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Arnold-Chiari Malformation (ACM) and Syringomyelia (SM): A literature review & survey of ocular signs and symptoms

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Arnold-Chiari Malformation (ACM) and Syringomyelia (SM): A literature review & survey of ocular signs and symptoms

Abstract
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Thesis

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ARNOLD-CHIARI MALFORMATION (ACM) AND SYRINGOMYELIA (SM):
A LITERATURE REVIEW & SURVEY OF OCULAR SIGNS AND SYMPTOMS.

By
PAMELA M. BURTON
DEBRA J. McNAMARA

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Pacific University
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Advisor:
Willard Bleything, O.D., M.S.
ARNOLD-CHIARI MALFORMATION (ACM) AND SYRINGOMYELIA (SM):
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Pamela M. Burton

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Willard Breything, O.D., M.S.

December 17, 1997
BIOGRAPHIES

Pamela M. Burton

Pamela has a special interest in this topic as an Arnold-Chiari Malformation and Syringomyelia patient herself. Pamela was diagnosed while working towards her Bachelor of Science degree in Chemistry which she received from Fort Lewis College in Durango, Colorado. She will graduate in May of 1998 from Pacific University College of Optometry with a Doctorate of Optometry. Upon graduation, Pamela will be working for the Indian Health Services at a site that has yet to be determined.

Debra McNamara

Debra received her Bachelor of Science degree in Biology from Pembroke State University in Pembroke, North Carolina. She will also graduate in May of 1998 from Pacific University College of Optometry with a Doctorate of Optometry. Upon graduation, Debra will be an Optometrist for the United States Army.
ABSTRACT

Arnold Chiari Malformation (ACM) and Syringomyelia (SM) are being diagnosed more frequently with the use of Magnetic Resonance Imaging. Clinicians must consider ACM or SM in any patient that presents with unexplained sensory deficits, vertigo, headaches, nystagmus, oscillopsia, diplopia, motor reflex changes, transient vision loss and/or blurred vision.

Studies involving the neurological and ophthalmological signs and symptoms of ACM and SM were reviewed. A survey was conducted among 38 patients with either ACM, SM or both to determine what signs and symptoms are most frequently reported.

Optometric management requires recognizing when a patient presents with these symptoms, particularly downbeat nystagmus to refer them to for a neurological assessment with a recommendation for an MRI. In addition, management requires alleviating the patient’s symptoms and frequent monitoring of the patient’s extra-ocular muscles and evaluation of their optic nerve heads and visual fields.

KEY WORDS:
Arnold-Chiari Malformation, Hydrocephalus, Hydromyelia, Meningomylocele, Syringomyelia
INTRODUCTION

Arnold-Chiari Malformations (ACM) are a series of defects of posterior fossa anatomy, ranging from minor displacement of cerebellar tissue to gross bony defects or cerebellar hypoplasia.\textsuperscript{1} ACM, once thought to be a disorder of clinical importance only in the teen years and beyond, is now recognized as significant in the pediatric population as well.\textsuperscript{2} The clinical presentation of this condition is largely dependent upon age of onset, associated pathology, and the presence of syringomyelia. Considerable controversy exists within the literature concerning pathogenesis, signs and symptoms, diagnosis, and successful treatment of ACM.\textsuperscript{3}

Varying degrees of deformation and displacement occurs in ACM and is characterized by a displacement of the tectal region of the mesencephalon, caudal cerebellum and the medulla oblongata, (cerebellar tonsils, brainstem and the fourth ventricle) into the foramen magnum or upper cervical canal.\textsuperscript{4} Differential diagnoses between Chiari malformation types depends on the following two factors, how far the cerebellar tonsils are herniated and the amount of hydrocephalus present.\textsuperscript{5} If tomography scans show that the cerebellar tips have exceeded a distance of 5 mm below the foramen magnum, the diagnosis of ACM is confirmed and can be further defined by the exact millimeter of brain stem that is herniated through the foramen magnum.\textsuperscript{6}

ACM can be classified into four categories. ACM Type I has the least amount of herniation and is not associated with Spina Bifida.\textsuperscript{6} In ACM type I, there is cerebellar displacement into the spinal canal, but hydrocephalus and syringomyelia are variable.
Type I malformations pose diagnostic challenges because they often produce bizarre and vague symptoms. ACM type II is a disorder of embryological development and characterized by an opening of the spine and spinal cord on the lower back. These structures descend into the cervical spinal canal, often extending to the level of the fourth or fifth vertebrae. Type II typically manifests with severe hydrocephalus and myelomeningocele in infancy. Type II also consists of a downward displacement of the cerebellum, lower pons and medulla oblongata.

ACM types III and IV are present at birth and very rare. In ACM type III, the cerebellum, which is smaller in physical appearance, the lower brainstem, and 4th ventricle are pushed into the upper cervical canal. ACM type IV is present at birth and is lethal because the maldeveloped cerebellum and brainstem are herniated down through the foramen and into the posterior fossa.

The caudal cerebellum and medulla can be pushed outside the foramen magnum by pressure caused by an overproduction of cerebrospinal fluid or when the base of the skull is too small which forces the cerebellar region lower. Another theory is when the occurrence of an overgrowth results in a compression. A careful history and physical examination, coupled with neurological evaluation, especially MRI, can lead to the correct diagnosis.

Table 1 illustrates the characteristic signs and symptoms associated with the more common ACM Types I and II.
Table 1: Characteristic Signs and Symptoms of ACM Type I and II patients.

<table>
<thead>
<tr>
<th>CHARACTERISTIC SIGNS AND SYMPTOMS</th>
<th>ACM TYPE I</th>
<th>ACM TYPE II</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age of onset</td>
<td>Late childhood to adult</td>
<td>At birth</td>
</tr>
<tr>
<td>Hydrocephalus</td>
<td>Occasionally</td>
<td>Progressive</td>
</tr>
<tr>
<td>Nystagmus</td>
<td>Typically downbeat</td>
<td>Variable DBN / upbeat</td>
</tr>
<tr>
<td>Spinda bifida</td>
<td>Never</td>
<td>Usually</td>
</tr>
<tr>
<td>Syringomyelia</td>
<td>Between ages 20-40</td>
<td>Usually</td>
</tr>
</tbody>
</table>

Hydrocephalus is often an accompanying feature of ACM and this in turn is associated with scoliosis and motor weakness.\(^2\) Hydrocephalus develops from an obstruction of the CSF outflow from the fourth ventricle. This may be due to a membrane over the foramen of Magendie or by a fibrovascular net around the cerebellum and brainstem, at the level of the foramen magnum. The downward displacement of the spinal cord may cause the cervical roots to assume a rostral course rather than travel in a caudal direction. Mechanical distortion and an abnormal transmission of cerebrospinal fluid pressure in the preductal region may also induce CNS deformities. This may explain why so many of the children with ACM have strabismus and other ocular motility defects.\(^5\)

Hydrocephalus has been postulated as being directly responsible for ocular disorders. There are frequent cases of ACM without Hydrocephalus that show an abnormal transmission of CSF pressure which influences the midbrain and pretectal regions. This indicates that in the absence of hydrocephalus, local pulsations and hydrodynamic pressure changes at the level of the collicular cisterns occurs with the deformity, and may lead to disproportionate neurological deficits. Ventricular size and volume is probably not the most significant factor, but rather the local effect appears paramount.\(^10\)
Syringomyelia (SM) is a disorder in which an abnormal collection of fluid occurs in the spinal cord and is referred to as a syrinx. This fluid filled syrinx can expand and elongate over time destroying the center core of the spinal cord. Since the spinal cord connects the brain to the nerves in the extremities, this damage may result in pain, headaches, weakness, and stiffness in the back, shoulders, arms or legs. Other symptoms and signs of this disorder are often vague and may include weakness, fatigue, stiffness, or a disruption in body temperature with a loss of ability to feel extremes of hot or cold, especially in the hands and feet. SM may also adversely affect sweating, sexual function and bladder or bowel control.

Syringomyelia can be classified according to etiology.\textsuperscript{11}

1. Communicating Syringomyelia: Developmental anomalies at the foramen magnum and in the posterior fossa e.g. Arnold-Chiari I and II, Walker malformation, basilar invagination.
2. Acquired basal anomalies such as posterior fossa tumors, cysts.
3. Post traumatic syringomyelia as a late consequence of spinal injury.
4. Syringomyelia as a result of spinal arachnoiditis.
5. Syringomyelia associated with spinal cord tumors.
6. Idiopathic Syringomyelia.

The symptoms of SM can also occur as a result of a car accident or other trauma with ACM or spinal tumors.\textsuperscript{8} Frequently misdiagnosed as Multiple Sclerosis, muscular dystrophy, amyotrophic lateral sclerosis, cervical spondylosis and a variety of psychiatric diseases can create a confusing clinical picture.\textsuperscript{11}
Recent studies with MRI have shown that children with ACM II also suffer not only from hydrocephalus and spinal-cord lesions, but that they also have Meningomyelocele (MMC).\footnote{5}

Meningomyelocele is a major developmental malformation and the most severe type of spinal bifida which occurs in 2-5/1,000 live births in Great Britain. A cystic swelling formed by dura and arachnoid mater protrudes through a defect in the vertebral arches and this herniated sac contains spinal cord tissue or spinal nerve roots that are attached to the fundus of the sac. As a result there is usually a marked loss of motor and sensory function below the level of the lesion. Eighty-two percent of Meningomyeloceles are lumbar or sacral in site so that there is paralysis and anesthesia of the lower limbs. As a result there is usually a marked loss of motor and sensory function below the level of the lesion.\footnote{12}

**PHYSIOLOGY**

The mid-brain is primarily made of a large bundle of fibers called the cerebral peduncles. In these fibers lies the major motor pathway of the cortico-spinal tract. Traversing the midbrain is the cerebral aqueduct that connects the third and fourth ventricles. The midbrain also contains the nuclei of the oculomotor (VIII) and the cranial nerves.\footnote{13}

Inferior to the midbrain is the pons that functions as the conduction pathway for motor and sensory fibers from the cerebrum to the spinal cord. The nuclei of the trigeminal (V), abducens (VI), facial (VII) and vestibulocochlear (VIII) cranial nerves are located in the pons which makes it an important reflex center for cranial nerve function.\footnote{13}
On the basolateral surface of the pons are the middle cerebellar peduncles which provide extensive connections between the cerebral cortex and cerebellum. The nuclei of the trigeminal (V), abducens (VI), facial (VII) and vestibulocochlear (VIII) cranial nerves are located in the pons. Inferior to the pons is the most caudal brainstem structure, the medulla, which acts as a conduction pathway for motor fibers. Nuclei of the glossopharyngeal (IX), vagus (X), spinal accessory (XI), and hypoglossal (XII) cranial nerves are contained in the medulla, which also contains a group of neurons that influence respiration, vomiting and vasomotor responses. The medulla extends through the foramen magnum and gradually becomes the spinal cord.13

The cerebellum overlies the posterior aspect of the pons and medulla and extends laterally under the tentorium to fill the greater portion of the posterior fossa.14

The cerebellum is responsible for muscle synergy. It coordinates the action of muscle groups and times contractions so that movements are performed smoothly and accurately. It is also responsible for the control of the muscle tone and maintaining equilibrium. The fourth ventricle is posterior to the pons and medulla that extend from the central canal of the upper cervical spinal cord to the cerebral aqueduct of the midbrain. CSF flows from lateral ventricles through the foramen of Monro to the third ventricle. From the third ventricle, it travels through the cerebral aqueduct into the fourth ventricle where it can exit through one of the three foramina.13

Lesions of the upper brainstem might affect areas involved in the control of conjugate vertical eye movements. Structures in the upper brainstem are involved in vergence movements and vertical versional movements of the eyes.15
Lesions of the pons and lower brainstem also affect eye movements since the neural circuitry for conjugate horizontal movement is localized in these areas. Since the lower brainstem is more involved in control of horizontal eye movements, lesions in this area showed a better correlation between the defects of horizontal gaze and saccadic eye movements. The degree of hydrocephalus and less clearly to the extent of lower brainstem deformities were directly related to the amount of strabismus and spontaneous nystagmus. Ocular motility defects, with regard to oblique muscle functions, horizontal and vertical gaze and saccadic control, often correlated with lower brainstem lesions.\textsuperscript{5}

Abnormalities of eye movement in ACM are due to compression of the herniating cerebellum against the caudal brainstem and not a congenital aberrance of oculomotor pathways.\textsuperscript{9} The cerebellum is involved in control of saccadic and pursuit movements as well as fixation. Results suggest that horizontal convergent and divergent strabismus both manifest and latent nystagmus is not induced primarily by brainstem lesions but rather seems to be related to general CNS dysfunction in combination with hydrocephalus. Hydrocephalus may involve dilation of the third ventricle and cause damage to an upper part of the mesencephalon with the neuronal areas for control of vertical gaze.\textsuperscript{15,16,17,18} It has also been shown that MMC involves various developmental malformations, e.g. hypoplasia or aphasia of cranial-nerve nuclei, cerebellar dysphasia, thalamic deformations and agencies of the corpus callosum.\textsuperscript{19} These malformations may affect oculomotor development and also lead to oculomotor deficits commonly found in MMC.\textsuperscript{20,21,22}
OCULAR SIGNS

Neuro-ophthalmic signs and symptoms are important in the clinical diagnosis of ACM. Patients with advanced symptoms demonstrate oculomotor dysfunction, central vestibular nystagmus, abnormal vestibular visual interaction, and abnormal tilt suppression of post-rotatory nystagmus.\(^{23}\)

Nystagmus is generally the most common ocular presentation, particularly the down-beat variety with a fast down phase. The nystagmus frequently worsens with changes in head posture or changes in fixation. The presence or absence of oscillopsia can be used to make a distinction between an acquired and a congenital form of nystagmus. Patients who cannot compensate for the unstable eye movements and who perceive their visual world to be moving are more likely to have a developed nystagmus as opposed to patients with congenital nystagmus who perceive their visual world to be stationary.\(^{4}\)

Downbeat nystagmus (DBN) is pathological for a foramen magnum lesion. DBN enjoys a unique priority among the various forms of nystagmus because of its specific localizing value. Approximately 17% of ACM patients have DBN, 43% have some type of nystagmus.\(^{24}\) An acquired down-beat nystagmus is the classical finding although affected patients may reportedly have varying forms of nystagmus, skew deviations, VI nerve paresis, ocular dysmetria, ocular flutter, anisocoria, or spasm of the near reflex. DBN has been reported as the salient manifestation of ACM III. Down-beat nystagmus is reported as a common finding in acquired ACM without hydrocephalus.\(^{25}\) A common presenting feature of ACM type I is oscillopsia with nystagmus occurring in about 50%.\(^{1}\)
Pathological nystagmus degrades vision and causes illusory motion of the seen environment. These symptoms are due to excessive movement of images of stationary objects on the retina. The DBN patient may report symptoms as if the environment were jumping up and down. Oscillopsia patients present with visual disturbances that may run the risk of being unrecognized for some time as a result. Episodic downbeat nystagmus that is not associated with other ocular motor abnormalities due to cerebellar dysfunction can be an initial sign of a Chiari malformation.\textsuperscript{26}

Additional symptoms include motor reflex changes, oscillopsia, diplopia and blurred vision.\textsuperscript{4} Gait ataxia, dysarthria, and dysphagia are indicative of ACM/SM.\textsuperscript{23} Sixth nerve paresis, ocular dysmetria, ocular flutter are also common. The imbalance of the otolith-ocular reflexes, brought on by faulty communication between the vestibular neurons, the vertical sub nuclei of the oculomotor nucleus, and the interstitial nucleus results in vertical pendular eye movements.\textsuperscript{4}

Bisenman and Laguna reported that acquired, concomitant esotropia can be the presenting sign of type ACM I.\textsuperscript{25} Recent studies have shown that strabismus is also very common in these patients. The strabismus is seldom paralytic but mostly concomitant and commonly associated with and V patterns. Concomitant strabismus is common in MMC patients.\textsuperscript{18} Disturbances of convergence seem better correlated with lesions in these areas than in other brainstem regions.\textsuperscript{22} The mechanism is probably a brainstem compression since the convergent strabismus is combined with gaze-evoked nystagmus. Deformities of the tectal plate were common in patients with convergence defects and there was also a correlation with the clinically determined hernia level.\textsuperscript{5} No cases of paralytic strabismus were found. Strabismus and spontaneous nystagmus are
related mainly to convergence defects correlated with deformities of the upper brainstem. Such lesions could affect visual pathways, leading to defects of binocular fusion and subsequent development of strabismus. However, widening of the ventricles of the brain particularly the third ventricle, could affect oculomotor centers in the upper brainstem, particularly the vergence control areas located there. This in turn might lead to eso-deviation or exo-deviation of the eyes. The mechanism is probably a brainstem compression since the convergent strabismus is combined with gaze-evoked nystagmus. However, ocular motility disorders, including strabismus, were common, although no case of down-beat nystagmus was observed. Manifest strabismus was found in 11 of 28 ACM patients. Esotropia with or without A-pattern was seen in nine patients. Approximately 40 percent have manifest squint and 35 percent have latent strabismus. In normal population the prevalence of strabismus is 5 to 6 percent. Among manifest strabismus, the convergent type was more common. Most common single type of strabismus was A pattern esotropia.

The correlations were much stronger between concomitant strabismus and hydrocephalus than for any other combination of ocular motility problems and DNS deformities.

Visual disturbances were related to refractive errors and strabismus, but not to optic atrophy, and the most common neuro-ophthalmological abnormalities discovered are ocular motility abnormalities including strabismus. Oblique muscle overaction correlates with brainstem deformities which suggests an innervational rather than mechanical origin for such eye movement.
Superior oblique overaction is frequently found in patients with MMC, and MMC is uniformly associated with ACM type II which includes cerebellar and cervico-medullary region abnormalities. Overreaction of the superior oblique muscle is seen in A-pattern esotropia, and inferior oblique overaction in V pattern exotropia. Oblique muscle overaction correlated with the extent of changes is the tectal plate and medulla oblongata. The occurrence of strabismus in general correlated strongly with the degree of hydrocephalus.

Clinical evaluation of eye movements generally reveals that the saccade system is intact but smooth pursuits, optokinetic nystagmus and fixation suppression are all significantly affected.

Abnormalities of horizontal eye movements were more common than vertical eye-movements with slow saccadic movements. Rapid saccadic movements were absent in all directions. Gaze paresis restrictions of horizontal versional movements are noted in limitations equal to both sides and asymmetrical and strongest side for most patients. Loss of flocculus inhibition due to compression of the cerebellum frequently prevents fixation suppression, causing constant shifts of fixation as a patients eye tries to find a fixation point. Clinical evaluation of eye movements generally reveals that the saccade systems intact, but smooth pursuits, optokinetic nystagmus and fixation suppression are all significantly affected. Loss of flocculus inhibition due to compression of the cerebellum frequently prevents fixation suppression, causing constant shifts of fixation as the patients eye tries to find a fixation point.

All vertical gaze problems occurred when gazing upward and were most common in those who still had shunts and those with changes in the medulla oblongata region.
Progressive focusing difficulties were discovered in association with diplopia. Eighty percent up-gaze diplopia was noted and vertical gaze limitations were seen in upward gaze in 9 of 28.\textsuperscript{10}

A rise in intracranial pressure may cause papilledema leading to secondary optic atrophy or a dilation of the third ventricle, or a shift in the position of the brainstem giving rise to a primary optic atrophy from stretching of the optic nerves or optic chasm.\textsuperscript{12} No reduction of visual function was found that could be related to optic atrophy or other changes in the visual pathways. MRI is the imaging methods of choice for investigating patients with DBN.\textsuperscript{28}

Intermittent symptoms of intracranial pressure elevation is accompanied by mid peripheral retinal hemorrhages. Repeat computed tomographic scans and lumbar puncture showed normal result.\textsuperscript{29} Obstructive hydrocephalus may cause papilledema.\textsuperscript{10}

Type II malformations sometimes present with bilateral internuclear ophthalmoplegia (INO). INO is a manifestation of widespread brainstem or cerebellar dysfunction. Its origin is probably multifactorial, related to hydrocephalus, vascular compromise, direct neuronal distortion, or congenital neural malformation.\textsuperscript{24}

A 13-year-old girl who had Duane retraction syndrome associated with Chiari I malformation is reported. Midsagittal magnetic resonance imaging demonstrated cerebellar tonsillar herniation to 6mm below a line from the basion to opisthion. Taking into consideration the relative rarity of the two disorders, the association may not be coincidental. Referral for magnetic resonance imaging of the posterior fossa is recommended in Duane retraction syndrome.\textsuperscript{41}
SURVEY OF OCULAR SYMPTOMS

Considering many of the symptoms associated with both ACM and SM patients are visually related, a survey was conducted of ACM and/or SM patients to determine what symptoms these patients experienced. The purpose of the study is to determine the predominate symptoms experienced by ACM/SM patients, to provide healthcare providers with a list of the most frequently reported symptoms.

The survey was initially sent out on the internet to a group of subscribers to an ACM/SM mailing list. Once completed, the survey could be returned to us via e-mail or through the post office. Subjects could also request a copy of our survey to fill out and return via the postal service. The survey was also advertised in a newsletter that is distributed quarterly to ACM/SM patients and to physicians who care for ACM/SM patients. In that case, the readers could either request the survey by mail or over the internet. The participants were asked to indicate yes or no if they have ever experienced any of a list of symptoms, whether they occurred prior to or after diagnosis or surgery. Several of the respondents included as a side note that they had been frequently misdiagnosed several times prior to finally being told that they have either ACM, SM or both.

Of the 38 respondents, 17 have ACM alone, 18 have ACM and SM, and 3 have SM alone. Thirty-three of the respondents have ACM I, one has ACM II and one has ACM III. The mean age of the respondents is 39 with the range from 5-71 years old. The mean age of diagnosis was 33.5 years, with the range being birth-56 years. There were 5 males and 33 females respondents.
Table 2 illustrates the number and percentage of respondents who had experienced each of the symptoms. Headaches, blurred vision, and difficulty focusing on written material were the three most commonly reported symptoms. Eye movement problems, double vision and nystagmus were frequently reported.

**TABLE 2: Number and Percentage of 38 Respondents Reporting Symptoms**

<table>
<thead>
<tr>
<th>SYMPTOM REPORTED</th>
<th>NUMBER</th>
<th>PERCENTAGE</th>
</tr>
</thead>
<tbody>
<tr>
<td>HEADACHES</td>
<td>34</td>
<td>89.47</td>
</tr>
<tr>
<td>DIFFICULTY FOCUSING ON WRITTEN MATERIAL</td>
<td>31</td>
<td>81.58</td>
</tr>
<tr>
<td>BLURRED VISION</td>
<td>29</td>
<td>76.32</td>
</tr>
<tr>
<td>DIFFICULTY ADJUSTING VISION TO LIGHT CHANGES</td>
<td>24</td>
<td>63.16</td>
</tr>
<tr>
<td>ANISOMETROPIA</td>
<td>21</td>
<td>55.26</td>
</tr>
<tr>
<td>DIFFICULTY PERFORMING FINE EYE MOVEMENTS</td>
<td>21</td>
<td>55.26</td>
</tr>
<tr>
<td>DIFFICULTY TRACKING MOVING OBJECTS</td>
<td>20</td>
<td>52.63</td>
</tr>
<tr>
<td>PAIN ALONG EYE, EAR OR JAW ON THE SAME SIDE</td>
<td>19</td>
<td>50</td>
</tr>
<tr>
<td>DOUBLE VISION</td>
<td>18</td>
<td>47.37</td>
</tr>
<tr>
<td>EYES THAT BURN</td>
<td>17</td>
<td>44.74</td>
</tr>
<tr>
<td>NYSTAGMUS</td>
<td>17</td>
<td>44.74</td>
</tr>
<tr>
<td>UNEQUAL PUPIL SIZE</td>
<td>14</td>
<td>36.84</td>
</tr>
<tr>
<td>FOREIGN BODY SENSATION</td>
<td>13</td>
<td>34.21</td>
</tr>
<tr>
<td>HALOS AROUND LIGHTS</td>
<td>13</td>
<td>34.21</td>
</tr>
<tr>
<td>DRY EYES</td>
<td>12</td>
<td>31.58</td>
</tr>
<tr>
<td>EYE MOVEMENT PROBLEMS</td>
<td>12</td>
<td>31.58</td>
</tr>
<tr>
<td>VISION BLACKOUTS</td>
<td>11</td>
<td>28.95</td>
</tr>
<tr>
<td>EYELID THAT DROOPS</td>
<td>10</td>
<td>26.32</td>
</tr>
<tr>
<td>VISUAL FIELD LOSS</td>
<td>10</td>
<td>26.32</td>
</tr>
<tr>
<td>ITCHY EYES</td>
<td>9</td>
<td>23.68</td>
</tr>
<tr>
<td>OSCILLOPSIA</td>
<td>8</td>
<td>21.05</td>
</tr>
<tr>
<td>STRABISMUS</td>
<td>8</td>
<td>21.05</td>
</tr>
<tr>
<td>SUDDEN VISION LOSS</td>
<td>8</td>
<td>21.05</td>
</tr>
<tr>
<td>TINGLING SENSATIONS IN OR AROUND THE EYES</td>
<td>6</td>
<td>15.79</td>
</tr>
<tr>
<td>EXOPHTHALMOS</td>
<td>4</td>
<td>10.53</td>
</tr>
<tr>
<td>EYE MUSCLE PARALYSIS</td>
<td>4</td>
<td>10.53</td>
</tr>
<tr>
<td>INCOMPLETE LID CLOSURE</td>
<td>4</td>
<td>10.53</td>
</tr>
</tbody>
</table>

**OTHER SIGNS**

Refractive errors are common to ACM patients. The clinician must consider ACM I malformation in any patient with the most frequently associated non-visual symptoms which include hearing loss, headache, vertigo, ataxia, dysequilibrium,
dysphagia or cranial nerve symptom, especially if accompanied by the more classic symptoms of this disorder such as cervical pain or weakness.¹

ACM I is a cause of a variety of symptoms, and will be diagnosed even more frequently as the use of MRI increases. Unfortunately the condition is often not diagnosed by a neurologist until later in life. When syringomyelia is associated with ACM I, myelopathy and a sensory loss occur. It is characterized by weakness and atrophy of the upper extremities and loss of pain or temperature sensation around the shoulders and arms. Headache or pain in the cervical region is a common occurrence. It may increase with coughing or sneezing as alterations in cerebrospinal pressure occur. Painful occipital headaches can be elicited on coughing, loud yelling or laughing.⁴ Differential diagnoses for ACM include demyelination diseases, tumors or vascular disorders. Symptoms will generally worsen with time and may even be brought on during exercise or valsalva maneuvers. A correct diagnosis can lead to timely surgical intervention which can improve symptoms.⁴ A case study showed oscillopsia and nystagmus began in a woman two weeks after an inadvertent lumbar puncture during anesthesia for childbirth. Examination showed horizontal-torsional jerk nystagmus in all positions of gaze. Magnetic-search-coil oculography revealed accelerating slow phases, with an increase in nystagmus amplitude in darkness. MRI showed ACM I. Three months after occipital decompressive surgery, nystagmus had almost disappeared. Accelerating slow phases should not be considered diagnostic of congenital nystagmus, especially with an onset of oscillopsia in adult life. Imaging should be considered to exclude treatable hindbrain anomalies. Lumbar puncture in
patients with ACM may accentuate craniospinal pressure dissociation and precipitate neurological signs.\textsuperscript{30}

The signs and symptoms of ACM/SM are very diverse. Table 3 illustrates some of the more common signs and symptoms of ACM patients.

Table 3: Signs and Symptoms of Arnold-Chiari Malformation

<table>
<thead>
<tr>
<th>SYMPTOMS</th>
<th>SIGNS</th>
</tr>
</thead>
<tbody>
<tr>
<td>DEAFNESS</td>
<td>CENTRAL APNEA</td>
</tr>
<tr>
<td>EXTREMITY PAIN OR WEAKNESS</td>
<td>HYDROCEPHALUS</td>
</tr>
<tr>
<td>HEADACHES AND HEAD PAIN</td>
<td>LOWER CRANIAL NERVE PALSY</td>
</tr>
<tr>
<td>IMBALANCE</td>
<td>MENINGOMYEOLOCELE</td>
</tr>
<tr>
<td>PAIN SENSATION LOSS</td>
<td>SYRINGOMYELIA</td>
</tr>
<tr>
<td>SYNCOPE</td>
<td></td>
</tr>
<tr>
<td>TEMPERATURE SENSATION LOSS</td>
<td></td>
</tr>
<tr>
<td>TINNITUS</td>
<td></td>
</tr>
<tr>
<td>VERTIGO</td>
<td></td>
</tr>
</tbody>
</table>

Darlene Long’s ACM Symptoms List provides detailed descriptions of symptoms common to ACM patients. The most common symptoms include headache, syncope, eye movement problems, sensory losses, weakness, and cerebellar features such as ataxia.\textsuperscript{31} Dysphagia occurs in 5-15\% of patients, although only a few reports describe dysphagia as the only presenting symptom. Disordered esophageal motility and gastresophageal reflux are also reported.\textsuperscript{32}

The most frequently associated non-optometric symptoms include neural hearing loss, headache, vertigo, ataxia, dysequilibrium, dysphagia or cranial nerve symptom, especially if accompanied by more classic symptoms of this disorder, such as cervical pain or weakness.\textsuperscript{1}

In hydromyelia, scoliosis is the most common bony abnormality.\textsuperscript{11} Drop attacks, or sudden loss of body muscle tone with collapse to the ground is also common. Patients possibly have short necks.\textsuperscript{42}
In MMC, hiccups cause functional impairment of gag and cough. Respiration may also be a problem. Brainstem auditory responses may be decreased.

Among the protean manifestations of the Chiari type I malformation, headache is one of the major symptoms, reported to occur in up to 75% of patients. No headache type seems to be specific for the malformation. Best understood is the short-lasting cough headache due to the craniospinal pressure gradient created by the malformation. Longer cervicogenic-like headaches with occipital and neck pain, pain in the arm, restriction of neck movement and dizziness often accompanying the headache. Visual prodroma often accompanies the long lasting headaches to include visual field defects oscillopsia, diplopia, blurring macropsias, and skewed vision which can last anywhere from a few hours to several days.

Short lasting "cough headaches" that last less than five minutes, long lasting attacks from a few hours to several days and some with continuous attacks. Long lasting attacks were usually not precipitated by valsalvo-like maneuvers. A differential trace was performed and fluid was observed to surge up the spine in response to any sudden increase in abdominal thoracic pressure. This might be described as a pressure-dependent ballistic impulse. The human neuraxis has a large rostral capacitance, but only as small caudal capacitance. The caudal capacitance is affected by the external veins and by the elasticity of the dura. Valsalva's maneuver begins a fluid wave at the caudal end, which moves rostrally until it encounters a blockage. It may then rebound to produce an area of consternation, diffuse chaotic flow, which may be responsible for syrinx filling.
Headaches resembled cervicogenic headaches with occipital and neck pain, pain in the arm, restriction of neck movement, and dizziness often accompanying the headache. Recurrent syncope is a presenting symptom of ACM.  

Dizziness was the most distinguishing feature in the Chiari patients. Intermittent dizziness occurred with rotary vertigo unsteadiness. During headaches, visual field defects can be detected, as well as oscilloscopia, diplopia, blur, acropsias, metamorphopsia, and tinnitus. These features, together with a beneficial effect of surgical treatment in some patients, suggest a causal relationship between the malformation and headache. Functional impairment of gag and cough reflexes, respiration, brainstem auditory responses, and vocal cord paralysis may occur. It is important to recognize ACM as part of the differential diagnosis of balance disorders because patients may initially be seen with symptoms referable to the vestibular system, including ataxia, nystagmus, or vertigo.

SURGICAL PROCEDURES

Neurosurgical procedures can be grouped into six categories:

1. Decompression of the hindbrain malformation by suboccipital craniectomy and upper cervical laminectomy.
2. Laminectomy and syringostomy or myelotomy.
3. Terminal ventriculostomy.
4. Percutaneous aspiration of the syrinx.
5. European procedure posterior fossa reconstruction.
Nystagmus resolved more readily than pursuit or fixation suppression abnormalities, but most patients showed gradual improvement four months after operation. For patients with Chiari I malformation, there is an occlusion of the subarachnoid space at the foramen magnum. Before the dura was opened, the tonsils moved abruptly downward and the cervical portion of the syrinx constricted with each systole. After the dura was opened, but with the arachnoid still intact, there was no longer pulsatile motion of the spinal cord and downward movement of the tonsils ceased. In syringomyelia associated with hindbrain malformation, there is also a process of communication between the syrinx and the subarachnoid space. The therapeutic goal should be to alter this dynamic process to either prevent or collapse the syrinx.

Suboccipital craniectomy and upper cervical laminectomy was shown to completely relieve the visual disturbance of a patient with oscillopsia and DBN.

Posterior fossa and cervical decompression with dural grafting procedures, as well as various types of syringosubarachnoid shunts are used. Surgical treatment of the cyst has shown to end abnormal eye movements, however some nystagmus may still be observed in patients who have shunts.

SUMMARY

Optometric management of patients presenting with the symptoms presented, particularly downbeat nystagmus should be referred for a neurological consultation, with a recommendation to order an MRI. In patients that defer surgery, or remain symptomatic post-surgically, work to eliminate the patients visual complaints. It is important that the clinician treat any binocular problems that may be manifesting, including esotropia and hyperphorias. Educate the patient that their condition
progresses over time and can be exacerbated during periods of exercise or during a
valsalva maneuver. It is important to monitor the patients extra-ocular movements and
to frequently evaluate their optic nerve heads and monitor their visual fields as optic
atrophy and optic nerve head damage are potential complications.
REFERENCES


38. Stovner LJ. Headache Associated With the Chiari Type I Malformation. Headache 1993 Apr;175-181.


