Clinical Classification of Childhood Glaucomas

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Objective: An updated classification of the primary and secondary childhood glaucomas is offered for clinical use, and associated systemic diseases are included to enable their early recognition in children with known glaucoma.

Methods: Approximately 650 clinical records of patients with pediatric glaucoma were reviewed for type of glaucoma and associated systemic disease. A literature search was done for additional reported causes of childhood glaucoma. Previous classifications of pediatric glaucomas were also reviewed. Pertinent references to support inclusion of each clinical entity in the updated classification are included.

Results: A comprehensive and referenced classification of the pediatric glaucomas was enabled by this review.

Conclusion: A comprehensive, etiologically based classification of the pediatric glaucomas is now available to assist with the recognition of the many causes of primary and secondary glaucoma in childhood and to support the selection of specific treatment choices.


The childhood glaucomas have been classified by the age of onset, inheritance, associated systemic findings, and anatomy, according to the associated and responsible anterior segment anomalies. In this article, we offer a comprehensive classification of the childhood glaucomas to assist in the recognition and differential diagnosis of the reported clinically recognizable causes of primary and secondary pediatric glaucomas (Table).

Terminology

Historically, the childhood glaucomas have been labeled developmental glaucomas based on the associated presence of developmental defects of the eye. Primary childhood glaucomas will be classified as those caused by anomalies of the filtration angle. These glaucomas are often of genetic origin and may be associated with systemic diseases and other ocular defects. We have identified the systemic diseases that have been described in association with childhood glaucoma.

Congenital glaucoma denotes a glaucoma that occurs early in life related to a congenital anomaly. Newborn primary congenital glaucoma defines a glaucoma entity that is recognized immediately at birth with the presence of profound defects of the anterior segment. Infantile primary congenital glaucoma includes patients with evidence of glaucoma most often recognized in the first year of life. Late recognized primary congenital glaucoma indicates an entity diagnosed significantly after an age when ophthalmologic examination of the patient would have recognized the presence of abnormalities related to glaucoma.

Juvenile glaucoma has been used to describe glaucoma in childhood. We have continued its use specifically with juvenile open-angle glaucoma that characteristically develops during childhood.

The secondary childhood glaucomas are those that occur as the result of independent disease mechanisms that secondarily impair the function of the filtration angle.

Comment

All classifications of the childhood glaucomas have revealed the impressive number of clinical entities that may feature or be complicated by childhood glaucoma. Previously these diseases have been variably identified and classified. The term association in reference to glaucoma with systemic diseases does not require that the glaucoma is an essential aspect of the disease. In our classification, we list these...
entities together; however, the strength of the glaucoma relationship might be different. Glaucoma may be coincidental or strongly genetically related to the systemic disease. In the future, additional clinical experience and genetic testing may establish the relative importance of these relationships.
The use of this classification as a clinical aid can facilitate early recognition of glaucoma and identification of the specific glaucoma diagnosis as well as meaningfully influence the choice of treatment. Without recognition of specific types of childhood glaucoma, it is more difficult for the clinician to appropriately select treatment that has been found to be most beneficial. The secondary glaucomas are an important and large group of childhood glaucomas. When confronted with a child with glaucoma and atypical clinical findings, the clinician may be assisted in making an accurate etiologic diagnosis by reviewing the tabulated causes of both the primary and secondary glaucomas. Finally, this childhood glaucoma classification can enable improved communication between those who care for these particular patients.

Submitted for Publication: August 21, 2009; final revision received August 21, 2009; accepted November 12, 2009.

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Author Contributions: The authors had full access to all of the data in the study and take responsibility for the integrity of the data and the accuracy of the analysis and presentation.

Financial Disclosure: None reported.
tified in this clinical situation. Justification for a randomized clinical trial usually includes the following: (1) evidence of a significant public health problem, (2) a scientifically plausible intervention, (3) preliminary information that warrants the trial, and (4) the ability to enroll a significant number of patients, with an outcome obtained within a reasonable time frame. Regarding the criteria above, we believe a randomized clinical trial is not justifiable because (1) while intravitreal injections are common, endophthalmitis following injections may not be, (2) the intervention has not previously been proven to be scientifically plausible, (3) there is not sufficient preliminary information from endophthalmitis rates following this intervention or even other interventions such as those that occur during cataract surgery (and risks of endophthalmitis following cataract surgery would not be expected to be the same as risks following an intravitreal injection of a drug using a 30 gauge needle), and (4) enrollment for between 1.5 million and 15 million injections, depending on outcome assumptions, could take so many years that the antibiotic and intravitreal procedure being tested may no longer be relevant.

We agree that it is important to determine the risk of endophthalmitis after an intravitreal drug injection when topical antibiotics are not required either before, during, or after the injection because there is little scientific rationale to support topical antibiotic use in this situation. As Drs Ziemsen and Bartz-Schmidt state, omitting its use would avoid the cost, potential toxicity, and burden to patients following millions of intravitreal injections each year. To address this situation in the absence of randomized clinical trials, which may not be justified, we hope to continue to provide information that is of potential value related to the millions of intravitreal injections currently provided around the world for common retinal conditions.

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Group Information: A published list of the Diabetic Retinopathy Clinical Research (DRCR) Network investigators and staff who participated in this protocol can be found in Ophthalmology. 2008;115(9):1447-1449, with a current list available at www.drcr.net.

Financial Disclosure: A complete list of all DRCR Network investigator financial disclosures can be found at www.drcr.net.

Funding/Support: This study was supported by a cooperative agreement from the National Eye Institute and the National Institute of Diabetes and Digestive and Kidney Diseases, National Institutes of Health, and grants EY14231, EY14229, and EY018817 from the US Department of Health and Human Services. Allergan Inc provided the triamcinolone, and Genentech Inc provided the ranibizumab. Both companies provided funds for part of the clinical care costs of the DRCR Network laser-ranibizumab-triamcinolone protocols. Allergan, Inc also has provided unrestricted funds to DRCR Network for its discretionary use. As per the DRCR Network Industry Collaboration Guidelines (www.drcr.net), the DRCR Network had complete control over the design of the protocol, ownership of the data, and all editorial content of presentations and publications related to the protocol.

Role of the Sponsors: The funding organization participated in oversight of the conduct of the study and review of the manuscript but not directly in the design of the study, the conduct of the study, data collection, data management, data analysis, interpretation of the data, or preparation of the manuscript.

Trial Registration: clinicaltrials.gov Identifier: NCT00444600 and NCT00445003


Corneal Thickness Changes in Very-High-Altitude Mountaineers

Boesch and colleagues performed a very nice study of corneal thickness at high altitude. However, some statements may benefit from clarification. Specifically, “Besides AMS [acute mountain sickness], corneal changes during high-altitude climbs may also be a dangerous hazard owing to a potential significant decrease in visual acuity. The often-quoted experience of Dr Beck Weathers, a Mount Everest climber who had undergone radial keratotomy prior to ascent and incurred severe vision loss during the climb, is such an example.” The authors correctly imply that corneal thickening plays a major role in radial keratotomy visual changes at high altitude. However, the mechanism of these changes is a bit more complicated and warrants a more precise explanation.

It is well known that any cornea thickens with exposure to hypoxia. However, when the normal corneal architecture is weakened by radial incisions, the hy-
poxic cornea remains clear but the incisions may allow swelling to occur in an anterior direction. This anterior elevation in the midperiphery causes central corneal flattening and a resultant hyperopic shift.\(^2,3\) Thus, alterations in corneal structure caused by radial keratotomy incisions, combined with increased corneal thickness, account for the induced refractive changes. Although plus lenses were required for clear vision, a climber with bilateral radial keratotomy successfully ascended Mount Everest (8 850 m).\(^4\) Similarly, owing to structural change from flap creation, corneas that receive Lasik and are exposed to hypoxia are thought to have a preferential central expansion, causing a slight myopic shift.\(^5\) This contrasts with normal and photorefractive keratectomy corneas that do not undergo refractive change with hypoxia because they thicken uniformly, thus preserving the shape of the anterior corneal surface.

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Financial Disclosure: None reported.


In reply

We welcome the comments regarding our article\(^1\) from Drs Mader and White, who have published several articles on refractive changes induced by hypoxia in subjects after refractive surgery. We value their additional account of the mechanism of the corneal changes in Dr Beck Weathers’ eyes during his climb on Mount Everest that led to severe vision loss. This is an interesting amendment to our manuscript, which focuses primarily on mountaineers with healthy corneas.

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Financial Disclosure: None reported.


Correction

Error in Table. In the Clinical Sciences article titled “Clinical Classification of Childhood Glaucomas” by Yeung and Walton, published in the June issue of the Archives (2010;128[6]:680-684), the Table was formatted incorrectly. In the right column, the subheading “Trabecular Meshwork Endothelialization” should not have been a subheading. It should be aligned below “Iris bombe with pupillary block” under “Angle-blockage mechanisms,” and there should have been no line above it. The only two column subheadings should have been “Primary (Developmental) Glaucomas” and “Secondary (Acquired) Glaucomas.”

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(CORRECTED)ARCH OPHTHALMOL/VOL 128 (NO. 9), SEP 2010 WWW.ARCHOPHTHALMOL.COM

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