

Congenital Cataracts: Classification and Association With Anterior Segment Abnormalities

Manuel B. Datiles and J. Fielding Hejtmancik

National Eye Institute, National Institutes of Health, Bethesda, Maryland, United States; datilem@nei.nih.gov

The American Academy of Ophthalmology and Otolaryngology (later the American Academy of Ophthalmology/AAO) published a manual, titled "Cataract Types," by Frederick C. Cordes, MD, as part of its Continuing Education Program in 1961,¹ which served as a practical guide for the classification of congenital cataracts for generations of AAO members. It contained a complex and confusing scheme to describe myriad types of congenital cataract that was difficult to apply clinically. Jules Francois published in 1963 a monumental textbook on congenital cataracts,² which contained similar descriptions of myriad types of congenital cataracts. More recently, Amaya et al.,³ Reddy et al.,⁴ and Hejtmancik⁵ published excellent reviews on correlating the lens phenotype with known molecular genetic abnormalities of congenital and childhood cataracts. The lens phenotypes were quite varied and were categorized based on a combination of anatomic location, etiology, and descriptions of the shapes of the lenses, which were still complex and cumbersome to use in the clinic. In this issue of *IOVS*, Lin et al.⁶ propose a simplified clinical classification based on anatomic location with correlation to coexisting anterior segment abnormalities discovered using Scheimpflug imaging, performed under chloral hydrate sedation, of 598 eyes of 428 patients. These findings suggest that genetic and other factors that caused the cataracts may also affect the cornea, iris, and other anterior segment tissues. Hence a more thorough eye examination using newly available technology should be conducted to allow correlation of genetic abnormalities, not only to the lens but also to abnormalities of the cornea and iris. This is a pilot use of this classification system, and further development of a simple but comprehensive clinical classification system for congenital cataracts that is easy to use in practice should be undertaken. Perhaps it would be useful to have a Consensus Conference on this issue at some point.

References

1. Cordes F. *Cataract Types*. 1st ed. Rochester, MN: American Academy of Ophthalmology and Otolaryngology; 1961.
2. Francois J. *Congenital Cataracts*. 1st ed. Assen, The Netherlands: Charles C. Thomas; 1963.
3. Amaya L, Taylor D, Russel-Eggit I, et al. The morphology and natural history of childhood cataracts. *Surv Ophthalmol*. 2003;48:125-144.
4. Reddy MA, Francis PJ, Berry V, et al. Molecular genetic basis of inherited cataract and associated phenotypes. *Surv Ophthalmol*. 2004;49:300-315.
5. Hejtmancik JF. Congenital cataracts and their molecular genetics. *Semin Cell Dev Biol*. 2008;19:134-149.
6. Lin H, Lin D, Liu Z. A novel congenital cataract category system based on lens opacity locations and relevant anterior segment characteristics. *Invest Ophthalmol Vis Sci*. 2016;57:6389-6395.

