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**Mirrored Bilateral Lens Zonule Colobomas in Identical Pediatric Twins**

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Disciplines
Optometry

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Mirrored Bilateral Lens Zonule Colobomas in Identical Pediatric Twins

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BACKGROUND
- Colobomas are the result of incomplete optic fissure and most often present inferior-nasally. 
  - Lens colobomas are an indentation of the lens periphery that occurs due to a localized absence of lens zonules or ciliary body.
  - The lack of tension of the developing lens capsule results in contraction and subsequent notching of the lens.
- Lens colobomas are rare: few unilateral cases reported, two bilateral cases. No cases of twins with lens colobomas in the literature.
- Implications on refractive status and accommodative amplitude

CASE HISTORY
- Monozygotic 12 year old male, Latino twins presented for comprehensive vision exams.
- Chief complaint: Headaches with prolonged near work.
- Medical history:
  - Premature birth: 27 weeks
  - Low birth weight: 2.0 lbs
  - Asthma
  - Mild reading disabilities - possibly dyslexia, reading at grade 1 level

PERTINENT FINDINGS
- Refractive Error (Table 1)
  - Twin A
    - OD +0.50 -0.75 x 011
    - OS PL -1.25 x 163
  - Twin B
    - OD +0.50 -1.50 x 173
    - OS +1.00 -5.75 x 170

  - Slit Lamp Examination (Figure 1)
    - Bilateral asymmetric lens zonule colobomas inferiorly in both twins
    - Twin A: OD>OS Twin B: OD<OS
    - Iris coloboma, all other anterior structures within normal limits
  - Binocular Vision Testing
    - Cover Test
      - Twin A: Far Ortho/Near 4 Exophoria
      - Twin B: Far 4 Exophoria/Near 6 Exophoria
    - Stereopsis with Randot 3
    - Local & Global absent in both twins
  - Accommodative Testing
    - Positive Relative Accommodation
      - Twin A: -1.00/-0.50
      - Twin B: +0.50/+1.50
    - Reduced accommodation
  - Dilated Fundus Examination (Figure 2)
    - Bilateral tortuous blood vessels in both twins
    - Intact retina 360 (○) posterior coloboma
    - Visual fields with screening Matrix N-30-5
    - Non-repeatable shallow scattered defects OU

DIAGNOSIS & DISCUSSION
- Lens Zonule Colobomas
  - Embryologically, colobomas occur due to an incomplete closure of the optic fissure which affects the developing optic cup.
  - The adult derivations of the optic cup include the optic disc, retina, ciliary body, and iris.
  - The lens itself is derived from the lens placode, and develops separately from the optic cup (Figure 3).
  - In this case, a coloboma of the ciliary body, and subsequently the lens zonules, caused an incomplete development of the lens.
  - Mild Meridional Amblyopia (Twin A: OD, Twin B: OS)
    - Refractive error not diagnosed early on, uncertain of age when refractive correction was initiated
    - Suspected Accommodative insufficiency
      - Possible correlation with high astigmatism
      - Accommodation was not tested monocularly
      - High Astigmatism
        - In both twins, the eye with the larger lens zonule coloboma was associated with higher astigmatism

TREATMENT & MANAGEMENT
- Updated Spectacle Rx released
- Near add was trialed with mixed responses and ultimately not prescribed
- Vision therapy recommended for accommodative insufficiency but deferred by parent at time of exam
- At next annual comprehensive exam
  - Repeat visual field
  - Corneal topographies may be useful in determining full extent of lenticular astigmatism for learning purposes

CONCLUSION
- Lens zonule colobomas
  - High genetic predisposition, as seen by its presentation in our twins
  - Possible correlation with high astigmatism
  - Zonular changes may be a concern for cataract surgery later in life.
  - Most often asymptomatic, but binocular dysfunction and amblyopia can occur if refractive errors are not diagnosed and treated early on.
  - Pre-senile cataract extraction surgery is additional treatment option.

REFERENCES
5) Remington, Lee Ann; Clinical Anatomy and Physiology of the Visual System; Chapter 7: Ocular Embryology, 2012.)