s Fitted on Infant with Congenital Limbal Dermoid as Prevention of Refractive Amblyopia

Ivan Hare Dip. Optom., D.O.B.V., **Eli Savyon** F.I.D.O., **Adina Fried** B.Optom., **Judith Raz** MD., **Fanny Segev** MD.

Pediatric Ophthalmology Department, Meir Hospital, Kfar Sava, Israel.

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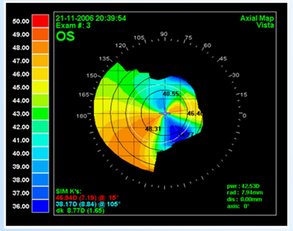


Corneal Topography depicting the high corneal astigmatism.

Pathophysiology

Limbal dermoids are benign congenital tumors containing choristomatous tissue (tissue cells atypical to the organ in which they are found). They appear most frequently at the inferior temporal quadrant of the corneal limbus. Limbal dermoids consist of thick collagenous tissue and may also contain elements of a variety of histologically aberrant tissues, including epidermal appendages, connective tissue, hairs, skin, fat, sweat gland, lacrimal gland, muscle, teeth, cartilage, bone vascular structures and neurological tissue including the brain. The surface generally consists of simple corneal or conjunctival epithelium.
Malignant degeneration is extremely rare.

Dermoids may represent:

- An isolated finding
- May be seen in conjunction with other ocular or oculosystemic disorders, including eyelid and iris colobomas, microphthalmos, and retinal or choroidal defects.
- 30% associated with systemic abnormalities (eg., Goldenhar syndrome) nevus flammeus and neurofibromatosis.

Most limbal dermoids represent sporadic occurrences and are not caused by known exposure to toxins or mechanical irritants.

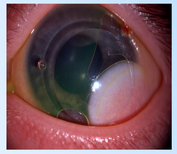
Frequency:
Internationally: The estimated worldwide incidence of limbal dermoids is 1 per 10,000 to 3 per 10,000

Mortality/Morbidity:
Visual morbidity may result from encroachment of the lesion into the visual axis, development of astigmatism, or formation of a lipid infiltration of the cornea, which obstructs the visual axis.
Large limbal dermoids can be cosmetically disfiguring.

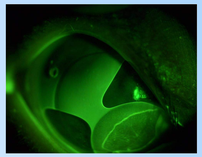
This case demonstrates the most common limbal dermoid, in which the tumor straddles the limbus. This is the most common dermoid, involving deeper ocular structures. The second most common type involves only the superficial cornea, sparring the limbus, the Descemet membrane and the endothelium. The third type of dermoid involves the entire anterior segment, replacing the cornea with a dermolipoma that may involve the iris, ciliary body and lens.

Treatment Program

Surgical removal by performing lamellar keratoplasty was considered. However, due to the young age of the infant and the risk of corneal perforation, it was decided that if a contact lens could be successfully worn for 6 to 8 months, this would be the treatment of choice and surgery re-considered at a later stage of development. Different scleral contact lens designs were manufactured until a satisfactory fit was achieved. Initially a scleral lens was fitted which presented with an excessive tear reservoir and air bubbles adjacent to the dermoid, as the lens pivoted on the dermoid apex.

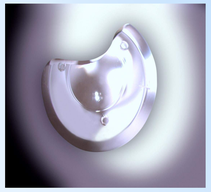


Scleral lens pivoting on the limbal dermoid, illustrating the large tear reservoir and air bubbles adjacent to the dermoid.



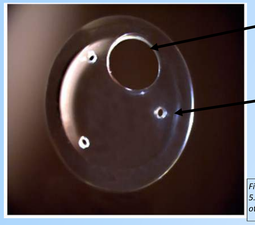
Fluorescein under lens shows large air bubbles adjacent to dermoid

Next, a scleral with a formed edge to fit adjacent to the Dermoid was manufactured. The lens on the eye had insufficient stability and continually rotated with one edge elevated onto the dermoid after blink. The lens would continually fall out after a few blinks.



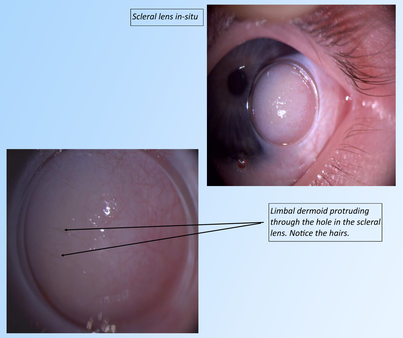
Scleral lens with edge ground away designed to fit adjacent to the dermoid.

Finally, a complete circular scleral lens was designed, 17.0mm in diameter, with a 5.5mm hole, situated in alignment with the position of the dermoid, equidistant between two fenestrations. The inner surfaces of the hole were well polished and blended so as not to cause insult to the dermoid. The lens, when on the eye would then lock onto the dermoid such that the dermoid would protrude through the hole. Three other fenestrations, which are standard in the production of a scleral lens of this type, were included in the lens design, to facilitate air ventilation and tear exchange under the lens.

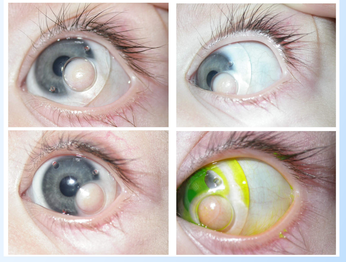


Final scleral lens demonstrating the 5.5mm hole and additional three other standard fenestrations.

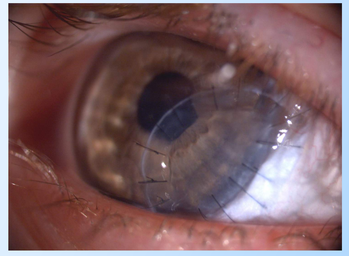
Scleral lens in-situ



All lenses were manufactured in Harmony material with a DK/t 50, (the highest DK/t available at that time), center thickness of 0.5mm by Gelflex Laboratories in Perth, Western Australia. The infant successfully wore the lens for all the mothers waking hours from the age of 2 months to 17 months, coupled with patch therapy of the right eye for half the hours the child was awake. The Retinoscopy over the lens was +1.00D sphere with no apparent astigmatism.
Weekly re-assessments of the corneal physiology were monitored and no adverse corneal insult was recorded throughout the 15 months of lens wear.
Partial lamellar keratoplasty was performed at the age of 17 months.
Cycloplaegic refraction 2 months post op:
OD: +1.50
OS: +3.00/-1.50 X 160
Spectacles were prescribed for both optical correction and protective purposes:
OD: Plano 6/6
OS: +1.50/-1.50 X 160 6/6-1



Scleral Lens in Situ

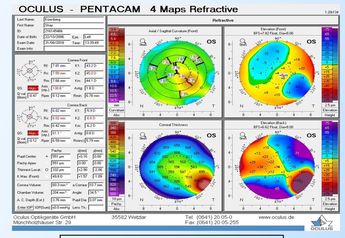


Partial lamellar keratoplasty performed at age 17 months.

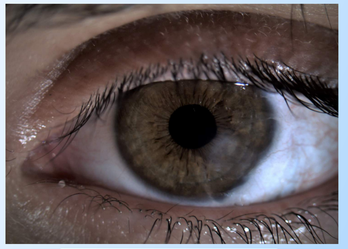
Post operative follow-up examinations were continually undertaken over the years until the patch therapy was discontinued due to improvement in visual acuity.

The child is 12 years old today and presented with 20/15 unaided acuity in this eye at his last recent follow-up examination in August 2018.

Current refraction:
OD: Plano
OS: Plano/-0.25 X 15



Recent Topography Maps



Recent corneal photograph depicting scar

Conclusion

A scleral contact lens may be worn in the presence of a limbal dermoid, compensating the refractive error, thereby preventing the development of amblyopia in a newborn infant. The lens enables the eye and cornea to grow and normal visual function to develop until the child reaches an age where he/she is old enough to undergo a partial lamellar keratoplasty, removing the dermoid.

Treatment of the dermoid in this manner allows the team sufficient time to arrange for surgery eliminating the danger of developmental refractive amblyopia.

References

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