ABSTRACT
A 2-year-old boy with Goldenhar syndrome had a limbal dermoid removed and covered with a lamellar corneoscleral patch graft that was attached with fibrin glue and no sutures. The graft healed and attached well. A sutureless technique is beneficial due to decreased scarring and chance of infection. [J Pediatr Ophthalmol Strabismus. 2016;53:e22-e25.]

INTRODUCTION
Goldenhar syndrome, also known as oculo-auriculo-vertebral spectrum, is a disorder that involves developmental abnormalities of structures derived from the first and second pharyngeal arches. It has a heterogeneous presentation and can be prevalent in up to 1 in 3,500 births. The abnormalities generally affect the ear (ear tags, hearing loss), face (hemifacial macrosomia), eyes (dermoids, microphthalmia, colobomas), and vertebral column. Syndrome severity can range from mild to severe; symptoms usually only affect one side but can be bilateral in some cases. Goldenhar syndrome is usually sporadic, but there is also data to suggest genetic transmission. The most common ocular abnormality is an epibulbar dermoid, which can be found in 4% to 35% of patients with Goldenhar syndrome. We present a case of limbal dermoid removal using a novel technique involving a sutureless lamellar corneoscleral patch graft with fibrin sealant.

CASE REPORT
Exemption was granted by the institutional review board and HIPAA regulations were followed. A 2-year-old boy with Goldenhar syndrome presented to an ophthalmology clinic with a large inferotemporal growth on his left eye that approached the central visual axis. The growth was opaque, flesh-colored, and measured approximately 6 mm in height by 7 mm in width (Figure 1A). It appeared that significant astigmatism was being induced by the lesion. Retinoscopy showed a refraction of +3.50 diopters (D) sphere +3.50 D cylinder at an axis of 045° on the left eye. His right eye refraction was +3.00 D sphere, with no cylinder. With the growth approaching the center of the visual axis, the decision was made to remove the growth to prevent amblyopia.

Prior to surgery, ultrasound biomicroscopy was performed under anesthesia to show the depth of the lesion (Figure 1B). The ultrasound showed the lesion extending into the corneal stroma with the possibility of a full-thickness lesion, but analysis was limited by shadowing behind the lesion. The lesion was 9 mm in diameter measured by calipers. Subsequently, a guarded diamond blade was used to demarcate the corneal border of the dermoid and enable start of dissection (Figure 1C).

Lamellar keratectomy was performed to remove the entire depth of the dermoid (Figure 1D). Removal of the dermoid resulted in a corneal micropерforation due to the extent of the lesion, which was sealed intraoperatively with hydrogel sealant (ReSure; Ocular Therapeutix, Inc., Bedford, MA) and air in the anterior chamber. The donor corneal button was then hand-cut into a 250-micron anterior lamella using an artificial anterior chamber, a guarded diamond blade, and a crescent blade. The
defect left by the dermoid removal was traced onto a transparent dressing (Tegaderm; 3M, Two Harbors, MN) and was then used to mark the borders of the lamellar corneoscleral patch graft, which was prepared from corneoscleral donor tissue mounted on an artificial anterior chamber. The donor tissue was trimmed to match the area from which the dermoid was removed and the graft was attached with fibrin glue (Tisseel; Baxter International, Inc., Deerfield, IL) and no sutures (Figure 1E). Multi-layer amniotic membrane was used to cover the graft and exposed sclera and was secured with 8-0 sutures (Vicryl;
Ethicon, Inc., Somerville, NJ) and hydrogel sealant. Absorbable sutures were used for the amniotic membrane to maximize the presence of the graft.

One day after surgery, the graft and amniotic membrane appeared to be holding well. The patient was given moxifloxacin and prednisolone topical drops four times a day and eight times a day, respectively. Oral vitamin C 15 mg per day was also prescribed. Three weeks after surgery, 3+ edema was noted within the graft on slit-lamp examination, but the graft borders were still holding well and in good position. To relieve the edema, sodium chloride and albumin topical drops were given four times a day. Prednisolone topical drops were decreased to six times a day. Moxifloxacin and vitamin C were continued. Five weeks after surgery, the graft edema had resolved and the graft was still holding. Moxifloxacin was stopped, prednisolone was decreased to four times a day, and the remaining interventions were continued. At 10 weeks after surgery, the graft appeared to be integrating well, with some remaining haze but no edema (Figure 1F). At 3 months after surgery, retinoscopy showed a refraction of -1.00 D sphere, +6.50 D cylinder at an axis of 050 on the left eye.

DISCUSSION

Limbal dermoids are generally classified into three grades. Grade I dermoids are superficial with a size less than 5 mm. Grade II dermoids are larger (cover most of cornea) and extend into the corneal stroma without involving Descemet’s membrane. Grade III dermoids cover the entire cornea and extend into the anterior chamber. Our patient appeared to have a Grade II limbal dermoid, with a size greater than 5 mm and extending into the corneal stroma.

Indications for surgery depend on the grade of the lesion. Grade II and III limbal dermoids require surgery because they can cause refractive or occlusive amblyopia. Grade I limbal dermoids can be managed medically with spectacles, occlusive therapy, and close monitoring to prevent amblyopia. If amblyopia occurs despite medical management, then surgery is indicated. Other indications include chronic eye irritation or recurrent conjunctivitis, progressive dellen, growth approaching pupillary area, aesthetics, inadequate eyelid closure, and induction of irregular astigmatism.

Surgical technique varies depending on the depth, size, and location of the limbal dermoid, and can include superficial keratectomy, simple excision, lamellar keratoplasty, and penetrating keratoplasty. Nevares et al. suggested superficial keratectomy after analyzing 50 patients with ocular dermoids, but noted that an opacification would remain in the cornea. More recently, Cha et al. used superficial keratectomy with corneal tattooing to achieve a good cosmetic result and reported no complications.

Panton and Sugar analyzed simple excision of limbal dermoids. Robb analyzed no complications but Panton and Sugar reported persistent epithelial defects and peripheral corneal neovascularization and opacity in some patients. More recently, new techniques have been developed to augment simple excision and decrease complications. Lazzaro and Coe added a pericardial patch graft, Hong et al. added an autologous limbal stem cell transplant, Pirouzian et al. added a sutureless amniotic membrane transplant, and Lang et al. added mitomycin C. All of these authors reported having no complications such as persistent epithelial defects, corneal vascularization, pseudopterygium, and scar formation.

Mader and Stulting described lamellar keratoplasty as a method of removing limbal dermoids. Since then, Scott and Tan, Panda et al., Watts et al., and Shen et al. retrospectively analyzed cases using the lamellar keratoplasty technique. Complications included microperforations during dermoid excision, opacification of the graft, haze in the graft, graft rejection, interface neovascularization, steroid-induced glaucoma, and reepithelialization. The increased number of complications with lamellar keratoplasty in comparison to simple excision and superficial keratectomy is likely related to the grade of the limbal dermoids that were operated on. Grade I limbal dermoids are more likely to be successfully treated without a graft, and thus have fewer complications.

The histology from our patient’s tissue shows stratified squamous epithelium with underlying dense irregular connective tissue and some adipose tissue. Multiple hair shafts and follicles and eccrine sweat glands were observed.

We were able to demonstrate successful removal of a limbal dermoid using a lamellar corneoscleral patch graft technique with fibrin glue instead of sutures to attach the graft. To our knowledge, this is the first reported case of a pediatric limbal dermoid removal with a sutureless attachment of the graft with fibrin glue. With no sutures on the cornea,
there is less scarring and less chance of infection at the suture sites. If there is a documented increase in size or depth of the lesion over time, we suggest early removal of the dermoid to avoid complications, such as microperforation seen in this case because the lesion had grown deep into the corneal stroma. In this case, the parents reported the lesion increasing in size. Earlier intervention may also decrease the size of the graft necessary or avoid a graft altogether and can prevent progression to refractive or occlusive amblyopia.

REFERENCES