.22
Location: Vitrious
Associated Conditions: None

S: The patient is usually asymptomatic except for complaints of floaters.

O: Upon observation with slit lamp or ophthalmoscope, free floating opacities can be observed in the vitreous. These opacities have a flat, angular, crystalline appearance. These crystals settle on the bottom of the vitreous cavity upon lack of eye movement.

A: This is a rare condition which occurs after inflammation, chronic degeneration, trauma, and hemorrhage. Cholesterosis bulbi typically is associated with a fluid vitreous but may be seen with an intact vitreous as crystals on the iris and retina. This condition is usually bilateral and occurs in patients younger than 40. It is not associated with such lipid abnormalities as juvenile arcus or xanthelasma.

P: There is no known effective treatment.
General Name: Liquification of the vitreous, Syneresis
Specific Name: Syneresis
ICD 9-CM#: 379.21
Location: Vitreous
Associated Conditions: Myopia, injuries and inflammation of the eye.

S: Floaters.

O: Floating vitreous fibrillar structures in the vitreous are seen with ophthalmoscopy or with the slit lamp.

A: By clinical appearance. The vitreous humor becomes partially or completely fluid, creating the appearance of membranes or strands floating freely in the fluid. The condition is caused by release of water by the vitreous fibrillar structure, which then float in the water. These particles float across the visual line, impair vision, and may be extremely vexing. They are easily visible with the ophthalmoscope or slit lamp.

P: The symptoms may be relieved by wearing an appropriate lens for any ametropia present, but no treatment will restore the integrity of the vitreous. The patients must be monitored for vitreal detachments following liquification.
**General Name** Floaters  
**Specific Name** Muscae Volitantes  
**ICD 9-CM#** 379.24  
**Location** Vitreous  
**Associated Conditions**  
Floaters, embryonic remnant, entoptic phenomenon

**S:** Patient complains of floaters which drift in and out of the view and dart away with attempted fixation. It is especially noticeable on viewing a bright uniform field such as a cloudless sky or a pastel-shaded wall.

**O:** Floaters maybe observed in the vitreous.

**A:** Muscae volitantes is an entoptic phenomenon. Their presence is attributed to minute remnants of embryonic structures in the vitreous humor. Most vitreous opacities are harmless and the patient needs nothing more than reassurance. However, it may be of serious importance when patient complains of vitreous opacities accompanied by flashes of light which may indicate retinal detachment.

**P:** Reassure patient that it is harmless and a common occurrence. Correction of ametropia makes the opacities more difficult to observe.
General Name: Hemorrhage
Specific Name: Vitreous Hemorrhage
ICD 9-CM#: 779.23

Location: Vitreous

Associated Conditions:
Diabetic Retinopathy, Retinal Tear, Posterior Vitreous Detachment, Retinal Vein Occlusion, Sickle Cell Retinopathy, Congenital Retinal Vascular Anomalies, Trauma, Disciform Macular Lesions, Choroidal Malignant Melanoma, Subarachnoid Hemorrhage.

S: Symptoms depend upon the location of the vitreous hemorrhage. Peripheral hemorrhage causes floaters, whereas blood in the visual axis markedly reduces vision and may cause a red haze. Visual loss may result because of persistent opacification, inflammation, glaucoma, siderosis, of fibrous tissue proliferation and contraction that causes retinal detachment.

O: Ophthalmoscopically, the blood may be evenly dispersed throughout the vitreous humor, localized, or distributed in sheets. Blood in longer standing vitreous hemorrhage often becomes converted to a white opaque mass resembling an inflammatory exudate, endophthalmitis, or an intraocular tumor. A detailed history along with complete ocular examination combined with ultrasonography usually establishes the diagnosis.

A: Vitreous hemorrhage and membrane formation are seen in a number of ocular conditions. Diabetic retinopathy is the most important cause, followed by retinal tear.

P: In a younger person resorption may be rapid, but persistent or recurrent hemorrhage are followed by yellow or white debris and fibrous membrane formation. Intravitreal injection or urokinase is the preferred method of treatment in diabetes and hypertension. If the injection does not work then vitrectomy is the next treatment of choice because nonresorbing vitreous blood carries a high complication rate. Vitrectomy through the pars plana can remove the diseased, opacified vitreous and membranes.
General Name Hyperplastic Vitreous
Specific Name Persistant Hyperplastic Primary Vitreous
ICD 9-CM# 379.25
Location Vitreous
Associated Conditions
Strabismus, Microphthalmia, Secondary Glaucoma, Secondary Cataract

S: Parents report that the full term infant has little or no vision in one eye and that the pupil appears white in the affected eye.

O: Signs include:
- Strabismus and microphthalmia of the affected eye.
- Pinkish-white mass immediately behind the lens which gives a leukocoric appearance. the mass may vary in size from a small plaque to one covering the entire posterior lens surface.
- Elongated ciliary processes extending into the mass are visible upon dilation.
- Anterior chamber appears shallow and the lens appears smaller than normal.
- Tissue behind the lens is well vascularized and vessels radiate outwards.

A: The etiology of the condition is a failure of the hyaloid artery to regress, with associated hyperplasia of the tunica vasculosa lentis. It is usually noted at birth or shortly after. 90% of the cases are unilateral. The early onset, elongation of the ciliary processes, and microphthalmia in a full term normal weight infant differentiates PHPV from retinoblastoma which has a later onset and no associated microphthalmia; and from retrolental fibroplasia which is present in low birthweight premature infants. Late complications include rupture of the posterior capsule leading to cataract and secondary glaucoma and buphthalmos.

P: Early removal of the lens via extracapsular extraction followed by excision of the retrolental membrane is recommended by several authorities to save the eye. Although useful vision is rarely obtained the lens must be removed in order to avoid the need for enucleation due to secondary glaucoma. The surgery should be done as early as possible (1 day to 4 weeks of age) and patients should be fit with a soft contact lens post surgically in order to attempt to prevent amblyopia.
General Name  Vitreous Prolapse
Specific Name  Vitreous Prolapse
ICD  9-CM#  379.26
Location  Vitreous
Associated Conditions  Bullous Keratopathy, Secondary Glaucoma, Retinal Detachment, Vitreous Detachment, Post-ICCE surgery.

S: Asymptomatic. Vitreous prolapse describes a condition where the vitreous pours into the anterior chamber via the pupil. If the vitreous were to touch the endothelium, endothelial damage would result which can lead to bullous keratopathy of the cornea; blur may, therefore, result due to a hazy cornea.

O: Another problem which can result is secondary glaucoma when the vitreous impedes the normal flow of aqueous. Retinal detachment may also manifest if the protruding vitreous is still attached to the ora seratta, iris damage may also result.

A: In previous years when intracapsular cataract extraction had been the procedure of choice for the removal for most cataractous lenses, this was a very common secondary complication.

P: Today, the incidence of vitreous prolapse has greatly decrease due to the change in the procedure of choice for the removal of cataractous lenses from the former intracapsular cataract extraction to the extracapsular cataract extraction which leaves behind the posterior capsule of the cataractous lens. As a result, there is now a "retaining wall" to decrease the chance of the vitreous from pouring into the anterior chamber. Surgery may be indicated if any of the above complications manifests.
General Name: Retinitis Proliferans
Specific Name: Retinitis Proliferans
ICD 9-CM#: 362.29
Location: Vitrious
Associated Conditions: Retinal Hemorrhage

S: The patient complains of reduced vision. In the severest case, the patient will present himself with total blindness.

O: Examination reveals a proliferation of the blood vessels and capillaries into the vitrious. A fibrous response of the connective tissue is observed from the retina into the vitrious. Evidence of a retinal hemorrhage is often present.

A: Retinitis proliferans is usually associated with trauma. It results from a retinal hemorrhage which ruptures into the vitrious. Similar neovascularization and fibrosis into the vitrious occurs in proliferative diabetic retinopathy and retinopathy of prematurity.

P: Once a vitreal hemorrhage and the resulting fibrosis takes place, the prognosis is poor. A vitrectomy is often performed with limited success.
General Name: Granulomatous uveitis
Specific Name: Granulomatous uveitis
ICD 9-CM#: 364.10
Location: Uveal tract
Associated Conditions:
Trauma or infections of the eye, systemic conditions, intraocular abnormalities such as hypermature cataract or uveal tissue necrosis, or because of an immunologic response of a previously sensitized region, endophthalmitis phacoanaphylactica.

S: Visual loss, pain (usually with acute uveitis), photophobia and lacrimation.

O: Anterior uveitis: ciliary injection, exudation into the anterior chamber, iris changes such as constriction or mid-dilation, edema, nodules, diffuse iris atrophy, loss of iris pattern, posterior synechiae.
Posterior uveitis: vitreous opacities and choroiditis.

A: Granulomatous uveitis has an insidious onset and is chronic. The KP's are of the muttonfat type and are large. There are frequent Koeppe and Busacca nodules. On the fundus, nodules will also be found as well as areas of retinitis and vascular sheathing. Diagnosis is usually confirmed by biomicroscopic and ophthalmoscopic examination. The endogenous etiologic factor may be immediately evident when associated with systemic disease, but in most instances, it is impossible to demonstrate. This is complicated by the inability to secure uveal tissue for study and often the cause is presumed.
Some systemic conditions associated with uveitis include: Sarcoidosis, Arthritis, Genitourinary tract disease, Behcet disease, Reiter disease, Histoplasmosis, and Pars planitis.

P: If possible specific treatment is carried out. For inflammation of unknown cause that is confined to the iris and the ciliary body, the pupil is dilated and accommodation paralyzed by topical instillation of 1% atropine solution. Full pupillary dilation is desired to prevent posterior synechiae. Corticosteroid preparations are instilled at frequent intervals. Subconjunctival injections may be used. IOP must be monitored and acetazolamide and timolol must be used in individuals with a genetic sensitivity to corticosteroids. Systemic corticosteroids are the main treatment for posterior uveitis. Retrobulbar or sub-tenon injection of corticosteroids and, rarely systemic cytotoxic anti-inflammatory agents may be used. Early syphilitic inflammations respond readily to antibiotic. TB responds to ethambutol and similar agents, but there are few cases in which they are indicated. Antibiotics are not administered empirically.
General Name: Uveitis
Specific Name: Non-granulomatous uveitis
ICD 9-CM#: 364.32
Location: Uveal Tract
Associated Conditions: Acute angle closure glaucoma, trauma, conjunctivitis, post surgery

S: Symptoms include pain (due to ciliary spasms), photophobia, lacrimation, blurred vision (due to aqueous flare and keratic precipitates), and red eye. Uveitis is most commonly seen in patients ages 20-50 with marked decrease in individuals over 70 years of age.

O: There are two types of non-granulomatous uveitis: Acute and Chronic.

Slit lamp evaluation shows:

<table>
<thead>
<tr>
<th>Type</th>
<th>ACUTE</th>
<th>CHRONIC</th>
</tr>
</thead>
<tbody>
<tr>
<td>aqueous</td>
<td>many cells and flare</td>
<td>moderate # of cells, few flare</td>
</tr>
<tr>
<td>keratic deposits</td>
<td>lymphoid cell deposits</td>
<td>lymphocytic cell deposits</td>
</tr>
<tr>
<td>posterior synechiae</td>
<td>none, or easily broken</td>
<td>slowly forming</td>
</tr>
<tr>
<td>iris changes</td>
<td>acute capillary dilation</td>
<td>slight capillary dilation</td>
</tr>
</tbody>
</table>

Other clinical findings:
- pupil: irregular

A: Differential diagnosis granulomatous vs. nongranulomatous in orienting toward the proper therapy. Other causes of red eyes must be ruled out. e.g. conjunctivitis, scleritis, episcleritis, acute angle glaucoma, and keratitis.

- In conjunctivitis, vision is not blurred, pupillary responses are normal, a discharge is present, and there is no pain, photophobia, or ciliary injection.
- In scleritis, the injection is more toward the fornices.
- In episcleritis, there is no photophobia, the anterior chamber is clear, and the injection is more toward the fornices.
- In acute angle glaucoma, the pupil is dilated, the cornea is steamy, and there is a sudden decrease in vision.
- In keratitis, vision may be blurred and photophobia may be present. Pain is relieved with topical anesthetic. There is also corneal staining.

P: Treatment consists of:

1) Cycloplegics (1% Atropine, 5% Homatropine, or 1% Cyclopentolate) for comfort by relieving iris sphincter and ciliary muscle spasm and to prevent posterior synechiae.
2) Topical steroids for controlling inflammation.
3) Symptomatic measures:
   a) warm compress for 10 minutes 3-4 times a day
   b) systemic analgesics
   c) dark glasses for photophobia
4) Counsel and advise patient on the nature, severity, and duration of the pain, and also the clinical course of the disease.
General Name Uveitis
Specific Name Anterior Uveitis
ICD 9-CM# 364.0
Location Iris and Ciliary Body
Associated Conditions Sarcoidosis, Arthritis, Genitourinary Tract Disease, Toxoplasmosis, Histoplasmosis, Necrosis of Intraocular Tumor, Trauma, Inflammation of Adjacent Tissues, Disease of the Lens, Blind Eye with Degenerative Changes

S: Ocular pain due to ciliary spasms, photophobia, lacrimation, hyperemia and blurred vision due to aqueous flare. Pain is more common in acute than chronic iridocyclitis and is particularly severe when associated with keratitis. Photophobia varies in severity.

O: Perilimbal flush. Cells and flare in the anterior chamber with occasional hypopyon formation. Keratic precipitates on corneal endothelium and trabecular meshwork. The iris may be edematous with engorged capillaries and muddy appearance. Koepppe and Busacca nodules. The pupil is miotic or irregularly shaped due to posterior synechiae.

A: The diagnosis of anterior uveitis is usually confirmed by bimicroscopic examination and observation of keratic precipitates, aqueous flare and cells. A variety of inflammations and diseases involving the body, eye or its adnexia may be associated with the uveitis.

P: If the uveitis is associated with a systemic or a local disorder disease, then specific treatment should be addressed to the disease process. For inflammation to unknown cause that is confined to the iris and ciliary body, the pupil is dilated to prevent posterior synechiae and accommodation is paralyzed to prevent ciliary spasms. There is a spectrum of drugs that can be used for the therapeutic purpose of cycloplegia depending upon the severity of the uveitis. It is useful to utilize a cycloplegic that will provide the iris with some movement so that the pupil can be somewhat mobile and prevent a dilated pupil posterior synechiae. For this reason atropine is oft avoided in favor of 5% homatropine. A spectrum of steroids can be administered depending upon the degree of severity of the uveitis. Subconjunctival injections may be used. Ocular tension must be measured regularly and acetazolamide and timolol must be used in individuals with a genetic sensitivity to corticosteroids.
**General Name** Posterior Uveitis

**Specific Name** Acute Posterior Uveitis

**ICD 9-CM#** 363.20

**Location** Uvea

**Associated Conditions**
Toxoplasmosis, Histoplasmosis, Toxocariasis

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**S:** Patient suffers a loss of vision, and may complain of a red eye with little or no pain.

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**O:** Acute Posterior Uveitis- Patients show:
- Decreased visual acuity
- Posterior chamber flare.
- Vitreous opacities: consist of fibrin, cells and exudates and are visible with the biomicroscope in combination with a concave lens. Vitreous involvement is usually slight or intensely generalized.
- Chorioretinitis: appears as a nebulous grayish white area surrounded by normal fundus. May have single or multiple lesions often seen with marked retinal and subretinal edema. After healing has occurred there is little residual damage.

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**A:** Uveitis is usually seen most frequently between the ages of 20-50. It is usually more common in males than females, with the posterior type being 1/4 as common as the anterior type. Posterior uveitis is most frequently associated with endogenous causes such as toxoplasmosis, histoplasmosis, and toxocariasis. Diagnosis is based on fundus appearance and diagnosis of associated conditions using appropriate lab tests. Differential diagnosis from anterior uveitis is straightforward since posterior uveitis is relatively asymptomatic as compared to anterior uveitis.

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**P:** Treatment for posterior uveitis consists of periocular and oral steroids since topical steroids don't have the required penetration. They can be administered by injection in a retrobulbar fashion. The advantage to administration by injection is that the steroids are placed close to the area of inflammation. Systemic steroids are less desirable since they produce numerous side effects. In either case the patient must be monitored while of steroid treatment for increased IOP and posterior subcapsular cataract formation. Referral to a physician is in order.
General Name: Posterior Uveitis
Specific Name: Chronic Posterior Uveitis
ICD 9-CM#: 363.20
Location: Uvea
Associated Conditions: Toxoplasmosis, Histoplasmosis, Toxocariasis

S: Patient suffers a loss of vision, and may complain of a red eye with little or no pain.

O: Chronic Posterior Uveitis—Patients show:
- Decreased visual acuity
- Vitreous opacities: With the chronic type the vitreous opacities are usually heavy, and veil-like opacities are frequent.
- Chorioretinitis: In chronic uveitis, heavy massive choroidal exudates with blurred edges due to accompanying slight retinal edema are present. Often there is secondary retinal involvement which results in retinal destruction. After the condition resolves glial scars remain with massive pigment epithelial proliferation around the lesion.

A: Uveitis is usually seen most frequently between the ages of 20-50. It is usually more common in males than females, with the posterior type being 1/4 as common as the anterior type. Posterior uveitis is most frequently associated with endogenous causes such as toxoplasmosis, histoplasmosis, and toxocariasis. Diagnosis is based on fundus appearance and diagnosis of associated conditions using appropriate lab tests. Differential diagnosis from anterior uveitis is straightforward since posterior uveitis is relatively asymptomatic as compared to anterior uveitis.

P: Treatment for posterior uveitis consists of periocular and oral steroids since topical steroids don't have the required penetration. They can be administered by injection in a retrobulbar fashion. The advantage to administration by injection is that the steroids are placed close to the area of inflammation. Systemic steroids are less desirable since they produce numerous side effects. In either case the patient must be monitored while on steroid treatment for increased IOP and posterior subcapsular cataract formation. Referral to an ophthalmologist is in order.
<table>
<thead>
<tr>
<th>General Name</th>
<th>Panuveitis</th>
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<tbody>
<tr>
<td>Specific Name</td>
<td>Panuveitis</td>
</tr>
<tr>
<td>ICD 9-CM#</td>
<td>360.12</td>
</tr>
<tr>
<td>Location</td>
<td>Uveal Tract</td>
</tr>
<tr>
<td>Associated Conditions</td>
<td>Anterior Uveitis, Posterior Uveitis.</td>
</tr>
</tbody>
</table>

**S:** Patient's subjective symptoms are the same as those in both anterior and posterior uveitis combined. Occasionally, panuveitis may occur in the pediatric age group, although usually in children the inflammatory signs of uveitis are localized to anterior, posterior, or peripheral portions of the uveal tract.

**O:** Patient's objective signs are the same as those in both anterior and posterior uveitis combined.

**A:** A collective term used when the entire uveal tract is inflamed.

**P:** Treatment of this condition is the same as those for both anterior and posterior uveitis.
General Name Uveitis
Specific Name Syphilitic Uveitis
ICD 9-CM# 091.50
Location Uveal Tract
Associated Conditions
Systemic Syphilis

S: In congenital syphilis, the parents will often notice a bilateral red eye in the child. In secondary syphilis, the patient will complain about an acute onset red, painful eye which can be bilateral or unilateral.

O: The anterior segment upon examination demonstrates a circumcorneal ciliary flush. The pupil is usually miotic and cells and flare are seen in the anterior chamber. Fine to course KP bodies can be seen on the corneal endothelium.

A: The uveitis associated with syphilis presents the signs and symptoms of a typical anterior uveitis. The diagnosis of syphilis comes with the observation of the uveitis with a chorioretinitis and interstitial keratitis in the congenital case. In an adult case, an unresponsive, chronic, anterior uveitis should be suspect. Also the typical genital lesions or symptoms of syphilis may be present. In both cases, the definitive diagnosis can be determined by a positive FTA-ABS test.

P: The underlying syphilis should be treated with penicillin once diagnosed. The uveitis is usually treated with cycloplegics and if necessary topical steroids.

mild ant. uveitis: 1% tropicamide, 1% dexamethasone NaP
moderate: 5% homatropine, 1% prednisolone NaP or .1% fluorometholone
severe: 1% atropine, 1% prednisolone acetate
General Name  Iridocyclitis
Specific Name  Herpes simplex iridocyclitis
ICD 9-CM#  054.44
Location  Uveal Tract
Associated Conditions  Panuveitis, Herpes simplex keratitis, Synechiae

S: Symptoms include:
- red eye
- pain
- photophobia
- lacrimation

O: Slit lamp examination reveals:
- keratic precipitates
- aqueous flare and cells
- clouding of the vitreous.
Ophthalmoscopic observation reveals:
- chorioretinal inflammatory lesion.

A: Iridocyclitis and active keratitis often occur concomitantly in herpes simplex infection. Occasionally panuveitis may also occur. It is an inflammation of the iris and the ciliary body. The etiology is unclear but it is thought to be an immune inflammatory reaction against a viral antigen. Diagnosis is based on evidence of Herpetic lesions elsewhere on the body e.g. vesicular facial lesions and the cornea (dendritic ulcers, stromal interstitial keratitis, stromal immune disciform keratitis).

P: Treatment consists of:
1. Topical antivirals: Although this is the best therapy available, it is far from ideal due to the fact that it does not penetrate in sufficient therapeutic titer.
2. Cycloplegic mydriatic therapy: e.g. atropine 1.0%-4.0% bid or scopolamine 0.1% bid to prevent synechiae formation and relieve pain.
3. Nonsteroidal anti-inflammatory drugs: e.g. indomethacin or aspirin to help avoid structural damage.
4. Steroids: If the patient has never been treated with steroids, the drug should be avoided. However it is indicated in severe cases e.g. to prevent synechiae formation. Prophylactic antivirals and antibiotic agents should be used along with steroids.
General Name: Herpes Zoster Iridocyclitis  
Specific Name: Herpes Zoster Iridocyclitis  
ICD 9-CM#: 053.22  
Location: Iris and Ciliary Body  
Associated Conditions  

S: The onset of symptoms is usually acute with pain, photophobia, lacrimation, hyperemia and blurred vision. Symptoms may be minimal however and a chronic course is usual especially with recurrences.  

O: Perilimbal flush. Anterior chamber cells, flare and keratic precipitates that are not well formed and tend to be pigmented. Hemorrhage may occur and elevated IOP is frequently a complication.  

A: Herpes zoster uveitis may present as a nongranulomatous (vasculitis) iridocyclitis or as a diffuse chorioretinitis and iridocyclitis. Hypopyon, hemmorhage into the anterior chamber, iris sector atrophy, heterochromia iridis, sympathetic ophthalmia, and phthisis bulbii may all result from severe zoster dermatitis and ischemia. Diagnosis is based on the clinical findings of zoster dermatitis with or without neuritis.  

P: Therapy is controversial with regard to systemic corticosteroids. In general, therapeutic guidelines are:  
1) mild or no pain requires no drug except a mydriatic-cycloplegic such as cyclopentolate or homatropine bid. 2) Topical steroids as necessary to control corneal edema and iridocyclitis are used. Prednisolone or dexamethasone may be instilled every 3 hours to once daily, as required. Antiviral idoxuridine or vidarabine ointments qid should be used when topical steroids are in use. 3) Topical antibiotics such as erythromycin of bacitracin should be used if ulcerative keratitis is present. 4) Non-narcotic or narcotic analgesics for neuralgia may be used during the first 10 days. If there is no relief or if uveits or pain is not controlled, after chest x-ray, complete blood count, and an immune status evaluation, the basically healthy patient may be started on systemic steroids while continuing mydriatics and topical steroids as needed. Frequent monitoring for dissemination of disease should be carried out as well as screening for occult malignancy.
General Name Cytomegalo Inclusion Disease
Specific Name Adult Cytomegalo Inclusion Disease
ICD 9-CM# 078.5
Location Uvea

Associated Conditions
AIDS, Organ transplants

S: In immunologically normal adults the symptoms are non existent or a mild mono-like symptoms. No ocular symptoms are present. However, in immunologically suppressed adults such as transplant patients or AIDS patients the main ocular symptom is loss of vision.

O: Ocularly a retinitis is present. It starts out as a few yellow-white spots that look like crumbs. They become confluent in about 4 weeks and hemorrhagic patches appear, giving the involved retina a "crumbled cheese and tomato sauce" appearance. The patches are due to retinal necrosis and they leave behind an atrophic gray colored patch. Areas of loss are permanent. Posterior hyaloid precipitates may also be found on the back of the vitreous. Other ocular involvement may include swelling of the disc, retinal detachment, and iridocyclitis.

A: Adult infection is fairly common, in fact 80% of the population over 35 have positive complement fixation titers. The best bet in diagnosis is to isolate the virus from the throat or urine. Cultures may take 4-6 weeks to show positive reactions. Electron microscope examination of the urine gives a more rapid diagnosis.

P: Prognosis for vision is poor since damage is irreversible. As in the neonatal, vidarabine should be given and immunosuppressive agents should be decreased. Immunosuppressive therapy may be of benefit. Referral to an internist is in order.
General Name: Cytomegalic Inclusion Disease
Specific Name: Congenital and Perinatal Cytomegalic Inclusion Disease
ICD 9-CM#: 771.1
Location: Uvea
Associated Conditions:
Mental retardation, Optic atrophy, Retinitis, Cataract, Glaucoma

S: Most infants are asymptomatic although long term effects are possible which include deafness and slow development. Symptomatic infants present with fever, jaundice, clotting problems, mental retardation, and subnormal ocular function.

O: Symptomatic Infants:
- Liver and spleen problems which affects the clotting system
- Intracranial calcification which produces brain damage
- 25-30% of the time ocular complications are present which include: optic atrophy, retinitis, cataract, glaucoma, and other associated congenital anomalies.

A: CMV inclusion disease must be differentiated from hemolytic disease, congenital syphilis, toxoplasmosis, herpes simplex or pigmentary retinopathy of muscles. Diagnosis can be made if a diffuse periventricular calcification is present and if toxoplasmosis is ruled out. A complement fixation test is also of value.

P: Vidarabine should be used although antiviral chemotherapy has not been very effective. Immunosuppressive agents should be decreased since cell mediated immunity is important to the control of the virus. In addition, immunotherapy may be successful.
Behcet's syndrome (recurrent uveitis of young adults) is typified by uveitis with hypopyon, aphthous lesions in the mouth, and genital ulcerations. Reported principally in Mediterranean countries, but today being most common in Japan. Almost always bilateral, sometimes simultaneously, sometimes separately. Usually male. Age of onset is usually 17-37 years. Acute symptoms disappear after 5-10 days, but recurring crises can result in blindness, either from atrophy of the optic nerve or phthisis bulbi. Characteristically recurs at intervals of 1 wk.-3 yrs., avg. being 2 mos.. As the disease progresses, the intervals between attacks lengthen so that after 15-20 yrs. there are no further attacks.

O: Avg. period from onset to blindness is 3.4 yrs.. The 4 major signs are: 1) oral: aphthae 2) dermal: erythema nodosum, thrombophlebitis folliculitis, cutaneous hypersensitivity 3) ocular: iridocyclitis, retinitis and 4) genital: ulceration. Oral lesions are tender and may be white single or multiple, pinhead to pea-sized or larger. These erosions or ulcerations are sharply circumscribed with a dirty base and surrounding halo. Acute polyarthritis is common. It involves large joints and resembles rheumatoid arthritis, but x-rays and rheumatoid tests are negative. Vitreous clears slowly and incompletely. At an early stage, there is a narrowing of the vessels of the retina, and there are usually signs of periarteritis and endarteritis. Poliomyelitis is the most serious complication, esp. if Behcet's syndrome involves the brainstem. The loss of visual field from widespread vasculitis results in a characteristically reduced ERG. Ocular lesions occur in about 75% of the cases, and complications include cataract formation, glaucoma, and eventually blindness. Half of all retinal involvements will lead to blindness in 4-8 yrs..

A: The etiology is unknown but, outside of the U.S., HLA-B5 typing was found in 69% of patients with the complete syndrome. Difficulties in the diagnosis of Behcet's disease are due to the variability in clinical manifestations, the frequent long delay between the onset of the disease and the appearance of lesions in a new target organ, and the absence of pathologic or immunologic features useful in diagnoses. Diagnosis can be made only on the basis of the clinical manifestations. There are 4 types: complete--4 main signs appear at some time during the course of the disease; incomplete--3 signs appear; suspect--two signs appear; possible--one sign only. In complete and incomplete types, CNS symptoms may appear. The appearance of a pustule within 24-36 hours after an intradermal needle puncture (Behcet's skin puncture test) with or without the injection of 0.1 ml. of normal saline, is almost diagnostic for Behcet's syndrome.

P: Patients in whom the gamma-globulin values show an increase during the period of acute attacks have a better prognosis for vision than those with subnormal values. Chlorambucil is the treatment of choice, 6-8 mg should be administered daily and increased to 20 mg/day in patients with acute disease, which includes rapidly deteriorating vision or CNS involvement. It should be started early in the course of the disease. Complete remissions have been obtained in 80-100% of the patients in these series. The period of treatment has ranged from 6 mos. to 2 yrs.. The best that can be expected is to arrest the disease at the stage at which treatment is started. Cases which are diagnosed early in the course of the disease and cases of acute disease usually respond quickly to treatment. Treatment should be continued for 6 mos. after the complete arrest of the disease. Blood transfusion have proved of value in some patients. Cataract extraction has been found effective even in the presence of a nonrecordable ERG.
General Name: Vogt-Koyanagi-Harada Syndrome
Specific Name: Vogt-Koyanagi-Harada Syndrome
ICD 9-CM#: 364.24
Location: Uvea
Associated Conditions: Glaucoma, cataract, phthisis bulbi

S: The patient complains of poor vision in both eyes often accompanied by headaches and nausea.

O: The following ocular signs are seen in the syndrome: poliosis; vertigo-depigmentation of the skin; alopecia-focal baldness; exudative retinal detachment; peripheral retinal sea fan neovascularization. Other secondary ocular changes include glaucoma from PAS, cataract, and phthisis bulbi.

A: VKH must be differentially diagnosed from sympathetic opthalmia. Sympathetic opthalmia has in its case history trauma or penetrating injury to the eye. The diffuse choroiditis and granulomatous panuveitis of VKH is usually accompanied by a cranial neuropathy. This creates the signs of hearing problems, headaches, nystagmus, vertigo, and muscle paralysis.

P: Visual prognosis is generally poor but an aggressive steroid therapy is usually instituted. The typical regimen is prednisone 200mg with breakfast plus periocular injections of 4mg dexamethasone 1-2x/wk depending on the severity of symptoms. After 7-10 days, prednisone may be reduced to every other day. Two weeks later the dosage is 100mg p.o. q.a.m. for 1-2 weeks.
**General Name** Sympathetic ophthalmia

**Specific Name** Sympathetic uveitis

**ICD 9-CM#** 360.11

**Location** Uveal tract

**Associated Conditions**
Traumatic eye injuries, Intraocular surgery.

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S: See under General Name: acute anterior uveitis, acute iridocyclitis, granulomatous uveitis.

O: See under General Name: acute anterior uveitis, acute iridocyclitis, granulomatous uveitis.

The injured eye (exciting eye) has a torpid, persistent, granulomatous type of uveitis that is then followed by a similar uveitis in the fellow eye (sympathizing eye).

A: The history of trauma or injury to one eye and later onset of inflammation in the other eye should alert one to the possibility of this diagnosis. It is an uncommon, diffuse, granulomatous inflammation of the entire uvea, affecting one eye (exciting eye) and then spreading to the other eye (sympathizing eye). It can occur days, months, or years after penetrating ocular injury or intraocular surgery. The cause is unknown, but the inflammation may be caused by an unidentified virus, an auto immune inflammatory response to uveal pigment, or a protein component associated with the membranes of the photoreceptor outer segments. Clinically and histologically, the inflammation resembles that occurring in Vogt-Koyanagi syndrome although the inflammation has no antecedent ocular penetration.

P: The inflammation may be suppressed by corticosteroids or other immunosuppressive agents, which may have to be continued for many months or years. Enucleation of the exciting eye is of no benefit once the fellow eye has become inflamed, but blind, exciting eyes should be enucleated. Sympathetic ophthalmia does not occur if the injured eye is removed within 7 days after the injury. Because sympathetic ophthalmia does not occur immediately after an injury, irrespective of the severity of the ocular damage, it is never necessary to enucleate an injured eye before skilled evaluation of the potential for repair with restoration of some function.
General Name: Histoplasmosis  
Specific Name: Histoplasmosis  
ICD 9-CM#: I15.02, I15.12  
Location: Uveal Tract  
Associated Conditions: Histoplasmosis, circumpapillary choroiditis, macular lesion

S: Over 99% of patients with histoplastic infections are asymptomatic. However, in mild cases, metamorphopsia with little or no decrease in vision may be experienced. In severe cases, it may lead to blindness.

O: Clinical finding consists of the triad of Histoplasmosis:  
1. Histo spots: localized punched out lesions which are areas of pigment epithelial atrophy. They are yellow, 0.2 to 0.7 disc diameter in size, and vary in number.  
2. Circumpapillary choroiditis: atrophic changes around the optic disc.  
3. Macular lesion: an active stage typified by a pigment ring and detachment of the underlying sensory retina.

Laboratory test indicates a positive histoplasmin skin test.

A: Presumed ocular histoplasmosis syndrome describes a pattern of fundus abnormalities related to Histoplasmosis capsulatum infection which is endemic in the Mississippi river valley and the great river valleys of South America, Africa, and Asia. The condition is usually bilateral. The diagnosis is based on the clinical picture of the triad and a positive skin test. Differential diagnosis is indicated from other similar condition of maculopathies (e.g. Fuch's spot of high myopia, disciform degeneration, or maculopathy from angioid streak) and hemorrhagic circumpapillary histoplasmosis (e.g. drusen of optic disc with hemorrhage).

P: 1. Fluorescein angiography is indicated to assess neovascularization and any presence of blood or elevation of the sensory retina.  
2. Treatment:  
   a. After the disease has become established the patient may be started on 50 to 150 mg. of prednisone and retrobulbar injection of long standing steroid. Steroid tapering is done over the following weeks proportionate to the improvement of the maculopathy. Fluorescein angiography may assist in the decision to taper off medication.  
   b. Photocoagulation is used to eradicate the exudation from the neovascular net that may produce serous hemorrhagic detachment.
General Name: Aspergillosis
Specific Name: Aspergillosis
ICD 9-CM#: 117.3
Location: Orbital
Associated Conditions: Paranasal Sinus Disease, Proptosis

S: External and internal ophthalmoplegia, decreased vision, decreased sensation, ptosis as well as chemosis, proptosis and facial pain. Symptoms may occur over a span of 6 months to a decade.

O: In up to 75% of the patients, proptosis is the first sign of the disease followed by signs of paranasal sinus disease. The optic nerve may become damaged with nerve head swelling and engorgement of the central retinal vein.

A: The Aspergillus infection develops in a sinus and spreads to the orbit and, as such, it is not a primary orbital infection. Rhinitis, polyps, recurrent sinusitis, and a tropical humid climate appear to be conducive to aspergillosis of the sinuses. Fungus infection in the orbit is usually a granulomatous, fibrosing disease, but it can be a nongranulomatous disease with abscess formation.

P: Vision can be salvaged if the disease can be interrupted before the granulomatous inflammation encircles the optic nerve. Surgical drainage appears to be the treatment of choice. Wide surgical drainage with iodoform packing can eradicate the infection.
**General Name** Toxoplasmosis  
**Specific Name** Ocular Toxoplasmosis  
**ICD 9-CM#** 130.2  
**Location** Uvea  

**Associated Conditions**  
Vision loss, Retinal Detachment  

**S:** Often patients with ocular toxo are asymptomatic. If the lesions involve the fovea they present with vision loss.  

**O:** Ocular toxo patients present with a characteristic type of lesion. It starts off as a translucent or gray area of the retina with an accompanying vitreous inflammation over the lesion. The lesions progress to a white inflammatory section of the retina and has the appearance of a cotton wool spot, but only larger—up to 1 DD. Posterior pole lesions are more commonly seen. The inflammation is so severe that venous sheathing and hemorrhage may also be present. The lesions last 8-10 weeks and when the active inflammation clears, a scar remains which is a combination of the white of the sclera and the hyperpigmentation of the PE cells. The area is an absolute scotoma. Any new lesions appear as satellites off the old. Most of the damage is done due to the secondary inflammatory response and not the organism. If the macula is involved VA’s may be as bad as finger counting or light perception.  

**A:** In the early stages ocular toxo must be ddx from macular colobomas, herpes simplex virus chorioretinitis and retinoblastoma. Some factors to consider are that toxo is common in the teen years but is rare over age 40. An active attack lasts from 1 week to 2 years but the average duration is 4.2 months. The Sabin Feldman methylene blue dye test is the diagnostic test most often used in toxo. Another useful lab test is the complement fixation test for determining if children have been infected during the first 6 years of life.  

**P:** After diagnosis is made treatment should be instituted in active cases as the the visual prognosis depends on the time between activation of retinitis and the initiation of therapy. If the necrosis stage is reached (5-6days) before therapy is started, any type of treatment will only limit the extent of damage. Indications for treatment are: any active lesions near the papillomacular bundle or near the macula, lesions which either threaten or involve the optic nerve, severe lesions that could possibly cause vitreous traction or retinal detachment, any peripapillary lesions especially ones close to the disk. Systemic therapy includes tx with Pyrimethamine and corticosteroids to control the inflammatory response. Sulfa drugs can also be utilized for their synergistic effects. Other drugs: Chloretetracycline and Clindamycin. Refer for treatment.
General Name: Toxoplasmosis

Specific Name: Congenital Systemic Toxoplasmosis

ICD 9-CM#: 130.1

Location: Uvea

Associated Conditions:
Mental Retardation, Convulsions, Chorioretinitis

S: The oocysts can penetrate the placenta and are transmitted in utero during the first 7 months of pregnancy. About half the infected infants are born without clinical signs of toxo but often develop a loss of vision between 10-20 years. In those that exhibit symptoms they range from mental retardation, convulsions, failure to thrive, and appearance of a floppy baby.

O: Often congenital toxo is reactivated and its effects are manifested in the nervous tissue in a triad: 1) Convulsions, 2) Intracranial calcification, 3) Chorioretinitis. In addition to the chorioretinitis ocularly there may be an additional vitritis and anterior uveitis which is probably due to an immune response and not a direct infection by the organism.

A: Toxoplasmosis gondii is the causative agent in toxoplasmosis. The organism invades and multiplies in host cell cytoplasm. Once immunity develops the organism goes into a cystic form which have a predilection for the eye, brain and muscle tissue. Based on the signs and symptoms and a positive Sabin Feldman methylene blue dye test the diagnosis is congenital systemic toxoplasmosis.

P: Refer to an internist or physician for systemic therapy consisting of a combination of an antitoxoplasmic agent such as Pyrimethamine (Daraprim) and a corticosteroid to control any inflammatory reactions. Often triple sulfa drugs are used in combination with pyrimethamine synergistically since each affects a different point in the organisms cycle. The sulfa drugs also help prevent some unwanted side effects caused by the antitoxo agent. Other drugs used instead of Pyrimethamine are Chlortetracycline and Clindamycin. Corticosteroids are never used alone in the treatment of toxo as it suppresses the immune response and allows the release of cysts all over the body.
**General Name**: Ankylosing Spondylitis

**Specific Name**: Marie-Strumpell's Disease

**ICD 9-CM**: 720.0

**Location**: Uveal Tract

**Associated Conditions**: Chronic Arthritis.

**S**: Usually English male (85%) between 15-30 yrs. A stooped posture from a chronic arthritis affecting the spine, sacroiliac joints, and adjacent soft tissues. Ankylosing spondylitis typically begins in the sacroiliac joints and has a tendency toward subsequent spread up the spine. Its onset is insidious with intermittent episodes of low back pain and stiffness. The pain occasionally has a sciatic distribution. Symptoms are usually worst a.m., are exacerbated by inactivity, and may be partially relieved by mild exercise. Tenderness, muscle spasm, straightening of the spine, and limitation of motion of the involved are often present. Chest pain & pain on breathing, sneezing, coughing, & peripheral arthritis are present.

**O**: The associated iridocyclitis (35%) is a mild to fairly severe nongranulomatous type involving the anterior segment almost exclusively. It may occur several years before the onset of the spondylitis, and it usually affects only one eye at a time. The occurrence of the anterior uveitis is unrelated to the severity of the systemic disease, although it is more common in patients with long-standing spondylitis. Ankylosing spondylitis has also been found in a significant percentage of patients with anterior uveitis. Severe disease is easily diagnosed by the patient's stooped posture. Verhoeff's test is positive when the patient cannot bend his cervical vertebrae backward to bring his chin into the cup for slit lamp examination.

**A**: Patients with acute nongranulomatous iridocyclitis should have an x-ray of the lumbosacral spine and an HLA-B27 test. The x-ray will give rise to a "bamboo spine" appearance. HLA-B27 test is especially important in women since an x-ray of the pelvis may be dangerous in the childbearing years. A positive HLA-B27 test indicates a hereditary predisposition. The ESR is usually elevated at times of systemic disease activity.

**P**: Systemic treatment includes analgesics and antiinflammatory agents. When possible, specific therapy directed toward the underlying disease should be instituted. Systemic corticosteroids are usually reserved for cases of advanced disease unresponsive to other medication. Topical steroid drops are used to treat the anterior uveitis, instilled every hour in severe cases. Mydriatics and cycloplegics provide symptomatic relief in patients with anterior uveitis and allow healing by putting the eye at rest, thus relieving the spasm of the pupilloconstrictor and ciliary muscles. Relaxing the ciliary muscle spasm decreases pressure on the ciliary arteries, resulting in increased blood flow to the affected structures. Cycloplegics also decrease vascular permeability and thereby curb exudation. They also dilate the pupil so that the formation of posterior synechiae is lessened. Phenylbutazone may be used when systemic corticosteroids is contraindicated, but routine blood checks and medical supervision should be employed with extended periods of use.
General Name: Sarcoid
Specific Name: Sarcoidosis
ICD 9-CM#: 135
Location: Uvea, Adnexa
Associated Conditions: Chorioretinitis, Periphlebitis

S: The patient complains of a painful red eye in early cases to decreased vision in later cases.

O: 25-50% of sarcoid patients present an anterior uveitis. This may either be granulomatous or nongranulomatous. Nodules are often seen on the iris. Ophthalmoscopic examination reveals a chorioretinitis, a candle wax dripping appearance on the equatorial retina, possible optic nerve edema, and snowball opacities in the vitreous. Inflammation of the equatorial veins (periphlebitis) is also a common sign. Granulomatous bodies are often found on the lids and conjunctiva. Granulomatous bodies in the lacrimal gland often give the lid margin a "S" shaped appearance.

A: Sarcoid is a systemic disease which usually occurs between the ages of 20 to 50 years. It is much more prevalent in the Black-American population, especially females. Chest X-rays and Kviem biopsy of the granulomas often are used to give a definitive diagnosis.

P: Patients diagnosed in the early uveitic stages have a good prognosis. The course of the disease is variable and frequent remissions in the first three years are common. The uveitis is treated with steroids and cycloplegics (see Anterior Uveitis). Severe cases of sarcoid usually require stronger systemic steroid therapy. Oral prednisone of 60 mg daily dosages, which is tapered as the inflammation decreases, is usually used in those cases with retinal involvement.
General Name: Endophthalmitis
Specific Name: Endophthalmitis
ICD 9-CM#: 360 (general)
Location: Intraocular tissues of the eye
Associated Conditions:
Bacterial infections, fungal infections, surgical trauma, secondary to trauma

S: Classifications: postoperative, trauma induced, bleb associated, and endogenous.
Postoperative: Pain and reduced vision are the primary symptoms. Bacterial: pain, hyperemia, lid edema and spasm, rapid loss of vision and light perception. Fungal: some pain and redness, good light perception. Bleb associated: Sudden onset of ocular pain and redness without warning. Trauma induced: Pain, decreased vision. Endogenous: Reduced vision in one or both eyes.

O: Postoperative: lid edema, conjunctival hyperemia with chemosis and exudate, a hazy cornea with epithelial and stromal edema, anterior chamber reaction with fibrin exudates and often a hypopyon, and vitritis. Fungal: smoldering inflammatory reaction, mild external involvement, progressive iridocyclitis and vitritis (most pronounced at the iris-pupillary border or anterior vitreous. Bleb associated: purulent material and an apparently intact conjunctiva. Endogenous: variable

A: When intraocular inflammation presents after surgery or trauma or in patients with blebs and the inflammation is more severe than expected in that clinical setting, an infectious endophthalmitis should be suspected. Microbial isolation helps in determining the causative agent. This can be obtained by fluid samples taken from the vitreous or anterior chamber.

P: The goal of management in endophthalmitis consists of intensive antibiotic administration to sterilize the eye. Nonspecific anti-inflammatory therapy is frequently used to limit the damage caused by the inflammation stimulated by the infection. In certain selected cases therapeutic vitrectomy is combined with antibiotic and anti-inflammatory therapy to eliminate the infectious organisms more rapidly, to aid in the antibiotic delivery to the infected site, and to eliminate areas of sequestered vitreous.
General Name: Iridocyclitis
Specific Name: Fuchs' heterochromic iridocyclitis
ICD 9-CM#: 364.21
Location: Uveal Tract
Associated Conditions: Uveitis, Glaucoma, Posterior Subcapsular Cataract, Iris depigmentation, Neovascularizations

S: Symptoms:
- blurred vision (major complaint)
- iris depigmentation
Seventy percent of Fuch's heterochromic cyclitis is unilateral. However bilateral cases are seen although they are more difficult to diagnose.

O: In unilateral heterochromia, the affected eyes show brown eyes becoming less brown and blue eyes becoming more saturated blue.
Slit lamp evaluation:
- Chronic anterior uveitis in a white and quiet eye with no synechiae formation.
- Keratic precipitates scattered all over the entire corneal endothelium with fluffy featherly borders, which is distinct from the usual well circumscribed KP borders.
- Iris atrophy with a characteristic moth-eaten appearance on the iris stroma.
- Along with iris atrophy and keratic precipitates, in established cases, there is also posterior subcapsular cataract, glaucoma, and fine neovascularizations.

A: The etiology of Fuch's heterochromic iridocyclitis is suspected to be some form of vascular degenerative process, possibly involving sympathetic nerve fibers and iris pigment. This condition is found in 2% of uveitis patients. The major complaint of blurred vision is due to the development of a posterior subcapsular cataract or vitreous veil. Furthermore, glaucoma is not uncommon and is caused by the fine neovascularization of the angle.

P: Treatment consists of short term topical steroids. However, caution is advised since both cataract and glaucoma may be aggravated by steroid therapy.
S: The patient is a young adult who might complain of floating spots. Most patients are asymptomatic.

O: Observation with the bimicroscope reveals punctate opacities in the retrolental space and anterior vitreous. There are soft round spots over the peripheral retina - "snow bank" appearance over the inferior ora and pars plana. This may be observed with the Goldmann three mirror or with the binocular indirect ophthalmoscope.

A: Pars Planitis is a chronic inflammatory lesion affecting the pars plana of the ciliary body and the peripheral retina. The disease starts as a mild chronic cyclitis and peripheral vasculitis which progresses to exudation into the vitreous base, which progresses to exudation into the vitreous base, which is usually inferior and forms the snowbank appearance over the ora and pars plana. The disease may be benign, but it often leads to many vitreous hemorrhages, cataract and glaucoma. Posterior cortical or subcapsular cataract is most common complication of pars planitis. This disease of children and young adults is typically bilateral in 2/3 to 4/5 of the affected patients. Unilateral conditions should be suspect of similar conditions such as sarcoid, toxocariasis, peripheral toxoplasmosis, and intraocular foreign body.

P: Studies indicate that the projected 5 year visual prognosis is excellent in 80% of the afflicted persons, without any treatment. It is only when vision is reduced that treatment is indicated. The reduction of vision is usually due to cysts in the macula, best observed with fluorescein angiography. Therefore, most of the remaining 20% of the afflicted persons do well with alternate day administration of prednisone or periocular steroids. Treatment is guided mainly by the visual acuity and the response to therapy is slow, usually taking months; but these cystic maculas usually improve with adequate therapy. Macular damage is related to the severity of the pars planitis rather than to its duration. In cases that do not respond to treatment, transcleral cryotherapy of the inflamed uvea may be helpful.
General Name  Choroidal Atrophy
Specific Name  Senile Choroidal Atrophy
ICD  9-CM#  363.41
Location  Uvea
Associated Conditions
Normal ageing change

S: Usually changes are gradual so patient doesn't complain of symptomology. They may report reduced vision in advanced cases.

O: Ophthalmoscopic exam reveals:
- Thickened arteries and veins which may appear white and opaque in late stages.
- Peripapillary halo due to changes occurring around the disk.
- Fields may reveal and enlarged blind spot

A: Senile changes are part of a continuous process. Observable changes usually first appear in the 5th decade and progress throughout life. The choroidal arteries show hyaline thickening and sclerosis of the media without general vascular disease. Initially there is fatty deposits in the intima and degeneration of the muscular coat. The muscular coat is then replaced by fibrous tissue and later by hyaline material. Eventually the whole vessel is hyalinized and the vessel becomes white and opaque. Changes may also be seen in the capillaries. Changes in the veins are also seen and appear as increases in connective tissue. It is especially evident around the optic disk and appear as a senile peripapillary choroidal atrophy. The degeneration occurring in the choroid also affects the pigment epithelium and is associated with hyaline or drusen deposition on Bruch's membrane.

P: It is hard to separate changes due to age and pathologic degeneration. In most cases senile choroidal atrophy is a normal ageing change seen in most individuals if they live long enough.
**General Name:** Choroidal Sclerosis  
**Specific Name:** Choroidal Atrophy  
**ICD 9-CM#** 363.50-363.57  
**Location:** Uveal Tract  
**Associated Conditions**

- **S:** Patient is a 33 year old Oriental who complains of loss of vision which is painless. The patient noticed that he was bumping into objects and that he was losing his side vision. He states that he was always healthy. The central form of choroidal sclerosis can start as early as 15 years of age and appears as edematous and exudative changes beginning in the macula or posterior pole region, and the changes resemble uveitis. This eventually leads to progressive atrophy of the overlying RPE, and the larger choroidal vessels acquire the appearance of broad white streaks.

- **O:** Observation of the anterior segments of the eye were normal. Observation of the fundus showed massive choroidal sclerosis extending centrally from the periphery. There were some areas of blackish and brownish pigment accumulation in the retina. The retinal vessels and the optic nerve heads appeared normal. The visual fields were constricted with both eyes to 5 degrees with large targets at 1m. The ERG was completely extinguished. The dark adaptation curve showed only minimal cone adaptation. The visual acuity was 20/20 with either eye. Choroidal sclerosis occurs predominantly in 3 regions of the fundus: the extreme periphery (most common in the elderly), the peripapillary zone, & the posterior pole. Rarely will it occur diffusely throughout the entire fundus. The condition is probably caused by fibrosis, hyalinization, and occlusion of the choroidal vessels, especially of those of the choriocapillaris. These changes are primary and the P.E. changes and other retinal changes are secondary. It is not yet known whether these 3 types are the same entity with different locations or whether they are separate entities.

- **A:** Diffuse choroidal sclerosis is usually rare and commences in the 4th decade of life. It causes generalized atrophic changes throughout the choroid and the larger choroidal vessels can be seen as a prominent whitish network. The condition starts with an edematous appearance of the fundus, a pigmented migration, and small yellow and cream-colored spots. The condition slowly advances with age until it becomes generalized in the 6th decade of life. A diffuse atrophy develops, and the fundus develops a brownish tigeroid appearance which is associated with extensive destruction of the RPE, clumps of pigment within and even overlying the retina sometimes are seen. The larger choroidal vessels are exposed and stand out as a prominent whitish network. The condition usually begins in the central or peripapillary area. The final result is an exposure of the sclera which has the appearance of complete atrophy. The symptoms depend on the area of the fundus involved. When the macula is affected (central choroidal sclerosis), visual acuity may be reduced to hand motion or worse. The visual fields are usually contracted and can become tubular; however, depending on topography other field defects such as ring scotoma or central scotoma may develop. Color vision, night blindness, and dark adaptation are affected as well as the ERG which can be subnormal or extinguished. Rule out R.P., O. Neuritis, Choroidal Hypopig.

- **P:** The usual hereditary mode of transmission is as an autosomal dominant; however, this is variable. Recessive inheritance and even sex-linked transmission have been recorded. There is no known treatment or prevention of this condition. Documentation of field defects with low vision aids is used as therapy.
General Name: Gyrate Atrophy of the Choroid
Specific Name: Gyrate Atrophy of the Choroid
ICD 9-CM#: 363.57
Location: Choroid and Retina
Associated Conditions: None

S: The primary complaint is that of having difficulty seeing things at night. Other complaints may include blurred vision and at late stages, tunnel vision.

O: Ophthalmoscope examination reveals oval areas of atrophy at the equator with scalloped borders. The atrophy eventually looks pale as it reaches the sclera. These atrophic holes eventually coalesce and spread toward the macula. In later stages, a fine velvety pigmentation with glittering crystals is seen.

A: Gyrate atrophy is a recessive hereditary progressive atrophy of the choroid and retina. Lab test indicate increased plasma levels of orthinine indicating an amino acid metabolism deficiency. Without the lab test it is very difficult to differentially diagnose from choroideremia.

P: If untreated visual prognosis is very poor. The choroidal and retinal atrophy will slowly constrict the field of view until central vision is lost. If the atrophy is diagnosed in early stages, a special diet low in the amino acid argenine, restores plasma levels to normal and prevents progression of the disease. Referral to a nutritionist is indicated.
General Name Choroideremia/Choroidemia
Specific Name Choroideremia/Choroidemia

ICD 9-CM# 363.55
Location Choroid
Associated Conditions

S: Males: Night blindness and contracture of visual field.
   Females: Asymptomatic.

O: Males and females: increased pigmentation and depigmentation of the fundus that is most marked in the equatorial region. The pigment granules have an irregular, square appearance similar to chunks of coal and are about 100 micrometers in diameter. Under or adjacent to the clumps of pigment are depigmented areas up to 0.5 disk diameter in size. They may appear paler than the rest of the fundus or have a bright yellow color. Males only: Eventually, atrophic changes dominate, and the white sclera becomes exposed in the equatorial region. At this time, night blindness occurs and an annular scotoma is present. The atrophy then spreads centrally and peripherally until all vision is lost. The retinal vessels are normal although some leakage from the choroid occurs through degenerate pigment epithelium and around the areas of remaining choroid.

A: Family history, clinical appearance, visual fields, ERG (extinguished in males), dark adaptation testing.

P: No proven treatment is available. Genetic counseling is of known value since it is clear cut that the disease is X-linked.
General Name  Choroidal degenerations
Specific Name  Angioid Streak
ICD 9-CM#  363.43
Location  Uveal Tract
Associated Conditions
psuedoxanthoma elasticum (Gronblad and Stradberg syndrome), sickle cell anemia, osteitis deformans (Paget's disease), fibrodysplasia hyperelastica (Ehlers-Danlos), hypertensive cardiovascular disorders, acromegaly, hypercalcemia, and lead poisoning.

S: Patient is asymptomatic.

O: Ophthalmoscopic evaluation reveals angioid streaks as breaks in Bruch's membrane. They appear as linear fractures which arise from the optic disc with offshoots extending toward the periphery mimicking retinal blood vessels. However, unlike blood vessels, they do not branch dichotomously and maybe several times wider than retinal veins. Their appearance is much like cracks in dry mud. Subretinal neovascularization is a frequent complication occurring in the papillomacular region or along side the optic disc.

A: The condition is most commonly seen in middle aged males. It is almost always bilateral but the ocular involvement is often asymmetrical. Ocular changes are asymptomatic unless a subretinal vascular membrane form. This is caused by the breaks in the Bruch's membrane which allow the ingrowth of capillaries from the choroid that leak and produce a disciform degeneration similar to that seen in senile patients. Angioid streaks signify the frequent association of wide spread degenerative changes of similar nature involving elastic tissue elsewhere in the body. Angioid streak occur most commonly in psuedoxanthoma elasticum (Gronblad and Stradberg syndrome). Other common systemic conditions associated with angioid streaks are sickle cell anemia, osteitis deformans (Paget's disease), fibrodysplasia hyperelastica (Ehlers-Danlos), hypertensive cardiovascular disorders, acromegaly, hypercalcemia, and lead poisoning.

P: Treatment consists of photocoagulation of the neovascular net.
General Name: Myopic Retinopathy  
Specific Name: Myopic Retinopathy  
ICD 9-CM#: 360.21  
Location: Retina, Choroid  
Associated Conditions:

S: Progressive myopia that continues on into adulthood. The degree of myopia is high, usually 6 diopters or greater.

O: Temporal crescent at the optic disc that may spread to become a circumpapillary zone of atrophy. Staphyloma of the posterior pole leading to degeneration of Bruch's membrane and lacquer cracks. These are branching, irregular, yellow-white lines that take a horizontal course and may cause foveal hemorrhage, which later leads to a Fuch's spot. Melanocytes disappear in the choroid and there may be widespread loss of the choriocapillaris. The peripheral retina may show lattice degeneration and retinal breaks.

A: In pathologic progressive myopia, a primary choroidal atrophy develops in which the sclera is thin and the choroid becomes atrophic. This degenerative myopia is usually associated with axial lengthening of the globe. The degree of myopia is usually genetically determined. It is more common in women and in some national groups such as Chinese, Japanese, Arabs and Jews.

P: The length of the eyeball should be determined with an A-Scan ultrasonography. If lengthening is documented, support for the stretching sclera can be gained by surgical insections of autographs of fascia lata or homographs of sclera.
General Name Nevi
Specific Name Uveal Nevi
ICD 9-CM# 224.0
Location Uvea
Associated Conditions
Malignant Melanoma

S: None. Patient is asymptomatic.

O: Uveal Nevi consist of a cluster of normal melanocytes. They have a flat appearance. They are congenital, but may not become obvious until puberty when it acquires pigment. Visual fields are normal to losses of predicted size. IOP is normal and photo documentation reveals no change. Ultrasound reveals no mass and fluorescein angiography produces hypofluorescence at the site of nevi due to absorption of dye by the pigment.

A: Uveal nevi can convert to a malignant melanoma of the choroid which is the most common intraocular tumour. Therefore it is important to differentiate it from a malignant melanoma.

P: Due to the possibility of conversion to a malignant melanoma it is important to monitor uveal nevi and document with fields and fundus photos if possible. Patient should be seen every six months. If changes are observed referral to an internist should follow.
General Name: Uveal Malignant Melanomas
Specific Name: Uveal Malignant Melanomas
ICD 9-CM#: 190.0 (190.6 = choroid)
Location: Uveal Tract
Associated Conditions: Lung Cancer, Breast Cancer.

S: Carcinomas of the breast in females and of the lungs in males are the most common primary tumors which metastasize to the eye. Tumors may also metastasize to the eye from the kidney, gastrointestinal tract, pancreas, and testicle, but these are less common. Some authorities believe that tumors metastatic to the uvea are the most common intraocular tumors. Most metastatic tumors involve the posterior choroid, but occasionally the ciliary body or the iris may be the focus of involvement; an iris melanoma may be either pigmented or nonpigmented; pigmentation and vascularity may vary from case to case. Patient usually experiences pain and decreased vision.

O: A tumor metastatic to the iris presents as a pink or yellow mass which may be located anywhere on the iris. A metastatic tumor to the choroid usually presents as a placoid or oval amelanotic choroidal mass. Although it may be located anywhere, it is usually in the macular region. It may have characteristic mottled-brown pigment clumping on the tumor surface. There is usually extensive serous detachment of the retina with shifting subretinal fluid.

A: There are no definitive tests for this condition.

P: Patients with metastatic tumors should be referred for treatment with external beam irradiation as a palliative measure to preserve their remaining vision. Enucleation is contraindicated unless painful secondary glaucoma occurs. Chemotherapy or immunotherapy should be used in conjunction with irradiation in most cases.
General Name  Hemangioma  
Specific Name  Uveal Hemangioma  
ICD 9-CM#  228.00  
Location  Uvea  

Associated Conditions  
Malignant Hemangioma, Sturge-Weber syndrome, Serous Retinal Detachment.

S: As a single entity, the hemangioma usually presents no symptoms.

O: The hemangioma presents itself as a focal, yellowish, slightly raised lesion usually in the region of the posterior pole. The width of the lesion is usually a few disc diameters.

A: Uveal hemangiomas are most often found in the choroid. Differential diagnosis from the malignant melanomas can be accomplished by transillumination, ultrasonography, and fluorescein angiography. Fluorescein angiography reveals a course vascular pattern followed by diffuse leakage and localized mottling. The vascular bed is highly reflective to ultrasound. A hemangioma will normally transilluminate while a malignant melanoma will appear as a dark area. Uveal hemangiomas are very common in those with Sturge-Weber syndrome.

P: The biggest threat to sight is that from secondary serous retinal detachment. Hemangiomas are usually treated with photocoagulation if potential posterior pole involvement is possible.
General Name Uveal coloboma/choroidal coloboma
Specific Name Uveal coloboma/choroidal coloboma
ICD 9-CM# 743.59
Location Inferior nasal fundus, usually beginning at optic disc.
Associated Conditions

S: Decreased visual acuity.

O: In the areas of the coloboma, lack of RPE, visibility of the sclera through the transparent retina, possible scleral ectasia. If the ciliary body is affected, a notching defect of the lens corresponding to the deficient zonule in the area may be present.

A: The appearance of a coloboma may be simulated by prenatal retino-choroiditis caused by toxoplasmosis. These inflammatory lesions, however, are usually not present in the inferior nasal quadrant, and there is proliferation of the retinal pigment epithelium. Typical congenital colobomas of the iris involve the inferior nasal portion and cause a defect in the shape of the pupil. The margins of the coloboma show the pupillary frill with pigment epithelium, unlike surgical iridectomies.

P: Ranges from no treatment to use of a contact lens to simulate a pupil to surgery to remove a deformed lens.
**General Name**: Coat's Disease  
**Specific Name**: Retinal telangiectasia  
**ICD 9-CM#**: 362.12  
**Location**: Peripheral Retina  
**Associated Conditions**: Glaucoma, cataract, leukocoria, retrolental fiberplasia, large chorioretinal coloboma, and persistent hyperplastic primary vitreous.

**S**: Patient reports a unilateral blurring of vision, leukocoria (white pupil), and cataract. The condition is nonfamilial and for the most part occurs in otherwise healthy children. It affects predominantly boys, usually appearing in the first decade.

**O**: The disease is characterized by the following ophthalmoscopic findings:
- Telangiectasis of retinal blood vessels
- Leakage of plasma to form intraretinal and subretinal exudates
- Retinal hemorrhages and detachment

**A**: Coat's disease is an exudative retinopathy of obscure etiology. The basic anomaly of Coat's disease is a congenital malformation of the retinal blood vessels leading to aneurysmal dilations which leaks, leading to accumulations in the subretinal space. Massive subretinal lipid exudate and serous retinal detachment can cause a leukocoria simulating retinoblastoma. Differential diagnosis is aided by the vascular anomalies and the yellowish green lipid exudate in contrast to the whitish pink appearance of retinoblastoma. Coat's disease must be differentiated from other common causes of white pupillary reflex such as congenital cataract, retrolental fiberplasia, large chorioretinal coloboma, and persistent hyperplastic primary vitreous.

**P**: Treatment with photocoagulation or cryotherapy may be helpful.
General Name  Soft Exudates

Specific Name  Cotton-Wool Patches

ICD 9-CM#  362.83

Location  Retina

Associated Conditions  Retinal Trauma, Severe Arterial Hypertension, Severe Anemia, Papilledema, Diabetic Retinopathy, Generalized Carcinomatosis, Acute Systemic Lupus Erythematosus, Dermatomyositis.

S:  Variable depending on underlying disease process.

O:  Cotton-wool patches appear ophthalmoscopically as indistinct, white retinal patches with a hazy, irregular outline and rounded off edges. They are usually ovoid in shape, variable in size and number, and are mainly seen in the posterior segment.

A:  Cotton-wool patches occur in the nerve fiber layer of the retina as the result of capillary infarction. They must be differentiated from nerve myelinization which tend to have feathering out edges in the direction of the nerve fibers.

P:  Treatment should be directed at the underlying disease process.
General Name: Retinal Exudates

Specific Name: Hard Exudates

ICD 9-CM#: 362.82

Location: Retina

Associated Conditions: Hypertension, Macular Star, Diabetes, Papilledema

S: Patients are ocularly asymptomatic. They may have systemic symptomatology related to a specific systemic condition.

O: Signs include:
- Hard yellow waxy areas that surround an area of retinal edema and microaneurysms. They are well defined and often coalesce and form a ring.

A: Hard exudates are usually found in patients with hypertension and diabetes. They are protein and lipid deposits left behind after reabsorption of areas of localized retinal edema. They are generally located in the outer plexiform layer and associated with involvement of the deep capillary bed. Hard exudates form only after chronic edema. A macular star is a special formation of hard exudates that results after a papilledema resolves. Drusen is the only differential diagnosis for hard exudates.

P: No specific treatment for hard exudates exists. The underlying condition—diabetes or hypertension must be treated.
General Name Drusen
Specific Name Drusen
ICD 9-CM# 362.57
Location Retina
Associated Conditions
Drusen of the optic nerve head, "Giant drusen".

S: Patient is healthy and asymptomatic.

O: Drusen of the pigmented epithelium are deposits of basement membrane material between the pigmented epithelium and Bruch's membrane. This calcification of Bruch's membrane that occurs with age is not visible ophthalmoscopically when the pigmented epithelium is intact. The presence of basement membrane material atop Bruch's membrane attenuates the overlying pigmented epithelium, thinning out the normal distribution of melanin in these cells. Usually, these basement membrane deposits appear yellow-white, but when they calcify, they may appear ophthalmoscopically as white elevated dots. Should the number of the drusen dots increase and coalesce, a field loss may manifest.

A: Parenthetically, it would be advantageous to detect drusen of the pigmented epithelium because the brittleness that it induces in Bruch's membrane may lead to breaks in this barrier, contributing an access path to the subretinal space by choroidal neovascular membranes in the pathogenesis of exudative, disciform macular degeneration (assuming it is occurring in the macular region).

P: Basically, there is no treatment or prevention of this condition. However, with regards to drusen that causes aging macular degeneration, three clinical trials have proven that argon laser photocoagulation of subretinal neovascular membranes 200u or more from the foveal center significantly reduces the central visual loss over a period of at least 2 years.
General Name: Flame Shaped Hemorrhage
Specific Name: Superficial Retinal Hemorrhage
ICD 9-CM#: 362.81
Location: Retina
Associated Conditions: Hypertension, Diabetes, Retinal Vein Occlusions, Anemia, Hemoglobinopathies, Blood Dyscrasias

S: Usually none other than those from the underlying disease process.

O: Observation of the fundus demonstrates a flame shaped hemorrhage or small splinter looking hemorrhages. The flame shaped hemorrhage may have a white spot in it.

A: The hemorrhage is located in the superficial nerve fiber layer. The blood tends to follow the nerve fibers giving it the flame appearance. A white dot in the center is called a Roth spot. It is an accumulation of white blood cells. These are usually seen in those with endocarditis, leukemia, and anemia. Flame shaped hemorrhages are a strong sign of systemic disease. It is very common in untreated hypertension. Other causes are anemia, hemoglobinopathies, blood dyscrasia, and diabetes.

P: Visual prognosis is dependent on treatment of the underlying disease. A tentative diagnosis of the cause of the hemorrhage and a referral to the proper physician is dictated.
**General Name** Blot and dot hemorrhages  
**Specific Name** Punctuate, round, and blot hemorrhages  
**ICD 9-CM#** 362.81  
**Location** Deep capillary plexus of the retina  
**Associated Conditions**  
Blood dyscrasias, diseases affecting the vasculature of the body.

**S:** None

**O:** Tiny, dark, discrete hemorrhages that are frequently associated with microaneurysms.

**A:** Recognition of these alterations is necessary to evaluate the accompanying ocular manifestations, but it is equally important to realize that the retinopathy may be the initial manifestation of a blood dyscrasia. Although the alterations in the retina seldom are sufficiently characteristic of any specific hematologic disorder to permit a definitive diagnosis, descriptions of the important retinal changes and their pathologic basis may provide important clues concerning the patient and his disease.

**P:** Treat the overlying cause of the hemorrhages.
General Name: Hemorrhage  
Specific Name: Preretinal hemorrhage (Subhyaloid hemorrhage)  
ICD 9-CM#: 362.8  
Location: Peripheral Retina  
Associated Conditions: Diabetes, vascular occlusions, hypertension, increased intracranial pressure, trauma, glaucoma, and blood dyscrasias.

S: Symptoms vary according to the cause and the stage of each of the following conditions:  
Diabetes: Fluctuating vision in mild case to blindness in severe case.  
Vascular occlusion: Sudden loss of vision (central retinal vein or artery occlusion) and field loss (branch occlusion.)  
Hypertension: Increased blood pressure.  
Increased Intracranial pressure: Loss of vision and exophthalmos.

O: Ophthalmoscopic evaluation shows a characteristic large meniscus of blood with distinct borders in the subhyaloid space at the posterior pole. A preretinal hemorrhage adjacent to the optic disc may be a sign of subarachnoid hemorrhage or glaucoma.

A: The most common cause of preretinal hemorrhage is diabetes and vascular occlusion. Other causes include hypertension, increased intracranial pressure, trauma, glaucoma, and blood dyscrasias. Although rare, a bilateral preretinal hemorrhage is pathognomonic ocular sign for subarachnoid hemorrhage.

P: Treatment consists of referral to the proper specialist:  
1. Diabetes and hypertension: Referral to an internist.  
2. Vascular occlusion: Referral to an ophthalmologist.  
3. Glaucoma: Referral to the ophthalmologist.  
4. Intracranial pressure: Referral to a neurologist.
General Name: Hemorrhage
Specific Name: Subretinal Hemorrhage
ICD 9-CM#: 362.81
Location: Retina
Associated Conditions: Drusen, Disciform Degeneration, Myopia, Angoid Streaks, Presumed Ocular Histoplasmosis

S: None

O: Hemorrhage between the choriocapillaris and RPE is dark brown, well circumscribed, and sometimes elevated and may simulate a neoplasm. The blood may rupture into the sensory retina and appear as a bright red crescent at the margin of the subretinal neovascular membrane.

A: Subretinal hemorrhage is usually preceded by proliferation of neovascular nets from the choroid as a result of breaks in Bruch's membrane. Subretinal neovascular membranes develop between Bruch's membrane and the RPE and between the RPE and sensory retina. The membrane appears as a dark green circular or oval area. The overlying retina may be detached, infiltrated with hard yellow deposits or swollen with cystoid edema.

P: Treatment should be directed at the neovascular membrane. Photocoagulation of the entire membrane can be instituted as long as it does not occur beneath the fovea centralis. Hemorrhage in the RPE which bursts into the sensory retina is followed by fibrous metaplasia of the RPE.

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General Name: Occlusions
Specific Name: Arterial Occlusions
ICD 9-CM#: 362.11
Location: Peripheral Retina
Associated Conditions: Vision Loss, Stroke

S: Patients report a sudden painless loss of vision. If the central artery is affected the loss is central, and if a branch is affected the defect is usually quadrantic.

O: Ophthalmoscopic exam reveals an opaque white inner layer of the retina, like a giant cotton wool spot secondary to ischemia. The fovea stays relatively red due to its supply from the choriocapillaris and appears as a cherry red spot. Arteries appear as thin red threads and the blood column may appear segmented. After ~1 week the tissue is dead and edema has resolved. The retina appears normal again. Arteries stay thin and eventually become white threads due to sheathing. The optic nerve atrophies and becomes white due to the lack of blood supply. In the case of a branch occlusion the same observations are present but only in the area of the retina that the vessels supply.

A: Need to decide if the occlusion is caused by an emboli or by a vasospasm. An emboli produces a sudden, complete loss of vision without forewarning. A vasospasm produces numerous transient episodes of decreased vision until finally vision doesn't return.

P: If the occlusion can be relieved within the first couple minutes to an hour the retina can be saved. In the case of vasospasm this is accomplished by the use of drugs. In the case of an emboli, it should try to be dislodged by massaging the globe or by using the anticoagulant drugs. In either case referral to an internist or physician is in order.
General Name: Venous Occlusion  
Specific Name: Venous Occlusion  
ICD 9-CM#: 362.35-362.37  
Location: Retinal  
Associated Conditions: Arteriosclerosis, Arterial Hypertension, Neovascular Glaucoma, Rubeosis Iridis, Peripheral Anterior Synechiae.

S: A mechanical blockage of a vessel that transports blood out of the eye, e.g., central retinal vein occlusion. Nonischemic CRV occlusion is a much more variable disease in appearance, symptoms, and course than ischemic CRV occlusion. Patients with nonischemic are an average of 5 yrs. younger. Complaints vary from none to transient blurred vision. The VA may range from normal to counting fingers, but the majority of patients have an initial VA of 20/50 or better. Patients with an ischemic pattern are usually aware of a sudden painless decrease in VA ranging from 20/400 to hand movements. However, the onset is generally not as rapid or the visual loss as extensive as in CRA occlusion. Average age is 68.5 yrs.

O: A CRV occlusion can fall into 2 groups, one study says one with a dramatic "blood and thunder" ophthalmoscopic appearance, loss of vision, and a poor prognosis and the other with mild ophthalmoscopic changes, generally good visual acuity, and a good prognosis. Other investigators describe the 2 categories as a nonischemic type, or venous stasis retinopathy. Engorgement of the venous tree is prominent with increased tortuosity and dilation and a darker appearance of the blood column. Retinal hemorrhages vary markedly. The IOP is frequently lower on the side of the occlusion. The natural course of nonischemic CRV occlusion is relatively benign (but it does appear that some eyes with a nonischemic CRV occlusion do go on to develop a more ischemic type of CRV occlusion). The majority of patients will have a final visual acuity of 20/40 or better.

A: Occlusion of the CRV is often a result of both local and systemic causes. It can be roughly divided into those conditions that produce a physical blockage at the level of the lamina cribrosa and those conditions in which hemodynamic factors result in an obstruction to the flow of blood. However, a combination of these mechanisms probably occur. The precise etiology of CRV occlusion remains unclear. Based on studies of histopathologic findings, a primary thrombus within the intralaminal portion (at or just behind the level of the lamina cribrosa) of the CRV was the most common cause of occlusion. At the lamina cribrosa, it may also be further impinged upon by arteriosclerosis of the adjacent CRA. Arteriosclerosis is undoubtedly the most important systemic condition related to CRV occlusion. Closely related is arterial hypertension. The most important ocular condition is open-angle glaucoma. The intravenous fluorescein angiogram pattern is usually characterized by a delayed filling time and extensive bleeding of fluorescein into the retina, particularly in the macular area and in the area adjacent to the large venous trunks and capillary nonperfusion. Microaneurysms are present or will shortly manifest.

P: Generally poor prognosis because of decreased visual acuity and neovascularization (the most serious complication of CRV occlusion) which may lead to neovascular glaucoma. Patients with neovascular glaucoma complain of tearing, irritation, pain, and further blurring of vision as the IOP in the affected eye begins to rise. The pain may become excruciating. Cornea is hazy, pupil is dilated, and rubeosis iridis is present. Peripheral anterior synechiae eventually develop which will lead to bullous keratopathy. Dense cataract eventually form. Retinal edema usually gradually subsides except in the macula, where it may persist for many months or years. Macular holes or cysts may form. Once developed, neovascular glaucoma responds poorly to any type of treatment. Treatment of the underlying systemic condition, if one is found, is indicated, although only rarely will this reverse the vein occlusion. A wide variety of therapeutic agents have been used, but there are significant ocular and systemic bleeding complications associated with anticoagulation therapy due to the administration of heparin. Panretinal photocoagulation is the accepted method of treatment of ischemic CRV occlusion because it is effective in both the prevention and the regression of neovascularization.
General Name   Retrolental Fibroplasia
Specific Name   Retinopathy of Prematurity
ICD 9-CM#    362.21
Location    Retina
Associated Conditions
Vitrious Hemorrhage, Neovascular Glaucoma

S: None

O: The primary sign is that of neovascularization. The neovascularization starts at the disc and progresses toward the periphery. In severe cases microaneurisms may be visible as well as neovascular proliferation on the surface of the retina and into the vitrious. Traction marks are usually seen on the retina from tugging of scarred tissue. Severe cases of traction will result in retinal detachment.

A: The risk of retrolental fibroplasia occurs whenever a baby is put in an incubator with increased oxygen. These babies are often premature or born with respiratory difficulties. The length of stay in the incubator and risk of retrolental fibroplasia is often weighed against that of brain damage or death. Assessment involves that of the degree of neovascularization. Besides the resulting scarring and retinal traction, dangers of vitrious hemorrhage and neovascular glaucoma are also of concern.

P: Up to the point of scarring, retrolental fibroplasia can regress leaving only minimal peripheral changes and normal vision. However the severity usually is dependent on the length of stay in the incubator and the amount of time the baby is born premature. If severe scarring takes place, very poor prognosis is given and the child will most likely be left blind.
General Name: Eales' disease
Specific Name:Periphlebitis retinae
ICD 9-CM#: 362.18
Location: Peripheral retinal veins
Associated Conditions:

S: Gradual or sudden decrease in vision. Spot or floaters in the field of vision.

O: Segmental, dilated, beaded, occluded veins, with sheathing or exudation, and blood in the vitreous body and the retina. The hemorrhages tend to absorb rapidly but repeated hemorrhages result in vascularization of the vitreous humor, chronic uveitis, and glaucoma.

A: Eales' disease is a nonspecific peripheral periphlebitis that mainly affects men between 15 and 30 years of age. It is characterized by recurrent retinal hemorrhage adjacent to the involved veins and by vitreous hemorrhage. Both eyes are involved in about 50% of the cases. The cause is unknown, but the condition may result from a nonspecific reaction to various antigens. Although the hemorrhages tend to absorb rapidly, repeated hemorrhages result in vascularization of the vitreous humor, chronic uveitis, and glaucoma. Differential diagnosis includes ruling out Coats disease, angiomatosis retinae, branch vein occlusion, and diabetic retinopathy, but sheathing in the peripheral retinal vessels reveals the true nature of periphlebitis retinae. Rarely are the more central vessel affected but when this occurs, the differential diagnosis is against CRV occlusion.

P: Systemic steroid therapy in massive doses is indicated in the previtreous hemorrhage state when there is active periphlebitis and retinal edema. When hemorrhage has already begun, photocoagulation has proven to be very effective.
General Name: Retinal Dystrophies

Specific Name: Retinitis Pigmentosa

ICD 9-CM#: 362.74

Location: Peripheral Retina

Associated Conditions:
- Night Blindness
- Bassen-Kornzweig syndrome
- Ushers Syndrome
- Refsum's Disease
- Leber's congenital amaurosis
- Mucopolysaccharidosis (Hunter's Disease and Sanfilippo's disease)
- Psuedoretinitis pigmentosa
- Syphilis
- Bardet-Biedl Syndrome

S: There is progressive impairment of visual function:
- Poor night vision (most common symptom)
- Poor dark adaptation
- Field loss (beginning with ring scotoma it often leads to tunnel vision and eventually it may progress to total blindness)

O: Ophthalmoscopic evaluation shows:
- Attenuation of retinal vessels
- Bone spicules pigment proliferation in the midperiphery of the fundus
- Waxy pallor of the optic disc
- Hyperpigmentation and hypopigmentation of the retina

A: Retinitis Pigmentosa is a progressive retinal dystrophy. There are a number of tests which will establish RP: 1) ERG (scotopic) and EOG (scotopic & photopic) are abnormal while ERG (photopic) is spared. 2) Visual field starts with ring scotoma which contract inward and outward and may eventually continue to contract until total blindness results. Field loss varies with the type of RP: a) x-linked: (rarest, most destructive, and first to have field loss), b) autosomal recessive: (next to have field loss & more destructive at the end) c) autosomal dominant: (least destructive and may never lose central vision.) 3) Dark Adaptometry: tests to screen children with familial history of RP. 4) Fluorescein Angiography: RP has hyper fluorescence due to increased permeability in the vessels of decreased pigment in the fundus. Ddx RP from associated conditions e.g. Bassen-Kornzweig syndrome, Ushers Syndrome, Refsum's Disease, Leber's congenital amaurosis, Mucopolysaccharidosis (Hunter's disease & Sanfilippo's disease), Psuedoretinitis pigmentosa, Syphilis, & Bardet-Biedl Syndrome.

P: There is no known treatment to Retinitis Pigmentosa. The following is recommended for patients with Retinitis Pigmentosa:
1. Examination of eyes annually to determine the progression of the disease.
2. Genetic counselling.
3. Low vision aids e.g. field expanders.
4. Mobility training.
5. Dark glasses for photophobia.
General Name: Retinal Folds
Specific Name: Retinal Striae
ICD 9-CM#: 362.89
Location: Retina

Associated Conditions:
Retrolental Fibroplasia, Intraocular Penetration of Foreign Bodies, Granulomatous Uveitis, Toxoplasmosis, 13-15 Trisomy

S: Variable depending on the underlying disease process.

O: Fine retinal folds occurring only in the neural section of the retina and generally running in a horizontal direction.

A: Retinal folds may be congenital in origin or secondary to a disease process. The simple congenital fold is apparently an isolated anomaly in differentiation of the retina. These folds extend from the nerve head into the retinal periphery and are usually unilateral. Children with this condition are normal in other respects. Retinal folds secondary to scar formation in the retina occur in grade 3 retrolental fibroplasia, intraocular penetrations of foreign bodies and following granulomatous uveitis, especially toxoplasmosis.

P: None
General Name Phakomatoses

Specific Name Tuberous Sclerosis (Bourneville's)

ICD 9-CM# 759.5

Location Peripheral Retina

Associated Conditions
Mental retardation, Seizures

S: Patients with tuberous sclerosis present with symptoms in the first 3 years of life. They include mental retardation, subnormal vision, and ash leaf spots of depigmentation.

O: The diagnostic triad of tuberous sclerosis is mental retardation, adenoma sebaceum and seizures. Ocularly retinal lesions are present as glial hamartomas and may be large with white concretions. The lesions have a mulberry-like appearance and are flat or gelatinous. The adenoma sebaceum of the skin develops from 2-5 years of age and is a misnomer since it's really an angiofibroma. Ash leaf spots of depigmentation are particularly evident under UV light. Periungual and subungual fibromas often appear after puberty.

A: Tuberous sclerosis is an irregular autosomal dominant disorder and prognosis in these cases is poor. 75% of those afflicted die before the age of 20. Glial hamartomas of the cerebrum result in seizures and intracranial calcifications.

P: Patients should be under the care of an internist. OD's should take care of vision needs and can monitor the ocular manifestations.
General Name: Neurofibromatosis

Specific Name: Von Recklinghausen's Disease

ICD 9-CM#: 237.7

Location: Retina

Associated Conditions:
Skin Pigmentation, Multiple Skin Tumors, Ptosis, Proptosis, Elephantiasis.

S: Cutaneous manifestations are prominent in this disease. However, lesions involving the brain, bone, and optic nerve are common. Symptoms depend on the location and extent of the tumors. Although Recklinghausen's disease is a dominant inheritance (with very irregular penetrance) and congenital disease, signs and symptoms may not become apparent until late childhood or early adulthood. This disease is characterized by dermal pigmentation, multiple tumors arising from the schwann cells of the peripheral nerves, and neurofibromas of the CNS. Subcutaneous tumors stand out under the skin but do not adhere to the deeper layers. These sometimes have the consistency of a "bag of worms".

O: Solitary pedunculated cutaneous neurofibromas may be scattered over the body and are commonly found on the eyelids, resulting in thickened lid margins and perhaps ptosis. Enlargement of a particular part of the body with neurofibromatosis is called elephantiasis neuromatosis. Cafe au lait spots constitute the most striking cutaneous manifestation of this disease. They may occur anywhere on the body, are 1mm to many centimeters in size, and are light or dark brown in color. Cafe au lait spots are found in 10% of the population, but patients with 6 or more spots exceeding 1.5cm in diameter invariably have Recklinghausen's disease. Ophthalmologic findings include neurofibromas of the iris (i.e. iris nodules) which may result in heterochromia. The retinal tumors are indistinguishable from those found in tuberos sclerosis. Proptosis as a result of orbital neurofibromatosis is not uncommon. Increased ICP and resulting buphthalmos has been reported in younger patients with Recklinghausen's disease.

A: Glaucoma in adults has also been reported in association with neurofibromatosis. CNS neurofibromatosis usually involves the meninges, cerebrum, sympathetic nerves, and cranial nerves, resulting in thickened and prominent corneal nerves. Glioma of the optic nerve is present in 10% of patients, while retinal glioma is rare.

P: Referrals for differential diagnoses can be sent to a dermatologist, internist (adrenal gland tumor), neurologist, and ophthalmologist. Treatment, when necessary, consists of local incision of tumors. However, results are not good, and surgery should be avoided if possible. The prognosis is variable.
General Name: Wyburn-Mason syndrome
Specific Name: Wyburn-Mason syndrome
ICD 9-CM#: 362.13
Location: Retina and brain
Associated Conditions

S: Ocular: Nothing to a considerable decrease in vision.
   Neurological: Seizures, delirium, disorientation, hallucinations, somnolence, and sleep inversion.

O: Ocular: Decrease in vision, visual field defects, proptosis (pulsatile), nystagmus, A-V aneurysms of the retina, oculocutaneous telangiectasis, retinal exudate and edema, tortuous and dilated veins. The retinal arteriovenous malformations (AVMs) show a great deal of variability from patient to patient, involving vessels in one small area, one quadrant, or the entire fundus.
   Neurological: Intracranial AVMs, acute subarachnoid hemorrhage, various midbrain syndromes, hydrocephalus secondary to intrusion on the sylvian aqueduct. Skull x-rays may demonstrate thinning of the cranial vault where distended blood vessels are present.

A: Clinical appearance, fluorescein angiography, cerebral angiography (the definitive diagnostic procedure).
   The etiology of the syndrome is unknown, although both developmental and hereditary factors may play a role. The syndrome consists primarily of arteriovenous malformations involving the retina and brain, especially the midbrain. Symptoms usually occur before the fourth decade and the presentation may be either ocular or neurologic.

P: No ocular treatment.
**General Name** Emboli  
**Specific Name** Retinal emboli  
**ICD 9-CM#** 362.30  
**Location** Peripheral Retina  
**Associated Conditions** Central retinal artery and vein occlusion, Branch artery and vein occlusion, Amaurosis fugax, and Hollenhorst plaque.

**S:** Patient may be asymptomatic or more typically, notice a sudden, uniocular visual loss starting as a concentric dimming of vision or horizontal curtain coming over the eye depending on where the emboli is lodged. The attack may last anywhere from few minutes to two to three hours before vision returns to normal. The attack may occur once or periodically during the day.

**O:** Ophthalmoscopic evaluation can indicate different types of emboli:  
1. Cholesterol emboli (Hollenhorst plaque)  
   - shiny, yellowish fat crystals that reflect light as the ophthalmoscope is tilted.  
   - most commonly found at the bifurcation of the retinal arteries.  
2. Calcific emboli: pearly white appearance and larger than other types of emboli.  
   - usually lodged at the larger arteries at the optic disc.  
3. Platelet and Fibrin emboli:  
   - soft and breakable, the emboli may move slowly as attack evolves.  
   - produces amaurosis fugax.  
4. Occasionally other materials such as talc in drug addicts or fat emboli in patients with multiple fractures is observed.

**A:** Differential diagnosis of different types of emboli:  
1) Cholesterol: It is the most common and most dangerous type of emboli. It is mostly seen in the older population. Due to the planar shape of the cholesterol emboli, blood flow may not be affected and patient may be asymptomatic. However, these patients are at a high risk of stroke.  
2) Calcific: It is mostly seen in the younger population with cardiac valve problems e.g. mitreal valve prolapse and heart murmur. Sight loss can be temporary (few minutes <20 min); If vision loss is >20 min., there can be neural tissue death with appearance of cotton wool spots and sectoral field loss.  
3) Platelet and Fibrin: The most common case of transient visual loss (Amaurosis fugax). The emboli breaks up easily- usually panic breaks the emboli block with the sympathetic nervous system rush. Ddx other causes of amaurosis fugax e.g. temporary reduction pressure and hyperviscosity syndromes.

**P:** The prognosis is related to the cause, the degree of obstruction, and the length of time of the occlusion. Restoration of vision within one hour usually restores all vision, while a relief of vision within 3 to 4 hours may restore peripheral vision with a persistent defect of central vision. After 4 hours, the visual defect may be permanent. Treatment is directed toward dislodging the embolus by massaging the globe intermittently. Moderate pressure is applied for 5 seconds and released for 5 seconds and then repeated. Patient with retinal embolization has a high risk of developing a stroke over the next few months. A referral to an internist for a careful evaluation of the cardiovascular system is indicated.
General Name: Microaneurisms

Specific Name: Retinal Microaneurisms

ICD 9-CM#: 362.14

Location: Retina

Associated Conditions:
Central or Branch Vein Occlusion, Coats Disease, Periphlebitis, Hyperviscosity of the Blood, Carotid Artery Insufficiency

S: Variable depending on underlying disease process.

O: Ophthalmoscopically they appear as minute red dots of exchanging appearance that are unrelated to visible blood vessels. Microaneurisms leak plasma and are often surrounded by edema which gives the retina a hazy appearance. Sometimes hard yellow deposits surround the microaneurisms.

A: Microaneurisms are a common retinal abnormality. Large numbers form in diabetes mellitus and they are characteristically on the venous side of the capillary network. They may be seen in most of the conditions associated with retinal venous stasis. Microaneurisms can be identified with certainty by fluorescein angiography.

P: Treatment is directed towards the underlying disease process.
General Name: Phakomatoses

Specific Name: Angiomatosis Retinae (Von Hippel-Landau)

ICD 9-CM#: 759.6

Location: Peripheral Retina

Associated Conditions: Seizures

S: Patients may present with visual problems, or seizures but will have no cutaneous lesions.

O: Ophthalmoscopically, vascular hamartomas are seen. The hamartoma is composed of capillaries with proliferation of endothelial cells on both feeding and draining vessels. Through both hypertrophy and hyperplasia the lesions enlarge over time. The lesions ultimately affect the macular area due to abnormal permeability. Changes in the macular area can include; hard exudates, edema, serous detachment and cholesterol deposits.

A: Von Hippel's disease consists of angiomatosis retinae, while Von Hippel-Landau includes cerebral manifestations (present in 20% of the cases). It is an autosomal dominant condition but has incomplete penetrance. If a patient has angiomatosis retinae in one eye he has a 50% chance of having it in both eyes. The condition most commonly develops in young adulthood.

P: Photocoagulation of the tumour may reduce the exudation process, but it is difficult to treat areas near the temporal margin of the optic nerve without affecting central vision.
General Name Retinoblastoma
Specific Name Retinoblastoma
ICD 9-CM# 190.5
Location Retina
Associated Conditions

S: A highly malignant, congenital tumor which characteristically arises in one or both (30%) retinas. As long as retinoblastoma remains confined to the retina it does not present a great threat to life. However, it is a rapidly growing tumor. Once the tumor has breached the relatively resistant structure of Bruch's membrane and flourishes in the choroid, growth can accelerate dramatically. Significant invasion of BV transforms retinoblastoma into a virulent systemic malignancy, as well as spread via the CSF pathway throughout the CNS. Orbital spreading type comprise 8% and optic nerve spreading type comprise 13%. Avg. age of onset is 20 mos. (oldest at 11 yrs.). No sex predilection. Bilateral hereditary often fatal.

O: The most common characteristic of leukokoria is present in 56% of these cases, and strabismus is present in 20%. The tumor is usually creamy pink in color and frequently neovascularization is present on the surface. Microaneurysms and telangiectatic vessels are frequent in the vascular stroma of these tumors. Some tumors are precluded by vitreous hemorrhage, retinal detachment, or an inflammatory reaction. Orbital spread is signaled by proptosis, a palpable orbital mass, or a lid swelling with or without ocecmhymosis. There may also be an enlarged optic canal or bony erosion, simultaneous orbital, systemic, and/or CNS spread. Gross optic nerve involvement may be evident under B scan ultrasonography as a widened (swollen) optic nerve shadow.

A: Two features of retinoblastoma are almost pathognomonic. The first is the fluffy pattern of calcification which can be seen with the ophthalmoscope, and the second is the seeding of tumor cells into the vitreous. Retinoblastoma is inherited as an autosomal dominant characteristic of extremely high penetrance. Any involved child with bilateral retinoblastoma will have a 50% chance of passing the disease on to his offspring. 10% to 15% of patients with sporadic, unilateral retinoblastomas will transmit the disease to their offspring. When a child with retinoblastoma is born to normal parents with no family history of the disease, there is a 1% chance that subsequent children will develop retinoblastoma and a 5% chance that there will be an affected child somewhere in the ensuing family line. Rule out Toxocara, Granulomatous uveitis, Coat's, Angiomatosis retinae, Metastatic retinitis, Persistent hyperplastic primary vitreous, Retrolental fibroplasia, Retinal dysplasia, Retinal fibrosis, Dictyomas.

P: Orbital disease may develop following iatrogenic rupture of the globe at the time of enucleation. 10% survive clinically apparent or biopsy-proved orbital retinoblastoma, whereas 40% survive if unsuspected orbital spread was discovered only on microscopic examination of the tissues. Adjunctive chemotherapy appears to increase chances of survival. When the tumor grows back into the optic nerve 10-12mm behind the eye to the point where the CRA and CRV exit, the tumor almost always gains access to CSF and may then slowly progress into the meninges over the base of the brain and in the ventricles. When this occurs, the prognosis is hopeless. The greater in size and number and the more anterior the tumors are located in the fundus, the less favorable the prognosis. Spontaneous regression is particularly common in retinoblastoma, estimated as 1%. The reasons for this is not clear. Systemic chemotherapy (6 mos.-1yr.) and local orbital irradiation (3500-4500 rads, cobalt 60) should be utilized if there is evidence of 1) episcleral nodules of retinoblastoma 2) massive posterior choroidal involvement with evidence of perilemmisarial extension, or tumor cells in a serous detachment of the choroid, and 3) iatrogenic rupture of the globe at the time of surgery. Other techniques include: diathermy, radioactive applicators, light coagulation, and cryotherapy. Prompt enucleation is usually preferred over treatment.
General Name: Toxocara
Specific Name: Toxocariasis
ICD 9-CM#: 128.0
Location: Retina, Macula
Associated Conditions: Systemic infection

S: Toxocara may infest the human in several forms. In visceral larva migrans, complaints include wheezy cough, chest pain, intermittent fever, loss of appetite, and sometimes abdominal pain. Skin eruptions can occur in the trunk or lower extremities. Ocular involvement reveals complaints of a progressive decrease in vision.

O: Biomicroscope examination reveals a low grade iritis usually with peripheral anterior synchilia. Granulomatous lesions on the retina are common and appear as raised hemispheric, whitish, protrusions about 1 disc diameter in size. The lesion is usually surrounded by pigmentary migration. Fibrous bands often radiate from the lesion. A greyish area can often be seen in the granuloma.

A: An infection of toxocara is usually associated with contact with dogs or cats. It is most common in the South-Central United States. Visceral larva migrans usually occurs before the age of 3 years. The average age for ocular larva migrans is 7.5 years. Other than a case history of eating dirt or association with animals, the definitive test is the ELISA test.

P: Prognosis if granulomas have formed at the macular area is poor. Periocular depomethyprednisolone 40-60 mg/week is the standard ocular treatment. This should not be used if a live larva is in the macula in an attempt to get the larva to migrate. Photocoagulation and cryotherapy have also been used to kill the larvae. Referral for systemic treatment is usually indicated.
General Name: Microcystoid degeneration/Cystoid degeneration
Specific Name: Microcystoid degeneration/Cystoid degeneration
ICD 9-CM#: 362.62
Location: Periperal retina from the ora serrata to the equator.
Associated Conditions: Reticular cystoid degeneration of the peripheral retina

S: None

O: Decreased size of visual field, conspicuous vascular pattern of degeneration gray background, finely stippled surface pattern.

A: By clinical appearance.

P: No treatment necessary.
General Name: Retinal Degeneration
Specific Name: Pavingstone Degeneration
ICD 9-CM#: 362.61
Location: Peripheral Retina
Associated Conditions: High myopia, Vitreous Degeneration

S: Patient is asymptomatic.

O: Ophthalmoscopic evaluation shows an area of thinned retina with a whitish splotchy appearance where it is devoid of pigment epithelial cells. The area of degeneration is non-elevated and sharply demarcated with scalloped margins. It is usually found at the ora serrata to the equator zone. The lesion may sometimes be hyperpigmented at the edges.

A: Pavingstone degeneration is usually innocuous and its prevalence rate is about 27% of the population. It is most often seen in myopes. The condition is benign as long as vitreous and the inner retinal layers are intact. However, in rare cases, the vitreous degenerates and can cause detachment. The size of the lesion increases with age and the axial length of the eye. Differential diagnosis from other peripheral retinal lesion must be made.

P: In cases where the vitreous degenerates, refer to an ophthalmologist. In most cases, treatment is not necessary. Document and have patient come in for an annual eye exam.
General Name: Retinal Degeneration

Specific Name: Peripheral Cystoid Degeneration

ICD 9-CM#: 362.62

Location: Peripheral Retina

Associated Conditions:
Retinal Detachment

S: Patients are usually asymptomatic.

O: Peripheral cystoid degeneration is observable most easily through a dilated pupil using a BIO. Scleral depression may be necessary to get a view out to the ora serrata. Cystoid degeneration appears as stippled depressions with rounded domes interspersed. They are most frequently observed at the ora, usually at the base of a dentate process.

A: This is the most common degeneration of the peripheral retina. Spaces between the outer plexiform and inner nuclear layers form and coalesce. These spaces run form the inner to outer layers which accounts for the stippled appearance of the retina. The degeneration begins to encircle the eye and extend from the ora to the equator. It may be noted in individuals as young as 8 years old and increases with age. It is most commonly found in the superior temporal quadrant.

P: This is a normal ageing change and it is usually a benign condition. In high myopes however, careful monitoring of the periphery must be undertaken due to the increased risk of detachments.
General Name: Retinoschisis

Specific Name: Retinoschisis

ICD 9-CM#: 361.10-361.12, 362.62, 362.73

Location: Retina

Associated Conditions:
- Retinal detachment.

S: Corresponding relative scotomas are present for both Degenerative/Senile and X-linked Juvenile retinoschisis types. X-linked Juvenile Retinoschisis is a vitreoretinal dystrophy that affects only males. It is bilateral, x-linked recessive, and potentially serious.

O: Degenerative/Senile Retinoschisis is a round or ovoid area of retinal splitting with a smooth fusiform elevation at the level of the outer plexiform layer (fusiform type) for a distance of at least one disc diameter surrounded on all sides by typical cystoid deneneration (the Bullous schisis type is a benign condition which resembles retinal detachment in appearance but holes are not seen). Visual field testing often shows a relative central scotoma as well as peripheral scotomas corresponding to area of schisis.

A: Vision is variable even in the presence of foveal schisis. However, as the disease progresses, there is a gradual deterioration of central vision, generally decreasing to the level of legal blindness by the age of 60 or 70. Rule out Acquired retinoschisis, Cystoid macular edema, Acquired macular hole formation, Goldmann-Favre disease, Wagner's disease, Stickler's syndrome.

P: Photocoagulation with final visual acuity often approximately 20/200.
General Name: Lattice Degeneration
Specific Name: Lattice Degeneration
ICD 9-CM#: 362.60
Location: Peripheral Retina

Associated Conditions:
- Retinal Detachment

S: No symptoms are given unless an associated rhegmatogenous retinal detachment occurs.

O: Ophthalmoscopic examination reveals a retinal thinning located at the equator or anterior to it. A classic lattice degeneration will show a network of fine white lines across the retinal thinning. Pigment accumulation occurs along the border of the thinning. White particles can be seen in the vitreous adjacent to the lesion.

A: Lattice degeneration is found in about 10% of the population. It is most commonly found in the 11 to 1 o'clock and 5 to 7 o'clock position. It is most easily diagnosed using the BIC or MIO. Vitreous liquification usually occurs at the lesion causing strong vitreous fibrils to condense around the lesion. The tugging vitreous can cause a retinal detachment which resembles a horseshoe.

P: The prognosis for lattice degeneration is good. The degeneration is stable and only and only about 25% will go on to retinal detachment. Management includes routine dilated fundus exams as well as educating the patient on the signs of retinal detachment.
General Name Retinal detachment
Specific Name Many types
ICD 9-CM# 361.0 (gener)
Location RPE/sensory retina interphase
Associated Conditions
Too numerous to list.

S: Traction detachments (from vitreous traction): photopsia, or flashes of light without retinal stimulation by light caused by vitreous traction on the retina and a sudden shower of black dots in the peripheral visual field resulting from a minute vitreous hemorrhage at the point of a retinal break. Non traction detachments: Decreased vision and a progressive visual field defect corresponding to the area of detachment.

O: The detached retina is gray or translucent and the normal choroidal pattern cannot be seen. The retina may be thrown into folds that change in location or shape with shifts in position of the eye or the head. The retinal vessels are dark red in the area of detachment and have an undulating course over its surface. The arteries and veins may appear to have blood of the same color.

A: The diagnosis of retinal separation is based on the ophthalmoscopic appearance of the retina. The most important differential point is the solid detachment caused by tumor, particularly malignant melanoma of the choroid. Failure of the detachment to regress with bed rest, the darker color, yellowish infiltrates, associated drusen, failure to transilluminate, and usual absence of hole formation indicate tumor formation.

P: Retinal breaks are closed by producing an area of chorioretinitis in the region of the defect so that adhesions between the edge of the break and the retinal pigment epithelium will seal the opening. The chorioretinal reaction may be produced by cryosurgery with application of intense cold (-80 degrees C.) or diathermy to the sclera in the region of the hole. Diathermy is used when the sclera is thick with dark choroid, giving a slate gray color, or after scleral dissection. Cryosurgery is used when the sclera is of normal white color and or unknown thickness. A variety of procedures are used to indent the sclera and choroid (buckle) so that the retinal pigment epithelium will be in apposition to the retinal hole. Selection of a particular procedure depends on the type of hole and the number of quadrants of the retina that contain holes.
General Name: Melanosis of the Retina
Specific Name: Bear Track Fundus, (Grouped Pigmentation)
ICD 9-CM#: 743.53
Location: Peripheral Retina
Associated Conditions:
Retinal pigment epithelium hypertrophy, Retinitis Pigmentosa

S: Patient is asymptomatic.

O: Ophthalmoscopic evaluation shows an area of retinal pigment epithelium hypertrophy which morphologically looks like bear tracks.

A: Group pigmentation is a non-progressive retinal hypertrophy. Eighty five percent of the cases are unilateral. It is an autosomal recessive condition. Thus, it props up with no prior family history. The retinal pigment epithelial hypertrophy must be distinguished from intraretinal pigmentation which is always pathological. Differential diagnosis must be made from Retinitis Pigmentosa and other similar clinical conditions of retinal dystrophies, degenerations, or inflammations.

P: Treatment is not necessary.
General Name  Retinal Vasculitis
Specific Name  Retinal Vasculitis
ICD 9-CM#  362.18
Location  Retina

Associated Conditions
Periarteritis Nodosa, Rheumatic Arteritis, Temporal Arteritis, Hypersensitivity Angitis, Wegener's Granulomatosis, Multiple Sclerosis, Tuberculosis, Sarcoidosis, Syphilis, Bechet Disease, Cytomegalic Inclusion Disease, Eales Disease.

S: Variable depending upon the underlying disease process. May have reduced vision due to macular edema.

O: Ophthalmoscopic examinations reveals segmented, dilated, occluded vessels with sheathing and inflammatory exudates. Retinal edema and ischemia. Retinal and vitreous hemorrhage. Cells in the aqueous or vitreous.

A: This is a complex group of conditions on which there is evidence of retinal vascular disease along with signs of inflammation. The most frequent cause is extension of an adjacent chorioretinitis. Fluorescein angiography shows stain of the vessel walls with leakage, macular edema, and all the other signs associated with ischemic response of the retina.

P: Treatment is directed at the underlying disease process.
**General Name**: Hyperviscosity Syndromes  
**Specific Name**: Waldenstrom's Macroglobulinemia  
**ICD 9-CM#**: 273.3  
**Location**: Peripheral Retina  
**Associated Conditions**: Cotton Wool Spots, Venous Tortuosity

**S**: The patient reports fatigue, weakness, skin and mucosal bleeding, headaches, and visual disturbances.

**O**: Ophthalmoscopy reveals:
- Dilation and tortuosity of the veins
- Arteriovenous nicking
- Superficial and deep hemorrhages with associated cotton wool spots
- No retinal edema

**A**: The condition is a disease of the reticuloendothelial system due to an abnormal IgM globulin with a high molecular weight. It is usually seen in elderly patients. The increased viscosity causes a slower flow which creates an increase pressure and dilation and tortuosity. The condition may resemble a bilateral vein occlusion and can be differentiated by a blood workup.

**P**: The condition can be treated by decreasing viscosity with plasmaphoresis. Retinopathy has been reported to be reversed with this type of therapy, but the underlying disease process remains. In any case a referral to an internist is in order.
General Name: Hyperviscosity Syndromes

Specific Name: Cryoglobulinemia

ICD 9-CM#: 273.2

Location: Peripheral Retina

Associated Conditions:
Multiple Myeloma, Hodgkin's disease

S: Patient will complain of fatigue, weakness, skin and mucosal bleeding, headaches and visual disturbances.

O: Ophthalmoscopic findings are similar to that of Waldenstrom's macroglobulinemia and include:
- Dilation and tortuosity of veins
- Arteriovenous nicking
- Superficial and deep hemorrhages with associated cotton wool spots
- No retinal edema

A: Cryoglobulins are proteins that precipitate when exposed to cold, but redissolve when heated. The condition is often associated with macroglobulinemia, but can be a clinical entity of its own. 50% of cases are found with multiple myeloma. It is also seen with Hodgkin's disease and some collagen diseases.

P: Plasmapheresis can also be utilized as a form of treatment in cryoglobulinemia, but again the underlying disease entity remains. Referral to an internist is in order.
General Name: Hyperviscosity Syndromes
Specific Name: Polycythemia
ICD 9-CM# 238.4
Location: Peripheral Retina

Associated Conditions:
CRV obstruction, Papilledema, Iron deficiency Anemia

S: Patient complaints are usually of recent origin and include: fatigue, decreases efficiency, difficulty in concentration, headache, drowsiness, forgetfulness and vertigo. Itching is present in ~50% of patients especially after a bath. Some patients are asymptomatic.

O: Initially signs include venous dilation and tortuosity which progresses to crossing defects, hemorrhages and impending CRV obstruction. Severe retinal edema can also occur. Occasionally papilledema is present.

A: The condition is due to hyperplasia of the bone marrow's erythroblastic elements. It is a chronic disease which occurs in adults that have high RBC counts of 6-8 million. It is also accompanied by enlargement of the spleen and liver. Polycythemia is considered in males with a hematocrit of >52% and in females with a hematocrit of >49%.

P: A phlebotomy is the safest therapy and doesn't depress marrow function. Iron deficiency anemia usually develops after the procedure however. It is less effective in those with high iron absorption and greatly elevated platelet counts. Usually 3-6 phlebotomies are needed to reduce the hematocrit to <50% and return the RBC mass to normal. After a phlebotomy is performed high iron foods should be avoided. Treatment prolongs life--average survival time with treatment is 13 years. Death usually results from myelofibrosis, acute leukemia or diseases of old age. Referral to an internist is in order.
General Name  Hyperviscosity Syndromes
Specific Name  Leukemia
ICD  9-CM#  208.9
Location  Peripheral Retina

Associated Conditions
Cotton Wool Spots, Retinal Hemorrhages, Roth Spots

S: In acute leukemia symptoms take an abrupt onset and include high fever, joint pains, bleeding from the mouth, nose and kidneys and bowel, or it may be insidious with progressive weakness and pallor.

In chronic cases symptoms include fatigue, weakness, anorexia, and weight loss

O: Signs include fullness of veins which progresses to tortuosity and segmentation and it may appear like a partial vein occlusion. Retinal hemorrhages are also present with Roth spots and cotton wool spots.

A: The hyperviscosity is due to a high WBC count. The associated anemia and thrombocytopenia contribute to superficial and deep hemorrhages. There is a relationship between severity of the disease and extent of retinopathy, but the relationship has very little prognostic value. The WBC counts are elevated to 15,000-500,000.

P: Therapy is variable and is aimed at reducing WBC counts to normal levels. Types of treatment include: antineoplastic drugs, alkylating agents, splenic irradiation, and X-ray therapy. The course of the disease is progressive and is usually terminal. The length of survival varies with the type of leukemia. A referral to an internist is in order.
General Name  Hyperacute Bacterial Conjunctivitis
Specific Name  Hyperacute Bacterial Conjunctivitis
ICD 9-CM#  098.40
Location  Conjunctiva

Associated Conditions  Panophthalmitis, Neisseriae gonorrhoeae, Neisseriae meningitidis.

S: In the very beginning the symptoms are similar to acute mucopurulent conjunctivitis but quickly progress to include swelling of both upper and lower eyelids, excessive mattering, aching pain, and a definite tenderness on touching the eye. Usually included are lid edema, marked hyperemia, chemosis with or without inflammatory membranes, and the presence of a rather prominent preauricular lymph node on the involved side which is tender on palpation. The disease usually involves the right eye in a right-handed adult. The gonococcus has the capacity to invade intact corneal epithelium.

O: The appearance is that of an ulcerative gutter near the corneal limbus (more often than centrally) surrounded by an inflammatory stromal infiltrate. If treatment is continually delayed or inadequate, the ulcer may remain local, extend circumferentially to form a ring abscess, or progress centrally. In any case, deeper involvement is rapid and perforation with subsequent endophthalmitis may occur.

A: Most often implies infection of the outer eye by one of the pathogenic Neisseriae, N. gonorrhoeae or N. meningitidis. The gonococcus is by far the more common cause. Gonococcal conjunctivitis is seen chiefly during the neonatal period (contracted via the infant's passage through the mother's infected birth canal), adolescence (as the result ofomite spread or autoinoculation), and adulthood (via autoinoculation from the infected genitalia). Chocolate agar selective media are preferred for culture. Rule out Mucopurulent conjunctivitis, Chronic blepharitis (chronic bact. conj./blepharoconj.).

P: Frequent lavage is useful in the early stages. Penicillin is the mainstay of parenteral therapy in nonsensitive patients. Methods: 1) Penicillin  G IM divided into 2 doses (2.4 million units per dose) and injected at different sites at one visit; administer 1gm of probenecid orally before the penicillin injection 2) Ampicillin, 3.5gm orally and 1gm of probenecid given simultaneously at 1 visit 3) Spectinomycin, 4gm IM at 1 visit in 2 divided doses; each 2gm (5ml) injection should be in the upper outer quadrant of the gluteal muscle 4) If the patient is sensitive to the penicillin and spectinomycin cannot be used, then tetracycline hydrochloride 1.5gm may be given orally as an initial dose followed by 0.5gm q.i.d. for 4 days. Topical antimicrobial therapy should be accompanied: bacitracin ophthalmic ointment, 500 units per gm, should be instilled every 2 hrs. for 2 days, thereafter 5 times daily until resolution. Gentamicin sulfate (garamycin) (0.3%) or tetracycline (1%) ointments may also be used. Use of cycloplegics is dependent on the extent of corneal involvement. Appropriate therapy usually results in the cessation of ocular discharge within 24-48 hrs.. Lid edema, hyperemia, and chemosis are slower to resolve, taking 7-14 days. Corneal ulceration is equally slow in resolving and usually leaves stromal scarring. Cicatricial bands may occur beneath the tarsal conj. if inflammatory membranes were present.
General Name: Mucopurulent Conjunctivitis
Specific Name: Acute Bacterial Conjunctivitis
ICD 9-CM#: 372.00-372.05
Location: Conjunctiva
Associated Conditions

S: Following a variable incubation period, patient complains of watering and irritation in one eye followed shortly by redness and subsequent sticking together of the eyelids upon awakening. The other eye is generally involved 1 to 2 days after the first. This spread is usually by the hands. It can spread from 1 person to another by fomites.

O: Both the tarsal and epibulbar conjunctiva are hyperemic with small petechial hemorrhages particularly characteristic of the pneumococcus or Haemophilus species. The discharge is relatively scanty and mucopurulent and accumulates readily at the base of the cilia. At times, a diffuse punctate keratitis is present.

A: The exudate, if copious, and the lids are stuck together; the conjunctivitis is probably bacterial or chlamydial. Membranes &/or pseudomembranes can be found in any severe acute conjunctivitis. Hemorrhagic exudation or petechiae possibly indicate hypersensitivity reaction to the infection. The acute conjunctivitis with exudation is called catarrhalls and is usually a self-limited process of about 2 weeks duration. The exudative process, characteristic of bacterial conjunctivitis, is the most important feature and is the source for transmission of infection. Children having lymphadenopathy readily develop follicles in any bacterial conjunctivitis. Older individuals respond chiefly by papillary hypertrophy, particularly in chronic infection. Staphylococcus aureus is the most common pathogen for adult conjunctivitis. Pneumococci are more common in childhood conjunctivitis, particularly in the northern states and during the colder months. Haemophilus species is more often a cause of conjunctivitis in the warmer regions of the U.S., particularly in the Southeast. It usually appears in young children and has been described as occurring seasonally between May and October, peaking during the early fall. Rule out Chronic Blepharitis (chronic bacterial conjunctivitis, blepharoconjunctivitis), Hyperacute bacterial conjunctivitis.

P: The conjunctival sac should be irrigated with saline solution as necessary to remove the conjunctival secretions and to prevent spread. Personal hygiene should be emphasized. Topical therapy with sulfonamide or an antibiotic (i.e. 1gt garamycin qid x 4 days) should be started immediately. In purulent cases laboratory studies should be done. Self-limiting. Pneumococcal conjunctivitis is basically self-limiting and clears within 7 to 10 days without treatment. Haemophilus clears in 9 to 12 days. However, most cases tend to resolve spontaneously and even more quickly with antimicrobial treatment. Under treatment, it should be improving in appearance each day. Staphylococcus aureus is an exception. If left untreated, a chronic blepharitis or blepharoconjunctivitis may result. Topical antimicrobial drops instilled hourly with ointment at bedtime or ointment 4 to 5 times daily over a 5- to 7-day period should considerably shorten the course of the infection. If treatment is based on clinical diagnosis alone, a broad-spectrum antimicrobial such as sodium sulfacetamide should be used. Topically applied in 10% to 15% strength, it is active against staphylococci, pneumococci, Haemophilus, and the moraxellas. Have patient RTC in approximately 1, 3, & 7 days for follow-up evaluation appointments.
General Name: Chronic Bacterial Conjunctivitis
Specific Name: Chronic Bacterial Conjunctivitis
ICD 9-CM#: 372.10-372.12, 372.15
Location: Conjunctiva

Associated Conditions:
Chronic Meibomianitis, Seborrheic dermatitis, Phlyctenular conjunctivitis.

S: The symptoms elicited vary depending on specific etiologic factors. Uniform complaints are foreign body sensation, redness, and mild to moderate mattering of the lids on awakening. If the symptoms also include redness of the lid margins, "granulated eyelids", lash loss, recurrent hordeola, one should suspect Staphylococcus as causative.

O: Additionally, there is most often present a crusted yellowish exudate, much of which encases the base of the cilia. Removal of this crust may be difficult and often results in loss of the lash with exposure of its infected follicle. These ulcerative characteristics distinguish staphylococcal blepharitis from the more common squamous form with its greasy scales and lid margin deposits. Seborrheic dermatitis involves the scalp and eyebrows. Persistence may cause chronic meibomianitis. A variable finding with long-standing bacterial blepharitis is the presence of marginal corneal ulceration, with the sudden formation of grayish white infiltrates within the stroma. Segmental dilation of the limbal vessels adjacent to the marginal lesion is not uncommon. Seen less commonly is the occurrence of phlyctenular keratitis.

A: Staphylococcus is the most common microorganism singularly responsible for chronic bacterial conjunctivitis, either insidiously or following an acute infection. Other causes can be from gram-negative rods (i.e. enteric bacteria), diplococci (i.e. N. catarrhalis), and diplobacilli (i.e. moraxella). If the infection continues untreated, thickening of the upper lid may cause a restricted elevation suggestive of ptosis, or there may occur distortion of the lid margin, resulting in entropion or ectropion. Rule out Acute bacterial conjunctivitis, Hyperacute bacteria conjunctivitis.

P: In most cases the frequent use of ophthalmic antibiotic solutions such as gentamicin or chloramphenicol for a gram negative organism for 7-10 days will eliminate the pathogens. The patient should be instructed of daily lid margin hygiene at home. The use of slightly moistened cotton applicator sticks after the use of warm compresses to the area is usually sufficient. This should be followed by the application of an ophthalmic antimicrobial ointment to the same area, which can be more effectively accomplished just before sleep. Sulfacetamide or bacitracin have proved useful. This treatment should be performed daily for 3-4 weeks. Acute internal hordeola and chronic meibomianitis can be managed with warm compresses and massage. Sterile marginal corneal ulcers occurring with bacterial blepharitis are responsive to topical steroid solutions or suspensions. Prednisolone (0.12%) used 4-5 times daily for 4-5 days is usually sufficient for resolution.

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General Name Conjunctivitis
Specific Name Ophthalmia Neonatorum
ICD 9-CM# 771.60
Location Conjunctiva
Associated Conditions
Systemic infection

S: Parents may bring in baby while observing red eyes with varying degrees of swelling and mucous discharge.

O: Ophthalmia neonatorum is a conjunctivitis which is contracted from birth or from infected persons or instruments in the early post-partum period. Differential diagnosis may be made from the following:
1) Bacteria: The baby demonstrates a hyperacute red eye usually 3-5 days post-partum. Marked swelling of the lids and mucopurulent discharge are present especially with Neisseria.
2) Chlamydial: Presently the leading cause of opthalmia neonatorum. Lid edema and chemosis are much milder than bacterial infections. Unilateral infections are very common. Onset is from 24 hours to a few weeks after birth. If left unchecked, conjunctival membranes and pannus may occur.

A: 3) Viral: The most common viral infection is Herpes. At approximately 2 weeks after birth a nonspecific chemosis, hyperemic conjunctiva, mucoid discharge, and lid edema are present. Women with active herpes are often advised to give birth by Cesarian section.
4) Chemical: The baby shows a red, watery eye 24 hours after instillation of Crede's prophylaxis (silver nitrate).

* Conjunctival scrapings are usually used for definitive diagnosis.

P: The prognosis is usually excellent if diagnosed promptly and appropriately treated. The primary approach is that of prevention with the use of Crede's prophylaxis at birth. It is toxic and used primarily against Neisseria. The preventive measure that is becoming the preventive measure of choice is tetracycline since it is effective against both Neisseria and Chlamydia. Treatment of a developed bacterial conjunctivitis is bacitracin in 10,000 units/ml sln alternated with .3% gentamycin sln 4-6x/day. Viral is best managed by 1% trifluridine every 2hrs 9x/day for 3 weeks. Irrigation of any discharge is advised. Treatment of chemical conjunctivitis is supportive. Concurrent treatment of the other eye in unilateral infections is advised.
General Name: Allergic conjunctivitis, hay fever conjunctivitis, atopic conjunctivitis

Specific Name: Allergic conjunctivitis

ICD 9-CM#: 372.14

Location: Conjunctiva, cornea, lids

Associated Conditions: Hay fever, allergies, asthma, hives, sinus problems

S: Eye, irritation, severe itching, photophobia (due to a mild keratitis), teary, watery eyes, burning sensation, vision affected slightly, rapid onset.

O: Conjunctival chemosis, vascular congestion, milky-pale pink appearance of conjunctiva, clear whitish exudate (stringy exudate in chronic reactions), swollen lid(s), uticaria, mild epithelial keratitis.

A: By symptom and signs. Conjunctival scrapings will show many eosinophils. The vision is affected due to the mild epithelial keratitis. Allergic conjunctivitis may involve a family history or a personal history. The key symptom is the itching and pale conjunctiva with a mild papillary response. Do not confuse the condition with Vernal conjunctivitis which also has intense itching but also has GPC. With chronic reactions, follicles may appear in the lower fornix.

P: Remove the allergen. Cold compresses will decrease the lid edema and hyperemia and help with vasoconstriction. Sunglasses decreases the photophobia. Antihistamine type drugs can alleviate the symptoms of the allergic conjunctivitis.
General Name: Papillary Conjunctivitis
Specific Name: Giant Papillary Conjunctivitis (GPC)
ICD 9-CM#: 372.12
Location: Conjunctiva
Associated Conditions: Cobblestone papillary hypertrophy, Decreased contact lens wear

S: Symptoms:
- Itch
- Decreased contact lens wearing time
- Vision not as clear

O: Slit lamp evaluation reveals a cobblestone papillary hypertrophy primarily affecting the superior tarsal conjunctiva.
Lab smear indicates eosinophiles, basophiles, and mast cells.

A: Giant papillary conjunctivitis most often observed from reaction to soft contact lens wear which leads to chronic irritation. It has also been reported in patients using methymethacrylate ocular prosthesis.

P: In the early stages of GPC, with only mild symptoms, the reaction usually subsides with the discontinuation of soft contact lens wear. However, more developed GPC require the use of topical antihistaminic-vasoconstrictor preparations for symptomatic relief. Increased mucin production may be a prominent component of GPC. Thus a mucolytic agents applied topically in artificial tears 4-6 times daily can be effective in reducing the problem. Once the patient becomes asymptomatic, reinstitute contact lens wear using a more vigorous schedule of enzymatic and surfactant cleaning in attempt to prevent recurrence of GPC or at least to keep it at an asymptomatic level.
General Name: Vernal Conjunctivitis

Specific Name: Vernal Conjunctivitis

ICD 9-CM#: 372.13

Location: Palpebral Conjunctiva, Limbus

Associated Conditions: Asthma, Eczema, Allergic Rhinitis, Hay Fever, Shield Ulcer

S: The chief symptom is itching which may be nearly intolerable. In addition there is tearing, photophobia, and foreign body sensation.

O: Vernal conjunctivitis occurs in two forms. The palpebral form has cobblestone papillae on the tarsal conjunctiva and sometimes a pseudomembrane. There is also a thickropy mucous discharge. It may be associated with shield ulcers of the superior conjunctiva. The limbal form occurs with papillary hypertrophy on the limbal conjunctiva. There is inflammation of the circumference of the corneoscleral limbus with the formation of a gelatinous, elevated area about 4 mm wide. It is associated with white chalky concretions known as Trantas dots.

A: Vernal conjunctivitis is a seasonally recurrent bilateral inflammation of the conjunctiva with a predilection for the warm months of the year, particularly in warm climates. One of the main diagnostic features is the conjunctival scraping showing prominent eosinophils. In addition to the papillary changes, a thickropy mucous discharge is a hallmark of the disease. The condition usually occurs in young people and tends to run a course from 4-10 years before remission occurs. Vernal conjunctivitis must be differentiated from GPC associated with contact lens wear, hayfever conjunctivitis, other forms of allergic conjunctivitis, other forms of allergic conjunctivitis, and trachoma.

P: Treatment consists of local instillation of corticosteroids, acetylcysteine and weak solutions of epinephrine and applications of cold compresses. Recurrent attempts at treatment with disodiumcromoglycate 2% to 4% solution topically qid has shown promising results in control of symptoms. Some relief can be seen with a change of locale or environmental exposure. Extensive allergy workup and hyposensitization are usually unsuccessful.
General Name: Conjunctivitis

Specific Name: General Viral Conjunctivitis

ICD 9-CM#: 077.9

Location: Conjunctiva

Associated Conditions:
PCF, EKC, Herpes simplex and zoster, Measles, Mumps and Newcastle's Disease

S: Symptoms are generally mild. Patients may complain of mild discharge which is usually watery, accompanied by a burning sensation that started about 3-7 days ago. They may report recent systemic illness.

O: The conjunctiva appears light pink to purple increasing in hue toward the plica.
- Tear BUT is decreased
- Patient may be tearing and follicles are observable on the superior tarsal conjunctiva.
- Preauricular lymphadenopathy is present.

A: Viral conjunctivitis must be differentially diagnosed from bacterial and allergic conjunctivitis. Lab tests reveal monocytes, inclusion bodies and giant cells in viral conjunctivitis. Lymphadenopathy is most frequently found in viral infection also. Once bacterial and allergic conjunctivitis have been ruled out a diagnosis of the specific viral agent must be made. The most common possibilities are PCF, EKC, Herpes Simplex and Zoster, Measles, Mumps and Newcastle's Disease.

P: Therapy is usually supportive. Cold compresses and vasoconstrictors are useful in acute stages. Topical corticosteroids are important in zoster. Topical sulfonamides assist in preventing secondary bacterial infections.
General Name: Conjunctivitis  
Specific Name: Chlamydial Conjunctivitis  
ICD 9-CM#: 077.0  
Location: Conjunctiva  
Associated Conditions: Trachoma, Systemic infection

S: Patient complains of a red, irritated eye. Symptoms include foreign body sensation, photophobia, lacrimation, and some purulent exudate.

O: Unilateral conjunctival injection with mixed follicular-papillary response are observed in early stages. A tender preauricular node is also noticed on the same side. During the second week, punctate staining may be observed as well as infection of the other eye. Marginal and central infiltrates may be present as well as a secondary iritis.

A: The lack of respiratory symptoms and fever differentiate chlamydial conjunctivitis from PCF. The symptoms are usually acute. Case history often reveals a new sexual partner within 1-2 months. Serologic test to determine if antichlamydial immunoglobins are present as well as scrapings indicating inclusion bodies are definitive for chlamydia.

P: Chlamydia infections tend to be chronic with spontaneous exacerbations and remissions. Secondary bacterial infections are also common. If left untreated, it can cause pannus and scarring leading to trachoma. Management dictates oral tetracycline 1 gm/day or erythromycin 2 gm/day, in 4 doses. Clinical response may not take place for 3-4 months but prognosis is good.
General Name: Toxic conjunctivitis
Specific Name: Contact conjunctivitis
ICD 9-CM#: 372.05
Location: Conjunctiva
Associated Conditions: Molluscum contagiosum, prolonged use of certain eye medications

S: Itching, chemosis of the conjuctiva, photophobia.

O: Follicles on the tarsal plate, papillary hypertrophy, conjunctival scarring, keratitis and pannus.

A: Case history, signs and symptoms. This syndrome follows chronic exposure of the conjunctiva to some foreign substance for prolonged periods of time. It occurs with molluscum contagiosum nodules of the lid margin which spill their contents into the conjunctiva. It also follows prolonged use of certain eye medications, and since these preparations are frequently out of date, it has been postulated that the follicular conjunctivitis was due to degradation products in the eye drops, thus the term "toxic." The keratitis can result in a very fleshy vascular pannus, particularly in molluscum, with significant visual impairment. The most common drugs causing the disease are eserine, DFP, IDU, and atropine but many others are also causative agents.

P: Removal of the agent causing the problem, up-dating the medication, disuse of medication until the conjunctivitis clears, change of medication, specific treatment of current symptoms.
General Name  Acne Rosacea
Specific Name  Acne Rosacea Conjunctivitis
ICD 9-CM#  372.31
Location  Conjunctiva
Associated Conditions  Blepharitis, Conjunctivitis, Keratitis, Facial eruptions, Corneal ulcers

S: Symptoms:
- facial skin eruptions, often present as a butterfly configuration
- mild to moderate irritation
The patient often has a history of prominent blushing and facial erythema, particularly after drinking coffee or alcohol.

O: Clinical findings:
1. Blepharitis: The most common manifestation of acne rosacea. It is associated with prominent plugging of meibomian gland secretion, which can be expressed with pressure to the lid.
2. Conjunctivitis: Low grade conjunctivitis often associated with blepharitis.
3. Keratitis: Marginal infiltrate with fasicle of vessels from the limbus or as ulceration that progressively advances toward the cornea. These ulcers rarely perforate unless intensive steroid treatment is used. Once ulcer results, there is chronic scarring.
4. Multiple facial telangiectasia may be present. In advanced cases it presents as rhinophyma.

A: Acne rosacea is primarily a disease of the sebaceous glands of the skin. Diagnosis is based on the facial eruptions and the ocular findings. Chronic blepharoconjunctivitis is seen in 30% of the cases. Of those with ocular involvement, 7% will develop corneal ulceration.

P: Treatment:
1. Tetracycline 250 mg 4 times a day for a period of 10 days, tapering dose to bid, or qid dosage, to be maintained for 3 to 6 months. (Contraindicated for pregnant females and small children)
2. Lid hygiene with daily massage, hot compresses, and removal of crusts from the lid margins.
3. Mild topical antibiotic e.g. erythromycin or bacitracin ointment may be used to control superinfection.
4. Apply steroid treatment with caution. Limit to no more than 1.0% prednisolone. (Note: intensive steroid treatment may cause ulcer to perforate.)
General Name: Conjunctivitis
Specific Name: Conjunctivitis Associated with Psoriasis
ICD 9-CM#: 372.30
Location: Conjunctiva
Associated Conditions

S: Foreign body sensation, burning, itching, tearing and marked photophobia.

O: Conjunctival involvement is in the form of granulated lesions affecting both the palpebral and bulbar conjunctiva. Corneal neovascularization, infiltrates and erosions. Scaling of the lid skin and marked scaling along the base of the lashes. Deposition of scales in the conjunctival sac. Inflammatory ectropion and entropion with trichiasis or madarosis are not uncommon in severe ocular involvement.

A: Psoriasis typically involves the scalp and extensor surfaces of the knees and elbows. Itching and burning are characteristic symptoms. Psoriasis is a common chronic and recurrent disease characterized by dry, well circumscribed, silvery, scaling papules and plaques of various sizes.

P: The patient should be referred to a dermatologist for treatment of the generalized psoriasis which often results in reduction of the ocular involvement.
General Name  Hypersensitivity Reaction

Specific Name  Atopic Dermatitis

ICD  9-CM#  692

Location  Conjunctiva, Adnexa

Associated Conditions
Keratitis, Asthma, Hayfever, Sinusitis, Urticaria, Keratoconus/Keratoglobus, posterior polar cataract, Retinal detachment, Atopic uveitis

S: Symptoms include: Itch, tearing, light sensitivity, burning sensation with blink, foreign body sensation, ropey discharge. Patient may also complain of swollen eyelids.

O: Case history reveals that the condition is not seasonal and tends to occur year round. Slit lamp exam shows papillae on upper lid, and possibly on the limbal area. If it is a recurrent problem they may have trantas dots. Epithelial keratitis may also be present. Initially the adnexa may be hyperemic and swollen, but it proceeds to become dry and scaly.

A: Usually patients will have a family history of atopic dermatitis. They may report any of these associated conditions: asthma, hayfever, urticaria, migraine headaches and sinusitis. Upon a blood workup they are found to have a high level of IgE producing B cells. This finding distinguishes the condition from GPC or trachoma. The lid and adnexa involvement along with the high IgE count distinguishes it from vernal conjunctivitis.

P: Initially steps should be taken to eliminate the allergin from the environment. Cold compresses, vasoconstrictors, and decongestants aid in comfort. Vaseline helps the skin flaking. Cromolyn is effective in stopping the allergic reaction and is a good alternative to steroids. If necessary steroids may be prescribed.

OCULAR CONDITIONS ASSOCIATED WITH ATOPIC DERMATITIS
1) Keratoconus/Keratoglobus
2) Posterior polar cataract (bilateral)
3) Retinal detachment
4) Atopic uveitis

133
General Name: Pemphigoid

Specific Name: Benign Mucous Membrane Pemphigoid

ICD 9-CM#: 694.61

Location: Conjunctiva

Associated Conditions:
- Filamentary Keratitis
- Keratoconjunctivitis Sicca

S: The patient may complain of blisters on the mucous membranes. The patient usually complains of an irritated, scratchy dry eye.

O: In the early stages subepithelial bullae, inflammation, and ulcers are seen on the conjunctiva. These bullae later rupture leaving scar tissue. At this stage the practitioner will observe conjunctival scarring, ectropian, and trichiasis. The progressive obliteration of the conjunctiva and fornix lead to eventual exposure and dry eye signs on the cornea. It may at end stages totally obliterate conjunctival fornices, create symblepharon, and leave the cornea keratinized.

A: Pemphigoid is believed to be an autoimmune disorder. It is most often seen in women over the age of 70 with HLA-B12 labeling. Besides the conjunctival involvement, concurrent oral and anal mucous membrane involvement is seen.

P: Prognosis in general is poor. Inflammation is chronic and progressive until massive scarring has occurred. Early use of systemic steroids and immunosuppressant drugs may help to reduce scarring. Supportive treatment such as ocular lubrication, artificial tears, and therapeutic contact lenses have been used to preserve the cornea with minimal success. Prophylactic antibiotics have also been advocated to prevent a secondary bacterial infection.
General Name: Stevens-Johnson syndrome

Specific Name: Erythema multiform

ICD 9-CM#: 695.1

Location: Skin, mucous membranes, conjunctiva

Associated Conditions:
Systemic and ocular herpes simplex infections, toxic reactions to systemic and ocular drugs, mycoplasma infection and neoplasms.

S: Fever, malaise, myalgia, arthralgia, symptoms of upper respiratory tract infection, bullous erosions of skin and mucous membrane, symptoms of conjunctivitis and acute iritis.

O: Signs of conjunctivitis and acute iritis, swollen eyelids, possible pseudomembranes, possible perforated cornea, skin lesions pigmented and depigmented areas where skin lesions have healed.

A: By clinical observation and histopathological studies. Erythema multiforme is an acute skin and mucous membrane disorder characterized by bullae and papules. Stevens-Johnson syndrome is a more severe form with conjunctival involvement in younger patients although common use has made the terms interchangeable. The disease occurs most frequently in patients between the ages of 3 and 50. Both sexes may be involved and there is no racial predilection. It is fatal in approximately 1/3 or patients secondary to liver involvement. The problem is the healing of the conjunctival epithelial leads to scarring, causing tear deficiencies, eyelid distortion, symblepharons, corneal erosions, neovascularization, and scarring. The disease is not progressive like pemphigus. The etiology of the entity is unclear although it seems to be precipitated by numerous antigens, including bacteria, viruses, and drugs, particularly the sulfonamides.

P: Treatment during the acute conjunctivitis stage includes keeping the eye irrigated and the eyelids apart from the globe so that the raw mucosal surfaces cannot adhere. Lubricating ointments such as Lacri-lube or Duratears may be instilled at bedtime or continuously during the day according to the severity of ocular involvement. Administration of topical antibiotics is helpful in preventing secondary infections. The chronic problems involve loss of tear quality and quantity, as well as eyelid problems that often require corrective surgery. Scleral shells and bandage lenses are helpful for protection of the cornea and tear film. Systemic and topical steroids may be used in early stages to suppress inflammation and swelling and help reduce the scarring.
General Name: Collagen Disease
Specific Name: Reiter's Syndrome
ICD 9-CM#: 372.33
Location: Conjunctiva
Associated Conditions: Arthritis, Urethritis, Conjunctivitis, Uveitis, Iritis

S: Ocular symptoms include conjunctivitis which occurs in one-third of the patients and iritis which occurs in only 1% of the patients. The condition is most often seen in men and rarely in children. The post-venereal form occurs exclusively to men while the post-dysenteric form has been observed in families.

O: The nongonococcal triad of Reiter's syndrome consists of:
- arthritis
- urethritis
- conjunctivitis or uveitis
The conjunctivitis is usually mucopurulent with preauricular adenopathy, chemosis, micropapillary hypertrophy, eyelid edema, and conjunctival hemorrhage. The iritis is usually seen with chronic prostatitis, sacroilitis, inflamed joints in the feet, hands, and wrist, and a positive HLA B27. Ocular symptoms are usually preceded by genitourinary symptoms.

A: The diagnosis is based on case history and the Reiter's syndrome triad. The white blood cell count and sedimentation rate and temperature are usually elevated. Although the cause of the disease is unclear, organisms such as Bedosonia (Chlamydia) and Mycoplasma pneumoniae may have a possible etiologic role.

P: Treatment consists of:
1. Treatment of the underlying cause of the disease.
   Systemic tetracycline may be useful in Bedosonia-induced cases.
2. The conjunctivitis is self-limited, usually resolves in 8 to 10 days.
   Topical vasoconstrictors and antibiotics in prophylactic doses e.g. sodium sulfacetamide.
3. Uveitis-Topical corticosteroids for patients with uveitis.
General Name: Relapsing Polychondritis
Specific Name: Relapsing Polychondritis
ICD 9-CM#: 372.30
Location: Generalized
Associated Conditions:

S: Ocular symptoms are varied depending on the specific ocular complication involved which includes: conjunctivitis, episcleritis, anterior uveitis, optic neuritis, and extraocular muscle paresis.

O: Episcleritis is the most common ocular manifestation. Generally mild and nonspecific conjunctivitis with little exudation occurs in approximately 1/4 of affected patients. Inflammation of the cartilage of the ear, nose, trachea and the peripheral joints. Floppy ears and saddle nose occurs in patients with repeated attacks of cartilage inflammation. Laringotracheal inflammation causes coughing and loss of voice.

A: Relapsing polychondritis is a recurrent inflammation of cartilage, and involvement of the pinnae of the ears is the most common feature. The onset is typically in the third to sixth decades and males and females are equally affected.

P: Management of the concomitant conjunctivitis observed in relapsing polychondritis essentially takes the form of supportive therapy such as topical vasoconstrictors and prophylactic antibiotics according to the severity of ocular involvement. The primary therapy should be directed at the underlying systemic disorder. Relapsing polychondritis responds well to adequate early treatment with steroids.
General Name Polyarteritis

Specific Name Polyarteritis Nodosa

ICD 9-CM# 446.0

Location Conjunctiva

Associated Conditions
Vascular Hypertension, Myocardial Infarction, Papilledema, Cogan's syndrome, Horner's syndrome, Cotton Wool Patches

S: Patients may report abdominal pain, fever, weight loss, nausea, headache, vertigo, convulsions and decreased vision. If the cornea is involved they complain of foreign body sensation and photophobia.

O: Systemic signs include:
- Renal disease which may progress to vascular hypertension
- Peripheral neuritis
- Myocardial infarction
- Pulmonary infiltration

Ocular involvement is common and includes:
- Hypertensive retinopathy with papilledema and cotton wool patches
- Conjunctival hyperemia with occasional subconjunctival hemorrhage
- Cogan's syndrome
- 3rd and 6th cranial nerve palsy
- Horner's syndrome and homonymous hemianopsia in some cases

A: Since polyarteritis nodosa is an inflammation of the small and medium arteries it effects are widespread. The disease can be triggered by an allergic reaction to a variety of drugs such as sulfonamides, penicillins, phenytoin, and isoniazid. It is seen more commonly in men (~75% of the cases) from age 20-50. Diagnosis is made on the basis of ocular and systemic signs and symptoms and laboratory blood work up which reveals leukocytosis, with marked eosinophilia and increased sed rate.

P: Prognosis is generally poor and mortality may approach 90% due to the widespread nature of the disease. The kidney, liver, heart and GI tract are common sites of involvement. Clinical remission has been produced by administration of systemic steroids.
**General Name** Systemic Lupus Erythematosus

**Specific Name** Systemic Lupus Erythematosus

**ICD 9-CM#** 710.0

**Location** Retina

**Associated Conditions**
Patients receiving isoniazid, phenytoin (dilantin), sulfonamides, and penicillin drugs.

**S:** Peak onset in the third and fourth decades and is 5 to 10 times more frequent in women than in men. The duration may be weeks to years. Various cutaneous lesions often occur and are frequently symmetrical. Erythematous lesions involving the face, neck, and extremities. A characteristic "butterfly" malar erythematous rash is sometimes seen.

**O:** The retinopathy is primarily a diffuse arteriolar occlusive vasculitis. Cotton wool spots, the most frequent retinal finding, are observed in 3-29% of patients. Multiple areas of capillary nonperfusion, retinal hemorrhage, venous stasis, papilledema, and retinal edema may arise from obliteration of minute arterioles. Fluorescein angiography may show focal leakage from capillaries and arterioles, obstruction of larger vessels, delayed venous filling, microaneurysms, capillary nonperfusion, neovascularization, and optic disc leakage. Involvement of the CNS was reported in 73% of patients, which resulted in neurological disturbances. Bilateral interstitial keratitis, superficial keratitis, corneal scarring, and bilateral band-shaped keratitis may also be seen.

**A:** The observed conjunctivitis tends to be nonspecific in nature. The pathologic hallmark of SLE retinopathy is inflammation of small vessels.

**P:** Prognosis is generally poor since there is particular predilection for injury to the heart, kidney, and retroperitoneal tissues. The primary therapy should be aimed at treatment of the underlying systemic disorder. Aspirin, chloroquine, and corticosteroids have been quite effective in the management of SLE. Management of the conjunctivitis takes the form of supportive therapy such as topical vasoconstrictors and prophylactic antibiotic according to the severity.
General Name Conjunctivitis
Specific Name Phlyctenular Keratoconjuctivitis
ICD 9-CM# 370.31
Location Conjunctiva, Cornea
Associated Conditions
Tuberculosis, Filamentary Keratitis

S: The patient complains of mild irritation, tearing, and itching. Pain and foreign body sensation are also often symptoms.

O: The primary sign is pinkish-white avascular nodules which vary in size from pinpoint to several millimeters. The location is typically at the limbus. The nodules are usually surrounded by hyperemic vessels. Corneal nodules migrate from the limbus in a wedge shape. These nodules may ulcerate leaving scarring and pannus. The nodules typically occur bilaterally. Filamentary keratitis is often seen concurrently in the superior cornea.

A: PKC is a delayed hypersensitivity response of the cornea and or conjunctiva to a protein antigen. It is fairly uncommon and is often seen in young patients, especially immigrants. It is most often seen in populations of poor public health conditions. PKC can be differentially diagnosed from a pinguecula in that it ulcerates and itches. Marginal corneal ulcers are less vascular and have a clear space between the ulcer and limbus unlike that from PKC.

P: PKC is a chronic disorder of exacerbations and remissions. Prognosis is usually good if corneal involvement can be controlled. Management includes topical steroids (prednisolone 1% q1h-q2h) and oral tetracycline 1gm/day for 2-4 weeks in patients over 8 years old. Referral to a general practitioner for TB evaluation is strongly indicated.
General Name Pinguecula
Specific Name Pinguecula
ICD 9-CM# 372.51
Location Bulbar conjunctiva
Associated Conditions Pterygium

S: Cosmetic only: yellowish-white bump on bulbar conjunctiva.

O: Yellowish white, slightly elevated, oval shaped tissue mass on either side of the cornea in the palpebral fissure.

A: By appearance with slitlamp. It is a benign degenerative tumor of the bulbar conjunctiva that appears as a yellowish white, slightly elevated, oval-shaped tissue mass on either side of the cornea in the palpebral fissure. The lesions are usually bilateral and located nasally. They become more common with advancing age. They cause a cosmetic defect and in some instances appear to precede a pterygium.

P: Treatment is usually unnecessary but excision is simple. Keeping the eye moist and not irritated from dust and exposed to the sun may help to reduce the formation of pingueculas. A pinguecula has the same histologic structure as a pterygium but is limited to the conjunctiva.
General Name: Conjunctival Disorder
Specific Name: Pterygium
ICD 9-CM#: 372.4
Location: Conjunctiva
Associated Conditions: Pinguecula, conjunctivitis, dry eye

S: Symptoms include:
- Mild irritation from conjunctivitis
- Decrease in visual acuity if it is encroaching on the visual axis
- Cosmetic complaints

O: Clinical findings:
- A wing shaped fibrovascular connective tissue overgrowth encroaching on to the cornea from the conjunctiva in the interpalpebral fissure.
- It usually advances from the nasal side of the cornea.
- There may be basophilic degeneration of the bulbar conjunctival stroma that demonstrate secondary calcification.
- There may be signs of chronic conjunctivitis.

A: Differential diagnosis must be made between pterygium and pinguecula. Histologically, pterygium is identical to pinguecula. A pterygium is characterized with corneal involvement while a pinguecula is limited to the conjunctiva. Neither condition is premalignant. The cause of pterygium is unknown, but conjunctival irritations from excessive sun and wind exposure may have a significant etiologic role.

P: Treatment consists of:
1. Lacrilube ointments
2. Ocuserts
3. Wet cells (protection from desiccation with goggles)
4. Surgery is indicated only if the pterygium is encroaching the visual axis or if the other conservative measures do not work. Recurrence of pterygium after excision varies from 20% to 40%. Beta radiation and triethylene-thiophosphoramide (Thiotepa), an anti-miotic drug have been used to reduce this recurrence rate.
General Name  Keratoconjunctivitis
Specific Name  Superior Limbic Keratoconjunctivitis
ICD 9-CM#  370.40
Location  Cornea, Limbus, Conjunctiva
Associated Conditions

S:  Patient complains of moderate to severe foreign body sensation, photophobia and often sharp pain.

O:  Superior palpebral and bulbar conjunctival hyperemia with superior filamentary keratitis in approximately 1/3 of the cases. Rose bengal reveals prominent staining in the superior bulbar conjunctiva.

A:  SLK is usually a chronic disorder exhibiting periods of exacerbation and remission. The disorder is typically bilateral through often asymmetric and is more frequently seen in young Caucasian patients. SLK tends to run a course of several months to years and while often quite annoying, rarely does it result in significant sequelae. Pannus formation, however, is not uncommon in severe cases. A subset of the disorder probably exists in relationship to soft contact lens wear. Viral, bacterial and fungal cultures have not revealed an infective organism. SLK must be differentiated from limbal vernal conjunctivitis, marginal ulcers, phlyctenulosis and some presentations of atopic conjunctivitis.

P:  Management is aimed at maintaining the patient in a comfortable a state as possible. Artificial tears often improve the patient's comfort and a pulse of topical steroids may reduce inflammation. Application of 0.5% silver nitrate solution may provide relief from symptoms lasting from 6-8 weeks after treatment. Bandage soft contact lenses have also been effective in relieving symptoms. Surgical resection of the superior bulbar conjunctiva has produced permanent remission in many patients and is the treatment of choice in severe cases or in cases unresponsive to medical therapy.
General Name: Hemorrhage
Specific Name: Subconjunctival Hemorrhage
ICD 9-CM#: 372.72
Location: Conjunctiva

Associated Conditions:
Hemorrhagic bacterial conjunctivitis, Adenoviral conjunctivitis

S: Patient reports a unilateral bright red blotch of blood on his eye. Pain can vary from none to severe (in cases of head injury).

O: Signs include:
- Bright red flat hemorrhage under the conjunctiva usually unilaterally.

A: Etiology of the hemorrhage is determined via case history. Patient may report major, minor or no detectable trauma to the eye. Hemorrhage may occur spontaneously with a cough, sneeze or lifting of a heavy object. If discomfort or discharge is present there may be an associated pneumococcal or adenoviral conjunctivitis. A hemorrhage that involves the entire bulbar conjunctiva following a head injury is suggestive of posterior globe rupture or orbital fracture.

P: If there is no indication of trauma or infection no treatment is needed and the hemorrhage will clear in 2-3 weeks. If discomfort or discharge is present diagnosis and treatment of the particular pathogen is required. If associated with head injury, a serious situation exists and medical help should be sought immediately.
General Name: Retention Cyst
Specific Name: Retention Cyst
ICD 9-CM#: 374.84
Location: Adnexa
Associated Conditions:

S: Patient may notice an elevation on eyelid.

O: A water-containing blister on eyelid.

A: This cyst is from a chronic inflammatory disease. It is associated with the accessory gland of Krause. Rule out Dermoid cyst, Viral wart.

P: It resolves itself, but it can be removed surgically if there are changes in keratometry findings or if visual acuity decreases.
General Name: Choristoma
Specific Name: Dermoid Cyst
ICD 9-CM#: 224.4
Location: Sclera, Cornea, Conjunctiva

Associated Conditions:
Goldenhar's Syndrome

S: Usually no subjective complaints since it is a developmental anomaly. Parents may bring child in due to the appearance of the cyst.

O: Observation reveals raised, circumscribed, pale yellowish growths which are generally located at the lower temporal limbus.

A: A dermoid cyst is a congenital tumor involving mesodermal and ectodermal tissue. The cyst is benign and rarely enlarges. When palpated the tumor is solid but can be moved around because it is encapsulated.

P: Prognosis is usually good. If the dermoid involves the cornea, a corneal keratoplasty may be required to restore corneal thickness and reduce astigmatism. Management is usually surgical excision of the cyst for cosmetic purposes.
General Name Pseudocancerous lesions of conjunctiva
Specific Name 1) Conjunctival papilloma 2) Xerosis
ICD 9-CM# 224.3
Location 1) Medial canthus 2) Temporal canthus
Associated Conditions

S: Bump in eye.

O: 1) Smooth, nonpigmented, nonkeratinizing surface and a regular, vascular papillomatous pattern.
   2) Localized abnormal keratinization of dry conjunctiva that forms a characteristic Bitot spot. It has a foamy surface and chalky, avascular appearance.

A: Tissue biopsy, clinical appearance.
   1) True pedunculated mucous membrane papillomas, particularly common at the medial canthus and often recurrent, may be of viral etiology. A less common, broad-based, sessile variety, which may occur anywhere on the conjunctiva and more closely resembles carcinoma in situ, has the same benign, regular histologic appearance. Neither of these tumors becomes malignant.
   2) This is a type of leukoplakic lesion, but the foamy surface and chalky, avascular appearance distinguish it from carcinoma. Bitot spot are not necessarily related to vitamin A deficiency.

P: Surgical removal of the tumor.
General Name: Non-pigmented tumors of the conjunctiva
Specific Name: Bowman's Disease
ICD 9-CM#: 190.3
Location: Conjunctiva

Associated Conditions:
- papilloma, adenoma, kerato-acanthoma, adenoma, squamous cell carcinoma, and pleomorphic adenoma, conjunctival carcinoma

S: Patient may report an elevated reddish-grey growth on the conjunctiva. The condition is most often seen in elderly males.

O: Bowman's disease of the conjunctiva is a carcinoma whose growth is by lateral extension within the epithelium without invasion of the underlying tissue. It is a highly vascular reddish-grey mass which is often accompanied by an inflammatory reaction.

A: Histological features of Bowman's disease are a proliferation of the basal cells of the epithelium with partial loss of their normal polarity. The nuclei are hyperchromatic and mitotic figures are common. The basement membrane is intact and spread by lateral growth. Differential diagnosis from other types of non-pigmented tumors of the conjunctiva of epithelial origin e.g. papilloma, adenoma, kerato-acanthoma, adenoma, squamous cell carcinoma, and pleomorphic adenoma.

P: Treatment consist of local excision.
General Name  Non-pigmented tumors of the conjunctiva

Specific Name  Squamous cell carcinoma

ICD 9-CM#  190.3

Location  Conjunctiva

Associated Conditions
Epithelioma, nodules, papilloma, adenoma, kerato-acanthoma, adenoma, Bowman's disease, and pleomorphic adenoma.

S: Asymptomatic. Tumors occur particularly to compromised host.

O: Clinical Findings:
1. Squamous cell carcinoma starts as a small gray nodule which becomes almond shaped as it extends around the limbus.
2. In advanced stages there is an extensive progression around the limbus and an invasion onto the cornea.
3. Larger feeder vessels develops which, when associated with tumor of the eye, should always give rise to the suspicion of malignancy.

A: Squamous cell carcinoma of the conjunctiva is relatively a rare condition which develops most commonly at the limbus in the interpalpebral zone. Differential diagnosis from other types of non-pigmented tumors and cysts of the conjunctiva with epithelial origin e.g. papilloma, adenoma, kerato-acanthoma, adenoma, Bowman's disease, and pleomorphic adenoma.

P: Treatment:
1. Local excision in the initial stages.
2. Radiation therapy if excision biopsy fails.
   Recurrence is common.
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**Associated Conditions**

None

**S**: Parents usually bring in children because of a red strawberry mark on the inner eyelid or on the white of the eye.

**O**: Signs include a capillary hemangioma on the conjunctiva, possibly associated with an orbital hemangioma.

**A**: These tumours occur early in life and usually disappear by 5 years of age.

**P**: The treatment of conjunctival hemangiomas is to let it resolve on its own. The cosmetic results of spontaneous regression are better than with any type of intervention.
General Name: Lymphoid Hyperplasia
Specific Name: Lymphoid Hyperplasia
ICD 9-CM#: 229.0
Location: Conjunctiva
Associated Conditions:

S: Patient may notice elevations on conjunctiva which are slow growing.

O: Conjunctival elevations are follicle-like in appearance and are nonpigmented.

A: This condition is rare. Reticulo's origin. Lymphoblasts and lymphocytes are present. Rule out Lymphoma/Lymphosarcoma, Orbital cellulitis, Orbital pseudotumor, Graves' disease.

P: This is a benign condition.
General Name Lymphoma
Specific Name Lymphosarcoma
ICD 9-CM# 202.3
Location Conjunctiva
Associated Conditions

S: Patient may notice elevations on conjunctiva which are growing at a moderate rate (faster than lymphoid hyperplasia elevations).

O: Conjunctival elevations are follicle-like in appearance and are nonpigmented.

A: A rare malignant condition. Reticulosis origin. Lymphoblasts and lymphocytes are present. Rule out Lymphoid hyperplasia, Orbital pseudotumor, Orbital cellulitis, Graves' disease.

P: Refer patient to a surgeon for removal, especially if there has been a history of this condition.
General Name: Non-pigmented Tumors of the Conjunctiva

Specific Name: Peripheral Nerve Origin Tumor

ICD 9-CM#: 190.30

Location: Conjunctiva

Associated Conditions:
None

S: 1) Benign:
   a. Neurofibroma: No symptoms unless large.
   b. Neurilemoma: A deep pain in the globe.

2) Malignant:
   a. Schwannoma: A deep pain in the globe.

O: A unilateral ptosis and exophthalmos are usually observed. Ophthalmoscopy is normal. There may be some restrictions of gaze.

A: Assessment is made primarily by the determination that a growing mass is growing and displacing tissue in the orbit. A neurofibroma upon surgery appears as a stringy strand of proliferative nerve cells. Neurilemomas and schwannomas are encapsulated, firm, yellow masses. An acquired unilateral exophthalmos is of high suspicion.

P: Benign neurilemomas are easily removed with no recurrence after surgery. Neurofibromas are more difficult to remove since nonecapsulated and tend to reoccur. The prognosis for a malignant schwannoma is poor since extreme posterior growth and early invasion into the roof of the orbit and cranial fossa make it very difficult to surgically remove.
General Name: Conjunctival nevi
Specific Name: Conjunctival nevi
ICD 9-CM#: 224.2
Location: Conjunctiva
Associated Conditions:

S: Bump in eye.

O: Pigmented or nonpigmented growth of conjunctival tissue.

A: Tissue biopsy. Approximately one third of conjunctival nevi are not pigmented. More than one half of conjunctival nevi contain associated cystic inclusions of conjunctival epithelium, which are not found in skin. Many of these cysts are clinically evident and can help in the differential diagnosis of these lesions. Increased melanogenesis in nevi can result from pituitary stimulation (pregnancy, adrenal insufficiency), physical irritation (actinic radiation, trauma, inflammation), and coexisting malignant melanoma. Malignant transformation of nevi to melanomas is rare. However, many of these nevi are a cosmetic blemish. Excision corrects this and eliminates the remote chance of malignant change.

P: Excision.
General Name: Pigmented Lesions of the Conjunctiva

Specific Name: Congenital Melanosis Oculi

ICD 9-CM#: 372.55

Location: Conjunctiva

Associated Conditions: Nevus of Ota, Melanosis Oculi, Blue Nevus

S: Patient may complain of a diffuse slate grey pigmentation of the eye lids or a bluish area near the limbus.

O: Clinically there are two types of conjunctival melanoma:
   1. Focal nodular melanoma: appears as an isolated elevated lesion.
   2. Diffuse melanoma: extends radially with the edge of the lesion slightly mottled.

A: Congenital Melanoma Oculi is seen as an isolated condition or as a part of Nevus of Ota where there is an unilateral blue nevus of the skin of the face and eyelid. These patients have an increased risk of choroidal malignant melanoma but do not carry any extra risk of developing malignancy.

P: The tumor is usually benign and periodic histological study is indicated only when there is evidence of progression. However, due to higher than normal risk of developing choroidal malignant melanoma, patient should be observed annually.
General Name: Melanosis

Specific Name: Acquired Conjunctival Melanosis

ICD 9-CM#: 224.3

Location: Conjunctiva

Associated Conditions

S: Asymptomatic.

O: Diffuse, flat, slowly growing melanotic lesion of the conjunctiva. An inflammatory zone in the subepithelial tissue is typical. The lesion may wax and wane. Elevation in an area suggests malignant transformation but can also occur from an inflammatory infiltrate.

A: Primary acquired melanosis is a lesion with a definite potential for malignant transformation. Atypical, pleomorphic, hyperchromatic melanocytes proliferate in the epithelium but do not invade the subepithelial tissue until malignant transformation has occurred. This condition must be differentiated from secondary acquired melanosis related to irritation of the conjunctival epithelium from such things as foreign bodies and inflammation and to aging changes in the bulbar conjunctiva, particularly nonwhites.

P: The ideal treatment of acquired melanosis has not yet been determined. Many patients can be successfully treated by local excision of affected conjunctiva. The use of cryotherapy after excision may improve the prognosis.
General Name: Pigmented Lesions of the Conjunctiva

Specific Name: Malignant Melanoma

ICD 9-CM#: 190.3

Location: Conjunctiva

Associated Conditions:
Melanoma of Ciliary Body, Skin, or Mucous Membranes

S: Patient may report observation of a pigmented lesion that has become elevated or is producing irritation.

O: Signs include an increased pigmentation on the conjunctiva that has an elevated appearance and changes in size over time.

A: The conjunctival melanoma may develop from nevi, acquired melanosis, or without a pre-existing lesion. It can also arise from a ciliary body melanoma or melanoma of the skin or mucous membrane. Determination of malignancy and differential diagnosis from other pigmented lesions is obtained from a biopsy of the affected tissue.

P: If the lesion is determined to be malignant the treatment is extenteration or wide local excision. If the lesion arose from an acquired melanosis it will probably respond to radiation treatment.
General Name: Adrenochromes
Specific Name: Adrenochromes
ICD 9-CM#: 372.55
Location: Conjunctiva
Associated Conditions:

S: Also known as "melanin pigments". This patient is usually asymptomatic, but the granules sometimes cause irritation by scratching against the cornea during blinking. Patient has been using epinephrine.

O: Black oxidation deposits of epinephrine deposited in the crypts of Henle of the palpebral and fornical conjunctiva.

A: Adrenochromes may also blacken corneal scars, the eyelid margins, the caruncle, and soft contact lenses. Occasionally they obstruct the lacrimal canaliculus or fill the lacrimal sac.

P: The deposits can easily be removed by plucking them out with the tip of a hypodermic needle.
General Name Argyrosis
Specific Name Argyrosis
ICD 9-CM# 372.55
Location Cornea
Associated Conditions

S: Patient is asymptomatic.

O: Grayish discoloration in peripheral deep stroma and Descemet's membrane which is more noticeable in patients with light irides.

A: Deposition of silver salt in the cornea.

P: Discontinue use of drug containing silver and switch to another drug of choice if possible.
General Name  Pigmented Lesions of the Conjunctiva
Specific Name  Copper (Kayser-Fleischer Ring)
ICD 9-CM#  372.56
Location Conjunctiva, Cornea
Associated Conditions  Wilson's Hepatolenticular Degeneration

S: Usually none except cosmetic complaint.

O: A brownish-greenish ring in the corneal periphery or conjunctiva.

A: Upon slit lamp exam, a copper ring can be seen at the level of Descemet's membrane. It extends from the peripheral cornea into the trabecular meshwork. It is often only seen during gonioscopy in early stages of Wilson's disease.

P: Prognosis is good as it fades with the treatment of Wilson's disease. Management of Wilson's disease includes a low copper diet and treatment with the antichelating agent penicillamine.
General Name  Scleritis
Specific Name 1) Diffuse scleritis (379.03) 2) Nodular scleritis (379.02)
ICD  9-CM#  379.0
Location  Sclera and overlying episclera and conjunctiva
Associated Conditions  Connective tissue disease

S: Pain, photophobia, redness, tenderness, and lacrimation. The pain is the most dominant feature. It is boring in nature and severe enough to prevent sleep.

O: On examination the area of inflammation has a bluish red hue in contrast to the brighter red of episcleritis and may be sectorial or diffuse. Episcleral and conjunctival tissues are characteristically dilated. Topical application of 1 drop of epinephrine 1:1000 constricts the superficial vessels but not the deeper scleral vessels. Elevated nodules are noted in nodular scleritis. Observation with the biomicroscope will reveal an edematous sclera pushed forward and the deep episcleral network is congested more than the superficial network. The episcleral network is disturbed and preferential channels open up, and beading and aneurysm formation of this vascular network occur.

A: By signs and symptoms, presence of systemic disease (not always necessary). The two basic types of scleritis are anterior (nodular, diffuse, and necrotizing) and posterior. This section will cover only nodular and diffuse. The nodule or nodules consist of scleral tissue which is immovable and edematous episclera which is tightly adherent to the nodule and tender to the touch. The sclera may become transparent below the nodule; but it does not become necrotic nor does the condition extend below the nodule as in necrotizing scleral disease. The diffuse anterior type of scleritis consists of an inflammation that is more widespread that in the nodular disease, and it may involve either a small segment or the whole of the anterior segment. It has an insidious onset with generalized orbital aching. The blood vessels are intensely injected and appear as deep-seated, small, numerous radial vessels surrounded by numerous capillaries.

P: If the scleritis is associated with a connective tissue disease, then anti-inflammatory and other agents should be administered based on the overlying disease. It is a characteristic finding in all patients with scleritis that the pain is relieved as soon as the inflammatory reaction is suppressed, even though the external appearance appears the same. The principle anti-inflammatory drugs are oxyphenbutazone or indomethacin. If these drugs do not cause a favorable response, then avascular areas may appear in the sclera or episclera. Prednisone is used systemically in high doses (60 mg. to 80 mg.) daily or higher if need be. Milder cases can be controlled with topical ophthalmic corticosteroid solution or ointment (10% oxyphenbutazone).
General Name  Episcleritis
Specific Name  Episcleritis: Simple or Diffuse
ICD 9-CM#  379.00
Location  Episclera
Associated Conditions
Uveitis, Rheumatoid arthritis, Gout, Herpes Zoster Ophthalmicus, Puberty, Menopause.

S: Symptoms include:
- Pain
- Photophobia
- Tenderness
- Lacrimation (variable degrees)
The condition is more prevalent in women than in men.
The peak age of incidence is between the ages of 30 and 40.

O: Ocular examination reveals:
- Segmental or diffuse vascular engorgement that gives the eye a pink or purple color.
- Infiltration, congestion, and edema of the episclera, the overlying conjunctiva, and the underlying tenon capsule.
- In nodular episcleritis, there is a purple, tender, and elevated nodule 2 to 3 mm in diameter.
- Most episcleritis involves the area between the insertion of the rectus muscle and the limbus.
- About 15% of patients with episcleritis develop mild iritis.

A: Episcleritis is a localized inflammation of the superficial tissues of the sclera. There are two main types: simple and nodular. Most cases of episcleritis is in the simple form (75%) with a tendency for regression and recurrence of moderate to severe inflammation at 1 to 3 month intervals. These inflammations lasts 7-10 days and occur more commonly in the spring or fall than summer or winter. Etiology is often unknown, but attacks are often associated with family or occupational stress and hormonal factors e.g. menopause and puberty. In nodular episcleritis, there is no history of periodicity but rather a more protracted course of mild attacks of inflammation. About 30% of cases of nodular episcleritis is associated with medical problems e.g. rheumatoid arthritis, gout, and herpes zoster ophthalmicus. Episcleritis is a benign, self-limiting condition. However recurrence is common and may torment patient for years. Over 50% of the patients experience intermittent attacks that lasts 3-6 years, with some lasting up to 30 years. Differential diagnosis from conjunctivitis & scleritis.

P: Treatment is usually not required since the disease is self-limiting (1-2 weeks) with little or no damage to the eye. Symptoms can usually be relieved by topical decongestants e.g. 0.1% Dexamethasone which will resolve the inflammation in 3-4 days. More symptomatic cases, responds well with topical steroids e.g. prednisolone 1% tid. Occasionally, a clear history of an exogenous sensitization can be obtained and the removal of this will prevent future attacks.
General Name: Scleritis
Specific Name: Necrotizing Anterior Scleritis
ICD 9-CM#: 379.06
Location: Sclera
Associated Conditions:

S: The patient presents with a gradual onset of a painful, injected eye. The pain is severe and may be localized to the eye or it may be diffuse and described as radiating to the jaw, sinuses and temple. There is often associated photophobia, lacrimation and loss of visual acuity.

O: Inflammation is a prominent feature of anterior scleritis and produces a bluish red injection which is more easily seen in daylight. The scleritis begins in a localized area with acute congestion of the superficial and deep vessels which become greatly distorted or occluded. The episcleral congestion can be differentiated from that of the sclera by instilling phenylephrine 10% or epinephrine 1:1000. There are focal areas of avascular scleral dropout in which the sclera becomes transparent and the uveal layer observed when viewed in daylight.

A: Necrotizing scleritis is the most severe and unremitting and has the highest complication rate. Over 60% of the cases develop complications other than scleral thinning and 40% have a loss of visual acuity. 29% of patients with necrotizing scleritis may be dead within 5 years. The scleritis begins in a localized area but if the inflammation remains uncontrolled, the entire segment can become involved. Severe edema and acute congestion is known as brawny scleritis. Scleritis occurs more commonly in females with a peak incidence in the 4th-6th decades and the condition is bilateral in approximately 50% of the cases. Ocular complication of necrotizing scleritis include acute stromal keratitis, corneal guttering, keratolysis, open angle glaucoma, cataract and retinal detachment.

P: Topical steroids frequently increase comfort and occasionally maintain a remission but may not be sufficient to induce a remission. When topical steroids are ineffective, systemic treatment with nonsteroid anti-inflammatory agents is possible. For unresponsive cases or severe necrotizing inflammation the mainstay of treatment is systemic steroids in a dose of 80-120 mg prednisolone per day for the first week with rapid tapering to 20 mg daily, to be discontinued within 2 weeks. Topical steroids may be used to sustain a remission. Immunosuppressive treatment may be necessary in cases of necrotizing scleritis when high doses of steroids have failed. With successful treatment, necrotic areas may disappear or may leave a thin film of conjunctiva or episclera covering the uvea. Small defects are usually covered by new collagen but large defects may require a scleral graft.
General Name  Necrotizing Scleritis without Inflammation
Specific Name  Scleromalacia Perforans
ICD  9-CM#  379.04
Location  Sclera
Associated Conditions
Connective Tissue Disorders, Rheumatoid Arthritis

S: Patient may feel prickling or burning pain but no true pain. They may notice a grayish-yellow patch on their sclera. Patients may report a longstanding rheumatoid arthritis.

O: Grayish yellow necrotic patches or plaques on the sclera. Eventually the plaques slough off and reveal areas where the choroid is visible through the sclera. These areas are avascular and no inflammation is present.

A: Need to rule out epitheliomas. Scleromalacia perforans is bilateral more than 50% of the time in patients with rheumatoid arthritis. The condition is caused by arteriolar occlusion of the deep vascular network. In cases where the thinning occurs near the cornea, conjunctival ulceration and corneal guttering are common.

P: Refer patient to a rheumatologist if currently not under treatment for the connective tissue disorder. Treatment of scleromalacia perforans is ineffective unless it is instituted early on. Steroids may be given and in severe cases a scleral transplant may be done. Generally, perforation is rare and the damaged sclera need not be replaced. The anterior chamber should be monitored for any type of reaction however, as this is an ominous sign.
General Name: Posterior Scleritis
Specific Name: Posterior Scleritis
ICD 9-CM#: 379.07
Location: Sclera, Retina, Choroid
Associated Conditions:
Anterior scleritis.

S: A posterior scleritis usually occurs as one extension of anterior disease, but occasionally the majority of the inflammation is in the posterior segment, in which case the exudative retinal detachments and subretinal granulomas can be mistaken for malignant melanoma. If the granulomatous condition extends outward, the extraocular muscles become involved, producing proptosis, lower lid retraction, and ophthalmoplegia, with or without intraocular signs.

O: Posterior scleritis masquerades as many different conditions. It can cause quadrantal and even larger geographic choroidal inflammations and effusions, proceeding to retinal detachment. Optic disc edema and retinal venous congestion may occur when the scleritis and tenonitis impinge on the optic nerve. Since the posterior sclera is invisible, the diagnosis of posterior scleritis can be made only if the anterior sclera is also involved or some other signs in the orbit lead one to suspect it. The conditions may be a posterior extension of anterior scleritis, in which case inflammation of the pars plana, anterior and posterior uveitis, and cataracts develop.

A: Often posterior scleritis raises the prospect of a retrobulbar or choroidal tumor. Posterior scleritis is often patchy, sparing most of the posterior pole. It may be restricted to a circumferential equatorial zone, with no involvement of the posterior pole. The disease can move from place to place on the posterior sclera at different phases of resolution, resulting in confluent patches. Posterior and anterior scleritis may alternate in the same patient. Rule out Pars planitis, Retrobulbar or Choroidal tumors.

P: Nonsteroidal anti-inflammatory agents are effective in suppressing the inflammatory response in most diffuse and nodular scleritis. Although local steroid therapy increases the patient's comfort, it is not effective in suppressing the scleral inflammation. If the scleritis is severe or necrotizing or if areas of vascular closure are detected, then the use of systemic steroids is mandatory. A sufficient amount must be given to suppress the condition. The problem is to decide what dosage is appropriate. One effective scheme is oral prednisolone, 60 or 80 mg, given for 2 days and reduced over 1 week to 20 mg. Reduction by 2.5 mg every other day is continued until the pain recurs or signs of inflammation begin to recur. Surgery is rarely necessary.
General Name: Sclerokeratitis  
Specific Name: Sclerokeratitis  
ICD 9-CM#: 379.05  
Location: Sclera, Cornea  

Associated Conditions:  
Rheumatoid Arthritis, Polyarteritis, Wegner's Granulomatosis

S: The patient complains of pain, photophobia, lacrimation, and irritation.

O: Observation reveals a hyperemic, inflammed sclera. Adjacent to the scleritis there is corneal edema and stromal infiltrates causing the cornea to thicken and appear grey. The corneal involvement spreads toward the axis usually leaving the pupillary axis clear. Irregular vascularization usually follows the leading edge. Also trailing the leading border, spotty white opacities may develop becoming crystalline in appearance.

A: The disease usually occurs unilaterally. It can only occur secondary to a scleritis. A non-granulomatous uveitis is always present.

P: The prognosis and management depend on the underlying scleritis. Scleritis treatment usually includes such drugs as oxyphenbutazone, indomethacin, prednisolone, and azathioprine (see Scleritis). Warm compresses and topical steroids can be used to relieve discomfort. Dilation with 1% atropine should be used to prevent anterior synishiae.
General Name: Staphyloma
Specific Name: Staphyloma
ICD 9-CM#: 379.1
Location: Sclera

Associated Conditions:
Congenital Glaucoma, Pathologic Myopia, Trauma

S: General symptoms include a bulging of the entire eye or of a localized area of the white of the eye that has a darkened hue.

O: Total Staphyloma- If a total staphyloma or buphthalmos is present the entire eye bulges and appears larger than the fellow eye. Corneal edema and increases IOP are also present.

Localized Anterior Staphyloma- Anterior staphylomas are visible as localized areas of bulging lined with dark pigment of the uvea.

Localized Posterior Staphyloma- In posterior cases visual acuity is decreased and ophthalmoscopic exam reveals a central retinal degeneration. BIO exam shows a well defined edge of posterior scleral outpouching.

A: A staphyloma is an enlargement of the sclera lined by uvea that occurs secondary to embryological defects, localized degeneration or high IOP.
-Total Staphylomas- attributed to uniform stretching of the immature corneal and scleral fibers due to the increased IOP in congenital glaucoma. It is seen unilateral or bilateral and is more common in males.
-Localized Staphylomas:
-Ant. type- occur anterior to the equator and are most commonly due to inflammation of the uvea or after trauma to the sclera accompanied by elevated IOP.
-Post. type- occur in pathologic myopia where the post. sclera thins as the axial length increases. Only seen in funduscopic exam.
-Equatorial staphyloma- Involve areas near the vortex veins. It may contribute to retinal separation but is often undetected until exposed in surgery.

P: In buphthalmos the initial treatment is a goniotomy supplemented by topical therapy or filtering surgery. Eyes will have high refractive error so adequate refraction and prevention of amblyopia is necessary. In other types of staphyloma remedial measures are advisable if there is any hope of saving the eye. These measures vary and may include prophylactic treatment for retinal detachment, basal iridectomy, scleral buckling and scleral resection.
General Name: Ectasia
Specific Name: Ectasia
ICD 9-CM#: 379.11
Location: Sclera
Associated Conditions

S: Patient is asymptomatic.

O: A general term used for the description of outpouching/stretching/bulging of a structure.

A: This can occur with the nasal fundus in a patient with a tilted disc, with the sclera with or without the involvement of uveal tissue, and with the cornea (an axial ectasia is also known as keratoconus).

P: Treatment should be referred to each specific condition. However, nothing can be done in most cases.
General Name: Scleral Thinning
Specific Name: Scleral Thinning
ICD 9-CM#: 379.1
Location: Sclera

Associated Conditions:
Scleritis, high myopia, aging, rheumatoid arthritis, osteogenesis imperfecta

S: Usually none in itself unless the accompanying cause is active. Cosmetic concern may be a complaint.

O: Observation of the sclera will reveal bluish brownish patches rather than the normal white.

A: Any type of stress on the sclera can cause a thinning of the scleral tissue. The color is a result of visualization of the uveal pigment. It is most often seen in chronic scleral and uveal inflammations. Other common causes include high myopia, aging, RA, and osteogenesis imperfecta.

P: The prognosis depends on the cause and the amount of thinning. Management is treatment of the underlying cause. If thinning becomes severe scleral grafts may be required.
General Name  Senile hyaline palque
Specific Name  Senile hyaline plaque
ICD 9-CM#  379.16
Location  Sclera
Associated Conditions  Old age

S: None.

O: Darkish deposition located immediately anterior to the insertion of the medial or lateral rectus muscle in the sclera.

A: By clinical appearance and histological testing. It occurs only in individuals over 50 years of age. It appears as an area of translucency in the sclera. The overlying episclera remains unchanged. The periphery of the hyaline ring is often hard and white and calcareous. It may contain calcium sulfate (gypsum). They are caused by the tug of the recti muscle insertion on the scleral fibers; however, they never cause any trouble and require no treatment.

P: No treatment necessary.
General Name: Eyelid Cyst
Specific Name: Sebaceous Cyst
ICD 9-CM#: 374.84
Location: Adnexa
Associated Conditions: suderiferous cyst, dermoid, sebaceous cell carcinoma, and basal cell carcinoma

S: Patient reports a yellowish white lumps on the eye lid.

O: Signs include:
- Yellowish white lumps on the skin with a central punctum.
- Do not spontaneously drain, thus there is no seepage.
- No decrease in vision.
- No pain.

A: Sebaceous cysts are a benign tumor caused by retained cheesy secretions from ordinary skin sweat glands. Differential diagnosis from other types of cysts such as suderiferous cyst, dermoid, sebaceous cell carcinoma, and basal cell carcinoma are indicated.

P: Sebaceous cysts may be treated with simple excision. Recurrence of suderiferous cyst where it has been excised should make one suspicious of malignancy.
General Name: Suderiferous Cysts
Specific Name: Suderiferous Cysts
ICD 9-CM#: 374.84
Location: Eyelid
Associated Conditions:

S: Usually painless but occasionally can cause irritation or interfere with successful contact lens wear.

O: Small, round, translucent, elevated masses caused by blockage of the ducts of the gland of Moll. One or more lesions ranging in size from 1-2 mm in diameter may be observed on the eyelid margin.

A: Suderiferous cysts is caused by blockage of the ducts of the gland of Moll and must be differentiated from sebaceous cysts, inflammatory and benign tumors of the eyelid margins.

P: Suderiferous cysts tend to reform following puncture, but will rarely reappear if the dome of the cyst is excised. After cleaning the eyelid margin with an antiseptic and anesthetizing the surface of the cyst with a drop of local anesthetic, the thin overlying membrane should be dissected with the sterile tip of a 25 or 27 gauge needle. Material from the cyst can be then expressed with two sterile cotton-tipped applicators placed on each side of the base of the cyst. Application of an antibiotic ointment such as bacitracin or bacitracin-polymyxin B will prevent infection.
General Name: Cysts
Specific Name: Serous cysts
ICD 9-CM#: 372.75
Location: Adnexa
Associated Conditions: None

S: Patient notices a painless fluid filled bump on the eyelid.

O: Cysts appears as an accumulation of watery material under the skin of the eyelid. May also be found elsewhere on the body.

A: Must differentiate from a papilloma which is a true growth with keratinized cells over it. Serous cysts are usually seen secondary to burns and can be found anywhere on the body.

P: No treatment is required. The cysts may be excised and drained if the patient desires. Personal hygiene should be stressed.
**General Name** Papilloma

**Specific Name** Squamous Cell Papilloma

**ICD 9-CM#** 239.2, 216.1, 702

**Location** Adnexa

**Associated Conditions**
Transitional cell papilloma, Mixed cell papilloma.

**S:** Patient is asymptomatic but may notice an elevation on the skin around the area near the eyes with little concern. It is more frequent in the elderly.

**O:** A generic term used for epithelial tumors. Size and shape of tumors are variable. Pigmentation varies from non-pigmented to darkly pigmented. Papillomas can be found anywhere on the skin, but it is most frequent on the mucocutaneous lid border; often benign overgrowth of squamous epithelium i.e. 'cutaneous horn' (with keratin build-up).

**A:** Once grown, there is little change; therefore these skin covered lumps are quite benign. Papillomas are classified into 3 histologic types: squamous cell, transitional cell, and mixed cell. Degeneration of squamous cell papilloma may lead to squamous cell carcinoma.

**P:** There is no treatment to make these tumors dissolve. All are resistant to radiation therapy. If these tumors are unstable/changing, the patient should be referred to a dermatologist for removal. Removal is recommended if one is unsure.
General Name: Melanocytic Mole
Specific Name: Nevi
ICD 9-CM#: 216.1, 216.3
Location: Adnexa
Associated Conditions: None

S: None except possibly cosmetic concern.

O: A pigmented spot or freckle located on the lids.

A: A nevi is a cluster of normal melanocytes whose pigment proliferation create a dark patch of skin. Differential diagnosis is to rule out malignant melanoma. Nevi are usually flat with regular borders. They are usually stable in size unlike the rapidly growing melanoma. No ulceration or bleeding should occur from a nevi.

P: Prognosis is excellent. Very few nevi go on to become malignant. However careful documentation of the size and shape should be recorded. If an increase in size is noted or other potential signs of malignancy, referral to a dermatologist for removal is advised.
General Name Verruca/warts
Specific Name Verruca vulgaris
ICD 9-CM# 078.1
Location Skin of the body (lids)
Associated Conditions

S: Bumps on lid. Symptoms of epithelial keratitis.

O: One or more cutaneous masses that have rough surfaces made up of many fine projections. Mild epithelial keratitis.

A: By S & O and clinical appearance. Verruca are contagious viral tumors. When located on the eyelid margin, the lesions scrape the cornea or debris contaminates the tear film and causes a chronic epithelial keratitis and conjunctivitis. Children may develop conjunctival papillomas.

P: Removal of the nodules from the eyelid margin is necessary to heal the keratitis. This can be done by topical application of various keratolytic agents or removal by excision. Dichloroacetic acid (Bichloracetic Acid), a chemical cauterizing agent, is easily employed by applying small amounts to each lesion with a wooden applicator. The lesion will immediately turn white, then gray-white after several days, and within 7 to 10 days the wart will desquamate. Usually only one application is necessary. This treatment modality is limited to lesions on the surface of the lid and must not be employed for verrucae of the lid margin because of the potential injury to the conjunctiva and cornea.
**General Name**: Hypercholesterolemia

**Specific Name**: Xanthelasma

**ICD 9-CM#**: 374.51

**Location**: Adnexa

**Associated Conditions**: Arcus senilis, Hyperlipoproteinemia

**S**: A yellowish slightly elevated area on the superior and inferior eyelid. It is often seen in middle aged women who is cosmetically concerned about this condition.

**O**: A small, bilateral sharply demarcated plaques which run approximately parallel to the eyelid margin.
Arcus Senilis is often seen with this condition.

**A**: Histology of Xanthelasma shows large histiocytic cells filled with foamy cytoplasm which represent engulfed lipids.
Xanthelasma are cutaneous deposits of cholesterol which occurs most commonly near the medial canthus of the upper and lower eyelid. They maybe idiopathic or associated with hypercholesterolemia. This condition is most often seen in middle age patients. However, those with familial tendency manifests the condition earlier on around ages 20 to 35.

**P**: Patients with xanthelasma, especially those who have this condition in their early years should have further evaluation for hypercholesterolaemia. Treatment consists of excision or cautery. Although xanthelasma can be surgically excised, they do recur if the underlying cholesterol problems are not solved.
General Name: Hemangiomas

Specific Name: Capillary Hemangiomas of the Eyelid

ICD 9-CM#: 228.00

Location: Eyelid

Associated Conditions:

S: Functional, emotional and cosmetic implications.

O: The tumor is reddish purple in color, elevated and has an irregular dimpled surface. The superficial hemangiomas are sometimes called "strawberry nevi" because of their appearance.

A: Capillary hemangiomas usually appear before the age of 6 months, have a tendency to grow during the first year of life and then tend to regress spontaneously. Hemangiomas involving the eyelids pose special problems. The mass of the tumor may cause total occlusion of the eye and produce irreparable amblyopia, anisometropic amblyopia due to induced astigmatism, and strabismus. In addition ulceration, necrosis and infection are possible complications.

P: In terms of minimizing scarring and side effects of therapy, intervention should be delayed as long as possible. But if the eye is completely occluded by the hemangioma, the doctor must intervene however young the infant. Systemic corticosteroids, up to 2mg/kg/day have been beneficial in some individuals and should be tried. Irradiation and occasionally surgery may be necessary if steroids fail.
General Name: Cysts
Specific Name: Epidermoid and Dermoid Cysts
ICD 9-CM#: 376.8, 374.8
Location: Adnexa

Associated Conditions:
Ptosis, Diplopia, Irregular astigmatism

S: Patients are usually asymptomatic. Frequently parents report a soft to solid round growth around the eye area.

O: The cyst is usually found in the superior temporal orbit area. They also can be found on the conjunctiva also. It is nontender and non inflammatory. Palpation reveals a soft to solid, smooth totally encapsulated cyst.

A: Need to separate dermoids and epidermoids from malignant growths which frequently change, are tender and are rarely self contained. Dermoids are composed of one or more normal skin structures such as hair follicles and sebaceous glands. The cyst is lined with keratinized epidermis. Epidermoids consist of epidermal tissues only. They are most commonly cystic, with cholesterol crystals and epithelial debris found in the cavity.

P: It is important to reassure the patient and to photodocument the lesion. Often surgical excision is indicated as the mechanical displacement due to the tumour can cause diplopia, ptosis and irregular astigmatism.
**General Name** Actinic Keratosis

**Specific Name** Actinic Keratosis

**ICD 9-CM#** 702

**Location** Adnexa

**Associated Conditions**
Dysplasias, Carcinoma-in-situ.

**S:** Patient is asymptomatic but may notice dark spots on eye or skin upon observation.

**O:** Actinic (or solar) keratoses represent dysplasias of the conjunctival epithelium and epidermis of the skin which are caused by ultraviolet irradiation. Elastotic degeneration of the underlying connective tissues is also evident.

**A:** Other dysplasias occur without apparent solar damage.

**P:** When the abnormal cells of the dysplastic process replace the full thickness of the epidermic or epithelium but there is no evidence of invasion of the subepidermal or subepithelial connective tissues, a carcinoma-in-situ stage has been achieved. Dysplasias and carcinoma-in-situs will, in some cases, advance toward invasive carcinoma (i.e. squamous cell carcinoma). All invasive carcinomas start off as carcinoma-in-situs. At this stage, the patient should be referred to the dermatologist who will remove the lesion or take a biopsy and then decide course of action.
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**S:** None except cosmetic concern.

**O:** External observation reveals a nodular tumor with a pearly surface and telangiectatic vessels. The nodule is typically elevated, has an irregular surface, and is sharply demarcated. The centers are usually excavated or ulcerated. The tumor is sometimes pigmented.

**A:** Differential diagnosis is from other lid tumors. Basal cell carcinoma is by far the most prevalent lid tumor. They typically appear on sun exposed parts of the lid, most typically on the lower lid on the medial side. Differential diagnosis is sometimes difficult between a pigmented basal cell tumor and a melanoma. A tentative diagnosis is made from the general observation as listed above in most cases.

**P:** If caught early, prognosis is good. Metastatic spread is rare. However neglect can lead to the spread and destruction of the orbit and at worse the cranial cavity. Radiation therapy is usually contraindicated due to the destruction of surrounding normal tissue and retina. Cryotherapy has been found useful in many tumors involving the inner canthus to decrease lacrimal drainage damage. The most common treatment is surgical excision since microscopic confirmation of complete excision can be made.
General Name: Squamous cell carcinoma of lids
Specific Name: Squamous cell carcinoma of lids
ICD 9-CM#: 232.0
Location: Lids
Associated Conditions:
Possibly actinic keratosis.

S: None.

O: Nodular appearing lesion with elevated, irregular surfaces and sharply demarcated, pearly margins.

A: By histological studies, Squamous cell carcinoma can generally be easily distinguished from basal cell carcinoma. The former cells have more conspicuous, eosinophilic cytoplasm, larger nuclei with more prominent nucleoli, and intercellular bridges. Spindle cell variants of squamous cell carcinoma, however can be quite difficult to diagnosis. Squamous cell carcinoma is a truly malignant, invasive tumor that takes origin from the epidermis and displays evidence of keratinization. It is a very rare tumor. It is more common in fair-complexioned individuals who are liable to develop actinic keratosis. The lower lid is more commonly involved than the upper, and the lid margin is a frequent site of origin. They may metastasize, although death from a squamous cell carcinoma of the lid is very rare. Actinic keratosis is a precursor lesion with respect to the development of invasive squamous cell carcinomas. It is believed that squamous cell carcinomas originating from actinic keratoses have a more benign course than those which arise from other forms of carcinoma in situ.

P: The management of all these tumors is adequate surgical resection.
General Name: Malignant Eyelid Neoplasm
Specific Name: Meibomian Gland Carcinoma
ICD 9-CM#: 173.1
Location: Adnexa
Associated Conditions: Chalazion, Meibomian gland cyst, Hordeolum

S: Patient reports a non-tender lump on the eyelid.

D: There are two types of Meibomian Gland Carcinoma:
   1) Localized: presents as a persistent and recurrent meibomian cyst.
   2) Generalized: presents as a severe unilateral and persistent chronic blepharitis.

A: Recurrence of what is believed to be a chalazion at the site where it has been excised should make one suspicious of Meibomian Gland Carcinoma. Differential Diagnosis should be made from other types of lid cysts such as chalazion and dermoid. Meibomian gland carcinoma can metastasize in the orbit and can lead to a loss of an eye.

P: Preferred treatment is surgical excision.
General Name Malignant Melanoma
Specific Name Malignant Melanoma of the Eyelid
ICD 9-CM# 172.1
Location Eyelid
Associated Conditions

S: Patient notices a growing bump which is sometimes tender.

O: The lesion is usually pigmented and may appear very small. It is normally seen during external examination of the eye.

A: Malignant melanomas of the eyelids are uncommon and usually arise from melanomas of the conjunctiva. Hutchinson's malignant freckle occurring in older people on the skin of the cheek may creep up to involve the lid. De novo melanoma of the lids and a malignant melanoma originating in a nevus of the lid are extremely rare lesions. Malignant melanomas of the lid can metastasize to the regional lymph nodes as well as hematogenously throughout the body.

P: Melanoma arising in the melanotic freckle of Hutchinson has the best prognosis and extensive lesions in older individuals can occasionally be managed with radiotherapy. All other melanomas should be removed surgically with lymph node dissection in high risk patients.
General Name: Neurofibroma
Specific Name: Neurofibroma
ICD 9-CM#: 215.0
Location: Adnexa

Associated Conditions:
Von Recklinghausen's Disease

S: Symptoms may include a growth visible in the upper lid with a swollen "bag of worms" appearance. Parents may notice one eye bulges forward.

O: Signs include:
- A diffuse neurofibroma that dissects through all orbital tissues in the upper lid. If the orbit is involved pulsatile exophthalmos may be visible.
- Proptosis is common
- Cafe au lait spots and cutaneous fibromas are also visible elsewhere on the body.

A: A neurofibroma is a peripheral nerve tumour and usually presents in the first decade of life in neurofibromatosis. There are often associated orbital deformities. Dysplasias along with growing tumours create exophthalmos and proptosis.

P: The growths are difficult to manage especially in the orbit. They are very intertwined with orbital structures and removal risks losing the entire globe. When the growth becomes unmanageable it is excised with as little damage to surrounding tissues as possible. Regrowth is common and surgery is frequently repeated. Most surgery is attempted before the child is of school age.
General Name  Stye
Specific Name  External Hordeolum
ICD 9-CM#  373.11
Location  Lid
Associated Conditions
Blepharitis, Keratitis

S: Patient complains of an acute inflamed sore lid.

O: Observation reveals an elevated red nodule at the base of the lashes. The nodule is generally surrounded by localized edema. Within a few days of onset a yellow point is observed on the nodule. Most will spontaneously drain within 3-4 days.

A: The assessment is made primarily by the location of the infection and the suddenness of onset.

P: Prognosis is excellent. A hordeolum is an infection of the glands of Zeis or Moll, most often by Staph. Management is by application of hot compresses several times daily. Topical antibiotics such as sulfonamide may be used to prevent surrounding gland infection but will not affect the course of the existing hordeolum. Incisions are often made in the hordeolum which do not resolve with hot compresses. Topical antibiotic ointment should be used following incision.
General Name  Chalazion
Specific Name  Lipogranuloma of the meibomian gland
ICD 9-CM# 373.2
Location  Meibomian gland
Associated Conditions

S: Painless bump in lid near the margins, possible blurred vision.

O: Palpation indicates a small swelling resembling buckshot in the substance of the eyelid (this may be the only evidence). It may become secondarily infected and cause an acute suppurative inflammation that usually points on the inside of the eyelid.

A: The lesion is a lipogranuloma resembling that seen in sarcoidosis or tuberculosis with giant cells but without caseation. Some individuals tend to have a series of chalazia, apparently because of inspissation of the meibomian gland contents in the excretory ducts.

P: Asymptomatic chalazia do not require treatment and usually disappear spontaneously within a few months. Acute suppuration is treated with local hot compresses and a topical antibiotic or sulfonamide. Excision, usually through a conjunctival incision, is indicated when persistent or large. Some individuals tend to have a series of chalazia, apparently because of inspissation of the meibomian gland contents in the excretory ducts. If pressure on the eyelid expresses a viscous secretion from the glands, massage of the eyelids, sometimes with a glass rod, may be helpful. Recurrence of what is believed to be a chalazion at the site where it has been excised should make one suspicious of a meibomian gland carcinoma.
General Name Lice-eyelid infestation
Specific Name Phthirus pubis, Pediculus humanus
ICD 9-CM# 132.2; 132.9
Location Adnexa
Associated Conditions Blepharitis

S: Symptoms include:
- foreign body sensation
- itch

O: Slit lamp evaluation reveals eggs or actual pubic louse clinging to eyelash, eyebrow or pubic hair. On low magnification, it is similar in appearance to blepharitis.

A: Nits on eyelashes causes chronic blepharitis. The infestation spreads from contact, e.g. bedding, sheets, and towels. Itch is due to the saliva of the louse on the lid.

P: The following treatment is effective against Phthirus pubis:
1) 1-2% Yellow oxide of mercury or 3% ammoniated mercury ophthalmic ointment. Ointment should be placed around the eyelash.
2) Vaseline—works by smothering the lice without irritating the eye.
3) Eserine ointment-paralyzes the lice.
General Name: Blepharitis

Specific Name: Staphylococcal Blepharitis

ICD 9-CM#: 373.01-.02

Location: Lids

Associated Conditions:
Madarosis, Trachiasis, Epithelial Keratitis

S: Foreign body sensation, mattering of the lids upon waking, itching, tearing, and burning.

O: The more common, squamous type of staphylococcal blepharitis is characterized by the presence of hard, brittle, fibrinous scales surrounding the lashes and on the lid margin. The scales are less greasy than those observed in seborrheic blepharitis. There is also a characteristic hyperemia of the lid margin. The less common, ulcerative type is characterized by matted, hard crusts surrounding the individual lashes. When these crusts are removed, small ulcers can often be observed and bleeding may occur. When the blepharitis is chronic, associated findings may include madarosis, trichiasis, or thickened lid margins. A chronic papillary conjunctivitis is almost invariably associated and an epithelial keratitis is often observed as a superficial punctate keratitis affecting predominantly the inferior quadrant of the cornea.

A: Bacterial infection of the lid margins is caused almost exclusively by Staph. aureus and Staph. epidermis. Mites may serve as vectors of the staphlococcal organisms. Cosmetics are subject to contamination during use and may therefore be an important source of bacterial infection. If the blepharitis is unilateral the lacrimal drainage system should be carefully examined as the etiologic factor.

P: Since staphylococcal blepharitis can become chronic and more difficult to treat, it must be treated aggressively in order to be successful. The lid margin should be mechanically cleaned using a cotton tipped applicator moistened with mild baby shampoo. Following this, antibiotic ointment may be applied directly to the lid margin. In moderate to advanced cases, gentamicin, erythromycin, or bactracin ophthalmic ointments are effective against most staphylococcal organisms and should be considered as initial therapy. The antibiotic can be applied at bedtime in mild cases or 3-4 times daily in severe cases. Yellow mercuric oxide ointment can be applied to the lids when there is only minimal involvement or when the condition has already been brought under control in order to prevent exacerbation of the condition.
**General Name** Entropion

**Specific Name** Entropion

**ICD 9-CM#** 374.00

**Location** Adnexa

**Associated Conditions**
Steven-Johnsons, Pemphigold(gus), Trauma, Trachoma

**S:** Patients report irritation and foreign body sensation.

**O:** Signs include:
- Conjunctival injection
- Malpositioned lid margin (turned inward)
- Epithelial defects and fluorescein staining due to lashes rubbing on the cornea.

**A:** Must differentiate which of the four types is present:
1. Congenital-rare condition and is commonly accompanied by tarsal hypoplasia or microphthalmia.
2. Involutional-Related to ageing changes due to degeneration of orbital tissue which causes the eye to drop back. Since the lid lacks support it rolls inward. Usually the lower lid is involved.
3. Spastic-Periodic condition caused by irritation or vigorous closure of the lid. Ultimately due to spasm of the orbicularis.
4. Cicatricial-Due to tars conjunctival shrinkage that occurs in scarring. Often seen in a variety of conditions: Steven-Johnsons, pemphigold(gus), trauma, acid burns, trachoma, and other mucous membrane scarring.

**P:** Surgery is the preferred treatment in all cases except spastic. In spastic entropion the best treatment is to identify and remove the irritant and applying a warm compress to decrease the frequency of orbicularis spasm.
General Name: Ectropion
Specific Name: Ectropion
ICD 9-CM#: 374.10-374.14
Location: Adnexa
Associated Conditions:
Lid thickening, Tarsal inflammation, and Edema.

S: An outward turning of the lid margin that in most cases occurs in the lower eyelid is noticed. Epiphora may be present if the puncta is also turned away from the globe. Irritation may result.

O: Essentially an evertion of the eyelid and may vary from a mild degree to total eversion of the lid. Inferior keratitis may result.

A: The ectropion is usually categorized into various clinical types, including lazity involutional (senile), tarsal, cicatricial, paralytic, and complex ectropions. The anatomic causes are loss of tarsoligamentous elasticity (involutional); weakening of the inferior retractors combined with loss of tarsoligamentous elasticity (tarsal); shortening of external skin or skin muscle (cicatricial); and loss of orbicularis tone combined with some tarsoligamentous elasticity (paralytic). There may be a combination of all of the above in some cases of ectropion. In involutional ectropion, the lid eversion first takes place medially which often results in epiphora in patients with adequate tears and even conjunctival hypertrophy-keratinization. In tarsal ectropion, the lid is completely everted with the lower border of the tarsus at the lower limbal margin. Cicatricial can result from burns, irradiation, inflammatory conditions, or skin removal following blepharoplasty, xanthelasma removal, or skin cancer excision. Paralytic is due to a seventh nerve or facial palsy, which is more common in older persons versus younger people. Complex results following severe eyelid lacerations, multiple incisions in the eyelid from repeated surgery, or other unusual situations. Congenital is a rare condition with multiple causes in which the child may have underlying developmental anomalies. It may be a temporary nature, or it may require surgical correction.

P: The "lazy T" incision seems to work most frequently for medial involutional ectropion when hot compresses & massage & wiping lids upwards to increase muscle tone do not work first. In cases of most generalized involutional ectropion a horizontal tightening procedure at the lateral canthal angle can be performed (this may even remediate medial ectropian--one should avoid excising healthy tarsal plate whenever possible). For tarsal ectropion, a repair of the inferior capsulopalpebral fascia or retractors should also be combined with a horizontal tightening procedure. Skin graft must be combined with the horizontal tightening procedure for cicatricial ectropion. For paralytic ectropion, reanimation of the eyelid itself from reinnervation techniques can be achieved with facial nerve transfer and graft if the facial muscles have not undergone atrophy. Complex ectropion can be corrected only with staged procedures of grafting of the external and internal lamilla and many times separation of fibrous adhesions from the orbital rim, if they exist.
General Name  Ptosis
Specific Name  Ptosis
ICD  9-CM#  374.30
Location  Lid
Associated Conditions
Myasthenia Gravis, Trauma, 3rd Nerve Palsey

S: Patient complains of a droopy lid.

O: The vertical fissure is abnormally small giving the droopy lid appearance. It is especially noticeable if unilateral.

A: Ptosis can be either congenital or acquired. Congenital ptosis is an autosomal dominant disorder. It can vary from complete closure to a slight droop. Acquired ptosis is usually caused by abnormality of the levator or its innervation. Common causes are myasthenia gravis, trauma, or mechanical effects. Levator function can be accessed by having the patient look up and mechanically limiting the action of the frontalis. Neurologic lesions such as those affecting the third nerve can also cause ptosis.

P: Prognosis is generally good. Congenital ptosis is usually surgically treated by resection of the levator. Aquired ptosis involves treatment of the cause. Surgical treatment is also often required.
General Name: Trichiasis or misdirected eyelashes

Specific Name: Trichiasis

ICD 9-CM#: 374.0

Location: Lids

Associated Conditions:
Diseases that cause scarring of the eyelid margin.

S: irritation of the cornea and conjunctiva, tearing.

O: Eyelashes turned toward the globe. Possible secondary infection.

A: By appearance.

P: Surgery to correct the position of the eyelashes, epilation, destruction of the lash follicles with liquid nitrogen, therapeutic contact lens.
General Name  Alopecia
Specific Name  Alopecia
ICD 9-CM#  none
Location  Eyelids
Associated Conditions  Ulcerative blepharitis

S: Patient reports loss of eyelashes.

O: Ocular examination reveals an absence of eyelashes. In case of ulcerative blepharitis, suppurative lesions surrounded by yellow pus and crusts which are very difficult to remove. Lash loss and the presence of necrotizing inflammation causes distortion of the eyelid.

A: Based on the S and O, the diagnosis is alopecia due to ulcerative blepharitis.

P: Treatment of blepharitis consists of lid hygiene. (Lid scrubs with a dilute solution of baby shampoo 2 times a day.)
General Name Distichiasis
Specific Name Distichiasis
ICD 9-CM# 743.63
Location Eyelid
Associated Conditions

S: The patient may be asymptomatic or present with foreign body sensation, lacrimation, burning and photophobia.

O: An accessory row of eyelashes lying posterior to the normal lashes and arising from the meibomian gland orifices. There is often wide variation in the number of eyelids involved, the number of abnormal lashes, the diameter, length or pigmentation of the lashes as well as the direction to the shafts.

A: Distichiasis is an abnormality in which extra lashes arise from the lid margin behind the mucocutaneous junction, frequently from the meibomian gland orifices. This disorder occurs sporadically or as an autosomal dominant trait.

P: Distichiasis requires no treatment if the patient is asymptomatic but when the lashes cause corneal irritation, they should be removed. If only a few lashes are involved, they can be removed by epilation or electrolysis. When the condition is extensive, however, surgical correction or cryotherapy is indicated.
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**S:** Patients complain that lid sometimes closes when they don't want it to which may last from seconds to minutes. They also experience flashes of vision loss and lid twitch.

**O:** Signs include:
- Spasm of the lid if occurring at the time of the exam
- Visual acuity normal-loss of vision due to closure of the lid.
- May have spasms of other muscles of the face.

**A:** Blepharospasm usually occurs in those over 60 years of age. It may follow postencephilitic states, hemiplegia and Bell's palsy.

**P:** The treatment varies with the severity of the condition. If other muscles of the face are involved the spasm is not relieved by sleep whereas if it is limited to the orbicularis the spasm is relieved by sleep. Therefore if the involvement is limited to the orbicularis rest and warm compresses will relieve the spasm. Antihistamines and quinine can help to slow the nerve refraction time. In more severe cases known therapy includes psychotherapy, levodopa, and dissection of the 7th nerve.
General Name Keratoacanthoma
Specific Name Keratoacanthoma
ICD 9-CM# 371.45
Location Eyelids
Associated Conditions

S: Growth/ bump on lid.

O: Very variable. Some have a cup-shaped configuration with a central keratin-filled crater. They may be verrucous, papillomatous, nodular or cystic.

A: The clinical appearance of these lesions is not diagnostic because of the variability. Because of the resemblance between these benign inflammatory tumors and malignant neoplasms, biopsies should be taken.

P: If the tumor is small, an excisional biopsy is diagnostic and curative. For larger ones, the biopsy allows correct diagnosis and a plan of treatment. Keratoacanthomas may occasionally be self-healing tumors, but they can attain a large size. Therefore, surgical excision when the proliferations are small is often a prudent decision. Furthermore, metastases have occurred from lesions that were erroneously diagnosed as keratoacanthomas because of their typical clinical history (abrupt onset and rapid growth), configuration, and tissue diagnosis based on small inadequate biopsies that did not disclose the true carcinomatous nature of the tumors.
**General Name** Internal Hordeolum  
**Specific Name** Meibomianitis  
**ICD 9-CM#** 373.12  
**Location** Lid  
**Associated Conditions**  
Blepharitis, chronic conjunctivitis  

**S:** Patient complains of irritation, burning, stinging, foreign body sensation, and redness.

**O:** Observation reveals plugs at the orifices of the meibomian glands, hyperemia of the lid margin, mild papillary hypertrophy, and thickened lid margins.

**A:** Digital pressure on the lid margin cause expression of a thick creamy white material from the gland openings. SPK is usually seen from the toxins being placed on the cornea. Rose Bengal staining also occurs on both the cornea and palpebral conjunctiva. The decreased lipid in the tears also creates a low tear break up time.

**P:** Prognosis is excellent. A primary meibomianitis is best treated by expression of the gland with cotton tipped applicators and hot compresses in the morning and at night. In cases resistant to therapy, antibiotic ointment (anti-Staph) can be applied to the lid margins BID. Artificial tears and lubricating ointment are indicated when corneal involvement is present.
General Name Eyelid Cellulitis
Specific Name Preseptal Cellulitis
ICD 9-CM# 373.13
Location Adnexa
Associated Conditions Eyelid blepharitis.

S: Depending on the severity of the cellulitis there is hardly any pain with very mild cases to very severe pain with greatly inflammed lid involving the deeper meibomian glands of the tarsal plate.

O: Swelling and inflammation of lids, vesicles, pustules and superficial ulcers along with edema and closure of eyelids. In severe cases, the upper lid cannot be opened to examine the globe. Edema of the lower lid is usually not as extensive as that present in the upper lid. Despite the severity of the adnexal signs, the globe usually remains normal unless there has been direct involvement with the injury. Full motility and the absence of pain on motion help distinguish the infection from true orbital cellulitis.

A: Careful examination of S & O and observing the lack of extraocular muscle and pupil involvement, along with the absence of proptosis and pain on ocular movement directs us to the diagnosis of preseptal or eyelid cellulitis. A primary inflammatory process occurring in front of the orbital septum which usually occurs by direct inoculation following trauma or by spread of infection from the skin of the face. The bacteria most commonly responsible is Staphylococcus aureus. Rule out Orbital cellulitis.

P: Hot compresses for the mild cases as often as 10-15 minutes per application, 3-4 times per day. Incision and drainage of the preseptal space, microbiologic investigation of the drainage material, and proper antibiotic therapy. Cellulitis of the entire eyelid requires vigorous antibiotic therapy. The selection of initial antibiotics is based on a gram-stained smear and the likelihood of responsible bacteria. Intravenous administration of antibiotics is preferred in the presence of extensive edema of the lids, orbital fracture, or possible extension of infection through the orbital septum. Oral therapy is sufficient in less severe infections. In patients with penicillin allergy, cephalothin is the preferred alternate drug. The drug of choice for anaerobic bacteria is penicillin G. Therapy should be continued in all infections for a minimum of 5-7 days or until there is resolution of suppuration. Staphylococcal cellulitis requires 10 days of treatment to prevent the late complication of glomerulonephritis. Oral antibiotics may be substituted after 48-72 hours of intravenous therapy if there is sufficient clinical improvement. Repeat drainage of purulent material may be required in staphylococcal infections. Haemophilus influenzae cellulitis occurs most commonly in children between the ages of 6 and 36 months. The drug of choice is ampicillin, although many infections resolve with penicillin G alone.
General Name: Orbital Cellulitis
Specific Name: Orbital Cellulitis
ICD 9-CM#: 376.01
Location: Orbit
Associated Conditions

S: Patient reports pain on movement of the eye. Also tenderness and dull pain with mild cases. Depending on the severity there is mild fever and malaise to high fever and marked debility. Diplopia is reported at times. Vision usually remains normal during the early stages. The injury may be considered trivial by the patient.

O: Edema of the eyelid with erythema and swelling. The conjunctiva is chemotic, hyperemic and with vascular engorgement. The eye is proptosed and there is external ophthalmoplegia.

A: Review of S & O leads to the diagnosis of orbital cellulitis. The pain on rotation, the unreactive pupil, the loss of some extraocular muscle function and the observed proptosis clearly points to the diagnosis of orbital cellulitis and not eyelid cellulitis. The bacteria most commonly responsible is probably Staphylococcus aureus. Paranasal sinusitis is the most common cause of bacterial orbital cellulitis. The most frequent cause in children under age 10 years is ethmoiditis. Bilateral involvement is uncommon. Soft tissue x-rays should be obtained to determine the presence of a foreign body. B scan ultrasonography may help. Blood cultures should be obtained. Postoperative orbital cellulitis is a rare complication of intraorbital surgery. The organism most likely responsible is Staphylococcus aureus. Phycomycosis is a potentially lethal infection of the orbit and paranasal sinuses caused by fungal genera of the class Phycomycetes. The disease occurs primarily in patients with ketoacidosis due to diabetes mellitus but may develop in mild or unrecognized diabetes. Additional initial symptoms including unilateral headache and ipsilateral facial weakness may occur. Amphotericin B administered intravenously (to control acidosis) and debridement of necrotic tissue. Efficacy of therapy may be assessed by serial paranasal sinus x-rays.

P: In mild cases warm/hot compresses several times daily and analgesics. There should be no attempt to aspirate fluid from the orbital spaces. Parenteral antibiotic therapy with high doses is immediate and mandatory in serious cases, preferably by pulse dose intravenous administration. Intravenous methicillin and gentamicin are preferred agents if organisms cannot be demonstrated or specific material cannot be obtained. Penicillin G should be added if there is clinical or microbiologic suggestion of anaerobic infection. If infection follows placement of implant material, removal may be necessary to control the infection. Abscess formation within the central or peripheral surgical spaces is suggested by failure of improvement following 2 or 3 days of appropriate antibacterial therapy, a decrease in visual acuity, increased proptosis, horizontal or vertical displacement of the globe, or isolated muscle weakness. If an abscess has formed, immediate exploration is emphasized. The route of exploration can usually be accomplished by a superior or an inferior incision through the skin or orbital septum. Rule out Preseptal cellulitis, Retinoblastoma, Metastatic orbital tumor, Orbital Pseudotumor, Rhabdomyosarcoma, and Cavernous sinus thrombosis, Graves' disease, Benign lymphoid hyperplasia, Malignant lymphoma.
General Name: Congenital and developmental eyelid anomalies
Specific Name: Epicanthus
ICD 9-CM#: 701.9
Location: Eyelid

Associated Conditions:
Wolff-Hirschorn syndrome, Cri du chat, Trisomy 13 Syndrome of Patau, Trisomy 18 Syndrome, and Trisomy 21 syndrome (Down Syndrome), pseudostrabismus, chromosomal abnormalities, oriental eyes

S: Patient may notice the appearance of the eyes turning in due to the extra fold of the skin on the medial eyelid. It is seen normally in many Oriental and infant eyes.

O: Epicanthus is an extra fold of semilunar fold of skin of the medial eyelid. It is a vertical skin fold which occurs in the medial canthus that conceals the medial angle and the caruncle. This simulates the appearance of esotropia.

A: Epicanthus is one of the most common congenital variations. In Oriental eyes, the epicanthus along with a fold of skin overhanging the palpebral fissure gives its almond shaped eyes. In many infants, the epicanthus is present until growth of the nose and face occurs. Epicanthus is associated with some chromosomal abnormalities e.g. Wolff-Hirschorn syndrome, Cri du chat, Trisomy 13 Syndrome of Patau, Trisomy 18 Syndrome, and Trisomy 21 syndrome (Down Syndrome). The concealment of the sclera by the extra fold of skin gives an appearance of esotropia (pseudostrabismus). The presence or absence of esotropia can be assessed by means of cover test.

P: Infants may outgrow the epicanthal fold. However, in cases where they do not, surgical intervention is indicated.
General Name: Coloboma
Specific Name: Lid Coloboma
ICD 9-CM#: 743.62
Location: Lid
Associated Conditions: Limbal Dermoids, Facial Dermoids

S: Obvious congenital defect of the eyelid.

O: Usually full-thickness defects in the medial portions of the upper lids.

A: Congenital eyelid coloboma is a rare clinical entity characterized by absence of a portion of the eyelid. In 90% of cases the upper eyelid is involved and the most common position of the coloboma is at the junction of the medial and middle third of the lid. A variety of other orbital or ocular abnormalities may be associated, including limbal dermoids and facial dermoids.

P: The primary problem caused by congenital eyelid coloboma is exposure keratopathy, which occurs when 30% or more of the upper lid is absent. Management is thus dictated by the severity of the defect. Surgical correction is generally indicated for cosmesis but this can be delayed until the child is 3-6 months old, at which time general anesthesia is a less serious risk. In the meantime, the cornea should be protected with topically applied lubricating ointments as well as antibiotic ointments if infection is a significant risk. If 4/5 of the upper lid is absent, the possibility of corneal exposure and scarring is substantial and surgery may be necessary within the first 48 hrs. of life.
General Name: Viral warts

Specific Name: Molluscum Contagiosum

ICD 9-CM#: 078.0

Location: Adnexa

Associated Conditions: Viral conjunctivitis and keratitis

S: Patient reports a growth on the eyelid which has gotten bigger gradually. Multiple growths may be present.

O: Pale nodule(s) on the lid margins usually up to 2mm in diameter. Centers of the nodules appear umbilicated. Cheesy material is expressed from the lesion. Upon lid eversion follicles may be present. The cornea may stain due to epithelial keratitis.

A: The lesion must be differentially diagnosed from other lid tumours such as verrucae, chalazia, sebaceous cysts, small fibromas and keratoacanthomas. The virus in addition is very hard to cultivate.

P: Preferred treatment is surgical removal especially when conjunctiva and corneal involvement are present. Other alternatives are electrocoagulation, or cryotherapy. The associated conjunctivitis and keratitis clear once the lesions are removed. If left untreated the keratitis can progress to subepithelial infiltration and peripheral corneal vascularization.
General Name: Contact Dermatitis

Specific Name: Contact Dermatitis

ICD 9-CM#: 692.3-692.9

Location: Adnexa

Associated Conditions:
Allergies.

S: Contact allergy of the eyelids occurs more often in women, probably due to the use of cosmetics. The dermatitis may be either unilateral or bilateral.

O: Usually due to the use of cosmetics or ophthalmic drugs. Other antigenic substances include clothing, jewelry, metals, and plastics, plus numerous other chemicals such as vegetable or animal products.

A: The diagnosis is based on a carefully taken history, a positive patch test, and a ruling out of other causes of eyelid eczema. In addition, one should be certain that the contact dermatitis is due to allergy rather than to primary irritation. The mechanism is a delayed hypersensitivity reaction subsequent to constant exposure to the antigenic stimulus. After sensitization, continued exposure to antigen results in a scaly eczematoid reaction. Rule out Chronic blepharitis, Preseptal cellulitis.

P: The treatment consists of eliminating the sensitizing agent.
General Name: Poliosis

Specific Name: Poliosis

ICD 9-CM#: 704.3

Location: Cilia

Associated Conditions:
Chronic Blepharitis, Vogt-Kayanagi-Harada Syndrome

S: None in itself except those form the underlying condition.

O: External observation reveals white eyelashes.

A: Assessment is made by examining the cilia during external eye examination and slit lamp.

P: Prognosis is dependent on the cause of the poliosis. Prognosis with treatment of a chronic blepharitis (see Blepharitis) is good that the lashes will regain color. Visual prognosis with VKH is poor (see Vogt-Kayanagi-Harada Syndrome).
General Name: Blepharochalasis
Specific Name: Blepharochalasis
ICD 9-CM#: 374.34
Location: Upper eyelids

Associated Conditions:
Diseases where there is chronic or recurrent swelling of the eyelids occur.

S: Possible constriction of the superior visual field.

O: Loose folds of wrinkled and venuled skin overhanging the upper eyelid margin. Possible constricted superior fields.

A: By appearance of the lids.

P: Treatment is not indicated unless there is a constriction of the visual field or for cosmetic purposes.
General Name: Eyelid degeneration
Specific Name: Dermatochalasis
ICD 9-CM#: 743.63
Location: Eyelid
Associated Conditions: Blepharochalasis

S: Patient may complain of "baggy" or "hanging" eyelids. The condition is seen in the elderly population. In severe cases, the patient often has a head tilt to compensate for the superior field loss.

O: Upon examination of the eyelids, there is an redundancy of the skin of the eyelids that is often accompanied by herniation of fat through the orbital septum.

A: Dermatochalasis in an involutional degeneration of the eyelid. As one ages, there is a loss of skin turgor and the fold of the skin hangs down over the eyelid margins. Although there is no sex predilection, a familial predisposition is often seen.

P: Treatment: Blepharoplasty.
General Name  Terrien’s Marginal Degeneration
Specific Name  Terrien’s Marginal Degeneration
ICD 9-CM#  371.48
Location  Cornea
Associated Conditions

S:  Relatively asymptomatic but symptoms of mild irritation may occur. The most bothersome problem can be progressive astigmatism.

O:  Usually the superior peripheral cornea is involved. The process begins with opacification and progresses with thinning and superficial vascularization. The leading edge of the ulcer is white due to formation of lipid deposits. There is a clear space between the ulcer and the limbus.

A:  Terrien’s degeneration is an uncommon, noninflammatory thinning of the marginal cornea. The epithelium is intact but the stroma and Bowman’s layer is gradually lost. Spontaneous perforation is rare, but trauma can rupture the thinned cornea. The condition usually occurs bilaterally and the condition predominantly affects males in the late teens or older.

P:  Treatment often is not necessary but surgical reinforcement with corneal grafting may occasionally be required if severe thinning occurs.
General Name: Corneal Degeneration

Specific Name: Mooren's Degeneration

ICD 9-CM#: 370.07

Location: Cornea

Associated Conditions: Corneal Perforation

S: Patients report a red eye, intense pain which is longstanding and gets progressively worse, photophobia, and tearing.

O: Acuities may be reduced if involvement is sufficient. Slit lamp reveals a red eye with a superficial ulcer on the cornea that begins peripherally. The typical appearance of the ulcer is that of a gray overhanging edge over the base. Advancement is centrally and peripherally till it completely encircles the limbus. It progresses to stromal and scleral involvement in severe cases.

A: There are two forms of the condition:

1. Benign-which affects older males and is frequently unilateral and mild.

2. Malignant- which affects younger patients and is bilateral and rapidly progressive to major scarring and corneal perforation. A biopsy reveals PMN's, lymphocytes, and plasma cells. IgG and IgM are found bound to the epithelium and epithelial antibodies are sometimes found in the serum.

P: There is limited treatment that's successful such as Vitamin A therapy and radiation treatment. Removal of the conjunctiva in the ulcerated area is relatively successful, in that it stops immune cells from getting into the area. Prognosis in the malignant form is not good and it can be a debilitating condition.
General Name  Salzmann Nodular Degeneration
Specific Name  Salzmann Nodular Degeneration
ICD 9-CM#  371.46
Location  Cornea
Associated Conditions  Phlyctenular disease, Trachoma.

S: Patient is asymptomatic.

O: A noninflammatory degeneration creating multiple, bluish-white superficial corneal nodularities usually in the mid-periphery of the cornea. There is no vascularization.

A: The nodules may be related to previous inflammation, especially phlyctenular disease, trachoma, or lues. The nodules represent hypertrophic areas of collagen superimposed on a normal-appearing posterior stroma; they generally replace Bowman's layer. Keratoplasty specimens have revealed occasional areas of abnormal epithelial basement membrane over the areas of abnormal collagen. Rule out Terrien's degeneration, Mooren's degeneration.

P: The treatment is lamellar or penetrating keratoplasty as indicated for visual reasons.
S: Pain, photophobia, and foreign body sensation from the associated corneal infection.

O: Slit lamp exam reveals a white cloudy haze which originates from the limbus and surrounds the site of the corneal infection.

A: The Wessley ring is a neutrophilic migration into the site of the antigen antibody complexes from the vascular arcades at the limbus. Common infections include bacterial ulcers.

P: Prognosis of remission of the ring is good if the infection is controlled. Treatment of the infection with the proper medication (anti bacterial, viral, or fungal) usually results in the fading of the ring over a few weeks.
General Name: Amyloid degeneration/corneal amyloidosis
Specific Name: B-Fibrillosis (amyloidosis)
ICD 9-CM#: 277.3 (gener)
Location: Cornea

Associated Conditions:
Long standing diseases such as retrolental fibroplasia, trachoma, glaucoma, uveitis, bullous keratopathy, keratoconus, and leprosy. Lattice degeneration.

S: Symptoms associated with corneal erosions (pain, tearing etc.), decreased vision.

O: Recurrent corneal erosions, decreased vision, changes in corneal transparency, irregular astigmatism because of subepithelial and stromal accumulations of amyloid material. These corneal amyloid lesions consist of salmon-pink to yellow-white, raised, fleshy masses which create a nodular surface. The cornea may be vascularized, depending on other factors.

A: The deposits seen in lattice dystrophy are also amyloid in nature, but that condition is a primary, localized amyloid disease. The amyloid material in the cornea is identical to that present in other organs. It stains with Congo red, displays birefringence and two-color dichroism with the polarizing microscope, and is fluorescent with thioflavin-T stain and ultraviolet light.

P: Systemic amyloidosis is almost invariably fatal. In reactive amyloidosis, recovery depends on control of the underlying disorder. For corneal amyloidosis, the treatment is keratoplasty.
General Name: Edema
Specific Name: Corneal Edema
ICD 9-CM#: 371.2
Location: Cornea
Associated Conditions:
- Iridescent vision
- Acute angle closure glaucoma
- Congenital glaucoma
- Bullous keratopathy
- Fuch's dystrophy
- Sattler's veil
- Trauma
- Keratoconus

S: Symptoms include:
- Decreased vision
- Iridescent vision (halos)

There are two main types of corneal edema: stromal and epithelial. In stromal edema there is minimal decrease in vision while in epithelial edema there is significant visual impairment and painful surface breakdown.

O: Clinical findings:
- In minor case, the cornea is dull, uneven, thickened, and hazy appearance. There is also some opacities.
- In severe case it can lead to bullous keratopathy and the vision is severely depressed.

A: Corneal edema results when the integrity of the endothelium or the epithelium is compromised. There are several causes of corneal edema:
1. Elevated IOP: e.g. Acute angle closure glaucoma, Congenital glaucoma.
2. Endothelial damage:
   a. Trauma (e.g. penetration of foreign body, surgical trauma from cataract surgery, birth trauma, nonsurgical contusion injury).
   b. Dystrophy (e.g. Fuch's dystrophy, congenital hereditary endothelial dystrophy posterior polymorphous dystrophy, keratoconus).
   c. Endothelial dysfunction secondary to inflammation (e.g. uveitis, focal keratitis, graft rejections).
3. Epithelial damage:
   a. Hypoxia due to contact lens overwear (Sattler's veil).
   b. Toxic effects of medications and anesthetics.

P: Treatment consists of first restoring the physiologic balance of corneal hydration according to the underlying cause of the edema:
1. Lower IOP in cases of glaucoma.
2. Control inflammation with topical steroids if corneal edema is secondary to inflammation e.g. uveitis, focal keratitis, or graft rejection.
3. Hypertonic agents e.g. Sodium chloride 5%
4. Keratoplasty for those cases of corneal edema unresponsive to other conservative treatment.
5. If visual rehabilitation is not essential, other methods of treatment include, soft contact lens therapy, conjunctival flap surgery, and electrocautery.
General Name: Bullous Keratopathy
Specific Name: Bullous Keratopathy
ICD 9-CM#: 371.23
Location: Cornea

Associated Conditions:
Glucoma, Cataract Extraction, Anterior Synechiae, Vitreous Touch, Fuch's Dystrophy, Epithelial Downgrowth, Perforating Wounds, Anterior-Posterior Radial Keratopathy, Birth Trauma, Immunologic Reaction After Keratoplasty

S: Decreased visual acuity. Severe pain, irritation and discomfort when the bullae bursts exposing corneal nerve endings.

O: Epithelial bullae or blebs. Filamentous tags are observed along with epithelial changes. The condition is persistent with the bullae enlarging, bursting and reappearing. The epithelial changes may remain localized to a section of the cornea or diffuse, depending on the extent of endothelial involvement. Ultimately, degenerative changes may appear, and blood vessels will invade the cornea from the limbus to form degenerative pannus.

A: This nonfilimentary disorder is not an independent disease but rather represents later stages of intense or chronic edema. Consequently, bullous keratopathy is seen only in severely diseased eyes.

P: Most therapeutic measures are directed towards alleviating the symptom of irritation. Therapeutic soft contact lenses can protect against recurrent epithelial breakdown and provide comfort in many patients, although this rarely improves visual acuity. Topical hyperosmotic agents are generally unsatisfactory in reducing the corneal edema. Other methods of alleviating discomfort include chemical cauterization of the cornea, placement of a conjunctival flap over the cornea and penetrating keratoplasty which has been used with increasing frequency in recent years with excellent anatomic and visual results. However, the patient must be referred for penetrating keratoplasty before neovascularization of the cornea occurs.
General Name: Band shaped Corneal Deposit

Specific Name: Band Keratopathy

ICD 9-CM#: 371.43

Location: Cornea

Associated Conditions:
Juvenile Rheumatoid Arthritis, Hyperparathyroidism, Vitamin D poisoning, Multiple Myeloma, Sarcoidosis, Hypercalcemia, Renal disorder, Prolonged glaucoma, Phthalic eyes

S: Patient reports decreased visual acuity and a cloudy cornea.

O: Visual acuity is reduced in the affected eye. Slit lamp exam reveals an interpalpebral deposition of calcium salts in the epithelium, basement membrane, Bowman's layer and in the superficial stroma. A clear area separates the band from the limbus.

A: Band keratopathy is associated with almost any severe ocular disease. In Still's disease band keratopathy may add to the chronic iridocyclitis which is present along with the arthritis. It is also seen in hyperparathyroidism, vitamin D poisoning, multiple myeloma, sarcoidosis, hypercalcemia, renal disorders, prolonged glaucoma, phthalic eyes, toxic vapors and irritant exposure.

P: Treatment is via an application of a calcium binding agent such a EDTA. The procedure involves first anesthetizing the cornea with 4% cocaine and scraping off the corneal epithelium. The EDTA is then dropped into a round well held against the cornea. After the calcium has been removed an antibiotic and short acting cycloplegic should be instilled and mild pressure patch applied for 24-48 hours.
General Name Neurotrophic Keratopathy
Specific Name Neurotrophic Keratopathy
ICD 9-CM# 370.00, 370.35
Location Cornea
Associated Conditions Herpes.

S: This entity is sometimes also called neuroparalytic keratitis. Patient has generalized ocular pain.

O: A corneal ulcer is visible. All patients with trigeminal anesthesia get excess mucous ("sleep") in the morning to some extent. The discharge often clings to their lashes. It does not imply infection or appear to be related to keratopathy. The entire palpebral conjunctiva may stain with rose bengal after gasserian ganglion destruction. This is an index of increased conjunctival epithelial cell death. It does not mean the eye is dry and does not appear to be related to keratopathy. About 50% of the patients with trigeminal anesthesia have tear film abnormalities and corneal pathology as seen with fluorescein staining as geographic drying areas. Transient blurring of vision, secondary to irregular corneal surfaces of punctate epitheliopathy is common in Stage I of the disease. Stage II develops as an acute episode and is characterized by epithelial detachment, which is marked by hyperemia, decreased vision, pain, folds in Descemet's membrane. Stage III is characterized by corneal stroma lysis, aqueous flare and cells.

A: It is related to a deficiency in innervation from the first division of the fifth cranial nerve, which, it is supposed, may normally provide trophic influences to the corneal surface cells to aid in surface maintenance and repair. Three most common causes are surgery of the trigeminal neuralgias, surgery of the acoustic neuromata and herpes zoster ophthalmicus (8% of patients with herpes zoster ophthalmicus develop neurotrophic keratopathy). Sensation in either the cornea or conjunctiva spares the patient from the disease. Conjunctival sensation is tested by applying a hypodermic needle to superior and inferior palpebral conjunctiva. Cornea sensitivity is tested with a wisp of cotton wool. The differentiation of viral epitheliopathy of zoster from neuroparalytic keratitis is made by demonstration of sensation in either conjunctival or corneal which implies a viral etiology. Neuroparalytic keratitis is probably a disease of abnormal corneal cell turnover. It has been suggested that the cyclic nucleotides (second messengers of hormone action) are involved in the regulation of epithelial cell turnover and that there is an imbalance in the anesthetic eye leading to a creased cellular turnover. These influences are poorly understood at present in humans, but animal studies have revealed that epithelial cell function may be altered over and above the traumatic changes secondary to anesthesia.

P: Many of the so-called trophic changes in neurotrophic keratopathy can be prevented with judicious tarsorrhaphy. Therapy includes patching mucomimetic drops, hydrophilic contact lenses, central and temporal tarsorrhaphy, atropine and antibiotics. Systemic cortico-steroids are contraindicated because they potentiate collagenase activity. Many of the pathogenetic aspects of neurotrophic keratopathy remain poorly understood, despite decades of research.
General Name  Punctate Epithelial Erosions
Specific Name  Punctate Epithelial Erosions
ICD 9-CM#  370.20
Location  Cornea

Associated Conditions
dry eye, chemical or drug toxicity, viral infection, and hypoxia from C.L. wear.

S: Patient complains of pain, photophobia, foreign body sensation, and tearing.

O: Slit lamp exam reveals fine depressions or pits extending through or partially through the corneal epithelium.

A: Differential diagnosis is made primarily from punctate epithelial keratitis (PEK). Punctate epithelial erosions are very difficult to observe in direct illumination with the slit lamp. However the pits pool with fluorescein and can be easily observed with a cobalt blue filter. In contrast PEK can be easily viewed in direct illumination without fluorescein. Besides the pit, PEK has a distinguishing grey areas surrounding the pit.

P: The prognosis is good for full resolvement of the erosions. Management involves treatment of the underlying cause of the erosions. Common causes of PEE include dry eye, chemical or drug toxicity, viral infections, and hypoxia from contact lens wear. If the concentration or number of erosions is severe, broad spectrum antibiotics may be used to prevent bacterial infection of the compromised epithelium. Artificial tears and ocular lubricants may also be given to aid in epithelial healings and patient comfort.
General Name: Epithelial microcysts
Specific Name: Epithelial microcysts
ICD 9-CM#: None
Location: Corneal epithelium

Associated Conditions:
Wide range of conditions will lead to epithelial microcysts.

S: Varies with the associated condition.

O: Cystic spaces in the corneal epithelium.

A: By appearance. The cysts may take up dye depending on the existence of an external opening. These occur in isolated groups or confluenlly. As localized phenomena they occur in areas of epithelial healing or sometimes as part of a superficial dystrophic process associated with recurrent erosions.

P: Microcysts are a nonspecific response of the epithelium and occur in a wide range of conditions. Treatment is aimed at the overlying condition or disease.
General Name: Keratitis
Specific Name: Punctate Epithelial Keratitis
ICD 9-CM#: 370.21
Location: Cornea
Associated Conditions:
Superficial Punctate Keratitis of Thygeson, Viral Keratitis, Herpes Simplex, Staphylococcal & Chlamydial infections, Reiter's, Vaccinia syndrome, and Acne rosacea.

S: Symptoms include:
- photophobia
- foreign body sensation
- tearing
- intermittent burning
- blurred vision

O: Upon slit lamp evaluation, punctate epithelial keratitis (PEK) presents as an area of white or gray opaque spots on the cornea. The size, shape, and location is variable. Rose bengal stains well as compared to irregular staining seen in sodium fluorescein dye. PEK is often seen with punctate epithelial erosions (PEE).

A: Differential diagnosis from other types of superficial dermatitis e.g. punctate epithelial erosions, punctate subepithelial infiltrates, and filamentary keratitis. The coarse, granular infiltrates of PEK are characteristic of Thygeson's Superficial Punctate Keratitis. PEK is typically seen with viral keratitis especially with epidemic keratoconjunctivitis (adenovirus type 8). It may also be seen with Herpes Simplex, Staphylococcal & Chlamydial infections, Reiter's, Vaccinia syndrome, and Acne rosacea.

P: Treatment depends on the cause:
1. Thygeson's SPK: weak topical steroids
2. Viral Keratitis: Self limiting (3 weeks to 3 months).
   a. Topical application of antibiotic drops may soothe the eye and prevent secondary bacterial infection.
   b. For EKC, educate patient to practice good hygiene due to the contagious nature.
3. Staph infection:
   a. Good hygiene, irrigation of the exotoxin, and warm compress.
   b. Broad spectrum antibiotic e.g. Tobramycin, Neomycin, or Gentamycin.
General Name: Infiltrates
Specific Name: Subepithelial Infiltrates
ICD 9-CM#: 371.0
Location: Cornea
Associated Conditions: Keratoconjunctivitis

S: Asymptomatic to marked reduction on visual acuity.

O: The subepithelial infiltrates are nonstaining, may be small punctate infiltrates or large, dense well-marked opacities and may be located anywhere on the cornea.

A: Subepithelial infiltrates are something seen as sequelae of punctate epithelial keratitis after adenoviral, herpes simplex, herpes zoster, vaccinia, chlamydial, Reiter's and rosacea involvement. These opacities probably represent a local immune response to viral proteins of bacterial exotoxins. Staphylococal infection must be considered if the pattern appears in a marginal infiltrate distribution. Inferior peripheral limbal infiltrates can accompany acute Haemophilus influenzae conjunctivitis.

P: Marginal infiltrates caused by staphylococcal exotoxin respond to low doses of topical corticosteroids. Much controversy has surrounded the role of corticosteroids in the management of adenoviral infection. Suppression of the immune response with steroids may only interfere with the clearing of the viral antigen and prolong the disease. Some patients receiving steroids show the recurrence of opacities as much as two years after the original epidemic keratoconjunctivitis infection when the drug is stopped completely. Therefore, corticosteroids should be given only if the opacities are central and bother the patient subjectively or visually. Corticosteroids are contraindicated in infections with herpes.
General Name  Corneal Ulcer
Specific Name  Bacterial Corneal Ulcer
ICD  9-CM# 370.00
Location  Cornea
Associated Conditions  Infections by Pseudomonas aeruginosa, Strep pneumoniae, Moraxella, Klebsiella, Staph aureus and epidermidis.

S: Patients present with symptoms of pain, tearing, photophobia, reduced vision, mucopurulent discharge and a very red eye especially around the limbus.

O: Slit lamp exam reveals:
- Conjunctival injection
- Circumlimbal or sectorial flush
- Gray-white necrotic stromal infiltrate
- Corneal edema
- Hypopeon

A: Corneal ulcers are caused by a variety of organisms. Positive diagnosis is achieved by doing a scraping, cultures, gram stain and sensitivities.
In the uncompromised host the most common causative organisms are:
- Psuedomonas aeruginosa, Strep pneumoniae, Moraxella, Beta Strep, and Klebsiella pneumoniae.
In a compromised host the most common causative organisms are:
- Staph aureus, Staph epidermidis, Alpha strep, Beta strep, Psuedomonas aeruginosa, Proteus, and other enterics.
Some of the more common ddx are:
- Herpes simplex indolent ulcer which has a decreased corneal sensitivity and PMNs and multinucleated cells in the scrapings.
- Trophic keratopathy which is due to nutritional factors and is seen frequently with KCS there is no cellular infiltrates and the endstage is stromal lysis.

P: Initial treatment is dependent on the gram stain. A broad spectrum antibiotic such as gentamycin should be used until definitive identification is obtained. Once specific ID is made specific therapy can be initiated.
- Staph aureus: Gray-white round or oval central ulcer with indistinct borders. Tx. with gentamycin and warm compresses and lid scrubs.
- Strep pneumo: Attacks disrupted epithelium and produces a gray, well circumscribed ulcer that has a serpiginous form. The leading edge develops an undermined appearance. Tx. with ophthalmic bacitracin, lid hygiene and warm compresses.
- Pseudomonas: Most common gram neg. organism causing corneal ulcers. Frequently found as a contaminant of ophthalmic solutions. Capable of penetrating cornea in 24 hrs. due to a protease. Tx. must be immediate to prevent perforation. Use gentamycin.
General Name: Corneal Dellen

Specific Name: Corneal Dellen

ICD 9-CM#: 371.41

Location: Cornea

Associated Conditions:
Pterygium, Episcleritis, Limbal tumor, Allergic conjunctival edema.

S: Patient notices a redness of the eye right where the cornea meets the sclera. There is little symptoms other than the observation by the patient.

O: Observation with the biomicroscope reveals a shallow saucer-like depression near the limbus on the cornea, usually on the temporal side, with the base of the lesion hazy and dry. Instillation of fluorescein reveals epithelial breakdown within the confines of the lesion.

A: Attention to S & O yields the diagnosis of corneal dellen. It may be associated with pinguecula or pterygium. It is a definite factor in initiating a recurrent pterygium. The process in the formation of dellen starts with elevated conjunctival tissue and resulting tear film defects. This disturbs the adjacent corneal tissue and dessication of the epithelial tissue results. Other predisposing conditions that can result in dellen formation is any condition that results in swelling of perilimbal tissues to set the stage. Episcleritis, limbal tumor, allergic conjunctival edema, limbal vasosclerosis which spontaneously occurs in the aged patient are all causes for dellen. As optometrists it must be understood that contact lenses can initiate this response. Rule out Terrien's degeneration Mooren's degeneration, Corneal ulcers.

P: Treatment is directed at the cause. However, artificial tears go a long way to prevent the dessication of corneal epithelial tissue while other treatment of the inciting factor e.g. episcleritis.
**General Name** Corneal Ulcer  
**Specific Name** Dendritic Ulcer  
**ICD 9-CM#** 054.42  
**Location** Cornea  
**Associated Conditions** Herpes Simplex

**S:** In early stages the patient may only complain of a foreign body sensation. As the infection progresses lacrimation, photophobia, and pain may be the main symptoms. A repeating ulcer may present little subjective symptoms.

**O:** The lesion usually begins as minute opacities in the corneal epithelium. As the disease progresses, the tiny vesicles rupture and appear as superficial dots. These dot erosions can coalesce forming the classic dendritic or tree branch shape. The borders of the ulcer are serrated and sharp. The ends of the lesion also usually end in the shape of a bulb.

**A:** The dendritic ulcer will stain with both flourosclen and Rose Bengal. The ulcer in herpes is also usually associated with facial and lid vesicular lesions. Corneal sensitivity is often reduced in the herpetic ulcer. Wisp test with a Q-tip can be used as a gross test of corneal sensitivity. Further questioning about sexual history and possible V.D. involvement should also be investigated for further differential diagnosis.

**P:** Prognosis is good. However the lesions are usually recurrent and can lead to a more severe stromal keratitis. Classical therapy involved debridement of the epithelium followed by pressure patching. More current therapy involves the use of antiviral drugs. Typical treatment is an IDU solution every hour while awake and IDU ointment every 4 hours after bedtime. If no improvement is noted in two days, treatment with Vira-A or trifluridine drops five times a daily and ointment every 2-3 hours after bedtime is indicated. Anti viral medication should be tapered 5-7 days post epithelial healing.
General Name: Stromal herpes
Specific Name: Disciform keratitis
ICD 9-CM#: 054.43
Location: Corneal stroma
Associated Conditions: Herpesvirus hominis type 1

S: Foreign body sensation, lacrimation, decreases vision.

O: Decreased corneal sensitivity, appearance of a disk-shaped gray area that may involve the full thickness of the cornea or merely the stroma adjacent to the epithelium.

A: Tissue culture. A cotton swab can be dipped into a test tube containing a prepoured 5 ml aliquot of Hank's modified balanced salt medium (Flow Laboratories). The wet swab is then rubbed on the ulcerated epithelium and subsequently broken into the same medium. The tube is then capped and sent to a virology lab. Disciform keratitis forms beneath an epithelial ulcer (dendritic keratitis) 5 to 10 days after the attack begins. The dendritic ulcer is an acute and chronic corneal inflammation that occurs in an individual who has had a primary infection with Herpesvirus hominis type 1. After repeated viral attacks, the disciform keratitis may appear as a chronic inflammation of the corneal stroma characterized by discoid edema and opacity. The stroma often becomes necrotic, and an accompanying iridocyclitis may be severe. Hypopyon is rare. However, if it is present, it usually indicates a secondary bacterial or fungal infection.

P: In immunocompetent individuals, the disciform keratitis is usually self-limited lasting from several weeks to several months. However, when immunosuppression, local or systemic, is severe, the lesion may persist for years. For viral drug treatment, see under Dendritic Ulcer. If a superimposed bacterial blepharoconjunctivitis occurs, an antibiotic should be prescribed. Cycloplegics are sometimes beneficial in those cases where associated anterior chamber inflammation has occurred (see anterior uveitis). Recurrent erosions may take place at some time following the clinical healing of the epithelial ulcerative keratitis. Management should include removal of the hypertrophic and loose epithelium. The eye should be patched with a bland lubricating ointment until healing occurs. The frequent use of an ocular lubricant throughout the day should be initiated; thereafter bedtime instillation may be sufficient. Avoid steroid use.
General Name: Corneal Trophic Ulcer, Epithelial trophic ulceration
Specific Name: Post infectious herpetic erosions
ICD 9-CM#: 370.00
Location: Cornea
Associated Conditions:
Herpes simplex, dendritic ulcer

S: Early in the herpes simplex infection, the patient complains of pain, lacrimation, and photophobia. As the disease progresses, a loss of corneal sensitivity and a loss of photophobia occurs.

O: A slit lamp evaluation reveals a nondescript, linear, or ovoid epithelial lesion with heaped up borders on the cornea.

A: Differential diagnosis from dendritic ulcer which is caused by a live virus as opposed to epithelial trophic ulcer which are not due to active virus replication. Trophic ulcers are due to severe damage to the epithelial basement membrane sustained during the acute infectious epithelial stage. Damaged membrane heals slowly over 8-12 weeks but the damage is so severe that epithelial cells are unable to regenerate normally and fail to cover the defective area, causing an ulcer.

P: The trophic ulcer commonly occurs until the damaged basement membrane can be protected by the use of continuously worn soft contact lens, patching, lid tapering, intermarginal lid adhesions, or surgically placed conjunctival flap. Artificial tears are also indicated. Steroids are contraindicated.
General Name: Keratitis
Specific Name: Stromal Keratitis
ICD 9-CM#: 054.40
Location: Cornea
Associated Conditions: Herpes Simplex

S: Apart from a complaint of blurred vision the symptoms are nonspecific. The eye feels uncomfortable and waters. The pain experienced varies considerably from patient to patient.

O: The stroma is the site of inflammatory reaction that is irregularly distributed, of varying intensity and accompanied by an anterior uveitis. The corneal lamellas are necrotic in places and the stroma is diffusely and locally infiltrated by inflammatory cells. In severe cases a broad, creamy, homogenous lesion develops and may occupy the entire corneal thickness. Occasionally the Wessely ring is seen. Vessels can penetrate the stroma from the limbus at all levels to invade the active lesion. The epithelium overlying the active lesion is edematous to varying degrees ranging from a fine superficial edema to grossly edematous with recurrent bullae. Punctate erosions that stain with florescin and rose bengal are frequently seen. The endothelium may also be edematous or infiltrated by inflammatory cells and especially beneath the stromal lesion, may be replaced by a coagulated film of fibrin and inflammatory cells. Keratic precipitates are prominent. The aqueous contains cells and flare. Involvement of the iris is variable, but it is frequently infiltrated by lymphocytes and plasma cells and thickened by edema. The anterior ciliary body shows similar involvement. Posterior synechiae and anterior lens changes are common, frequently in association with a fibrovascular membrane extending across the pupil. When ulceration occurs, Bowman's membrane and superficial lamellas are replaced by debris and inflammatory cells.

A: Patients exhibiting stromal keratouveitis have commonly experienced a number of previous attacks of epithelial herpes, although occasionally it can occur in the initial attack. The onset of stromal disease is of serious portent because it marks a new stage in the disease; deeper ocular structures are involved, vision is seriously threatened and morbidity is significantly increased. A history of herpetic epithelial keratitis is not absolute evidence the subsequent stromal keratitis is herpetic in origin. The differential diagnosis should include zoster, fungal, chemical, and staphlococcal keratitis.

P: The aim is to guide the patient through each episode as expeditiously as possible while seeking to minimize ocular damage, reduce morbidity, and avoid the side effects of treatment. Effective control of the inflammation sometimes demands steroid therapy, and the effective and safe administration of steroids demands close observation in reliable patients. The dosage of steroid requires some judgement. The principle must be to produce a clearly recognizable progressive reduction in inflammation while minimizing the undesirable side-effect of enhancement of viral replication. The preference is to use the lowest effective dose while keeping the patient under close observation and adjusting the steroid dosage accordingly. An antiviral agent must be administered concurrently to reduce the chance of recurrence of live virus in the epithelium. Antibiotic therapy as a prophylactic routine is discouraged.
General Name: Salmon Patch Keratitis
Specific Name: Interstitial Keratitis
ICD 9-CM#: 370.50
Location: Cornea

Associated Conditions:
Syphilis, Tuberculosis, Cogan's syndrome, Herpes simplex, Chemical burns

S: Ocular symptoms include: pain, tearing, red eye and reduced vision.

O: Ocular signs in the acute stages of the nonsuppurative infiltration of the stroma consist of:
-Cellular infiltration
-Endothelial edema
-Keratic precipitates
-Small stromal opacities
-Miosis
In later stages, gross vascularization of the cornea occurs which appears as a "salmon patch". Residual effects of inflammation include diffuse stromal scarring, opacification, residual vascularization and ghost vessels.

A: Interstitial keratitis is a vascularization and is nonsuppurative infiltration of the stroma that is associated with systemic diseases such as syphilis and tuberculosis. In congenital syphilis interstitial keratitis is usually seen bilaterally with the most cases occurring from 5 years of age to the late teens. Interstitial keratitis that occurs secondary to acquired syphilis is usually unilocular, milder and more amenable to treatment. Positive diagnosis is made by a positive FTA-ABS test. Interstitial keratitis secondary to tuberculosis is usually unilateral and only the peripheral inferior sector of the cornea is affected. It is accompanied by a ring shaped nodular opacity. It is differentiated from syphilis by a negative FTA-ABS test and a positive tuberculin skin test. Other causes include: leprosy, onchocerciasis, mumps, Cogan's syndrome, gold toxicity and herpes simplex.

P: Usually cases of interstitial keratitis require keratoplasty when visual acuity degenerates to an intolerable level. Acute stages are controlled with local steroids which sometimes are used for 1-2 years. Cycloplegics prevent posterior synechiae and improve comfort. Treatment for the underlying systemic disease must also be instituted. Penicillin is the drug of choice in syphilis as is rifampin and isoniazid in TB.
**General Name** Corneal Deposits

**Specific Name** Corneal Deposits

**ICD 9-CM#** 371.10

**Location** Cornea

**Associated Conditions**

See below for specific etiology.

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**S:** Corneal deposits in themselves usually cause very little subjective complaints. However if the deposits become concentrated or involve the optic axis, patients will complain of blurry vision.

**O:**
1) Hudson Stahli line: A horizontal brown line at the level of the epithelium which is commonly observed in the elderly. It is considered a normal aging change.
2) Stocker line: A brown vertical arc immediately anterior to a filtering bleb or at the leading edge of a pterygium.
3) Fleischer ring: A brown ring at the base of the corneal peak in keratoconus.
4) Keyser-Fleischer ring: A brownish-green ring in the peripheral cornea at the level of Descemet's membrane associated with Wilson's Disease.
5) Kruckenberg's Spindle: A vertical pigment band on the endothelial surface usually associated with pigment dispersion syndrome. The band can usually be seen in line with the pupil.

**A:**
6) Keratic Precipitates: Inflammatory cells adherant to the endothelium which are common in any inflammation of the anterior segment such as iritis. Large greasy KP bodies are known as Muttonfat KP's and are seen in chronic anterior segment inflammations.
7) Heavy Metals: Heavy metals such as silver (algerosis), iron (siderosis), gold (chrysalisis), copper (chalcosis), and mercury may be deposited in the stroma adjacent to Descemet's membrane. They are introduced by local medication (silver), foreign bodies (copper & iron), systemic therapy (gold), or toxic vapors (mercury).

**P:** Prognosis is usually good and concentration should be put on the underlying etiology.
General Name: Anterior megalophthalmos  
Specific Name: Anterior megalophthalmos  
ICD 9-CM#: 743.8  
Location: Cornea, lens zonules and ciliary ring.  
Associated Conditions: Marfan's syndrome  

S: Decreased visual acuity.  

O: Cornea is larger than 12 mm in horizontal diameter, enlargement of the lens, zonules, and ciliary ring, high astigmatic refractive error. The dilator muscle is attenuated leading to transillumination defects in the iris. Iridodonesis may also be found. Because of the abnormal spatial relationships of structures in the anterior segment, secondary glaucoma may develop as a result of lens dislocation. Premature cataract is more common than usual in anterior megalophthalmos.  

A: By clinical appearance. Rule out congenital glaucoma.  
In simple megalocornea as well as anterior megalophthalmos, there is no evidence of previous or concurrent ocular hypertension. The corneal thickness and anatomy are essentially within normal limits. In simple megalocornea, all modes of inheritance have been described, although X-linked recessive inheritance is most common. Female carriers may have slightly enlarged corneas. The differential diagnosis from congenital glaucoma rests in the findings that the cornea is clear and the intraocular pressure and optic nerve are normal.  

P: Treat the symptoms. Monitor IOP.
General Name Keratoconus
Specific Name Posterior Keratoconus
ICD 9-CM# 371.60
Location Cornea
Associated Conditions
Anterior Lenticonus, Aniridia, Fleischer's Ring, Cleavage Anomalies

S: The irregularity of the posterior cornea affects vision to some extent.

O: Discrete indentation of posterior cornea with a variable degree of overlying stromal haze. It tends to be sporadic, unilateral, and relatively central. In some cases, pigment surrounds the edges of the posterior depression.

A: Posterior keratoconus has no known relationship to anterior keratoconus. The anterior corneal surface is normal unless there is sufficient thinning to cause ectasia.

P: No available treatment.
General Name: Arcus Juvenilis
Specific Name: Anterior Embryotoxin
ICD 9-CM#: 743.43
Location: Cornea

Associated Conditions:
High cholesterol levels, Aniridia, Blue sclera, Megalocornea

S: None usually.

O: Slit lamp exam reveals a yellow white opacity in Descemets and Bowman's layer. It may either a partial or complete perilimbal opacity. There is usually a clear zone between the opacity and the limbus. Patients may also exhibit: aniridia, blue sclera and megalocornea.

A: When corneal arcus is seen in younger individuals it is known as anterior embryotoxin or arcus juvenilis. It is frequently associated with increase in blood lipid level, but there is no true correlation to vascular disease. A higher frequency of arcus has been found in blacks without raised cholesterol levels.

P: An arcus in a young individual requires further workup and tests to determine if a lipid metabolism disorder exists. Patient should be referred to an internist for further examination.
General Name: Corneal Keloid
Specific Name: Corneal Keloid
ICD 9-CM#: 371.10 (unspecified corneal deposit)
Location: Cornea
Associated Conditions: Ocular trauma.

S: Patient is asymptomatic, but may have had a history of ocular trauma.

O: A hypertrophic scan is seen on the cornea.

A: A hypertrophic scan following trauma. This condition may be found at birth (intrauterine trauma) or at any age following trauma. Rule out Lipid degeneration.

P: No treatment is necessary unless keratoplasty is indicated for visual reasons.
General Name: Peter's Anomaly
Specific Name: Peter's Anomaly
ICD 9-CM#: 365.41
Location: Cornea
Associated Conditions: Glaucoma

S: Usually none since it is a congenital anomaly.

O: Observation reveals a central corneal opacity which is due to lack of corneal endothelium. Iridocorneal adhesions at the edge of the central opacity are also seen. A prominent Schwalbe’s line, peripheral iris anomalies, and distortions in the pupil are also seen.

A: Peter's anomaly is a congenital malformation of the components of the anterior chamber. Assessment is made primarily from biomicroscope and gonio-lens evaluation of the infant.

P: Prognosis is variable depending if glaucoma is present. If the defects are detected early and the baby is put on appropriate glaucoma treatment, prognosis is dependent on the success of the penetrating keratoplasty.
General Name: Axenfeld's anomaly/syndrome
Specific Name: Iridocorneal dysgenesis
ICD 9-CM#: 743.44
Location: Anterior chamber
Associated Conditions:
Reiger's anomaly, Peter's anomaly.

S: None unless glaucoma sets in.

O: A prominent, widened Schwalbe's line. It is anteriorly displaced and is called a posterior embryotoxin. There are abnormal iris strands coursing across the anterior chamber angle which attach to the prominent Schwalbe's line (Axenfeld's anomaly). This blocks the trabecular meshwork and can lead to glaucoma (Axenfeld's syndrome). Combined with hypoplasia of the anterior iris stroma, this constellation of abnormalities becomes Rieger's anomaly, often associated with glaucoma.

A: By slit-lamp examination and by gonioscopy. The dysgenesis is thought to have dominant inheritance.

P: Approximately 50% of these patients will develop glaucoma therefore they must be carefully monitored. In infancy, trabeculotomy has been curative. In adults, medical therapy should be tried and filtering surgery used if needed.
General Name: Cornea Plana
Specific Name: Sclerocornea
ICD 9-CM#: 379.05
Location: Cornea & Sclera
Associated Conditions:
Posterior embryotoxin, iris coloboma and stromal abnormalities, abnormally shaped pupils, congenital cataract, retinal coloboma, glaucoma, bone and tooth abnormalities.

S: Symptoms include:
- Reduced visual acuity due to stromal opacification
- Abnormal refractive error
- Glaucoma
Bilateral condition.

O: Clinical findings:
- The corneal stroma is diffusely opacified, frequently more marked posteriorly and centrally.
- There is scleralization of the limbus and absence of defined corneoscleral interface so that the diameter of the cornea appears subnormal.
- The corneal curvature is flatter than normal which results in shallow anterior chamber.
- The lid may appear ptotic (Streiff sign.)
- The iris pattern may be absent or abnormal.

A: The etiology of sclerocornea is due to a failure of normal cleavage of the anterior chamber structures during embryonic life. Transmission is usually autosomal dominant and less commonly autosomal recessive.
Sclerocornea is one of the conditions associated with mesenchymal dysgenesis. Other associated condition of mesenchymal dysgenesis include posterior embryotoxin, Axenfeld's anomaly, Reiger's syndrome, and Peter's anomaly.

P: Treatment:
1. Primary prevention through genetic counseling.
2. Surgery for cataract, glaucoma, and corneal opacification as indicated.
Prognosis: Poor for vision.
General Name: Corneal Degeneration
Specific Name: Spherical (Keratinoid) Degeneration
ICD 9-CM#: 371.40
Location: Cornea
Associated Conditions: Lattice Dystrophy

S: Blurred vision and painful erosion.

O: Fine subepithelial yellow droplets starting in the peripheral cornea and later advancing to the central cornea causing blurred vision.

A: Spherical degeneration is an acquired keratopathy affecting male adults who work outdoors. Corneal degeneration is caused by ultra violet light exposure. It may affect the peripheral and central cornea. This condition is probably related to elastotic degeneration of collagen.

P: Treatment by penetrating keratoplasty.
General Name: Coats White Ring
Specific Name: Coats White Ring
ICD 9-CM#: none
Location: Cornea
Associated Conditions: Foreign Body

S: Asymptomatic patient reports a history of a foreign body. The patient may complain of blur if the defect is close to the visual axis.

O: Signs include:
- Small granular oval ring that usually contains iron subepithelially.
- Visual acuity may be decreased

A: A scar remains after the foreign body is removed only if Bowman's membrane was injured.

P: No treatment is needed, but you should draw or photodocument the ring
General Name: Lipid Degeneration

Specific Name: Lipid Degeneration

ICD 9-CM#: 371.49

Location: Cornea

Associated Conditions: Arcus senilis, Juvenile arcus, Varicella.

S: Patient is ocularly asymptomatic. Clinical findings include blurred vision and haziness and thickening of the cornea, particularly in the central zone. Symptoms and signs are slowly progressive until useful vision is lost.

O: This degeneration is characterized clinically by the accumulation of a yellow or cream-colored material in the corneal stroma, which may be abnormally thick or thin. There is extravasation of cholesterol and fatty acids from the vessels and non-commensurate resorption of these elements by the same abnormal vessels.

A: There is generally a history of past corneal inflammatory episodes with resultant stromal vascularization, as may follow varicella. The lipid accumulations are, therefore, of a secondary nature.

P: Lipid keratopathy has been reported following hydrops and as a finding with no clear antecedent corneal damage (corneal transplant improves vision significantly in most cases).
General Name  Dry Eye  
Specific Name  Keratoconjunctivitis Sicca  
ICD  9-CM#  370.33  
Location  Cornea, Conjunctiva  

Associated Conditions  
Lupus Erythematosis, Pemphigoid, Sjorgen's Syndrome, Erythema Multiforme, Scleroderma, Polyarteritis Nordosa, Sarcoidosis, Rheumatoid Arthritis

S: The patient complains of gritty, scratchy, burning, dry eyes.

O: The eye may be slightly hyperemic. Rose Bengal will stain both the cornea and conjunctiva. A tear break-up time (TBUT) of less then 10 seconds is seen. A Schirmer #1 (no anesthetic) and Schirmer #1 (with anesthetic) are both less then 10mm in 5 minutes.

A: Differential diagnosis for the cause of the dry eye can often be determined from case history. A dry eye of recent onset can often be linked to medication. Antihistamines, antianxiety drugs, phenylthiazines, anticholinergics, and birth control pills have been known to cause dry eye. Aging and the decreasing basal tear secretion can also cause dry eye. Systemic diseases as listed in associated conditions also affect the tear supply. Any exposure problem such as lagophthalmos, ptosis, scarring, and nerve palsies can cause dry eye.

P: Prognosis is good in mild to moderate dry eye. Artificial tears applied as needed during the day and a lubricating ointment at bedtime work well. More severe dry eye problems may be required to use lacriscerts. An alternative therapy is a therapeutic soft lens with ointments and artificial tears. However vision is usually compromised with ointments. Severe dry eye along with ointment at night may benefit from the use of wet cells while sleeping. If the dry eye is going to be long standing, appropriate education should be given.
General Name: Nutritional blindness
Specific Name: Xerophthalmia with keratomalacia
ICD 9-CM#: 264.4
Location: This file will only cover the cornea.
Associated Conditions

S: Corneal xerosis: mild photophobia, night blindness
Corneal destruction: pain, hyperemia, photophobia.

O: In order of appearance
Corneal xerosis: SPKs limited to the inferonasal quadrant. In time, they become more numerous, denser and cover a larger area. Stromal edema, mild superficial haziness (usually inferiorly), keratinization or the epithelium, dry pebbly appearance to the cornea. Corneal destruction: sharply punched-out partial or full-thickness defect, usually located in the periphery of the nasal quadrant. These are usually plugged by iris with the anterior chamber remaining formed. Larger, irregular shaped ulcers that extent toward the pupillary axis, possible secondary bacterial infection. Localized stromal destruction that may resemble keratomalacia; these are sharply demarcated lesions that have a swollen opaque grayish-yellow appearance. Stromal sloughing leaves a descemetocele.

A: Diagnosis requires a high degree of suspicion, especially in persons who are malnourished, who have severe systemic disease (eg. cirrhosis, tuberculosis, gastroenteritis) or xerotic changes in the conjunctiva, or who have atypical corneal lesions refractory to antibiotic therapy. Xerophthalmia is a clinical diagnosis. Lab tests may be helpful in unusual cases, but their value is limited. Most tests require specialized labs and have specific limitations. The tests available are: Serum Vitamin A Levels, Total RBP (retinol binding protein), and HOLO RBP (this is the most reliable but most complex test). Keratomalacia, which is less well understood and defined is commonly described as "liquefactive" or "colliquative" corneal necrosis. Often the last, most severe stage of xerophthalmia, it also occurs in other conditions like severe measles, in which general malnutrition is usually present but specific signs of vitamin A deficiency may be absent.

P: Oral admin. of 200,000 IU vit.A in oil repeated the next day. With severe corneal disease and potential malabsorption change the initial dose to 100,000 IU water miscible vitamin A intramuscularly. Oil-miscible vit. A should never be use parenterally. Children with severe protein deficiency, give an additional oral dose every 2 wks. until their protein status improves. Treat all systemic illnesses that may be causing the malnutrition. Prevent or cure secondary bacterial infections (broad spectrum antibiotics), protect the globe from undue pressure, and when indicated, speeding vit. A-dependent healing using retinoic acid, 0.1% in arachis oil, applied topically one to 3 x's/day. Large, less typical lesions require careful culture, smear and intensive antibiotic therapy. Surgery may be performed on lge. perforating ulcers.
General Name  Central Corneal Degeneration
Specific Name  Posterior Crocodile Shagreen
ICD 9-CM#  371.40
Location  Cornea
Associated Conditions
Endothelial dropout

S:  Asymptomatic.  
A bilateral condition.

O:  A slit lamp evaluation reveals a series of small, various sized, gray polygonal patches on the cornea. The patches are separated by dark regions at the level of the Descemet's membrane.

A:  Posterior crocodile shagreen is a central corneal degeneration. Minor degenerative changes in the cornea are common and are almost always benign.

P:  No treatment is generally required.
General Name: Corneal Dystrophy
Specific Name: Meesman's Epithelial Dystrophy
ICD 9-CM#: 371.52
Location: Cornea
Associated Conditions:

S: Usually asymptomatic although some patients may have mild ocular discomfort and slight decrease in visual acuity to the 20/40 range.

O: Corneal epithelium diffusely studded with minute flock-like opacities of variable density and distribution that on retroillumination with the slit lamp, appear to be minute collections of debris in an otherwise clear, spherical microvesicle. These spherical microcysts may elevate the corneal surface sufficiently to disturb the tear film. Superficial corneal staining is rare.

A: Meesman's epithelial dystrophy is a dominantly inherited dystrophy of the cornea. The epithelial changes have been demonstrated as early as 7 months and tend to increase with age. Pathologic changes are usually confined to the epithelium. This condition must be differentiated from bilateral microcystic epithelial changes that may be seen with corneal edema, vernal conjunctivitis, or in association with disturbed tear function.

P: Treatment is usually not necessary. If the discomfort is severe soft contact lenses can be considered. If visual impairment is unusually severe, lamellar keratoplasty may be indicated.
General Name: Dct Fingerprint Map Dystrophy
Specific Name: Cogan's Microcystic Dystrophy
ICD 9-CM#: 371.50
Location: Cornea

Associated Conditions:
Corneal erosions

S: Patients mainly complain of a mild foreign body sensation, but visual acuity and corneal sensation are generally not affected except during erosions.

O: Slit lamp exam reveals:
- Bilateral microcystic patterns in the corneal epithelium and adjacent basement membrane.
- Gray-white dot-like lesions or fingerprint or maplike designs formed by contoured lines may also appear.
- The opacities are best viewed with retroillumination.

A: The condition is seen mainly in females and is probably of dominant inheritance. It is thought to be caused by variations of changes in the growth pattern of the epithelial basement membrane. The cysts themselves contain cellular debris.

P: Treatment of the condition involves management of the recurrent corneal erosions by debriding the damaged epithelium and pressure patching. Therapeutic soft contact lenses may also be of benefit.
General Name: Reis-Bucklers' Dystrophy
Specific Name: Reis-Bucklers' Dystrophy
ICD 9-CM#: 371.52
Location: Cornea
Associated Conditions:

S: The dystrophy is usually bilaterally symmetric and becomes evident in the first and second decade of life as painful recurrent erosive episodes. Patients generally have decreased visual acuity due to anterior scarring and surface irregularity and, in time, decreased corneal sensitivity.

O: A superficial corneal dystrophy which affects Bowman's membrane. Slit lamp examination shows an irregular epithelium with diffuse, irregular, patchy geographic opacities in Bowman's layer. As time passes, central opacities appear as a reticulated pattern spreading into the midperiphery with a diffuse superficial stromal haze.

A: An autosomal dominant mode of transmission. The primary cause of this disease is unknown, but the primary lesion may be due to fragmentation of the collagen fibrils of Bowman's membrane, and the epithelial lesion may occur secondarily. Destruction of Bowman's layer and its replacement by fibrillar material are the relevant changes in the disease and distinguish it histopathologically from other anterior dystrophies. Rule out Fleischer (Vortex) dystrophy, Anterior mosaic crocodile shagreen (Vogt's), Idiopathic band keratopathy.

P: Superficial keratectomy has been found to be helpful in the management of this disorder and should be preferred to penetrating keratoplasty. Concomitant abnormalities in the epithelial basement membrane account for the recurrent erosions after keratoplasty that patients experience.
General Name: Corneal Dystrophy
Specific Name: Fleisher Vortex Dystrophy
ICD 9-CM#: 371.52
Location: Cornea
Associated Conditions: Fabry's Disease

S: None

O: Slit lamp exam reveals pigmented whorl shaped lines which travel in the epithelium and Bowman's layers of the cornea.

A: Vortex dystrophy is seen primarily in female carriers of the X-linked Fabry's disease. Other ocular findings in the Fabry's disease are star shaped lines in the posterior lens and tortuous vessels in the conjunctiva and retina.

P: Prognosis for female carriers is fair depending on to which extent the recessive gene disorder is expressed. Central retinal artery and vein occlusions often occur in males of the disease as well as skin lesions and pain in the limbs. Most males only live into the third to fourth decade without renal transplant.
General Name Anterior Mosaic Crocodile Shagreen (Vogt)
Specific Name Anterior Mosaic Crocodile Shagreen (Vogt)
ICD 9-CM# 371.50
Location Deep layers of the corneal epithelium and in Bowman's layer
Associated Conditions
Megalocornea, peripheral band keratopathy, iris malformation, post traumatic conditions.

S: None.

O: Bilateral, polygonal, grayish white opacities separated by clear tissue. The opacities are usually axial. Corneal sensation is somewhat decreased but vascularization is not present. V.A. is not affected.

A: By clinical appearance. This is a corneal dystrophy. Histological studies have revealed interruptions of Bowman's membrane and separation of the membrane from the epithelium by connective tissue. The so-called anterior mosaic pattern is a different entity in which a delicate mosaic pattern is seen after instillation of fluorescein. With mild pressure on the closed lid, a pattern of interconnecting lines which form a series of connected polygonal figures may be produced in the fluorescein sheen.

P: Unless the V.A. is affected, no treatment is indicated. On rare instances keratoplasty has been required.
General Name Corneal Dystrophy
Specific Name Idiopathic band Keratopathy
ICD 9-CM# 371.43
Location Cornea
Associated Conditions
Stromal dystrophies, Band keratopathy

S: Patient may complain of decrease in vision.

O: Slit lamp examination reveals a horizontal band of calcium deposits across the Bowman's membrane of the cornea. The distribution of the deposits is in the interpalpebral zone separated by a clear interval between the limbus and the ends of the band.

A: Band Keratopathy results from deposition of noncrystalline phosphate and carbonate calcium salts. Etiology is unknown. The condition is identical to the band keratopathy seen secondary to systemic problem.

P: Band Keratopathy may be removed for visual and/or cosmetic reasons with chelating agents e.g. sodium versenate or EDTA. Antibiotic ointment, short term cycloplegic, and mild pressure patching is indicated 24 to 48 hours post operatively.
General Name: Corneal Dystrophy
Specific Name: Granular Dystrophy (Groenouw type 1)
ICD 9-CM#: 371.53
Location: Cornea
Associated Conditions

S: Asymptomatic until late in the disease process where there is reduction in visual acuity.

O: Stromal opacities of dense, milky granular appearing deposits occurring in the axial portion of the cornea, more prominently in the anterior stroma. Between the "bread crumb" opacities the cornea remains clear.

A: Granular dystrophy is an autosomal dominant dystrophy. The lesions may be manifest in the first decade of life but visual acuity is usually not affected until late in the disease. The deposits are thought to be principally noncollagenous protein.

P: Occasionally when visual acuity is severely impaired, penetrating keratoplasty is indicated.
General Name: Macular Dystrophy
Specific Name: Gil or Bucklers II Dystrophy
ICD 9-CM#: 371.55
Location: Cornea
Associated Conditions:
Penetrating Keratoplasty

S: The patient reports loss of vision, episodes of irritation and photophobia. They may also notice a clouding of the cornea.

O: Signs include:
- Decreased VA
- Diffuse clouding of the central cornea involving most of the stromal thickness. Cloudy areas of cornea appear between the large irregular lesions.
- Increase in the density of the opacity leads to development of gray-white nodular deposits.

A: Macular dystrophy is the rarest and most serious of the stromal dystrophies. It is autosomal recessive and appears in the first decade of life. Visual acuity is affected by the late teens. The deposits in and near the keratocytes are accumulations of mucopolysaccharide resulting from a local enzyme deficiency.

P: There is often a serious loss of corneal transparency and lamellar or penetrating keratoplasty is required. The dystrophy has been known to recur in the grafted tissue.
General Name  Lattice Dystrophy
Specific Name  Biber-Haab-Dimmer
ICD  9-CM#  371.54
Location  Cornea
Associated Conditions  Amyloidosis.

S: Symptoms are in the first decade of life. Among them are recurrent erosions, decreased vision, changes in corneal transparence, and irregular astigmatism because of subepithelial and stromal accumulations of amyloid material appearing as pathognomonic branching lattice figures. The dystrophy advances inexorably, and by age 40 or earlier the above problems become markedly aggravated, causing considerable visual incapacity and, usually, discomfort.

O: In time, one sees opacities in the axial stroma, as well as in the superficial layers, leaving the limbus relatively free. The cornea exhibits a "ground glass" appearance with "pipestem" lesions scattered throughout the stroma and involving layers. At this stage, the superficial haze makes it difficult to diagnose lattice dystrophy were it not for examining younger affected family members. The accumulation of amyloid material will give rise to poor epithelial-stromal adhesion. As a result, the patient is periodically troubled with epiphora and photophobia due to recurrent erosions.

A: An autosomal-dominant familial form of amyloidosis (limited to the cornea). Staining with thioflavin-T, using fluorescence microscopy, is helpful in further characterizing the amyloid material, as are immunofluorescent studies using antihuman amyloid antisera. Although collagen degeneration may occur, perhaps from lysosomal enzymes elaborated by abnormal keratocytes, actually produce the abnormal amyloid substance, although this process is not ultrastructurally evident.

P: Treatment of this disorder is symptomatic, depending on visual acuity and patient discomfort. Penetrating keratoplasty in this condition carries an excellent prognosis, although recurrence of the dystrophy in the graft may take place.
General Name: Corneal Guttata
Specific Name: Fuch's Dystrophy
ICD 9-CM#: 371.57
Location: Cornea

Associated Conditions:
Nuclear Cataracts, Open Angle Glaucoma

S: The patient usually complains of blur and glare. Intense pain is found in advanced cases. Haloes can also be elicited in case history.

O: In corneal guttata, slit lamp evaluation reveals a beaten bronze appearance at the level of Descemet's membrane. Breaks in the hexagonal mosaic pattern of the endothelium are also seen. In Fuch's dystrophy the corneal endothelium takes on a ground glass appearance when viewed with retroillumination. High magnification also reveals wartlike expressions and pigment dusting of the endothelium. A diffuse corneal edema is observed in both cases.

A: These are both bilateral endothelial dystrophies. The unilateral exception is that seen in cases of endothelial compromise from cataract surgery. Guttata is often associated as an early stage of Fuch's. The condition usually occurs in middle to older aged patients. However it has been seen in contact lens wearers at earlier ages. In Fuch's dystrophy the guttata start centrally and progress toward the periphery. The dysfunction of the endothelium cause diffuse edema which is worse in the morning.

P: The prognosis is variable. If the edema is not controlled, epithelial bullae are formed. If these rupture, cornea abrasion symptoms occur as well as the potential for ulceration and infection. At this stage pannus can occur. Management involves controlling the edema. 5% NaCl ointment or drops is an effective hyperosmotic. A hair dryer at arms length has also been used to dehydrate the cornea. Therapeutic soft lenses have been used to decrease symptoms in bullous keratopathy. If pannus is reached, a penetrating keratoplasty is the treatment of choice.
General Name: Congenital Hereditary Endothelial Dystrophy
Specific Name: Congenital Hereditary Endothelial Dystrophy
ICD 9-CM#: 371.58
Location: Endothelium
Associated Conditions:

S: Autosomal recessive type: discomfort symptoms are not prominent.
Autosomal dominant type: progressive photophobia and tearing problems.

O: Diffuse, bilaterally symmetric corneal edema, corneal clouding which varies from a mild haze to a ground-glass opacification, microedema of the epithelium, swelling of the stroma (up to 3 fold), uniform thickening of Descemet's. No guttata, interstitial inflammation or its secondary vascularization are present.

A: By clinical appearance. Must rule out congenital glaucoma. This is done under anesthesia measuring corneal diameter and IOP. Rule out posterior polymorphous dystrophy in which the corneal thickness is normal. The recessive type is present at birth and is relatively stationary. The dominantly inherited form develops in the first of second year of life and progresses.

P: Use of 5% NaCl ointment of drops to decrease epithelial edema.
Hair dryer held at arms length 2-3 times/day to decrease epithelial edema.
Penetrating keratoplasty once progressive vision loss occurs.
General Name: Endothelial Dystrophy
Specific Name: Posterior Polymorphous Dystrophy (Hereditary Deep Dystrophy)
ICD 9-CM#: 371.58
Location: Cornea
Associated Conditions: Corneal edema, Fuch's dystrophy, Congenital Hereditary Endothelial Dystrophy.

S: Symptoms range from minimal to moderate effect on visual acuity. The effect varies even among family members.

O: Slit lamp evaluation reveals a variable number of round, elliptical, or irregular lesions often with vesicular appearance in the Descemet's membrane. These lesions may bulge into the stroma or project out into the anterior chamber.

A: Posterior Polymorphous Dystrophy is an autosomal dominant or recessive dystrophy of the endothelium and Descemet's membrane. The condition is generally benign and non-progressive. However, it may be associated with corneal edema, requiring penetrating keratoplasty to restore vision in severe cases. Differential diagnosis from other endothelial dystrophies such as Fuch's dystrophy and Congenital Hereditary Endothelial Dystrophy.

P: No treatment is generally required. However, keratoplasty is indicated in some cases.
General Name: Wilson's Disease
Specific Name: Wilson's Disease (Kayser-Fleischer Ring)
ICD 9-CM#: 371.14
Location: Cornea

Associated Conditions:
Hepatic Disease

S: Symptoms of the disease are mainly systemic if the condition is advanced. They occur between 8 and 40 years of age and usually consist of cerebral or hepatic signs such as rigidity, tremor, personality disorders, epilepsy, hepatic dysfunction or hepatic coma.

O: Ocularly patients exhibit a green to brown ring at the limbus at the level of Descemet's membrane also known as the Kayser-Fleischer ring. This is often the first sign of disease so it is important to recognize it so that early treatment can be instituted. The ring is composed of copper granules which accumulate and are only visible in Descemet's. The ring begins superiorly and extends circumferentially. It can only be seen with gonioscopy in the early stages. In a few cases a sunflower cataract that affects the anterior and posterior capsule may be present. With the slit lamp it appears as a powdery deposit of browns, greens, reds and yellows that is centrally located and extends into the periphery.

A: The disease is of autosomal recessive inheritance and involves a defect in copper metabolism. In children it usually presents as hepatic disease and in adolescents and adults neurologic involvement is more common. Definitive diagnosis is made by liver biopsy which is found to have 20-30 times the normal concentration of copper. Also serum levels of copper and ceruloplasmin a copper binding agent are low.

P: Treatment is essential in preventing liver and basal ganglion damage. Treatment involves a low copper diet and use of D-penicillamin, a copper chelating agent. If successful, the cataract as well as the corneal ring disappears and cerebral and hepatic conditions are relieved. Referral to an internist is in order.
General Name: Chandler's Syndrome

Specific Name: Chandler's Syndrome

ICD 9-CM#: (371.58, 364.51), 732.7

Location: Cornea

Associated Conditions:
Essential iris atrophy, Iris nevus (Cogan-Reese syndrome), Peripheral anterior synechiae.

S: Essential iris atrophy, Chandler's syndrome, and the iris nevus or Cogan-reese syndrome have recently been regarded as variations of a single disease process. Chandler's syndrome and the above associated conditions are non-familial, unilateral, and generally arise in early adulthood, usually in women. The term iridocorneal-endothelial syndrome (ICE syndrome) has been proposed.

O: A corneal edema secondary to endothelial abnormality, in the company of ipsilateral glaucoma. The degree of corneal edema is severe relative to the level of intraocular pressure. The various iris changes (stromal thinning, frank holes, "nevi", and peripheral anterior synechiae) are of controversial etiology.

A: It has been recently proposed that the primary problem is in the corneal endothelium, which, besides malfunctioning and allowing corneal edema, tends to grow across angle structures and iris surface, elaborating a Descemet's membrane-like tissue on these surfaces. Contraction of the membrane then leads not only to anterior synechiae but also to pupillary distortion and iris abnormalities. Differentiation from such disorders as Fuchs' dystrophy and posterior polymorphous dystrophy must be made. Specular microscopy may prove useful. Rule out Fuch's dystrophy.

P: Histopathologic study has been limited. Experience with three keratoplasty specimens has revealed typical evidence of "endothelial distress syndrome", as flat and discontinuous endothelial cells have elaborated excessive multilaminar basement membrane.
General Name: Posterior Polymorphous Dystrophy
Specific Name: Posterior Polymorphous Dystrophy
ICD 9-CM#: 371.58
Location: Cornea
Associated Conditions: None

S: Variable from no symptoms to complaints of blur, glare, and haloes.

O: Slit lamp exam reveals a number of round, elliptical, or irregular lesions of the endothelium and descemets's which bulge into the posterior stroma. In severe cases, bulging can be seen into the anterior chamber.

A: Posterior polymorphous dystrophy is a dominantly inherited dystrophy. Abnormal iris processes and peripheral anterior synichiae have been noted in some cases. It must be differentially diagnosed from Fuch's dystrophy where guttata and a ground glass appearance of the endothelium are seen.

P: The prognosis is good for asymptomatic patients. It is a stable and non-progressive dystrophy in most cases. However in cases of mild edema, 5% NaCl drops may be indicated to reduce edema. In cases of severe edema, keratoplasty has often been advocated.
General Name: Acanthamoeba
Specific Name: Acanthamoeba
ICD: 9-CM#: 370.0
Location: Cornea
Associated Conditions:

S: Ocular pain and discomfort and decreased visual acuity. Often the pain is out of proportion to the initial signs of infection.

O: Observation with the slit lamp shows minimal corneal epithelial changes on initial presentation. The epithelium is intact but has a mottled or dendritiform appearance similar to that of a Herpes simplex lesion. As the disease progresses, persistent epithelial erosions are a consistent, although variable, hallmark. The anterior chamber reaction is relatively mild although hypopyon has been reported in advanced cases. Stromal thinning occurs relatively late in the course but perforation can occur.

A: Diagnosis of Acanthamoeba by clinical or laboratory methods is often difficult. This is because clinically the condition resembles herpes simplex keratitis. Corneal scrapings sent to the laboratory for culture and sensitivity testing often fail to show a positive identification. Even when the more useful lab tests such as hemacolor, tricrome stain and calcofluor white are negative the attending doctor cannot rule out Acanthamoeba in the diagnosis. In the majority of cases, definitive diagnosis was obtained from histologic examination of the corneal button removed at the time of keratoplasty. A careful case history must be taken on patients suspected of having an Acanthamoeba infection. According to the literature, these infections are related to three events that can occur separately or in various combinations: 1) corneal trauma 2) contact lens wear 3) contact with one of Acanthamoeba's habitats.

P: Currently there is no good pharmaceutical therapy against acanthamoebic keratitis, as a consequence, adequate medical therapy is difficult to provide. Amoeba are resistant to most ocular antibiotics. Cysts appear to be more resistant than trophozoites and may persist in the cornea indefinitely, requiring prolonged topical medical treatment to ensure eradication before keratoplasty. Three of the most frequently tested amoebicidal and cysticidal drugs are propamidine isethionate, polymyxin-B-polymyxin, and miconazole. These drugs have shown only a limited success. The majority of patients infected with Acanthamoeba will eventually require keratoplasty. If the amoeba invades the sclera enucleation is the required medical procedure.
**General Name** Adenoviral Infections:

**Specific Name** Epidemic Keratoconjunctivitis (EKC)

**ICD 9-CM#** 077.1

**Location** Cornea, Conjunctiva

**Associated Conditions**

Gastrointestinal and upper respiratory problems, conjunctivitis, red eyes

**S:** Ocular symptoms includes pain, photophobia, red eyes, and watery eyes. Although it is usually not accompanied by systemic problems, complaints of fever, malaise, gastrointestinal and upper respiratory symptoms are often observed in children. The condition is initially unilateral, later becoming bilateral.

**O:** Slit lamp evaluation reveals:

- follicular conjunctivitis
- superficial punctate keratitis
- subepithelial infiltrates
- may have lasting infiltrates
- edema of the semilunar fold and caruncula
- pseudomembranes (occasionally)
- subconjunctival hemorrhage
- uveitis

Tender regional nodes is also observed.

**A:** EKC is due to Adenovirus type 8 and 19. The clinical triad of EKC consists of follicular conjunctivitis, keratitis, and adenopathy. Edema of eyelids, chemosis, conjunctival hyperemia with follicles and subconjunctival hemorrhages often appear within 48 hours of the onset of the virus. With in 5-14 days, photophobia, epithelial keratitis, and round epithelial opacities are observed. The conjunctivitis can lasts from 3-4 weeks in severe cases and can produce pseudomembranes and petechial hemorrhages of the conjunctiva. The rule of eight +/- two days:

- First 8 days: Follicular conjunctivitis
- Next 8 days: Superficial Punctate Keratitis
- Next 8 days following: Subepithelial infiltrates

**P:** 1. Strict hygienic measures must be undertaken due to the contagiousness of the disease.
2. Educate patient that the disease is self limiting and must run its course.
3. Treatment is mainly supportive management:
   a) Topical decongestant drops and cool compresses for symptomatic relief of inflammatory congestion for patient comfort.
   b) Cycloplegics e.g. atropine, may be used to decrease ocular pain, and manage resultant uveitis.
   c) Topical steroids are used primarily for patients with corneal opacities which interfere with vision. Long and gradual tapering of steroid dose may be indicated due to the recurrence of the opacities with discontinuation of steroids. Opacities may lasts for months but in time will clear without scars.
General Name PCF
Specific Name Pharyngo-Conjunctival Fever
ICD 9-CM# 077.2
Location Conjunctiva
Associated Conditions
Epidemic conjunctivitis

S: Patient usually presents with a bilateral burning, tearing, mildly red eye. Systemically complaints range from sore throat, fever, malaise, headache, abdominal discomfort and diarrhea. The symptoms are of fairly recent onset—usually 3-7 days.

O: The characteristic triad of PCF is:
- Fever of 101-104
- Pharyngitis
- Follicular conjunctivitis
   However not all three are always present. Exam with the slit lamp reveals follicles on the conjunctival mucosa. In some cases there may be a SPK or subepithelial opacities, but corneal involvement is rare. Palpation of preauricular nodes reveals a lymphadenopathy.

A: The causative agent is Adenovirus 3 and 7 which frequently causes epidemic outbreaks associated with contaminated swimming pools. In the early stages PCF can be confused with acute inclusion conjunctivitis, acute hemorrhagic conjunctivitis or herpes conjunctivitis. Differential diagnosis can be achieved by the associated triad of symptoms, along with culture on HeLa cells and neutralization tests.

P: The disease runs its course in 5-14 days and therapy is directed toward patient comfort, and prevention of complications and secondary bacterial infection. This is achieved by prescribing cold compresses, topical decongestants and ocular lubricants. Topical antibiotics are indicated in cases of secondary bacterial infection.
General Name: Arteriosclerosis

Specific Name: Arteriolar sclerosis

ICD 9-CM#: 362.10

Location: Cardiovascular

Associated Conditions: Hypertension, Attenuated blood vessels, Cotton wool spots, Papilledema, Hard exudates

S: Symptoms varies with the different hypertensive stages:
- Initial stages: Asymptomatic
- Accelerated hypertensive stage: Blurred vision, dizziness, headache
- Hypertensive encephalopathy: Coma

O: LONG DURATION MODERATELY SEVERE HYPERTENSION:
- Attenuation of the arteriole.
- Arteriovenous crossing changes: concealment, depression/elevation, deviation, stenosis.
- Changes in vascular light reflex: widening, copper-wire arteries, silver-wire arteries, sheathing, turouosity.
- Hard yellow exudates, Hemorrhage, and Microaneurysms.

RECENT ONSET SEVERE HYPERTENSION:
- Severe focal attenuation of arterioles.
- Retinal edema, Hemorrhage, Papilledema, and Cotton-wool patches.
- Ophthalmoscopic changes of replacement fibrosis may precede above signs or remain after condition is corrected.

A: To avoid confusing arteriolar sclerosis from clinically similar congenital anomaly, diagnosis should be made after the blood vessels have traversed one disc diameter away. Attenuation of the arterioles is best judged beyond the second bifurcation. Arteriovenous crossing changes and vascular light reflex changes are due to the replacement fibrosis of the retinal arteriole. Hard deposit (chronic edema residues or hard yellow exudates) are focal accumulations of macrophages in the outer plexiform layer or beneath the sensory retina. In severe hypertension, hard deposits may concentrate around the fovea creating a star-shaped appearance. Cotton wool spots occurs when the pressure exceeds 120 mm Hg. Papilledema occurs shortly after the appearance of the cotton wool spots. Patient reaching this stage will complain of headache, blurred vision, and dizziness. In severe, accelerated hypertension, hypertensive encephalopathy may occur resulting in coma due to the high arterial pressure.

P: Referal to an internist.
General Name Arteriosclerosis
Specific Name Atherosclerosis
ICD 9-CM# 362.10
Location Cardiovascular
Associated Conditions
Vascular hypertention, Turbulence eddies within blood vessels, hyperlipoproteinemia, embolus, amaurosis fugax

S: Asymptomatic in most cases. Transient vision loss "amaurosis fugax" may occur in cases when there is an embolus lodged in the bifurcation of the blood vessels. The condition is rare in puberty, but thereafter its extent and severity increases with age.

O: Ophthalmoscopic evaluation reveals:
1. Focal necrosis and thickening of the arterial intima.
2. Atherosclerosis involves the central retinal artery (CRA) within the optic nerve or its branches immediately adjacent to the optic disc. An atheroma in the CRA in the optic disc causes the retinal vessels to become straighter and the bifurcation angle more acute. An atheroma in the papillary branches of the CRA causes reduction in the caliber of the lumen and whitish opacification in the blood vessels due to fibrosis.
3. Embolis: Cholesterol (glistening, crystalline body). Fibrin (dull white to yellowish white body with ill defined borders).

A: Arteriosclerosis is a group of disease characterized by thickening and loss of elasticity of the arterial walls e.g. atherosclerosis and arteriolar sclerosis. In artherosclerosis, there are hyperplastic and degenerative changes in the arterial wall, especially of the internal elastic lamina. The disease affects arteries of all diameters, but has a predilection for major arteries. A cholesterol embolis dislodged from atheromatous plaque can cause amaurosis fugax or retinal ischemia. Atheromatous plaque formation in the artery may compress the vein, causing venous obstruction at the lamina cribosa (central retinal vein occlusion) or at one of the bifurcations (branch vein occlusion).

P: Referral to an internist.
General Name Diabetes

Specific Name Background and Proliferative Retinopathy

ICD 9-CM# 362.01-.02

Location Retina

Associated Conditions

S: Fluctuating vision and early presbyopia are the initial visual symptoms in diabetics. Overt diabetics may have high urine outputs, polydypsia, nocturia, weight loss, itching skin problems, sleepiness, pins and needles sensations, impotence, cyclic episodes of edema, and loose teeth. Children may have episodes of involuntary uriination or bed-wetting. Diabetics also tend to be overweight. Latent diabetics usually have no symptoms. Their blood sugar levels rise only after they’ve eaten.

O: Rapid shift in refractive error. Small conjunctival and subconjunctival hemorrhages and aneurisms. Corneal erosion is a frequent problem. Abnormal pupillary responses such as pseudo-Argyll-Robertson and Marcus Gunn pupil. Snowflake or punctate lens opacities under the anterior or posterior capsule. The iris pigment epithelium may form vacuoles of pigment granules that migrate to the corneal endothelium. Background retinopathy has dilated veins, microaneurisms, dot and blot hemorrhages, cotton-wool patches, hard exudates, intravascular microvascular abnormalities (shunt vessels), venous beading or loops, macular edema, sheathed or white-line arterioles. Proliferative retinopathy has new vessels developing in clinically distinguishable stages. Initially, red, bare, fine vascular trfts or fronds with little connective tissue form flat loops on the retinal surface. In the second stage, vessels proliferate more rapidly; the caliber and fibrous connective tissue also increase causing the new tissue to look pink rather than red. In the final stage the lesion presents with a whiter hue. Most neovascularization occurs at the optic disc but it may appear elsewhere. New blood vessels growing on the retinal surface are usually attached to the posterior hyaloid surface of the vitreous body. Vitreous contraction may cause vitreous hemorrhage, retinal holes, tears, and detachments. Late stage complications include rubeosis irides, glaucoma, and vitreal opacification.

A: There are two different causes of diabetes. One is pancreatic insufficiency which is usually diagnosed before the age of 30. The other is a hormonal imbalance in the regulation of the production of insulin which is usually diagnosed after the age of 30. The prevalence of associated retinopathy varies within the diabetic population and is influenced by many factors including: age, sex, age at diagnosis, and duration of the disease. Because of the insidious nature of adult onset diabetes, eye changes may be the first clinical evidence of its presence. The definitive diagnosis of diabetes is made via blood sugar testing.

P: Proper medical management of blood sugar levels is necessary to prevent the fatal consequences of diabetes and help delay its complications. Depending on its severity, diabetes can be treated with diet, exercise, oral hypoglycemic drugs or insulin. For known diabetics, a stable refraction on a least 2 visits should be obtained before prescribing glasses. Background retinopathy which isn’t sight threatening can be followed with bi-yearly exams, but refer the patient if he has signs that seem too severe. Intraretinal microvascular abnormalities and venous beading represent an advanced stage of background retinopathy. Diabetics with these severe changes will probably develop neovascularization within the next two years and they need frequent, detailed monitoring. Refer to an ophthalmologist any patient with proliferative retinopathy. Panretinal photocoagulation is beneficial in delaying blindness from disc and retinal neovascularization accompanied by vitreous hemorrhage. Vitrectomy is used to clear up vitreous hemorrhages of long standing duration and managing traction retinal detachments.
General Name Hypertension

Specific Name Ocular response to Hypertension-Normal vessels with sudden inc. BP

ICD 9-CM# 401.9

Location Retina

Associated Conditions
Heart attacks, Retinal hemorrhage, Cotton wool spots, Macular star, Hard exudates

S: Symptoms include:
Swollen face and hands may occur with toxemia of pregnancy or drug reactions.

O: Signs include a sudden rise in blood pressure of 30mmHg systolic or 15mmHg diastolic,
edema of face and hands, and straight narrow pale arterioles in the retina.

A: Monitor pressure. If blood pressure drops back to normal arterioles will normalize in a
few weeks if pressure was taken off artery walls before permanent damage was done.
In cases where the pressure remains elevated other measures need to be taken such as
drug therapy.

P: Non drug therapy-- 70-90% can be managed by cutting salt intake and exercising to
lose weight.
Drug therapy
1-Hydralazine (Apresazide, Apresoline, Unipres) Directly relaxes vascular smooth
muscle.
2-Chlorothiazide (Diuril) Diuretic-reduces plasma volume . One of the mildest
treatments.
3-Methyl dopa (Aldomet) One of the most widely Rx'd. Works by reducing cardiac
output and peripheral resistance.
4-Guanethidine (Esimil) Action is the same as Methyl dopa
5-Propanolol (Inderal, Inderide) Beta adrenergic blocker. Reduces heart rate.
General Name Hypertension
Specific Name Benign Hypertension
ICD 9-CM# 401.1
Location Retina
Associated Conditions
Heart attacks, Retinal hemorrhage, Cotton wool spots, papilledema, Hard exudates, Macular star

S: Symptoms may not present until pressures reach high levels, at which time the patient may experience headaches, weakness, nervousness, nosebleeds.

O: Signs include continued moderate elevation of blood pressure. Vessels build up in order to contain pressure and leads to arteriosclerosis. Hyaline deposits in the intima of the vessels leads to concealment or sheathing of the vessels. The blood column gets narrower and a more prominent light reflex is given off. As arteries get thicker there is more concealment of venules at the vessel crossings which appear as banking, sheathing and nicking.

A: Based on continued moderate elevation of blood pressure and vessel changes diagnosis is benign hypertension. It should be noted that the vessel changes are permanent even if on treatment and normal treatment is is achieved. Treatment simply stops the progression.

P: Non drug therapy-- 70-90% can be managed by cutting salt intake and excercising to lose weight.
Drug therapy
1-Hydralazine (Apresazide, Apresoline, Unipres) Directly relaxes vascular smooth muscle.
2-Chlorothiazide (Diuril) Diuretic-reduces plasma volume. One of the mildest treatments.
3-Methyl dopa (Aldomet) One of the most widely Rx'd. Works by reducing cardiac output and peripheral resistance.
4-Guanethidine (Esimil) Action is the same as Methyl dopa
5-Propanolol (Inderal, Inderide) Beta adrenergic blocker. Reduces heart rate.

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General Name Hypertension

Specific Name Malignant Hypertension

ICD 9-CM# 401.0

Location Retina

Associated Conditions
Heart attacks, Cotton Wool Spots, Papilledema, Retinal hemorrhages, Macular star, Hard exudates

S: Symptoms include: Morning headaches, weakness, nervousness, may experience nausea and vomiting if pressure is extremely high and precedes a MI.

O: Usually patients have onset in the 6th to 7th decade of life. It is characterized by rapidly increasing blood pressure that doesn't come back down. The change is so rapid that vessels don't have time to structurally compensate. The inability of the vessels to compensate leads to appearance of cotton wool spots, hemorrhages, retinal edema and papilledema. The venous system appears as very dilated, dark and tortuous. If papilledema is not reversed it leads to irreversible optic atrophy. Vessels should be assessed at least 1 disk diameter away from the optic disk.

A: Based on the rapid increase in blood pressure and bilateral whole eye involvement the diagnosis is malignant hypertension. Treatment should be immediate.

P: Refer to an internist or physician.
Drug therapy
1-Hydralazine (Apresazide, Apresoline, Unipres) Directly relaxes vascular smooth muscle.
2-Chlorothiazide (Diuril) Diuretic-reduces plasma volume. One of the mildest treatments.
3-Methyl dopa (Aldomet) One of the most widely Rx'd. Works by reducing cardiac output and peripheral resistance.
4-Guanethidine (Esimil) Action is the same as Methyl dopa
5-Propanolol (Inderal, Inderide) Beta adrenergic blocker. Reduces heart rate.
General Name: Rheumatoid Arthritis
Specific Name: Rheumatoid Arthritis

ICD 9-CM#: 714.0

Location: Uveal Tract, Sclera, Cornea.

Associated Conditions
Extraarticular manifestations: pericarditis, pleuritis, nodular pulmonary disease, leg edema, splenomegaly, diffuse pulmonary interstitial fibrosis, muscle atrophy, lymphadenopathy, rheumatoid vasculitis and nodules.

S: RA is a chronic, progressive polyarthritis collagen disease with many extraarticular manifestations. It is 3 times more common in females, although this sexual predilection is far less pronounced in older patients. Average age at onset is 30 to 40 years and appears to be some tendency for familial involvement. Early systemic manifestations include fatigue, weight loss, fever, and a migratory progressive arthritis. Almost any joint in the body may be involved, although most often involved are the fingers, knees, elbows, ankles, and feet. Skin changes include atrophy, moist cold extremities, and "liver palms" (erythema of the palms).

O: The American Rheumatism Assoc's set of diagnostic criteria include: 1) morning stiffness 2) pain on motion or tenderness in at least one joint 3) swelling in at least one joint 4) swelling of at least one other joint 5) symmetric and simultaneous joint swelling on both sides of the body 6) subcutaneous nodules 7) typical x-ray changes 8) positive agglutination test for rheumatoid factor 9) poor mucin clot from joint fluid 10) characteristic histologic changes in synovial membrane 11) characteristic histologic changes in nodules. Definite RA is diagnosed in the presence of any five criteria. Ocular manifestations (uveitis, iritis, iridocyclitis) of adult RA are usually seen in more severe cases. Exacerbations of the ocular problems often occur at times of increased activity of the systemic disease.

A: Its pathogenesis is unknown. The onset of scleritis may be a warning sign of the development of more severe systemic disease with widespread vasculitis (episcleritis has also been reported). Keratoconjunctivitis sicca (KCS) of Sjogren's syndrome is the most common ocular manifestation of RA. RA patients with coexisting Sjogren's syndrome have a much higher incidence of drug allergy (esp. penicillin) than those with RA alone. Classic IgM rheumatoid factor is found in 70% to 90% of patients with RA. It is an "autoantibody" which reacts with the heavy chain of IgG molecules. The erythrocyte sedimentation rate (ESR) is elevated at times of disease activity.

P: Systemic treatment includes resting inflamed joints, physical therapy, salicylates, phenylbutazone, oxyphenbutazone, ibuprofen, tolmetin, gold salts, corticosteroids, antimalarials, and immunosuppressive agents. Some produce a variety of ocular problems. Surgical reconstruction of deformed joints, ranging from arthroplasty to total joint replacement, may result in significant clinical improvement in joint function. Treatment of rheumatoid ocular disease is often less than satisfactory, while use of specific drugs may be contradictory. Topical steroids are of value in treating rheumatoid episcleritis. They decrease symptoms and shorten the course of the disease. Rheumatoid scleritis is not generally responsive to topical steroid therapy alone. Systemic oxyphenbutazone can be used in divided doses of 600 mg/day for 4 days, with tapering to 400 mg/day for 1 week after all symptoms disappear. Indomethacin can be given in divided doses of 100 mg/day, with a reduction to 75 mg/day when a response is seen. When these two drugs are ineffective, systemic corticosteroids in very high doses is used (e.g. prednisolone 80 mg/day), tapering the dose when the condition is controlled. Hydrophilic soft CL have been used effectively in the treatment of rheumatoid corneal ulcers. If perforation has occurred, a corneal or corneoscleral graft may be necessary.
General Name  Rheumatoid Arthritis
Specific Name  Juvenile Rheumatoid Arthritis
ICD 9-CM#  714.30
Location  Bones, Joints, Anterior Chamber
Associated Conditions
Still's Disease, Scleritis, Glaucoma

S: The child presents with a red eye with little discomfort. The child may also complain of stiffness, swelling, and pain in the joints. The manifestation of the disease will often accompany a fever and concurrent skin rash.

O: The red eye is a chronic, bilateral iritis which is often recurrent. Cells and flare in the anterior chamber and KP bodies are present. Secondary to the iritis may include band keratopathy, cataract, and synchia.

A: Juvenile RA typically manifest before 16 years of age. An iritis in a child is a strong indicator in the diagnosis of juvenile RA. Still's disease is a combination of iritis, RA, cataract, and band keratopathy in a child. Several lab test are used to access juvenile RA. The sedimentation rate is usually high but the antinuclear antibody may or may not be present during early stages.

P: Aggressive treatment is required for normal growth of bones and joints. The first line of treatment used by a rheumatologist is aspirin or a similar anti-inflammatory agent. If this is ineffective steroids are used. In very serious cases gold salts are prescribed. Normal optometric management of the iritis with cycloplegics and steroids should be performed.
General Name: Graves' disease  
Specific Name: Hyperthyroidism (with ocular signs)  
ICD 9-CM#: 242.0  
Location: Widespread throughout body  
Associated Conditions: Myasthenia Gravis, rheumatoid arthritis, diabetes mellitus, Hashimoto's thyroiditis, pernicious anemia, idiopathic thrombocytopenic purpura, Addison's disease, vitiligo, Sjogren's syndrome, thymic hypertrophy, lymph adenopathy, splenomegaly.

S: Corneal irritation from rapid drying of precorneal tear film, keratitis from failure of eyelids to cover cornea adequately in sleep. Other symptoms vary with severity of the disease.

O: Eyelid retraction (Dalrymple), eyelid lag on downward gaze (von Graefe), infrequent blinking (Stellwag), globe lags behind upper eyelid on upward gaze (Means), lower eyelid lags behind globe on upward gaze (Gifford), increased pigmentation of skin (Jellinek), orbital congestion, exophthalmos, EOM contracture, EOM weakness, papillitis or retrobulbar neuritis, papilledema, neuroretinal edema.

A: Ocular involvement in Graves' disease is always ultimately bilateral. Even in a clinically uninvolved orbit will show hypertrophy of the EOM by CT scan, echography or NMR unit studies. The symptoms and signs in Grave's come on slowly as compared to inflammatory orbital syndromes which usually come on rapidly and are painful. In these cases also, remission and relapses occur more rapidly and respond well to corticosteroids whereas Graves' doesn't. Furthermore, tissue biopsy of acute diseases tends to cause an acute inflammatory exacerbation. CT scan will also show a more diffuse involvement than that seen in Graves' in which early changes are more confined to the EOMs.

P: Mild supportive measures, corticosteroids, radiation therapy, minor to major surgery. If only signs are present, no treatment may be necessary. When symptoms appear it becomes necessary to treat each symptom as needed.
**General Name** Sickle cell Retinopathy

**Specific Name** Sickle Cell Retinopathy

**ICD 9-CM#** 282.60

**Location** Blood

**Associated Conditions**
Sickle cell trait, Sickle cell Anemia, Sickle cell disease, Hemoglobinopathies

**S:** Symptoms varies with different types:

**SICKLE CELL DISEASE:** Asymptomatic, but have the most severe ocular complications

**SICKLE CELL TRAIT:** Painless hematuria

**SICKLE CELL ANEMIA:** Unrelenting anemia, recurrent abdominal & joint pain, fever, and infarction of many organs. In older patients, chronic leg ulcers, cirrhosis of liver, and cerebral vascular accidents are common.

**O:** **SICKLE CELL DISEASE:** (Five Progressive Stages of Retinopathy)
1. Peripheral arteriolar occlusion
2. Peripheral arteriovenous anastomosis
3. Neovascularization (Sea Fans is most frequent at the equatorial plane)
4. Vitreous hemorrhage
5. Retinal detachment

**SICKLE CELL TRAIT:**
1. Cotton wool spots (sickle cell plugs up the capillaries and causes ischemia)
2. Conjunctival changes (rare)

**SICKLE CELL ANEMIA:**
1. Most severe conjunctival changes

**A:** Differential diagnosis between the different types:

**SICKLE CELL DISEASE (S/C):**
1. 0.13% of the Black population.
2. It has few systemic and has the most ocular problems.

**SICKLE CELL TRAIT (A/S):**
1. 10% of the Black population.
2. It has few systemic and few ocular problems.

**SICKLE CELL ANEMIA (S/S):**
1. 0.4% of the Black population.
2. It has a lot of systemic and few ocular problems.

**P:** Referral to an internist and ophthalmologist.

Treatment for Sickle Cell Retinopathy:
1. Fluorescein Angiography is valuable in detecting early "sea fan" neovascularization.
2. Photocoagulation to obliterate neovascular tissue.
3. Vitrectomy to treat prolonged vitreous hemorrhage.
General Name Marfan's Syndrome

Specific Name Marfan's Syndrome

ICD 9-CM# 759.8

Location Generalized

Associated Conditions

S: Patients tend to be myopic. Ectopia lentis which occurs in approximately 80% of the patients may cause high degrees of astigmatism requiring frequent refractions. They may also experience diplopia.

O: Ectopia lentis. Dislocation is usually upward but may be dislocated in any direction. Other ocular abnormalities include iridotonia, strabismus, blue sclera, filtration angle anomalies, pupillary block glaucoma, peripheral retinal degenerations that can cause rhegmatogenous retinal detachment, heterochromia irides, translucence of the iris, megalocornea and keratoconus.

A: Marfan's Syndrome is an autosomal dominant systemic abnormality of connective tissue of variable penetrance and expressivity. Patients exhibit skeletal abnormalities including arachnodactyly, laxity of joints, tall stature, and sternal deformities. They may suffer from dissecting aneurysms of the thoracic aorta and aortic and mitral valvular disease. They are of normal intelligence.

P: In general, surgical removal of the subluxated lens is not indicated. Complications are common and surgery should be deferred unless necessitated by secondary glaucoma due to lens dislocation or cataract formation. There is some risk in dilating the pupils to examine the lens and fundus because of the danger of dislocating the lens into the anterior chamber.
General Name: Myasthenia Gravis
Specific Name: Myasthenia Gravis
ICD 9-CM#: 358.0
Location: Adnexa

Associated Conditions:
Ptosis, Fatigue, Muscle Weakness, Diplopia

S: 15% of patients complain of sagging lid and inability to move eyes into extreme positions of gaze, accompanied by double vision. Patients may also complain of lid twitches and quivering of the eyes. In an additional 40% of the cases these ocular symptoms are accompanied by muscle weakness and difficulty swallowing. Women are involved in 75% of the cases where onset occurs before age 35. No sex predilection occurs after age 35.

O: Signs include:
- Ptosis, either unilateral or bilateral
- Ophthalmoplegia: if not profound there may be twitching of the upper eyelid which is a strong indicator of Myasthenia Gravis. It is elicited by having the patient change his gaze from downward to straight ahead.
- Rapid movement of the eyes described as quiver, lightening twitches or oscillations may also be present.

A: Diagnosis is made based on the history of fluctuations in strength and confirmed by administering edrophonium (Tensilon). 2-10 ml is administered I.V. If myasthenia gravis is present, relief of ptosis and and muscle strength improvement will occur within 30-60 seconds and last for 2-3 minutes. The extraocular muscles aren't very responsive to cholinergic effects however, and diplopia may decrease and electromyocardogram may show increased activity. Atropine is given to counter cholinergic toxicity. Differential diagnosis must include hyperthyroidism which occurs in 3-8% of patients with MG, and oat cell carcinoma of the lung which is a syndrome resembling MG.

P: Classic treatment of MG is anticholinesterase medication such as Neostigmine. Blepharoptosis responds well but EO muscle paresis may be resistant. 25% eserine into the cul-de-sac decreases ptosis.
**General Name** Syphilis

**Specific Name** Syphilis

**ICD 9-CM#** 090 (congenital), 091.50-091.52 (uveal), 094.94-094.89 (neural).

**Location** Cornea, Optic Nerve, Choriocapillaris, Retina, Iris, Ciliary Body, Conjunctiva, Adnexa.

**Associated Conditions**
Hutchinson's triad, Optic neuritis & atrophy, Argyll Robertson pupil, Pseudoretinitis Pigmentosa.

**S:** Poor resistance to drying and changes in temperature. Like *N. gonorrhoeae*, syphilis can be transmitted either by venereal contact or from mother to fetus. Unlike gonorrhoeae, which is acquired during the birth process, *T. pallidum* can cross the placenta to infect the fetus early in gestation, and the disease may be far advanced by the time of birth. Congenital syphilis can be severe and overwhelming, causing abortion, miscarriage, or neonatal death. It can also be mild or even not apparent at birth, later becoming manifest as coryza, a bullous skin eruption, hepatosplenomegaly, meningitis, and osteochondritis. Interstitial keratitis, deafness, and malformed incisor teeth is the classic 'Hutchinson's Triad'.

**O:** Adult acquired syphilis first appears as an ulcerated, painless chancre accompanied by regional lymphadenopathy. Systemic signs are rarely present. The chancre will heal spontaneously. Secondary syphilitic manifestations follow and can take many forms including a variety of lesions of the skin and mucous membranes as well as alopecia, hepatitis, nephritis, and meningitis. Ocular involvement can include iridocyclitis, optic neuritis, or retinal vasculitis. Tertiary phase evolves after the lesions of secondary syphilis subside, including neurosyphilis and cardiovascular syphilis. In either congenital or adult-acquired syphilis, the histologic appearance of the lesions is the same: a vasculitis of the smaller blood vessels, which can be found in many organs. There are many ocular manifestations of the early stage of congenital syphilis which include optic neuritis and atrophy, chorioretinitis (the nonprogressive and non-vision-impairing 'salt and pepper' fundus), iridocyclitis, chronic dacryocystitis, conjunctivitis, paesthesia of ocular muscles, and thickening and redness of the eyelids.

**A:** Almost every ocular structure may be affected by syphilis, in both its congenital and acquired forms. The Argyll Robertson pupil is only rarely seen in congenital syphilis. Scattered pigment clumping in a "bone-corpuscle" pattern (pseudoretinitis pigmentosa). Congenital syphilis usually enters a latent phase during childhood. Interstitial keratitis usually occurs late in the first or early in the second decade of life and is usually bilateral. There is usually severe pain, photophobia, tearing, blepharospasm, and prelimal injection which results in vascularization and a "salmon-patch" appearance of the stroma. After a period of several wks. to several mos., the inflammation subsides, leaving a variable degree of scarring which may or may not impair vision. The blood vessels do not recede but remains as "ghost vessels". The diffuse chorioretinitis of Forster usually occurs in the late secondary stage of syphilis. Scattered flame-shaped hem. & edema are common. It is often impossible to demonstrate the presence of Treponema pallidum; therefore, the diagnosis of syphilis often depends on the presence of positive serologic tests. There is a predilection for the outer layers of the retina & the choriocapillaris. 90% of all cases of interstitial keratitis are secondary to syphilis. Either congenital or acquired syphilis can precipitate uveitis. The most common ocular finding in congenital syphilis is chorioretinitis (50%).

**P:** Many of the ocular manifestations of tertiary & secondary syphilis are the same. The A. R. pupil can only be diagnosed in the presence of good VA. A. R. pupil is usually bilateral, irregularly contours, and less than 2.5mm in diam. even in the dark. Anisocoria is often present, & dilates poorly to atropine, iris atrophy may abolish near reaction miosis. Penicillin has a very good bacteriocidal effect but only against growing organisms & not against latent syphilis. Erythromycin is an alternate choice to penicillin for the treatment of all stages of syphilis. For neurosyphilis, intramuscular injection of penicillin G, 500,000 IU every 6 hrs. for 17 days, accompanied by oral probenecia, 500 mg every 6 hrs. will produce treponemical levels in the cerebrospinal fluid. If the VDRL test is negative and syphilis is still suspected, it is extremely important that the fluorescent treponemal antibody absorption test be ordered. This test may detect syphilis earlier than the VDRL test; it is just as specific and more sensitive than the T. pallidum immobilization (TPI) test, and the test permits the diagnosis of late syphilis. It is the most sensitive test in all stages of syphilis. There are at least five ophthalmic indications for the FTA-ABS test: 1) any pupillary abnormality not otherwise explained 2) optic atrophy 3) dislocated lenses not due to Marfan's syndrome or homocystinuria 4) apparent retinitis pigmentosa and 5) chronic uveitis.
General Name: Albinism
Specific Name: Oculocutaneous and Ocular Albinism
ICD 9-CM#: 270.2
Location: Melanocytes
Associated Conditions: High Myopia, Nystagmous, Strabismus

S: Typically the patient complains of photophobia and reduced vision.

O: Oculocutaneous albinism manifest several tell tale signs. Hair and skin are pale or white. Ocular albinoes do not exhibit the obvious external lack of pigment. Ophthalmoscopy demonstrates what resembles a blond fundus. The foveal reflex is usually absent and areas of hyperpigmentation may be present. The iris is usually grey translucent and may appear pink.

A: Albinism is a hereditary disorder characterized by lack or reduction of melanin. Oculocutaneous albinism is divided into the categories of Tyrosine positive or negative. Ocular albinism is an X linked disorder. Ocular albinos may or may not have color vision defects. Pendular nystagmous is usually present. Visual acuity is often in the 20/200 range. Strabismus is also often present. Transillumination of the iris is seen using a retro illumination off the fundus.

P: Treatment is directed toward correction of the refractive error and strabismus. Sunglasses or tinted lenses are usually prescribed to increase patient comfort. Low vision may be indicated for those with decreased acuity potential. Previously undiagnosed ocular albinoes should also be given genetic counseling.
BIBLIOGRAPHY


