Treatment of Epibulbar Limbal Dermoids

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Moderator: Rudolph S. Wagner, MD, Editor

Wagner: Treatment for many congenital defects or abnormalities of the cornea involves a combined approach with pediatric ophthalmology subspecialists and corneal specialists. I would like to discuss the case of a child with Goldenhar’s syndrome. This is a 3-year-old boy referred by a pediatrician because of the findings known to be associated with Goldenhar’s syndrome—recognizable epibulbar limbal dermoids.

When the patient was examined initially, he had bilateral hearing loss, which is associated with the syndrome, to the point that he needed to communicate with sign language. There was no significant family history.

Using a picture chart, we were able to get about 20/100 in each eye, although with the hearing impairment and the age of the child, he may have had better vision, but we could not prove it at that first visit. On examination he had bilateral epibulbar dermoids (Fig 1).

Much more involvement was noted in the right eye, with extension of the lesions centrally (Fig 2). In the left eye, the lesion was most marked inferiorly with a slight point where it was actually on the corneal surface inferiorly (Fig 3).

Elements of typical dermoids were noted and in Fig 4, one can see the small hair lashes extending from it.

Other critical information concerns the boy’s refraction: cycloplegic retinopathy in the right eye was +5.5 diopters (D) at 175° axis in the right eye, and the left eye was +8.25 D of cylinder at axis 165.

So, we have a 3-year-old boy who is seeing about 20/100 in both eyes. We are unsure if he could be refracted to better vision, but an initial attempt did nothing to improve his vision. The retinal examination was essentially normal: no abnormalities other than those noted were present.

Our dilemma, of course, is the degree of the astigmatism that he has. Should surgery be undertaken in one eye or both eyes to remove these dermoids? If so, what are the best tech-
niques to do this?

Medow: In my experience, dermoids cause a flattening of the meridian that involves the cornea. Even if you remove the dermoid, you do not remove the astigmatism. That astigmatism is always going to be there. He is probably amblyopic, but because he is 3 years old, our visual assessment may not be as accurate as we would like. He probably functions better than 20/100 because he probably sees better than 20/100, at least at near.

The dermoid causing the most astigmatism is in the right eye, and that would be the only eye I would do. The surgeon must be very careful not to remove the entire dermoid because it can be deep into the corner down to Descemet’s membrane, and you can perforate the anterior chamber.

I normally shave them by dissection. I used to take a graft with me to the operating room just in case, but I have become more conservative, and know I am not going into the anterior chamber.

Results have been disappointing from other than a cosmetic standpoint with that type of dermoid. It is unlike the dermoids in the center of the cornea or the dermoids that require corneal transplants.

Zaidman: Yes. The only indications for removal of a peripheral dermoid would be progressive astigmatism. Despite what parents say, most of us do not remove them for cosmesis. Also, if you superficially shave them off, you do not need a graft.

I have done corneal topography on two 4 year olds with dermoids. Both of them were able to cooperate. I determined that most of the cylinder on these children resulted from the dermoid. If topography proves that most of cylinder is of corneal origin, excision of the dermoid might be helpful.

But, in this case, you may not be able to get this child to cooperate for topography.

Wagner: We were able to use an autorefractor. We have a portable hand-held Nikon instrument, and I was able to get that reading.

How long does a corneal topography procedure take?

Zaidman: In an adult, about 15 seconds.

If I found 5 D of cylinder secondary to the dermoid, I would consider shaving it down. If you could decrease the cylinder, the chances of amblyopia would be less.

Kaufman: Yes. This particular dermoid is quite large, elevated, and keratinized, and may contribute an exposure problem—another reason to remove it. I, too, am conservative. I actually use a Vannas (Storz, St Louis, Mo) scissors rather than a rounded knife for the initial dissection, and follow that with a diamond burr for polishing. This technique ensures that I have not gone too deep, especially with a nearly full-thickness dermoid. This technique contours nicely and promotes healing.

Phillips: I agree. These are unique cases, and the basic consideration—especially with the corneal dermoid—is whether the dermoid is progressive. If it appears stable, an advancing lip line may identify the dermoid’s progress. A progressive dermoid must be removed.

Exposure does become a
problem if the dermoid becomes more elevated. You do not get wetting of the peripheral cornea, which eventually may cause dryness, ulceration, and scarring.

If the dermoid is stable, the issue then becomes one of removing it for cosmetic reasons. In my experience, parents want to do something because the cosmesis is not attractive, especially when the kids go to school.

I do remove them for cosmesis, but with a great deal of information—topography, if I can get it, to know whether the cylinder is regular or irregular. Just as with pediatric grafts, I like stable, supportive parents who are going to help with follow-up and who understand that getting to a clear cornea is not the goal.

Our goal is to do a superficial keratectomy and try to make the peripheral topography as normal as possible. And for that to happen, we have to know beforehand if the dermoid extends back into the orbit.

A number of papers1-3 discuss the surgical removal of dermoids, a procedure that is not terribly complicated as long as you can be reasonably certain they do not extend into the cornea. We perform an examination under anesthesia prior to surgery with a gonio lens to ensure the absence of a dome-shaped convexity extending into anterior chamber.

If you see [the convexity] and begin to debulk the dermoid with sclerectomy or keratectomy, it is nice to have a piece of tissue there—either corneal or scleral—to do a patch graft in the event of a perforation, which although unusual, does happen.

Another option is a corneal tattoo for the peripheral corneal scar when parents are not satisfied with the cosmetic result.

Wagner: How do you use the corneal tattoo and what do you use for that?

Phillips: I have used two different methods. I have the tattoo machine used by the ocular plastic surgeons to apply permanent lid liner. The pigment is actually tattoo pigment, which is mixed in various ways.

In the past, I did not tattoo eyes with vision. I worried about pigment toxicity, which has been addressed in the literature. For the most part, however, if the parents are dissatisfied with the cosmetic result and a tinted contact lens does not work, the tattoo pigment has been relatively safe and fairly inert.

When I do not want to use the tattoo machine, I apply the pigment manually with a blade and mix it in the operating room. The pigment comes in different colors, and you want to try to match the iris one eye to the other. A medical illustrator comes to the operating room to help me get the color right.

Tattooing is helpful to darken a really leukomatous scar for psychosocial reasons.

If you are going to do this for cosmetic reasons, you need to go ahead and try to make it look nice if you can. It is an available modality to consider.

Medow: From an historic standpoint, you may find a tattoo needle that was available throughout the ’20s, ’30s, and ’40s in ophthalmic kits. It is multiprunged and you can use that to apply the stain in a non-vibrating manner.

Phillips: I actually have one and it works well. I order pigment from a tattoo company and, amazingly, the company representative is excited about what he thought was a new indication for tattoo pigment.

Using a blade to impregnate the pigment into the anterior stroma also works.

Wagner: Could we expect a reduction in the astigmatism in this case if we were able to do more than a superficial keratectomy?

Medow: Although I have performed surgery on three or four patients, I have never substantially decreased the amount of astigmatism. These dermoids are in the cornea, which
is being drawn taut; that is the flattest meridian. You cannot correct it unless you could do a relaxing incision in the cornea's periphery and proceed as you would with a cicatricial ectropion: put in an element of sclera perhaps, if you could be that creative, and relax the meridian that is so flat.

For all the reasons mentioned, considering doing it is certainly appropriate.

Kaufman: There are two possible components of the astigmatism. The first is a congenital structural component, which would not be removed by shaving the dermoid. The second is a tractional component similar to a pterygium.

Topography would be valuable, and may help differentiate the two types of astigmatic effect. Similar to pterygium excision, removing the dermoid may reduce the irregular configuration.

Phillips: That is an excellent analogy because the steep axis is 90° away from the longitudinal growth of the pterygium. Even 0.5 or 1 D of astigmatism induced by pterygium is usually irregular. On topography, those patients get a greater improvement in their vision than you would expect based on the quantity of the cylinder.

You just do not know if the child is bilaterally amblyopic or if that cylinder will remain stable. Perhaps you could make a case that its progression might lessen. But it is difficult to know at this child's age—the refraction, the responses, what you will be doing exactly. In my cases, the cylinder just sits there. In an adult, you might get some comment as to the qualitative improvement in the vision, but a child cannot offer that information.

Medow: I have not done this but have thought about it: Pinhole these kids with pilocarpine and see how they function. If they have a large degree of compound or irregular astigmatism, and you get them to really be pinholed, they might see much better, have a greater depth of focus, and a good axial ray of light.

I considered putting pilocarpine in and patching the other eye, treating it amblyopically. I would not be changing the eye, but I might be helping the amblyopia. The refraction would not change, the corneal component would be decreased to the smallest axial raise that we could get. But, I have not had enough courage to try it.

Wagner: What if the lesion were removed and you got significant flattening of the cornea but continued with a residual astigmatism? Would a contact lens help correct some astigmatism?

Zaidman: If the child is cooperative, I do not see why you could not put a contact lens on. You would have to use a hard or RGP contact lens. Success depends on both parental and patient cooperation.

You do not know how deep and extensive a dermoid is until you try to remove it. If it is fairly superficial, you might get some decrease in the astigmatism. After removal you may be able to fit a contact lens over the eye.

Kaufman: If the child can tolerate a contact lens, use one. But, conjunctivalization may occur because of the lack of stem cells in the region where the dermoid was. This conjunctivalization may affect the child's ability to tolerate a contact lens. With a large dermoid and a large area of stem cell defect, a pseudopterygium or a conjunctival ingrowth may occur, preventing a contact lens from fitting well.

Significant flattening with a deep dermoid also may occur in the specific site where the dermoid was removed. This flattening also may adversely affect the fit of the contact lens.

Zaidman: You could customize a hard contact lens to 7 or 7.5 mm. I suspect that if you excised the dermoid even with conjunctival ingrowth, you could probably still get a small hard contact lens on the eye.

Medow: You could also use a soft lens with a hard lens car-
rier. In other words, a large soft lens to cover 11 or 13 mm with a hard lens in the center. This makes the surface of the cornea smoother so the hard lens sits better.

Wagner: What happens to an adult with a residual astigmatism who has this procedure? Is there a role for a laser corneal procedure to correct that type of astigmatism?

Zaidman: We would have difficulty getting a suction ring on to do [laser in situ keratomileusis] LASIK in an eye with a big dermoid like that.

Wagner: Following removal of the dermoid?

Zaidman: Well, you would have to remove a great deal of the dermoid over the sclera and you might thin the sclera.

For astigmatism, [photorefractive astigmatic keratectomy] PARK might be possible, depending on the amount of clear cornea after the excision. You would need a 6-mm clear axis to do PARK.

If you try LASIK, once suction was present, the temporal position of the dermoid would create additional, possibly insurmountable technical problems.

Kaufman: In an adult, the key component after the eye had fully healed would be topography. With today's technology, the excimer laser cannot treat irregular astigmatism. Perhaps in the future, with improved scanning technology and coordination with topographical analysis, irregular astigmatism will be treatable.

If this were an adult, perhaps astigmatic keratotomy could be performed at the axis of the irregular astigmatism in an effort to make it more regular.

Phillips: Those are both excellent comments. As I mentioned earlier, the key point is whether astigmatism is regular or irregular. That fact alone will determine whether you might do a LASIK or similar procedure.

If you removed the growth and you had some improvement in cylinder and cosmesis, you may leave well enough alone. Although tempting, the cylinder is there and the child is amblyopic. These are case-by-case decisions. It is hard to generalize and be dogmatic.

That brings up another point. When you remove these, how far posteriorly do you want to go and how lucky are you feeling that day?

My first goal is to use a diamond burr to smooth the cornea.

Then, if the goal is cosmesis, debulk the lesion as much as possible, at least to the point where it is not visible from a conversational distance.

I make a conjunctival incision and debulk the dermoid. The vacuum ring then would have a much better chance of sealing.

First, define your goal. Then try to abide by the initial plan and not go beyond that point.

Medow: As ophthalmologists, we should consider this surgery from a psychosocial standpoint rather than from a cosmetic one.

I believe, especially with children, that this rather large, ugly defect should be corrected for psychosocial reasons.

Wagner: I agree. In Dr. Kraft's article in the Canadian Journal of Ophthalmology, when discussing strabismus surgery, he used the term "reconstructive." He used the term technically, because "cosmetic" surgery means to alter something that is essentially normal in structure. Reconstructive surgery, on the other hand, attempts to improve a condition or structure that is abnormal.

Looking at ophthalmic surgery in that way, many procedures are reconstructive, rather than what we would have called cosmetic.

REFERENCES