Glaucoma Drainage Devices

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Introduction

The treatment of primary congenital glaucoma (PCG) and the many secondary forms of glaucoma in childhood was revolutionized in the 1940s with the introduction of targeted surgery of the anterior chamber angle: goniotomy *ab-interno* [1] and trabeculotomy *ab-externo* [2, 3], both of which are discussed elsewhere in this book. Retrospective studies of both trabeculotomy and goniotomy in patients with PCG demonstrate success rates as high as 75–90%. However, even in the best of hands, some 20% or more of primary angle surgeries eventually fail, due to the underlying structural defect, the severity of the glaucoma at presentation, or the underlying diagnosis. Secondary glaucomas presenting in infancy such as aniridia, Axenfeld-Rieger anomaly, Peters anomaly, and glaucoma following cataract surgery (GFCS) often respond poorly if at all to primary angle surgery. When angle procedures like goniotomy or trabeculotomy fail, cannot be performed due to abnormal anatomy, or are felt unlikely to succeed based on the underlying presentation, surgeons are then confronted with choosing an alternative. One increasingly attractive option is the implantation of a glaucoma drainage device (GDD). This chapter will review the current status of GDDs in the management of childhood glaucoma including general principles of these devices, surgical techniques, and a review of the current pediatric GDD literature. We hope to provide useful guidance to surgeons confronting this clinical challenge.

All GDDs share a common design – they employ a biocompatible silicone tube placed in the anterior chamber (AC) or vitrectomized posterior chamber in order to shunt aqueous humor to the subconjunctival space [4]. This potential space between the sclera and the overlying Tenon capsule and conjunctiva is then maintained by an external biocompatible "plate" made of silicone or acrylic which varies in surface area. Once healing has occurred, the IOP-lowering effect of a GDD is roughly proportional to the inner surface area of the capsule surrounding the plate [5, 6].

History

The first purpose-designed GDD was that of Molteno [7], introduced in the early 1970s. In the first iteration of his procedure, the circular acrylic

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plate was sutured to the equatorial sclera and the tube tucked out of the way for subsequent retrieval (Stage 1). Some weeks later, after a fibrous capsule had formed around the plate, the tube was then retrieved (without violating the plate capsule) and inserted into the eye to drain aqueous humor (Stage 2). Molteno subsequently described the use of a Vicryl® (polyglactin) (Ethicon Somerville NJ, USA) tie to temporarily occlude the tube long enough for a capsule to develop around the equatorial implant [8], thus avoiding a two-stage procedure. In either case, glaucoma medications are used to lower intraocular pressure (IOP) until the device is fully functioning some 6 to 8 weeks later.

Currently available GDDs are listed in Table 7.1 and can broadly be categorized based by whether they are valved (e.g., Ahmed glaucoma valve) or non-valved designs (e.g., Molteno™, Baerveldt®, and Aurolab aqueous drainage implant devices) and then further categorized by plate surface area. Non-valved implants must be temporarily occluded to prevent early hypotony until sufficient fibrosis has developed around the plate to prevent hypotony; valved devices allow flow immediately after surgical implantation.

Considerations for Surgeons Experienced with GDD Surgery in Adults

It's often stated that children are not simply little adults, and certainly their eyes don't behave like little adult eyes. Glaucoma surgeons experienced in placing GDD in adult eyes must modify their usual surgical technique for the pediatric eye. Covered in more detail in this chapter, the following is a partial listing of things to note when tackling these cases:

- *Consider ocular size when choosing a glaucoma implant.* An adult-sized implant can usually be placed in a buphthalmic eye, but in nanophthalmic or microphthalmic eyes, a shorter implant must be chosen to avoid impinging on the optic nerve.
- *The sclera of buphthalmic eyes is very thin.* Suture passes can easily perforate the sclera leading to a retinal detachment. A longer, shallow pass may be necessary to adequately secure the implant in place.
- *Ocular growth must be accounted for in selecting tube position and length.* Around 3 millimeters of tube length must remain in the AC to accommodate for progressive buphthalmos or normal ocular growth in young children. Tube retraction in the growing eye is a late complication that can generally be avoided.
- *Forward tube movement is common.* In the pediatric eye, tubes tend to straighten out over time and will erode through overlying sclera and peripheral cornea. It is generally best to position AC tube entry as posterior as possible away from the cornea (sometimes through a surgical iridectomy) to avoid late corneal complications.
- *Pars plana placement may be considered in aphakic and pseudophakic eyes.* In children, placement in the pars plana must be accompanied by a meticulous and thorough vitrectomy. Late occlusion with vitreous remnants and retinal detachment are common (~ 20%) [9].

Specific Glaucoma Drainage Devices

Molteno™ Glaucoma Drainage Devices

The Molteno drainage implant was the pioneering GDD first described in 1969 [7]. It provided the foundation on which all of the currently available GDDs are based. The Molteno™ implant is a non-valved device con-

Table 7.1 Currently marketed glaucoma drainage devices

Name	Type	Model	Materials	Valve/drainage mechanism	Plate surface area (mm^2)	Manufacturer	FDA/CE approval status	Website
Ahmed glaucoma valve	Valved	FP7 (flexible plate)	Medical-grade silicone	Elastomer membrane	184	New World Medical	FDA and CE	http://www.newworldmedical.com/product-fp7
		FP8 (flexible plate – pediatric)	Medical-grade silicone	Elastomer membrane	102	New World Medical	FDA and CE	http://www.newworldmedical.com/product-fp8
		S2	Medical-grade polypropylene	Elastomer membrane	184	New World Medical	FDA and CE	http://www.newworldmedical.com/product-s2
		S3 (pediatric)	Medical-grade polypropylene	Elastomer membrane	85	New World Medical	FDA and CE	http://www.newworldmedical.com/product-s3
Baerveldt® glaucoma implant	Non-valved	BG 103–250 glaucoma implant	Medical-grade silicone	Open tube	250	Johnson & Johnson Vision	FDA and CE	https://surgical.jnjvision.com/us/iols/other/baerveldt-glaucoma-implants
		BG 101–350 glaucoma implant	Medical-grade silicone	Open tube	350	Johnson & Johnson Vision	FDA and CE	
		Pars plana BG 102–350 glaucoma implant	Medical-grade silicone	Open tube with Hoffman elbow	350	Johnson & Johnson Vision	FDA and CE	
Molteno™	Non-valved	Molteno™ SS	Medical-grade acrylic	Open tube	185	Molteno Ophthalmic	FDA and CE	https://www.molteno.com/molteno-glaucoma-drainage-devices
		Molteno™ SL	Medical-grade acrylic	Open tube	245	Molteno Ophthalmic	FDA and CE	
		Pediatric/microphthalmic P1	Medical-grade acrylic	Open tube	80	Molteno Ophthalmic	FDA and CE	
Aurolab aqueous drainage implant (AADI)	Non-valved		Silicone	Open tube	350	Aurolab (a manufacturing division of Aravind Eye Institute, Madurai, India)	CE	http://www.aurolab.com/glaucoma-shunt.asp

FDA US Food and Drug Administration, *CE* European Commission

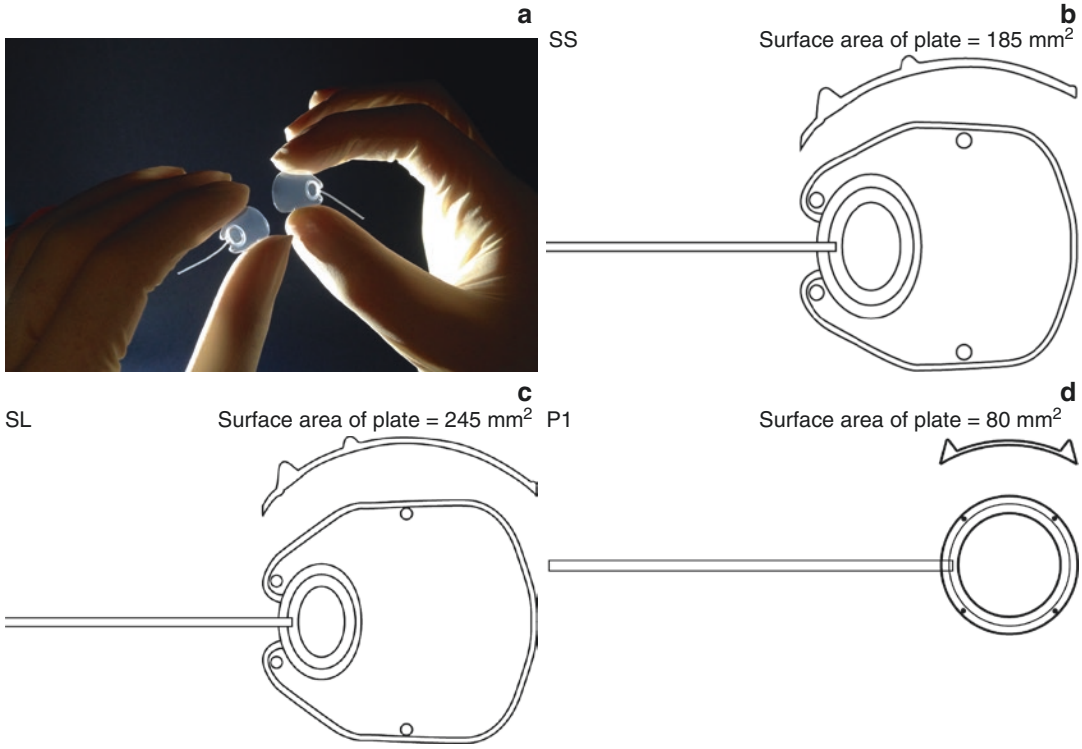

Fig. 7.1 (**a**) The Molteno3™ S-Series of glaucoma drainage devices; the model SS (right) has a surface area of 185 mm^2, the model SL (left) has a surface area of 245 mm^2; (**b**) diagram of model SS; (**c**) diagram of model SL; (**d**) diagram of model P1. Model P1 is designed for very small pediatric or nanophthalmic eyes and has a surface area of 80 mm^2. (Courtesy of Molteno Ophthalmic Ltd., Dunedin, New Zealand)

sisting of a silicone tube attached to an end plate placed 9–10 mm posterior to the limbus within the subconjunctival space. The plate is sutured to the sclera and covered by a thick flap of Tenon tissue and conjunctiva. A permeable fibrovascular bleb forms over the plate, the surface area of which contributes to the amount of aqueous drainage and the final level of IOP [10] along with the thickness of the bleb capsule.

The original Molteno™ implant consisted of a single 13 mm diameter plate molded from acrylic with a surface area of 135 mm^2. The single plate is inserted between two rectus muscles in the chosen quadrant. The double-plate Molteno™ implant was introduced in 1981 and consists of two plates, one of which is attached to the silicone tube in the AC, while a second tube connects the two plates forming a total surface area of 270 mm^2.

Currently marketed (Molteno Ophthalmic, Ltd., Dunedin, New Zealand) Molteno™ implants (Fig. 7.1) are the Molteno3™ S-Series, the SS (185 mm^2), and SL (245 mm^2) models, both designed for single-quadrant placement, and the Molteno™ P1 (80 mm^2) designed for implantation in eyes with axial lengths shorter than 17 mm.

Baerveldt® Glaucoma Drainage Devices

Introduced in 1990, the Baerveldt® glaucoma implant (BGI) is a non-valved device with a silicone tube attached to one of two sizes of external silicone plate (250 and 350 mm^2). The company (Johnson & Johnson Vision, Santa Ana CA, USA) also markets a 350 mm^2 version for implantation in the pars plana with the tube specially

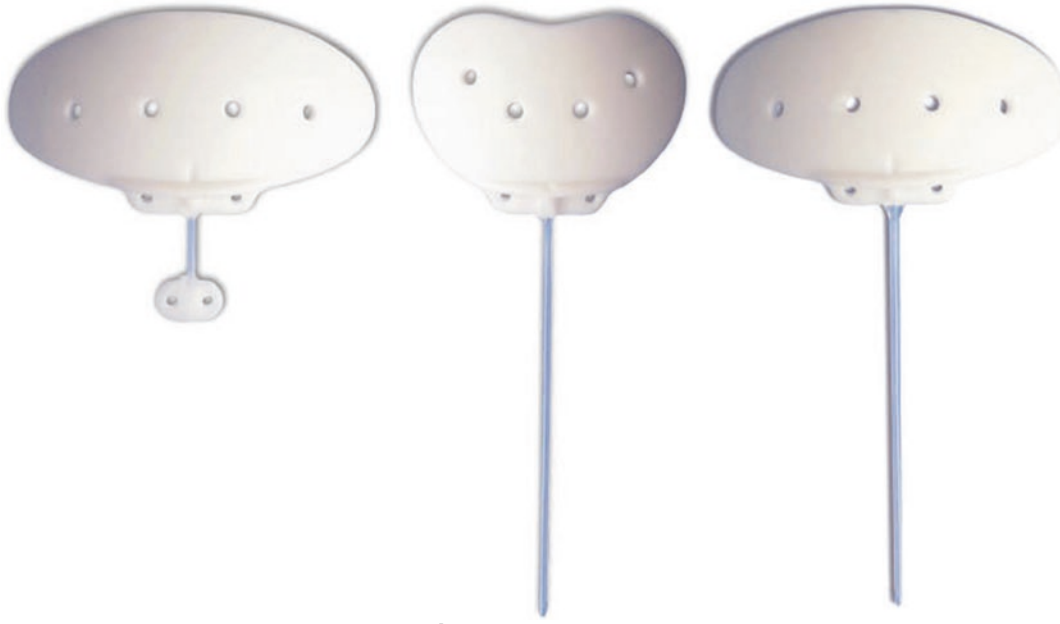

Fig. 7.2 The Baerveldt® glaucoma implant comes in three models: Model BG 101–350 (**a**) has a surface area of 350 mm^2; Model BT103–250 (**b**) has a surface area of 250 mm^2; Model 102–350 (**c**) has a surface area of 350 mm2 and is designed with a Hoffman Elbow for insertion into the pars plana. (Courtesy of Johnson & Johnson Vision, Santa Ana CA, USA)

modified with a Hoffman elbow for this purpose. All are designed for surgical implantation in a single quadrant (Fig. 7.2).

The "wings" of the plate are intended for placement under the adjacent rectus muscles. When positioned in this manner, the front edge of the implant rests approximately 8 mm posterior to the limbus. All BGIs are made of smooth, tumble-polished, pliable medical-grade silicone. Barium is incorporated into the silicone, which results in a white, radio-opaque device. The plates are designed with four holes to allow a tissue "bridge" to develop between the upper and lower surfaces of the eventual capsule to limit the size of the bleb and thus reduce the likelihood of restrictive strabismus and diplopia.

Ahmed Glaucoma Valves

The Ahmed glaucoma valve (AGV) received the US Food and Drug Administration (FDA) approval in November 1993. The implant consists of three parts: a plate made of medical-grade silicone, polypropylene, or porous polyethylene, depending on the model; a drainage tube fabricated of medical-grade silicone; and a valve mechanism (Fig. 7.3). The non-obstructive, self-regulating valve mechanism consists of thin silicone elastomer membranes 8 mm long by 7 mm wide enclosed within Venturi-shaped chamber. The membranes are pretensioned to open and close in response to IOP variations, in the range of 8–12 mmHg, and so reduce the rate of early postoperative hypotony [11, 12]. After implantation, aqueous humor flows into the trapezoidal chamber of the valve.

Aurolab Aqueous Drainage Implant (AADI)

The Aurolab aqueous drainage implant (AADI) was introduced in 2013 by Aurolab (a manufacturing

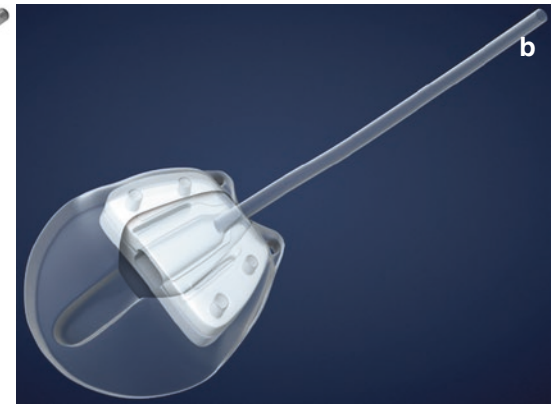

Fig. 7.3 Ahmed glaucoma valve FP7 (**a**) is a valved silicone glaucoma drainage device (GDD) implant with a surface area of 184 mm^2; the Ahmed glaucoma valve FP8 (**b**) is a GDD designed for small pediatric or nanophthalmic eyes, with a surface area of 102 mm^2. (Courtesy of New World Medical, Rancho Cucamonga CA, USA)

Fig. 7.4 Aurolab aqueous drainage implant (AADI) is a CE Mark approved, low-cost copy of the BG 101–350 Baerveldt® glaucoma implant (Fig. 7.2a above). (Courtesy of Aurolab, Madurai, India)

division of Aravind Eye Institute, Madurai, India). The AADI is a low-cost (~ US$50), non-valved GDD designed to replicate the BGI with a 350 mm^2 plate area (Fig. 7.4). Professor George Baerveldt authorized the use of his very successful design, and the device was manufactured in collaboration with the Bascom Palmer Eye Institute, Miami, Florida. Originally designed for use in India and other low-resourced countries, the device has received CE (European Commission) marking approval and is becoming broadly available in those countries that accept the CE mark for regulatory approval. It is not approved by the FDA and is therefore unavailable in the United States. Kaushik and colleagues [13] recently reported a prospective interventional study on 34 eyes of 31 children with refractory childhood glaucoma in which the AADI was implanted. Their results show an efficacy and safety profile that is comparable with published reports of the BGI and Ahmed glaucoma valve implants in children.

Indications and Contraindications

Glaucoma drainage devices are employed in childhood glaucoma when conventional angle surgery (goniotomy or trabeculotomy) has already failed or is believed unlikely to work. At such a point in clinical decision-making, most surgeons choose between a GDD and a trabeculectomy with anti-scarring agents. GDD surgery is also indicated when trabeculectomy is unlikely to work, e.g., in eyes with glaucoma following congenital cataract surgery or when trabeculectomy with anti-scarring agents has failed.

Advantages and Disadvantages

Table 7.2 broadly summarizes the pros and cons of GDD and trabeculectomy. There are no prospective randomized clinical trials comparing the two procedures in children. The Tube versus Trabeculectomy (TVT) study [14, 15] was a prospective randomized clinical trial comparing the Baerveldt® glaucoma implant to mitomycin C augmented trabeculectomy in adults greater than 18 years of age with prior failed trabeculectomy or prior cataract surgery. After 5 years of follow-up, the GDD group had a higher success rate than the trabeculectomy group with comparable complication rates, visual acuity outcomes, and medication burden. Although not directly applicable to the pediatric age group, GDDs are used most often in the eyes of older children with scarred conjunctiva, so the TVT provides at least some guidance to the surgeon considering a GDD or a trabeculectomy in such eyes.

Another consideration in balancing the decision between a trabeculectomy and a GDD is whether or not further surgical interventions are anticipated. The functioning of a well-established trabeculectomy will tend to diminish or even fail after further surgeries such as penetrating keratoplasty or even after uncomplicated cataract removal [16], e.g., in

Table 7.2 Pros and cons of glaucoma drainage devices compared to trabeculectomy in children

	Glaucoma drainage device		Trabeculectomy with MMC	
	Pros	Cons	Pros	Cons
Technique	Can be done with cloudy cornea	Violates conjunctiva Hardware in the eye	Can be performed with cloudy cornea No hardware left in the eye	Violates conjunctiva
Outcomes	Effective long-term IOP reduction, even after failed trabeculectomy Most likely to survive future intraocular surgery	Higher long-term IOP compared with trabeculectomy More likely to require supplemental medications Further surgery for complications more likely [73]	Lower long-term IOP Supplemental medications less likely	Poor results in glaucoma following cataract surgery even with MMC Less likely to survive future intraocular surgery
Complications	Lower risk of endophthalmitis	Risk of intra- and postoperative hypotony Risk of tube-related complications: corneal decompensation, cataract, tube erosion, migration, and obstruction Greater risk of postoperative motility disturbance	No tube-related complications	Risk of intra- and postoperative hypotony Lifelong risk of postoperative endophthalmitis if avascular bleb develops, especially if contact lenses are required
Quality of life	Contact lens wear possible (important for aphakic eyes with glaucoma) Post-op care involves fewer manipulations, reducing number of EUAs			Contact lens wear not recommended Post-op care requires frequent visits for close follow-up and possible suture adjustments/5FU injections which in turn might require more frequent EUAs

IOP intraocular pressure, *EUA* examination under anesthesia, *MMC* mitomycin C, *5FU* 5 Fluorouracil

uveitic eyes. In eyes likely to need additional surgery after glaucoma surgery, a GDD is much more likely than trabeculectomy to continue functioning postoperatively. For this and other reasons, we firmly believe that a team approach to complex childhood glaucoma is key to successful outcomes, especially one that engages cornea, pediatric, and vitreoretinal specialists early on to develop a long-term plan of care that includes the proper sequencing of interventions.

Preoperative Considerations and Preparation

Preoperative examination and planning are essential for successful surgical outcomes. In infants and young children, a thorough examination sufficient to plan surgery may not be feasible in the clinic, and the decision on surgical approach may only be made in the operating room following an examination under anesthesia (EUA). The EUA for patients with childhood glaucoma is discussed in detail in Chapter 3.

Important Considerations for GDD

Axial Length Measurement of the axial length is important in the baseline assessment and ongoing monitoring of children with glaucoma. Progressive increases in the axial length of an eye in excess of normal growth may indicate poorly controlled IOP. In the context of planning for a GDD, axial length measurement may influence device selection (see below). In buphthalmic eyes that are adult size or larger, adult GDDs are commonly used.

Conjunctival mobility, which may constrain which quadrant is best for GDD implantation or preclude a trabeculectomy if the choice between procedures has not yet been made. It is often useful to inject balanced salt solution (BSS) into the subconjunctival space with a 30-gauge needle to help delineate episcleral scarring.

Gonioscopy, which will help visualize the presence of iris strands, membranes, or peripheral anterior synechiae (PAS) which might interfere with the insertion of the tube into the AC. If implantation is planned in a quadrant with broad PAS, a surgical peripheral iridectomy can be performed through a small corneal incision in the area of planned tube insertion to avoid having the tube end up under the iris.

Anterior chamber depth and lens status, visualized clinically by slit lamp, under the operating microscope, gonioscopy, or by ultrasound. This is done to determine if tube insertion into the AC can be safely performed both avoiding tube-corneal touch and damage to the crystalline lens. In pseudophakic or aphakic eyes, it may be preferable to place the tube in the ciliary sulcus or pars plana in combination with a pars plana vitrectomy or through a surgical peripheral iridectomy to keep the tube as far away from the corneal endothelium as possible.

Size of the palpebral aperture and motility of the globe are important in providing surgical access to insert a GDD. It may be necessary to perform a lateral canthotomy in some cases of small palpebral aperture to gain sufficient access for surgery. Ocular motility considerations include how to handle the GDD placement when strabismus is present before surgery, especially if the eye to be operated has had prior extraocular muscle surgery. An additional topic, and one which is beyond the scope of this chapter, includes how to handle strabismus induced or worsened by GDD placement.

Scleral Integrity The eyes which have had previous surgical procedures, trauma, or transscleral laser may have areas of scleral thinning which will influence the choice of quadrant used for GDD placement.

Choice of GDD Patch Although many surgeons will use commercially available patches (e.g., Tutoplast® pericardium, dura, or fascia lata) or donor grafts (e.g., sclera or cornea), some surgeons prefer to fashion a long scleral tunnel to cover the tube.

Choice of GDD

The decision of which implant to use in a specific case is based on a number of factors, including

the underlying glaucoma diagnosis, level of IOP and ability to control IOP medically, ocular size, orbital anatomy, and finally surgeon preference.

Glaucoma Diagnosis GDDs lower IOP in all forms of childhood glaucoma, and there are no prospective randomized clinical trials comparing the different implants in children on which to base device choice. However, the specific form of disease may influence the choice of implant. In children with the potential for decreased aqueous production, e.g., eyes with uveitis or eyes that have already undergone cyclodestructive procedures, a valved or smaller surface area non-valved implant may be a better choice to avoid hypotony.

IOP Level The level of IOP and the ability to control the IOP with medications postoperatively can influence the choice between a valved or non-valved implant. The adult literature suggests that the non-valved 350 mm^2 Baerveldt® glaucoma implant may be superior to the Ahmed glaucoma valve in terms of long-term IOP-lowering and medication burden [17]. Hence, if the IOP in a child can be controlled with medications for 6 to 8 weeks while the non-valved implant is temporarily occluded, the surgeon may choose to use a Baerveldt implant in the hope of achieving slightly better long-term IOP control. In contrast, the Ahmed glaucoma valve lowers IOP immediately, which may be of prime importance especially if the child is in pain, has advanced glaucoma, and cannot tolerate medications or if corneal edema is causing amblyopia.

Ocular Size and Orbital Anatomy Often by the time pediatric eyes have already failed one procedure and require GDD surgery, the eyes have grown to adult size or larger. In such cases, adult-sized implants may usually be implanted without too much difficulty. As noted earlier, IOP-lowering is roughly proportional to plate size, so it is generally in the long-term interest of the child to implant an adult implant whenever possible. In contrast, when GDDs are used as primary surgery or in very young infants with small eyes (e.g., those with true microphthalmia), the physical size of adult implants comes into play.

GDDs are best implanted with the anterior edge of the plate at least 8 mm posterior to the surgical limbus. When an adult-sized GDD is placed in this position, the posterior edge of the implant plate may impinge on the optic nerve in young infants. Margeta and colleagues [18] measured the limbus to optic nerve distance in the superior temporal quadrant in 15 pediatric autopsy eyes. Figure 7.5 demonstrates that in a pediatric eye with axial length of 19 mm, an adult-sized Ahmed Model FP7 glaucoma valve, placed in the inferior nasal quadrant with the anterior edge sutured 7 mm from the limbus, overlaps the optic nerve to a significant degree. The Freedman-Margeta formula (http://people.duke.edu/~freed003/GDDCalculator/) offers a way to determine limbus to optic nerve distance in pediatric and small eyes in order to reduce the risk of optic nerve impingement by the posterior edge of the GDD plate. The Ahmed glaucoma valve is available in pediatric sizing (FP8 and S3 versions) as is the Molteno™ implant (P1 version); the posterior edge of the 350 mm^2 BGI and the AADI can be easily trimmed with a heavy surgical scissors to create a cutout to accommodate the optic nerve. The 250 mm^2 version of the BGI is designed with a posterior notch to avoid overriding the optic nerve.

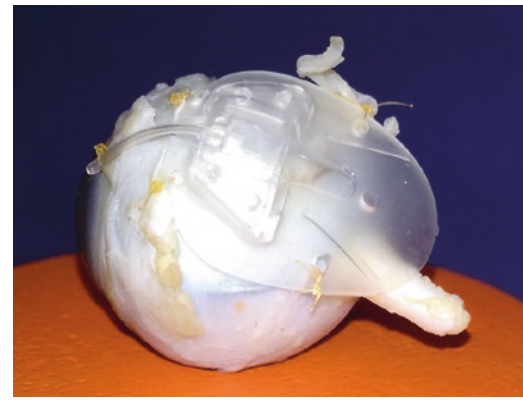

Fig. 7.5 Photograph of a pediatric autopsy eye (axial length 19 mm), showing an Ahmed Model FP7 glaucoma drainage device sutured in the superonasal quadrant. The anterior edge of the plate is located 7 mm from the limbus; note the extensive overlap of the posterior edge of the Ahmed device plate and the optic nerve. (Courtesy of Milica Margeta, MD, PhD and Sharon Freedman MD)

Operation

Intraoperative Preparation

The surgical field should be cleaned with sterile iodine or chlorhexidine-based preparation fluid, the area dried, and a surgical drape placed over the eye. Self-adhesive ophthalmic drapes with a transparent window and a pocket to collect irrigation fluid are desirable. This may be applied with the lids open and the lashes everted so that when the drape is cut open to apply a lid speculum, the lashes are retained underneath the sticky drape without straying into the surgical field.

With the lids held open by an ophthalmic speculum, a drop of 1:10,000 adrenalin or other topical ocular sympathomimetic drug may be instilled to promote vasoconstriction of the conjunctiva and episclera and minimize tissue bleeding.

The usual position for a GDD is in the superior temporal quadrant. The surgeon will generally be positioned superiorly or in the superior temporal position approximately 45° from the vertical; if the microscope permits, the assistant can be positioned superiorly.

Surgical Technique

A recommended set of instruments and suture materials for GDD surgery are listed in Table 7.3. Implantation of GDDs in pediatric patients requires special attention to the size of the eye and orbit, the thickness of the sclera, and the positioning of the tube in an eye that will continue to grow.

Traction Suture A corneal traction suture is placed in alignment with the intended quadrant to rotate the eye inferior-nasally to provide the best surgical exposure. This may be placed through partial-thickness cornea in the mid-periphery of the same quadrant or at the limbus on either or both side(s) of the planned entry site. The conjunctiva and surgical exposure are then evaluated for scarring, and sufficient room to place a GDD is confirmed.

Conjunctival Incision Most surgeons perform fornix-based conjunctival flaps (e.g., the incision is made at the limbus) for GDD implantation, but a limbus-based conjunctival flap (where the incision is placed approximately 6–8 mm posterior to the limbus) has several advantages in pediatric eyes. It facilitates placement of the sutures securing the plate to the sclera in the very tight pediatric orbit, and the incision can be closed confidently in a watertight manner that will not unravel if the child rubs the eye vigorously. And importantly in children with aphakia, contact lens use can often be resumed within days of surgery. In contrast, incisions at the limbus are more uncomfortable for children, and contact lenses cannot be resumed for several weeks in most cases. In a retrospective comparison of limbal-based to fornix-based incisions for GDDs in adults, Suhr et al. found no difference in IOP outcomes [19].

Another important consideration for incision type arises in children with aniridia. In these patients, limbal stem cell deficiency arises that leads to corneal conjunctivalization and opacity later in life. A limbal incision, cautery, and mitomycin C application are likely to be detrimental

Table 7.3 Recommended instruments and suture materials for glaucoma drainage devices implant surgery

Instruments and knives		Sutures and consumables	Device related
Eye speculum	Tenotomy scissors		GDD of choice
Caliper	Vannas scissors – straight, curved	7-0 Vicryl® or Mersilk for corneal traction suture	BSS on a 27- or 30-gauge cannula
Muscle hook	Mini-crescent blade	8-0 nylon on a spatulated needle	23- or 25-gauge needle
Conjunctival forceps	MVR blade/supersharp	8-0 or 9-0 Vicryl® for conjunctival closure	Patch graft material
Tying forceps	Conjunctival clamp	Viscoelastic	
Colibri forceps	Needle holder	Anterior chamber infusion	

GDD glaucoma drainage device, *BSS* balanced salt solution, *MVR* micro vitreoretinal

to the limited reserve of stem cells in these patients. For patients with aniridia, a limbal-based (limbus sparing) conjunctival technique is recommended, and the use of MMC is avoided.

Device Preparation and Placement Once the conjunctival incision is made, dissection and elevation of Tenon capsule from the episclera are carried out, and the adjacent rectus muscles are identified with a muscle hook. Tenotomy scissors are used to clear all adhesions and check ligaments posterior to the incision to clear the potential space for the implant and to facilitate the easy implantation of the selected device. Alternatively, blunt dissection of the correct plane may be started with scissors and completed with two squint hooks inserted back-to-back in this pocket and pulled gently apart, as demonstrated in the Video 7.1.

Before implanting an AGV, it is imperative that the surgeon "prime" the device with BSS. The silicone leaflets of the valve device stick together during manufacture, and if they are not primed with BSS, the device will fail. A 27- or 30-gauge cannula on a syringe filled with BSS is inserted into the tip of the silicone tube. BSS is gently injected into the tube, and flow is observed through the valve leaflets under the microscope [20]. Once this is done, the device is "primed," and implantation in the selected quadrant may proceed.

A muscle hook may be used to engage the superior rectus muscle; the AGV device can be grasped gently with smooth forceps with care taken not to crush the portion of the device housing the valve mechanism. With forward traction on the muscle hook, the device is placed between the two rectus muscles and pushed posteriorly. When adequate preparation of the quadrant and clearance of adhesions or check ligaments has been done, the device will achieve a resting position with the anterior suture holes approximately 8 mm posterior to the limbus.

In the case of a BGI or AADI, the superior rectus muscle is engaged with a muscle hook and the implant plate grasped with large non-toothed forceps (e.g., Nugent or Moorfields forceps) and the appropriate wing of the implant placed under the muscle. A second non-toothed forceps is then used to grasp the opposite wing of the implant, and the muscle hook removed from under the superior rectus and repositioned under the lateral rectus muscle. The plates of these devices are quite flexible, so by grasping and bending the plate, the temporal wing can be easily placed under the lateral rectus muscle. The implant is then gently pulled forward to verify that it has achieved a good position between and under the two rectus muscles. It is not pulled tightly up under the muscle but rather should rest gently behind the muscle insertions with the suture holes positioned some 8–10 mm posterior to the limbus.

Because the BGI and AADI devices are not valved, the tube portion of the device must be temporarily occluded. In adults, this can be done with absorbable sutures, sutures that can be lasered or with a rip cord that is removed later at the slit lamp in older children and adults and in the operating room in young children. The absorbable suture technique is appropriate in young children and is described here along with the optional modification of an intraluminal stent to reduce the chance of hypotony. A suture of 6-0 or 7-0 polyglactin suture is tied around the silicone tube a few mm anterior to the plate. A 27- or 30-gauge cannula on a syringe filled with BSS is inserted into the tip of the silicone tube, and complete occlusion of the tube is confirmed. Ligation of the implant tube can be performed based on the surgeon's preference either on the back table of the operating room before implantation or under the operating microscope after the device is in place or sutured to the sclera. Any flow through the tube must be avoided in children as a flat chamber is likely to result that will necessitate an early return to the operating room.

In addition to the extraluminal absorbable tie, an intraluminal stent may be used to partially occlude the lumen. A nonabsorbable suture thread (e.g., 3-0 Prolene® or 3-0 Supramid®) is used with one end fed into the GDD tube at the plate and the distal end tracked beneath the conjunctiva, often into the inferior fornix. Following the absorption of the extraluminal tie at 6–8 weeks, the presence of this intraluminal stent

mitigates the risk of hypotony. If, however, the IOP remains poorly controlled after the absorption of the Vicryl tie, the stent may be subsequently removed by making a small incision over its distal end in the lower fornix and the whole thread pulled out remotely from the device. The conjunctival incision is closed with a pre-placed Vicryl purse string suture.

Plate Suturing The next step in the procedure is to affix the plate to the sclera. A suture on a spatulated needle is used to make a deep partial-thickness pass just in front of the plate taking care not to penetrate the sclera, which can be quite thin in a buphthalmic eye. The suture is then passed through the fixation hole of the implant and tied down tightly to minimize plate movement. When possible, the knot should be rotated into the fixation holes to avoid the short lengths of suture eroding through to conjunctiva resulting in irritation or serious infection. Molteno described the use of 7-0 silk to secure the plate of his device to the sclera [7], and others advocate the use of nonabsorbable sutures, such as polyester (Mersilene®) or polypropylene (Prolene®). Current implants are designed with holes in the plate that allow tissue "rivets" to form between the sclera, through the hole, and to the capsule above. Once these tissue rivets are formed during the first few months of healing, the implant will not move.

Silk can cause significant local inflammation, and nonabsorbable sutures may cause the overlying conjunctiva to break down or erode, sometimes many years later. We recommend 8-0 Nylon to secure the implant in place on the sclera. Nylon has sufficient tensile strength to tie down the implant tightly; the material begins to degrade only after a year or so has passed, long after the tissue rivets have secured the implant in place. After several years, the nylon disintegrates, eliminating the risk of late conjunctival erosion.

Tube Implantation Insertion of the tube into the AC is the most challenging aspect of pediatric GDD surgery. The surgeon must account for an AC that may be shallow but also plan for the long-term growth of the eye.

An AC paracentesis is performed (e.g., using an MVR blade); if the child was dilated for an EUA or photography, a quick-acting miotic is first injected into the eye to constrict the pupil. A viscoelastic substance is injected, or an AC infusion inserted, to avoid intraoperative hypotony. The scleral surface anterior to the plate is cleared of adhesions, and gentle cautery is applied, if required, to dry the surface. When a limbus-based conjunctival flap has been created, the assistant can use a surgical conjunctival clamp (Khaw conjunctival clamp [Duckworth & Kent Ltd., Baldock, Hertfordshire, UK], Khaw/Shah 4 mm conjunctival clamp [Duckworth & Kent], or Lama cross-action 5 mm conjunctival forceps [Moria Inc., Doylestown PA, USA]) to grasp the flap and provide good surgical exposure (Fig. 7.6). The tube entry in children is usually created in an oblique direction so that the tube can be left long enough to accommodate growth without extending into the pupillary aperture. The tube is laid down on to the intended path of implantation and is trimmed to an appropriate

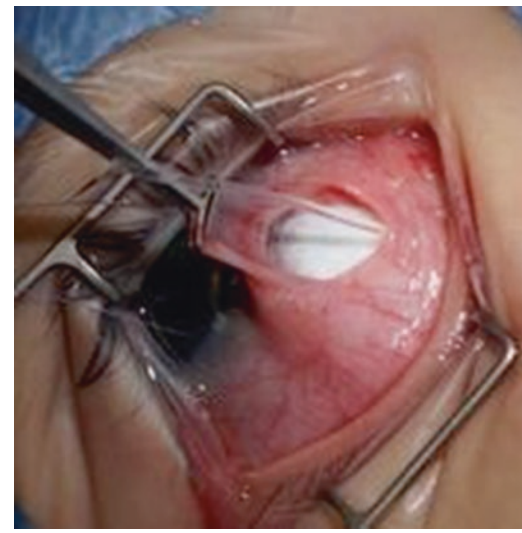

Fig. 7.6 Placement of an Ahmed glaucoma valve in the inferior nasal quadrant through a limbal-based conjunctival incision. The adult-sized plate (Model FP7) has already been sewn in place approximately 10 mm posterior to the limbus in this buphthalmic eye with a 25 mm axial length. Lama cross-action conjunctival forceps provide excellent exposure. (Courtesy of James D. Brandt, MD)

length creating an anteriorly oriented beveled tip. Before entering the eye with a needle to create a pathway for the tube, the AC is reassessed and the previously placed side-port incision used to deepen the chamber with viscoelastic as necessary. This side-port incision should be oriented in such a way that if the tube is misdirected in the eye, a Sinskey hook or iris spatula can be used to redirect the tube into the correct position. If the implant is positioned in such a way that the tube will enter the eye in an area of broad PAS or other obstructions, a surgical peripheral iridectomy can be created through a small corneal incision just inside the limbus. In this manner, the tube can be inserted more posteriorly and then rest in front of the iris but well back from the cornea. Any such corneal incision must be closed with a 10-0 suture of Nylon or Vicryl® as it will leak in a pediatric eye unless sutured.

In adults, a 23-gauge needle is often used to create the tube entry site; in children, the sclera is sufficiently flexible that a 25-gauge needle can be used and the tube can be inserted without too much difficulty. The use of a narrower gauge needle to fashion the tunnel also reduces the risk of leakage of aqueous around the tube and hypotony. Moving the tube entry site several millimeters back from the limbus avoiding a corneal track is important to avoid late erosions, but doing so can make it challenging to get the angle just right to avoid an anteriorly vaulted tube that touches the corneal endothelium. The beveled tip of the tube is then advanced into the needle tract and pushed forward with fine forceps. Alternatively, the tube may be inserted with the assistance of a fine blunt-tipped cannula by engaging the tip firmly onto the bevel of the trimmed tube and passing the cannula gently through the scleral tunnel taking the tube with it (Video 7.2). The surgeon should watch for the tube to enter the AC periphery where expected; if it does not, the tube may have migrated under the iris, in which case another pass can be tried or a surgical iridectomy created as noted above.

Once the tube has been inserted into the eye, the external portion should be secured to the sclera with a nonabsorbable suture (e.g., 9-0 Nylon) which helps to stabilize the tube. This both reduces the risk of migration out of the eye and also reduces outward bowing of the tube which may increase the susceptibility to erosion through the conjunctiva.

The tube must be covered to prevent its erosion through the overlying conjunctiva over time. The tube may be covered either with autologous tissue or with donor sclera or cornea obtained from an eye bank or dehydrated and preserved donor dura mater or pericardium [21]. This graft material is usually sutured into place with one or two sutures of fine (8-0 or 9-0) polyglactin; a permanent suture is not needed as graft materials rapidly incorporate into the surrounding Tenon capsule or conjunctiva.

Although autologous tissue may be obtained from fascia lata or temporalis fascia, many glaucoma surgeons now advocate using the patient's own sclera to support and cover the tube. This can be challenging in the thin sclera of a buphthalmic eye but can be done in a number of ways, the simplest of which is by creating a scleral flap. Alternatively, a long tunnel in the patient's native sclera may be created using a bent needle starting some 5 mm posterior to the limbus. Another method employs a small mini-crescent blade (1.25 mm in width) (Video 7.3) that can be used to tunnel in the sclera up to about 2 mm posterior to the limbus, then completing the entry into the eye with a 25-gauge needle.

Once the tube has been inserted, any viscoelastic remaining in the eye is expressed through the side-port incision or washed out with irrigation. While some viscoelastic may be left in the eye safely when a valved implant is placed, it should be aggressively removed from the eye when a non-valved implant is used, as any retained viscoelastic will cause a dramatic rise in IOP. The AC is then refilled to a physiologic level with BSS to determine where the tube will end up in relation to the iris or cornea. If the tube is vaulted too far forward and risks touching the endothelium with eye rubbing, it is far better to revise the tube position and/or length at the initial surgery than returning to the operating room months or years later after endothelial loss has occurred.

Incision Closure Tenon layer and conjunctiva are securely sutured to ensure adequate coverage of the plate, tube, and patch graft/scleral flap, preferably with absorbable sutures to avoid returning to the operating room and further anesthetic exposure to deal with irritating sutures. A sub-Tenon injection of long-acting local anesthetic performed during closure can help with early postoperative pain relief.

The sclera is often thin in pediatric eyes and the tissues less rigid than adult eyes. It is therefore frequently necessary to suture the paracentesis used for the AC infusion or viscoelastic in order to avoid postoperative leaks; 10-0 monofilament Vicryl® is used when available for the same reason described above.

At the end of the operation, the eye should be inspected to ensure that the implant plate, patch graft, and intraocular portion of the tube are in a good position, that the AC is well formed, and that a clear red reflex can be seen. Fluorescein drops or strips can be used to inspect the conjunctiva and cornea of leaks, and instillation of a cycloplegic such as atropine will assist in deepening the AC. A subconjunctival injection of corticosteroid and antibiotic is commonly performed at the end of the procedure.

Modifications to Standard Technique

Some modifications to the technique of GDD surgery have already been mentioned and relate to surgeon preference and experience; however, modifications to the standard technique may be dictated by scarring and tissue distortion from previous surgery and trauma or by the underlying diagnosis.

Conjunctival Scarring Although superotemporal placement of the drainage device is often preferred, circumstances may dictate that a superonasal or inferior placement of the tube is required. Examples include eyes where previous glaucoma surgery has taken place in the superior temporal quadrant or where PAS preclude entry of the tube into the AC at that site.

The use of explants for the repair of retinal detachments is infrequent now; however, if a GDD is required in this circumstance, placement of the plate may need to be behind or even on top of the explant. Before placing a GDD in such eyes, it is essential to ensure that adequate and sufficiently mobile Tenon capsule and conjunctiva are available to cover the hardware before placing such a device.

Prior Strabismus Surgery If a rectus muscle has been resected in the quadrant planned for the GDD placement, one needs to prepare for additional scarring around the muscle insertion, but usually careful technique will allow the GDD surgery to proceed without major modification. By contrast, a previous rectus muscle recession (e.g., lateral rectus for a planned GDD in the superior temporal quadrant) may call for a modified GDD procedure, especially in the case of a planned Baerveldt GDD. Basically, for a Baerveldt to be placed in a quadrant where an adjacent rectus recession has previously occurred, the surgeon must identify the insertion of the recessed muscle and then may either trim the front of the respective wing to allow that wing to be placed behind the recessed rectus muscle (personal communication, SFF) or may trim the back of the respective wing, such that the wing is entirely in front of the rectus muscle. This latter technique may lead to making the anterior portion of the bleb in front of the muscle much more visible, which is often cosmetically problematic, and is therefore not recommended. It may be preferable in this case to place the plate in the superonasal quadrant, that is, in the quadrant with the resected rather than the recessed horizontal muscle. In the case of a preexisting strabismus, such as an exotropia, the surgeon may elect to recess the lateral rectus muscle concurrent with the GDD placement (especially in the case of a planned Baerveldt implant), because the muscle will be much harder to access after the GDD surgery. As described above, it is recommended that the anterior portion of the, respectively, Baerveldt wing be trimmed to allow the recessed rectus to remain anterior to the Baerveldt wing.

Glaucoma Following Cataract Surgery (GFCS) GDDs are frequently the preferred primary procedure in the management of glaucoma following cataract surgery, particularly in aphakic eyes where contact lens is planned. In this circumstance, a trabeculectomy is contraindicated due to increased risk of bleb-related endophthalmitis.

A success rate for controlling the IOP following cataract surgery has been reported of up to 90% at 1 year [22], and in addition, GDDs allow children to more easily wear contact lens refractive correction which assists with management of amblyopia. Tube insertion may be performed in the sulcus or the pars plana to avoid complications of corneal tube touch [23]. Doing so avoids leaving hardware at or near the limbus where aphakic contact lenses can lead to late-onset conjunctival breakdown over the tube; furthermore, pars plana insertion of the tube protects the corneal endothelium from tube-corneal touch. However, eyes which have undergone cataract surgery may be particularly susceptible to occlusion of the tube tip by capsular remnants, "Elschnig pearls" of residual cortical material, or vitreous. To mitigate this risk, it is essential to consider performing excision of lens pearls, and/or further vitrectomy if the eye is aphakic, and the tube should be positioned away from the iris and capsule.

Vitreous incarceration into the tube can happen shortly after surgery or many years later; in a retrospective review of Baerveldt® implants placed through the pars plana, Vinod et al. [9] reported vitreous tube obstruction in 19% of their series, occurring 3–112 months after implantation.

Any mobile vitreous will eventually find its way to the tube tip and occlude it. It is absolutely necessary to perform a meticulous pars plana vitrectomy prior to tube insertion in order to prevent this late complication. If corneal clarity prevents a good view, a surgical endoscope can be used to perform the vitrectomy [24]. The anterior core vitrectomy routinely performed at the time of pediatric lensectomy is insufficient; even in an aphakic eye that was vitrectomized at the time of cataract extraction, the vitreous base must be shaved down aggressively under direct visualization and an attempt made to cause a posterior vitreous detachment to elevate and remove the posterior hyaloid face. In young children in particular, the posterior hyaloid face is difficult to detach during vitrectomy, and it is this posterior shell of vitreous and hyaloid that can detach years later.

Care should be taken to ensure that the tube is trimmed to a length which allows direct visualization of the tip, particularly when it is inserted through the pars plana as this assists in diagnosing or excluding occlusion as a cause of GDD failure.

Sturge-Weber Syndrome Patients with Sturge-Weber syndrome may have associated choroidal hemangioma, and it is important to diagnose this prior to surgical intervention. Precipitous reduction in IOP or hypotony may result in a suprachoroidal hemorrhage, which can be sight threatening. Choroidal effusions are more common and may even occur undetected in the early postoperative period (Fig. 7.7). It is therefore essential to take measures to avoid both intraoperative and postoperative hypotony. The use of an AC infusion is helpful to maintain the IOP while the tube tunnel is fashioned and the tube inserted. Care should be taken to ensure that the tube tunnel is tight and that there is no peri-tube leak following insertion. The use of extraluminal tube restricting ligatures and intraluminal stents in non-valved devices is essential; great caution is recommended if a valved device is chosen and implanted without ligating the tubing, as choroidal effusions may be severe in the early postoperative period. Choroidal effusions may occur at higher-than-expected IOP in the presence of choroidal hemangioma, and it is advisable to omit glaucoma medications until review on the first postoperative day confirms the IOP. B-scan ultrasound examination is useful in monitoring for choroidal effusion.

Uveitis Surgical intervention in an eye with uveitis risks exacerbating inflammation which, in turn, may threaten the success of surgery. It is therefore desirable to optimize both systemic and

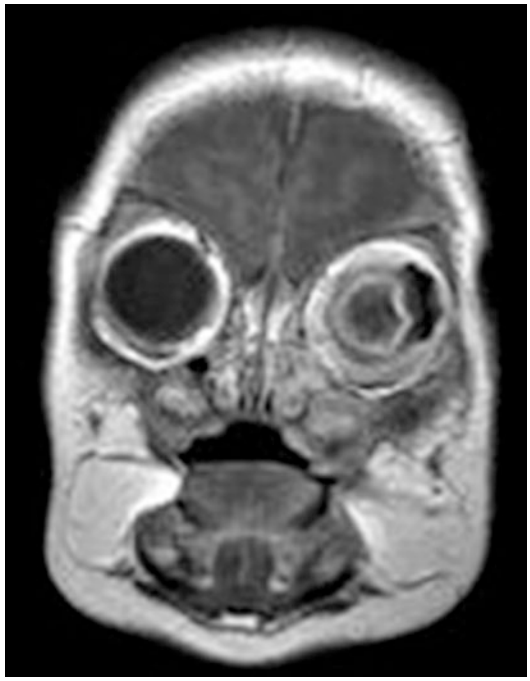

Fig. 7.7 Coronal reconstruction of an MRI performed on an infant with Sturge-Weber syndrome to evaluate central nervous system involvement in the disease. The scan was acquired 1 week after implantation of an Ahmed glaucoma valve in the superior temporal quadrant of the left eye. Note the lucent area representing the silicone implant, along with the annular choroidal effusion. The effusion had resolved by the next clinic visit and was never observed clinically. (Courtesy of James D. Brandt, MD)

topical immunosuppression before performing glaucoma surgery, and this may require collaboration with the pediatric rheumatologist. Complications of GDD surgery particularly related to poor control of intraocular inflammation include occlusion of the tube tip with fibrin, ciliary body shutdown, and hypotony. Hypotony is a risk in these eyes even in the absence of active inflammation as a consequence of poor ciliary function, and the risk is particularly high if the eye has previously undergone cyclodestructive procedures which are advised against in these eyes. Care should therefore be taken when performing GDD surgery to avoid over-drainage. For this reason, valved devices may be useful in these eyes, and if a non-valved device is preferred, the use of a restricting ligature and intraluminal stent is advisable. The choice of a small plate size in either a valved or non-valved device is also helpful.

Aniridia Aniridic eyes have limbal stem cell deficiency that leads to corneal conjunctivalization and opacity later in life. As previously mentioned, a limbal-based (limbus sparing) conjunctival technique is recommended, and the use of MMC is avoided in children with aniridia.

In addition, careful positioning of the tube in the AC is necessary to avoid it touching the lens, which is unprotected by the iris, as this results in cataract formation (Fig. 7.8). Tubes are commonly inserted into the eye in a radial or slightly oblique angle; however, a more aggressively tangential approach is used in cases of aniridia where it is desirable to avoid having the tube lying across the unprotected lens; a similar approach is sometimes taken in the setting of a corneal graft when it is desirable to have the tube lie beneath the host cornea rather than under the graft (Video 7.4).

The Role of Antimetabolites in GDD Surgery

Pediatric eyes frequently mount an exuberant healing response to the implantation of a foreign body such as a GDD; for this reason, some pediatric glaucoma surgeons advocate the use of antimetabolites at the time of surgery. Costa and colleagues performed a masked, randomized prospective clinical trial comparing the intraoperative use of MMC (0.5 mg/ml × 5 min] to BSS in adults older than 18 years of age undergoing AGV implantation (three of their subjects carried the diagnosis of "congenital glaucoma," but they were adults at the time of the study) [25]. They found no difference in short-term success at 1 year.

The role of antimetabolites in GDD success has yet to be established in the pediatric population. There are no prospective, randomized clinical trials comparing the intraoperative use of mitomycin C (MMC) in pediatric implants. Cui and colleagues performed a retrospective review of adjuvant treatment with antifibrotic agents during and after AGV implantation and reported

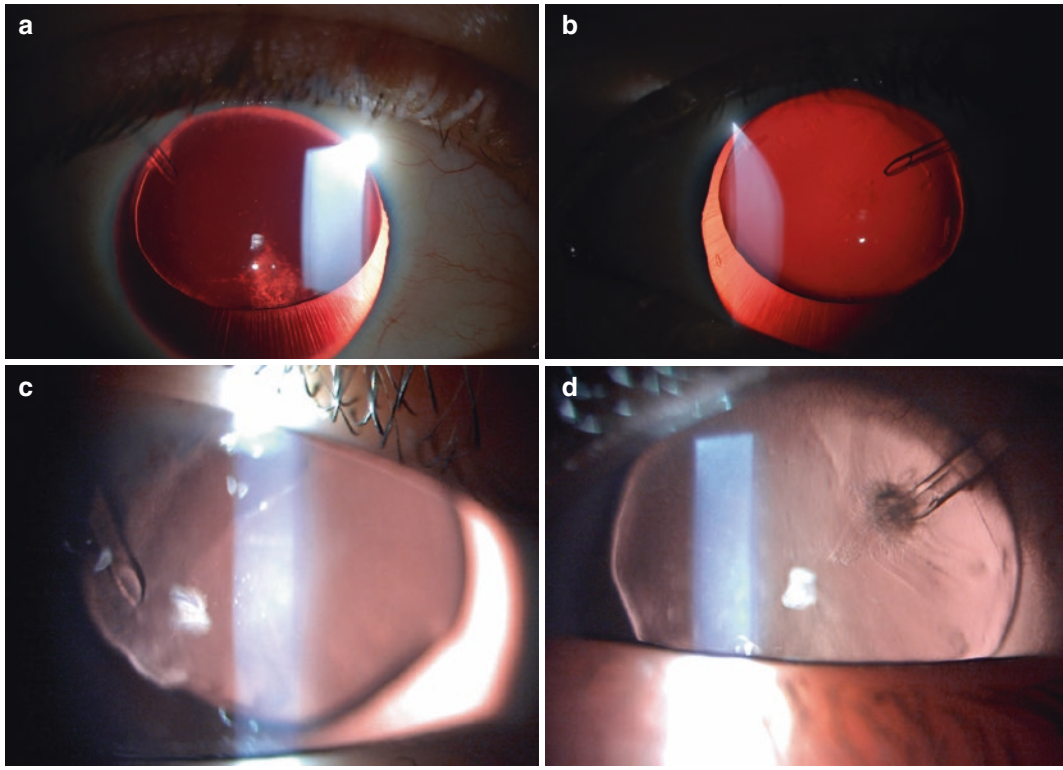

Fig. 7.8 (**a**, **b**) Bilateral glaucoma drainage devices placed at age 8 in a boy with glaucoma associated with aniridia. There is no tube-lens touch, but over the last 5 years, the lenses slowly dislocated upward, as shown, and developed early posterior subcapsular cataract with visual acuity in the 20/50 range bilaterally (patient does not have foveal agenesis). (**a**, **b** Courtesy of James D. Brandt, MD). (**c**) Tube of a Baerveldt glaucoma drainage device inserted at a tangent so that it lies near the equator of the lens (the thinnest portion of the lens) to avoid the tube touching the lens. (**d**) Tube of a Baerveldt glaucoma drainage device in an aniridic eye. The tube is inserted in a radial direction with the tip of the tube approaching the anterior pole of the lens (the thickest portion of the lens) resulting in the tube touching the lens and causing a localized opacity. (**c**, **d** Courtesy of Cecilia Fenerty, MD, FRCOphth)

decreased frequency of the hypertensive phase commonly observed with this device and improved surgical outcomes at 1 year [26]. A retrospective, nonrandomized study of AGV both with and without mitomycin C (MMC) in aphakic glaucoma showed no difference in IOP control between groups [27]. A larger retrospective study demonstrated worse outcomes after 2 years of follow-up in eyes receiving AGV plus MMC [28], though this may be due to differences in underlying diagnosis and previous number of surgical interventions between groups. A prospective randomized study looking at AGVs augmented with either Bevacizumab, MMC, or no MMC found higher success rate groups receiving both Bevacizumab and MMC at 1 year [29].

The bottom line is that the long-term safety and efficacy of the adjunctive use of antimetabolites in pediatric GDD surgery are not known. A surgical registry approach to capturing data on pediatric GDD surgery over many years may be the best way to determine the risk/benefit balance of antimetabolites in this population for whom long-term outcomes are so important.

Postoperative Management

Postoperative medication includes topical broad-spectrum antibiotic (e.g., chloramphenicol or fourth-generation fluoroquinolone), topical steroid (e.g., prednisolone acetate 1% or dexamethasone

0.1%), and often a topical cycloplegic (e.g., atropine 1% or cyclopentolate 1%). When a non-valved device has been implanted using an extraluminal tie and/or intraluminal stent, it may also be necessary to continue topical glaucoma medication until the extraluminal suture is absorbed and the tube opens. For those children with a valved tube, the device will function immediately, and often all the regular glaucoma medications can be stopped.

Pain relief is usually adequately managed with acetaminophen appropriately dosed by weight, with the addition of a systemic nonsteroidal anti-inflammatory drug if required.

Children are examined on the first postoperative day, and assessment should be directed to identify potential complications of surgery as detailed below. It is usually impossible to formally measure IOP in a young child on the first postoperative day, but if necessary the IOP may be estimated by gentle digital palpation over the closed lid. External examination will reveal any purulent discharge or bleeding, and ocular examination should aim to confirm the presence of a formed AC and clear red reflex. If necessary, a B-scan ultrasound examination can assist in assessment.

At 1 week after surgery, a more detailed exam is usually possible. In the presence of nonabsorbable sutures on the conjunctiva or cornea, antibiotic drops should be continued until they are removed. However, when absorbable sutures have been used for closure, antibiotics can be stopped within the first week or two postoperatively. The key issues during the first month or so after GDD surgery are infection surveillance, AC status, IOP, and tube positioning. If the IOP is very low, and the AC is very shallow or flat, reformation with viscoelastic in the operating room is urgently necessary to avoid corneal failure or cataract. B-scan ultrasound can help determine if choroidal effusions are present, and this can be done even in a crying infant in the clinic setting.

Topical cycloplegic, if used, may be stopped shortly after surgery if there is no hypotony and the AC remains deep. However, this medication may be temporarily reinstated around the time of opening of the extraluminal tie if there is a significant risk of hypotony. Topical steroid therapy should be tapered postoperatively according to the presence and grade of AC activity and external conjunctival hyperemia, usually over 1–2 months.

Management of valved versus non-valved GDDs diverges significantly in the postoperative period. In the case of a valved implant, preoperative glaucoma medications are usually discontinued to prevent hypotony. For those children who have had a non-valved device inserted utilizing an absorbable suture extraluminal tie, a critical time for review is around 6–7 weeks postoperatively when the tie usually spontaneously releases. In order to avoid hypotony at this time, it may be advisable to reduce or stop glaucoma medication a few days in advance. By this point, a fibrous capsule will have formed around the plate; this capsule offers resistance to outflow and thus avoids hypotony.

Further, EUA may be required to fully assess tube function and IOP control or for the removal of nonabsorbable sutures or the intraluminal stent. This should be planned at or beyond 6 weeks following surgery at which time any absorbable extraluminal tie will have spontaneously opened.

The "hypertensive phase" following GDD implantation is a widely recognized phenomenon, particularly with the Ahmed glaucoma valve [30], and is associated with high postoperative IOPs [31]. Approximately 4–6 weeks after implantation, the capsule forming around the AGV tends to thicken and offer more resistance to aqueous outflow. It is important to monitor for the onset of the hypertensive phase and if necessary reinstitute glaucoma medications at the earliest indication of a rise in IOP [32]. In a prospective study in adults, Pakravan and colleagues demonstrated that early aqueous suppression resulted in improved long-term IOP reduction and reduced the frequency of the hypertensive phase [33].

Complications

Although GDDs offer some benefits over other surgical procedures, they also carry the risk of significant complications. Complications which are particularly common or are unique to GDDs are described below.

Hypotony

Hypotony may be a serious and sight-threatening complication which may occur as an early, medium-term, or late complication of GDD surgery. It is often associated with a shallow or flat AC, maculopathy, choroidal effusion, serous retinal detachment, or suprachoroidal hemorrhage. Postoperative review should place an emphasis on assessing for the presence of hypotony and associated complications so that treatment can be initiated early. Further complications such as PAS, cataract, or phthisis may ensue if hypotony is not appropriately managed.

Buphthalmic eyes, which are large with little scleral rigidity, are particularly at risk of the complications of hypotony. Perioperatively, hypotony is avoided by the use of viscoelastic agents or AC infusion to maintain the AC. The use of appropriate-sized devices, small gauge needle entry, valved devices, and suture restriction of non-valved devices as described above reduces the risk of postoperative hypotony. In circumstances when hypotony occurs despite the above measures, assessment needs to be made as to whether this is due to over-drainage or under production of aqueous (ciliary body shutdown).

Over-drainage in the early postoperative period may occur due to:

- Leakage around the tube at the site of the scleral tunnel
- Failure of the valve mechanism in a valved device
- Lack of adequate restriction of a non-valved device
- Drainage via a different path, e.g., preexisting trabeculectomy exposed during GDD surgery, iatrogenic cyclodialysis cleft

Over-drainage may occur as a medium-term or late complication when a restricting ligature is released or an intraluminal stent removed in a non-valved device. Tube exposure (described below) may also result in leakage and over-drainage as a late complication. Management of clinically significant hypotony due to over-drainage will require surgical intervention to identify and remediate the underlying cause. The use of an AC infusion assists in identifying areas of aqueous leakage around the tube or through the tube when extraluminal restriction is not adequate. Suturing the tube tunnel on either side of the tube may stop leakage from this site; however, sometimes it is also helpful to use a small amount of Tenon capsule to plug the leak. Tubes which are draining because of inadequate restriction will require an additional tighter tie to be applied. In the extreme situation where these measures do not stop leakage, the tube is removed from the tunnel, which should be sutured closed. The surgeon should then decide whether to fashion a fresh tunnel or simply tuck the tube out of the way for later reinsertion, thus converting the surgery to the "two-stage" approach described at the beginning of the chapter.

Ciliary body shutdown as a primary cause of postoperative hypotony is most commonly encountered in eyes with glaucoma secondary to uveitis or in eyes that have previously undergone multiple ciliary ablations. Eyes which suffer a period of hypotony may additionally suffer ciliary body shutdown and aqueous hyposecretion as a consequence of ciliary body detachment with choroidal effusions. Primary ciliary body shutdown may require additional restriction or stenting of the tube to limit drainage while awaiting restoration of ciliary body function. In addition to aggressive management of any underlying inflammation, consideration should be given to artificially elevating the IOP by use of viscoelastic, BSS, gas (e.g., filtered air or 20% SF6), or a combination of these into the AC.

Tube Occlusion

Tube occlusion may occur early or late in the postoperative period, and for this reason, it is important in siting the tube and trimming its length to ensure that the tip is directly visible. In eyes having undergone cataract surgery, occlusion may be related to capsular remnants, lens pearls, or vitreous as described elsewhere in this chapter. Fibrin may also occlude the tube from intraocular inflammation postoperatively or blood in the presence of a hyphema.

Rates of tube obstruction in different pediatric glaucomas with different drainage devices range from 6% to 20% [34–44]. Surgical removal of the obstruction is often required. The options depend on the underlying cause but may involve a combination of tube flushing, AC washout, vitrectomy, iridectomy, or removal of the valve mechanism in an AGV.

Patients with a late-onset vitreous incarceration present with sudden, marked IOP elevation after months or years of good IOP control. B-scan ultrasound reveals no bleb over the plate, indicating blockage of the tube. If the patient can cooperate, vitreous incarceration can be seen in the tube tip at the slit lamp or with a gonioscopy prism in a dilated eye. Vinod and colleagues [23) recommend a surgical approach in such patients. It is not sufficient to simply perform a vitrectomy; these tubes are usually blocked with a plug of condensed vitreous that must be removed with end-grasping retinal forceps (Fig. 7.9); if only a simple vitrectomy is performed, this now amputated plug will be free to travel up the tube and cause permanent failure. Once the vitreous plug and remaining mobile vitreous are cleared from the tube, full function of the GDD is usually restored, and IOP returns to pre-occlusion levels; B-scan ultrasound will reveal the presence of a filtering bleb over the equatorial plate, confirming flow through the system.

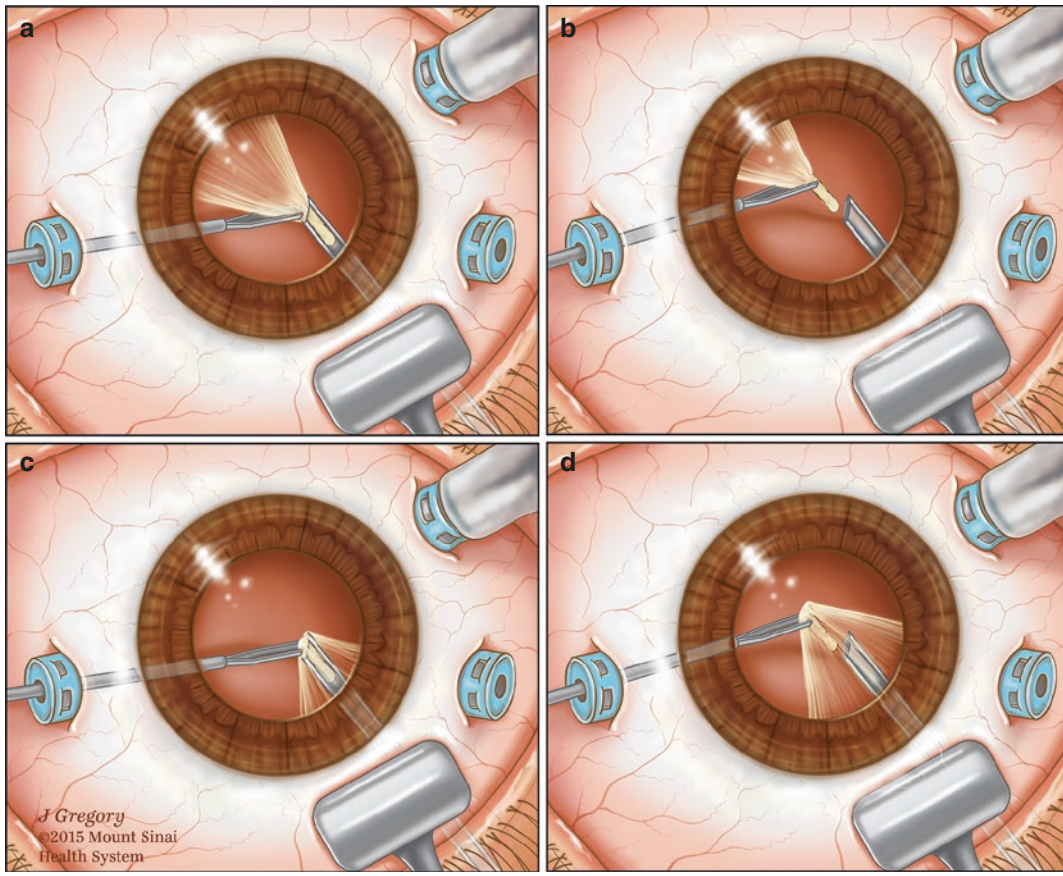

Fig. 7.9 Technique of pars plana vitrectomy to remove a vitreous plug occluding a tube as described by Vinod and colleagues. Note that a simple vitrectomy is insufficient – end-grasping forceps must be used to remove the condensed vitreous plug or the plug will be free to travel up the tube and cause permanent failure. (From Vinod et al. [30]. Illustration by Jill K. Gregory, CMI. Reprinted with permission from ©Mount Sinai Health System, New York NY, USA)

Tube Touch

Corneal touch and endothelial damage occurs when the tube is sited anteriorly in the AC and particularly if the intracameral portion of the tube is long. *Corneal opacification* occurs at the site of tube touch with endothelial damage resulting in *corneal edema* and *decompensation*. The tunnel for the tube should be constructed so that the tube enters the AC in front of and parallel to the iris. If the intracameral portion of the tube is too long, it may be observed at the slit lamp to sweep from side to side or forward and back when the patient blinks or when touching the eyelid on examination. In a young child, tube touch may be intermittent with eye rubbing, and a focal area of corneal edema near the tube tip seen at EUA may be the only clue that this is happening and may require shortening or repositioning of the tube.

Iris touch may occur if the tube is sited too far posteriorly in the AC. This may result in *chafing of the iris* with localized atrophy or *chronic uveitis*. This may also be associated with *corectopia,* particularly if the iris root is involved; corectopia on this basis is particularly common in eyes with Sturge-Weber syndrome (Fig. 7.10).

Lens touch results in cataract and may be a consequence of a tube which is too long, sited too far posteriorly, or in a radial rather than tangential direction.

The treatment of tube touch complications is ultimately surgical and is carried out by shortening and/or resiting the tube. In some cases of anterior segment tube-related complications, it is may be necessary to reposition the tube posteriorly through the pars plana [45].

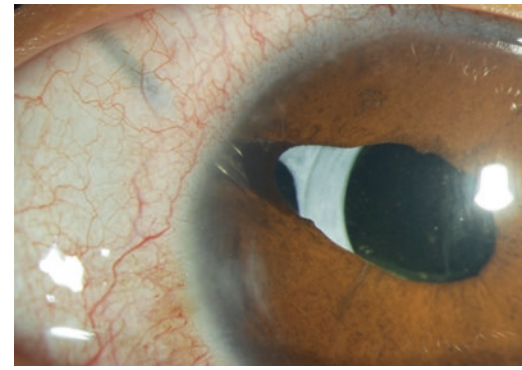

Fig. 7.10 Tube-related corectopia. Teenager with chronic uveitic glaucoma who underwent placement of an Ahmed Glaucoma Valve about 5 years earlier combined with phacoemulsification and injection of an Ozurdex™ dexamethasone implant (Allergan, Dublin, Ireland) who returns with maintained IOP control and good vision. Note the iris is dragged to the tube insertion. This may be avoided by performing a localized peripheral iridectomy and inserting the tube more posteriorly through the iridectomy so that the iris cannot adhere to the base of the tube. (Courtesy of James D. Brandt, MD)

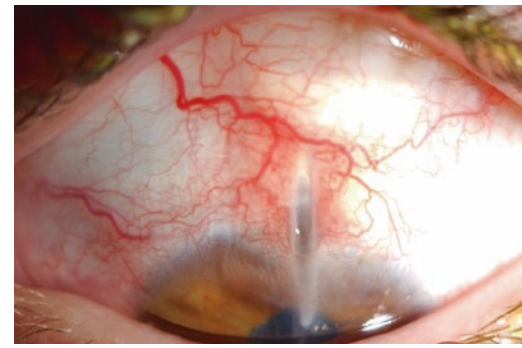

Fig. 7.11 Exposed tube. A teenage female with uveitis underwent Baerveldt tube surgery for secondary glaucoma. However, the tube tunnel was very superficial and anterior; note the long tunnel within the corneal stroma. Although the plate was sutured to the sclera, the tube was not sutured against the sclera (e.g., with a box suture) and as a consequence bowed anteriorly. Over time, the donor scleral patch graft overlying the tube melted away and the tube eroded through the conjunctiva, and the patient presented complaining of pain and discomfort. (Courtesy of Cecilia Fenerty MD, FRCOphth)

Erosion

Erosion of the GDD through the conjunctiva (Fig. 7.11) may result in infection *(endophthalmitis), leak, or hypotony*. Securing the plate and the tube with sutures as described above ensures the device does not sit raised over the underlying scleral plane with the overlying conjunctiva stretched over the device surface. The use of a patch graft cushions the conjunctiva from the ridge of the tube and the anterior plate and further reduces the risk of erosion. However, if the conjunctiva is under tension when replaced to cover the GDD (e.g., as a consequence of scarring from previous surgery or trauma), it is more likely to erode over the tube or plate.

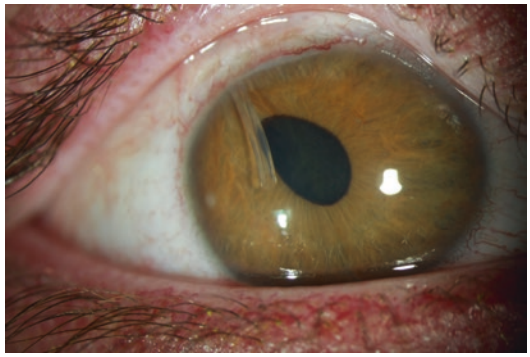

Fig. 7.12 Developmentally delayed teen with glaucoma associated with Sturge-Weber syndrome returned years later with a history of eye rubbing. IOP control was good and there was no erosion, but the plate had moved forward from its original position by about 3 mm so the tube was too long and risked causing corneal endothelial damage. The tube was everted through a small corneal incision 1 mm anterior to the limbus and shortened to about 2 mm in length. IOP control was maintained. Note that the pupil is drawn toward the tube entry point, a common late-term finding in eyes with Sturge-Weber syndrome. (Courtesy of James D. Brandt, MD)

Migration

Migration of the GDD may occur if the plate and tube are not securely sutured to the globe or the "wings" of a Baerveldt implant are placed above rather than behind the muscle insertions. As a consequence, the plate may end up advancing toward the limbus leading to "touch complications" or retracting toward the equator resulting in tube retraction out of the AC. With the device plate well secured, it is still possible for the tube to migrate out of the AC if it is not sutured securely to the sclera or with ocular growth (Fig. 7.12).

Changes in Intracameral Tube Length

Intracameral tube length may change with the IOP postoperatively. With a reduction in IOP, the dimensions of a child's eye may change with a reduction of the corneal diameter and the axial length. As a consequence, the intraocular portion of the tube may lengthen, and with that more "tube touch" complications are likely. Conversely, with elevated IOP following surgery, the corneal diameter and the axial length may increase, and the intraocular portion of the tube may retract from the anterior segment.

Ocular Motility Disturbances

Ocular motility disturbances may arise following GDD implantation, particularly when an eye has undergone prior strabismus surgery. Strabismus following GDD surgery should be managed by a surgeon familiar with both muscle surgery and pediatric GDD surgery, as these cases are very complex.

Outcomes

The World Glaucoma Association convened a Consensus Meeting on Childhood Glaucoma at its biennial global congress in 2013; the resulting monograph reported the results of a worldwide surgical consensus survey of glaucoma specialists and pediatric ophthalmologists with an interest in pediatric glaucoma surgery [46]. Almost half of experts (44%) preferred GDDs as the primary surgical procedure in glaucoma following cataract surgery (GFCS), with 30% preferentially using GDDs in uveitic glaucoma. Most stated they would use GDDs after failed trabeculectomy (82%) with the AGV being the most popular choice (63%) followed by the BGI (41%). Only 15% preferred to augment their surgery with adjunctive antimetabolites, and most preferred AC tube placement unless contraindicated.

Table 7.4 is a compilation of published studies on pediatric GDDs in the published literature from 1984 to 2017 which the reader may find useful [9, 12, 22, 35–44, 47–72].

The published success rates of GDDs in pediatric glaucoma vary widely between 54% and 90% [28, 40, 43, 73]. This is due largely to differences in the age of the child, underlying diagnosis, variation in surgical technique, and device employed as well as differences in the authors' definitions of success and failure. Despite these

Table 7.4 Published case series of glaucoma drainage devices in children, 1984–2017

	Study	Location(s)	Diagnosis	Implant	Eyes *(N)*	Mean follow-up (months)	1 year success (%)
1984	Molteno [47]	Otago (NZ)	Mixed	SP Molteno™	83	66	73
1988	Minckler [48]	Doheny Eye Institute (Los Angeles)	Mixed	SP Molteno™	13	22.8	54
1989	Billson [49]	Sydney	Mixed	DP Molteno	23	41.3	78
1991	Hill [35]	Doheny Eye Institute (Los Angeles)	Mixed	SP + DP Molteno	65	22.7	62
1991	Munoz [50]	KKESH (Riyadh)	Mixed	SP Molteno	53	18	68
1992	Lloyd [36]	Doheny Eye Institute (Los Angeles)	Mixed	SP Molteno	16	49.1	56
1992	Nesher [41]	Florida and Washington Univ.	Mixed	SP + DP Molteno	27	20	57
1993	Netland [51]	MEEI	Mixed	AGV + BGI	20	25	80
1995	Fellenbaum [37]	Doheny Eye Institute (Los Angeles)	Mixed	BGI	30	15	86
1995	Siegner [52]	MEEI and Univ California, Davis	Mixed	BGI	15	13.6	80
1997	Coleman [12]	UCLA	Mixed	AGV	21	16.3 ± 11.2	78
1997	Eid [38]	Wills Eye Institute (Philadelphia)	Mixed	Mixed	18	47.3	72
1997	Donahue [42]	Iowa	Mixed	BGI	23	19	61
1998	Cunliffe [53]	Otago (NZ)	Mixed	SP + DP Molteno	34	134.4	85
1999	Englert [39]	Duke University	Mixed	AGV	27	12.6 ± 8.2	85
1999	Hamush [54]	UCLA	Sturge-Weber	AGV	11	30.4 ± 19.1	79
1999	Huang [55]	Multiple	Mixed	AGV	11	13.4	91
2001	Djodeyre [43]	Madrid	Mixed	AGV	35	12.6	70
2002	Pereira [44]	Wills Eye Institute (Philadelphia)	Mixed	Mixed	10	50	80
2003	Morad [40]	Toronto	Mixed	AGV	60	24.3 ± 16	93
2004	Budenz [56]	Miami & Univ. of California, Davis	Mixed	BGI	62	23.4 ± 21.7	80
2004	Rodrigues [57]	Brazil	PCG	Susanna	24	24	88
2005	Chen [58]	MEEI	Mixed	AGV	52	26 ± 20	85
2005	Rolim De Moura [59]	Doheny Eye Institute (Los Angeles), Univ. of Southern California	Mixed	BGI	48	median 21 (4–95)	91
2005	Kafkala [60]	MEEI	Uveitic	AGV	7	36.8 (6–60)	69.9 IOP reduction rate
2006	Van Overdam [61]	Netherlands	Mixed	BGI	55	32 (2–78)	94

(continued)

Table 7.4 (continued)

	Study	Location(s)	Diagnosis	Implant	Eyes *(N)*	Mean follow-up (months)	1 year success (%)
2007	Autrata [62]	Czech Republic	Mixed	Molteno+ BGI	76	85 ± 78	91
2007	Souza [63]	UCLA	Mixed	AGV	78	61	80
2008	O'Malley Schotthoefer [22]	Duke Univ.	Mixed	AGV + BGI	70	66	92
2009	Sood [64]	Emory Univ.	Mixed	AGV + BGI	8	26.2 ± 9.5	75
2012	El Gendy [65]	USC	Mixed	BGI	20	46 ± 29	80
				AGV	11	33 ± 30	55
2014	Balekudaro [66]	India	Mixed	AGV	71	37.8 ± 32.1	97
2014	Razeghinejad [67]	Iran	PCG	AGV	33	32.6 ± 18.3	97
2014	Tai [68]	Univ. of Southern California	Mixed	BGI	45	??	87
2015	Chen [69]	UCLA	Mixed	AGV	119	73 ± 40	86
2015	Dave [70]	India	PCG	AGV	11	17.9 ± 9.3	91
2016	Mandalos [71]	United Kingdom	Mixed	Molteno+ BGI	69	45.7 ± 25.2	96
2017	Eksioglu [72]	Turkey	Uveitis	AGV	16	64.46 ± 33.56	56
2017	Vinod [9]	NYEEI	Mixed	PP BGI	37	78 ± 48.7	94.5

SP single plate, *DP* double plate, *AGV* Ahmed glaucoma valve, *BGI* Baerveldt® glaucoma implant, *PCG* primary congenital glaucoma, *KKESH* King Khaled Eye Specialist Hospital, *MEEI* Massachusetts Eye and Ear Infirmary, Boston, *NYEEI* New York Eye and Ear Infirmary

limitations, a few general conclusions can be drawn. Firstly, although many studies report success rates around 80% after 1–2 years of follow-up [37, 74], longer-term studies consistently report a steady decline to ~50% success after 5 years of follow-up requiring reinstitution of medical therapy [22, 56, 59, 61, 73]. Secondly, while it is difficult to directly compare success rates between different GDDs, a number of studies have demonstrated equivalent results among devices [22, 51, 73, 75].

In most reported case series, GDDs have been reserved for use in refractory pediatric glaucoma of mixed etiology where other medical and surgical procedures have failed to optimally control IOP. Primary congenital glaucoma and secondary glaucoma particularly associated with aphakia form the bulk of cases analyzed. A small case series of GDDs in aniridia showed a success rate of up to 88% at 1 year [76]. The use of BGI [77] and AGV [54] in Sturge-Weber syndrome has also been shown to successfully improve IOP control in refractory cases, though meticulous hypotony prevention in the postoperative period is strongly advocated. A number of studies looking at surgical outcomes of GDDs in pediatric uveitic glaucoma have shown between 80% and 100% success rates up to 40 months of follow-up [60, 78, 79]. In these cases, however, GDDs of smaller surface areas were used to minimize the risk of hypotony. GDDs associated with corneal grafting procedures have demonstrated higher failure rates both in terms of IOP control and graft longevity [66], with higher complication rates particularly in simultaneous combined procedures [80]. Ideally, IOP should be controlled prior to considering corneal surgery. And while a pars plana approach may reduce corneal complications, it risks tube occlusion with vitreous.

While GDDs have demonstrated a high cumulative probability of long-term success, sight-threatening complications may occur at any stage postoperatively, requiring lifelong follow-up [71]. The most common complications relate either to hypotony or to the tube itself [28, 35, 37, 38, 43, 73, 81, 82].

Hypotony may occur at any stage during the early or late postoperative period, with the pediatric population at greater risk due to reduced scleral rigidity. The reported incidence of hypotony and flat AC varies, ranging from 0% to 25.7% in pediatric patients [12, 22, 34, 35, 37–41, 43, 50, 51, 73]. The results of different GDDs vary widely and are dependent on surgical technique and underlying diagnosis with underestimation also likely due to paucity of reliable IOP assessment in children. Early hypotony has been reported in AGVs to be as present in up to 7% of cases [66]. Rates of choroidal effusion range from 0% to 22% [12, 34, 35, 38–41, 43, 44, 53, 66, 73], while the most devastating complication of suprachoroidal hemorrhage has been documented to range from 0% to 13% [12, 34–38, 53, 73].

Tube-related complications are a heterogeneous group, encompassing erosion, occlusion, migration, iris or lens touch, and corneal decompensation related to corneal endothelial touch [73]. Interestingly, a recent retrospective comparative study of 69 pediatric eyes and 145 adult eyes found higher rates of corneal decompensating in the adult population occurring earlier in this group [83].

Tube migration and retraction may occur secondary to normalization or elevation of IOP, respectively, and tend to occur in younger patients with buphthalmic eyes. Rates of tube-cornea touch vary from 5.7% to 20% [37, 39, 40, 50, 73], with most identified cases requiring revision. Conversely, if tube retraction occurs, then a number of surgical options have been described including the use of a tube extender, a segment of 22-gauge intravenous catheter [84], or angiocatheter material [85].

Erosion of the tube or the scleral plate carries a significant risk of endophthalmitis and should be repaired immediately using a patch graft or where required explantation of the device in cases of plate erosion [74]. The reported incidence of erosion or extrusion of the tube or scleral plate ranges from 0% to 13% in pediatric patients [12, 34, 35, 37–44, 50, 51, 53, 56, 73]. Gedde and colleagues [86] noted that exposure of the tube was present in all cases of late-onset endophthalmitis associated with the BGI. Early and late postoperative endophthalmitis rates associated

with GDDs in children have been reported between 0% and 5% [12, 34, 35, 37, 38, 40–43, 50, 51, 53, 56, 73].

Motility issues and strabismus should also be considered in children particularly where binocularity is present [22]. The incidence of this complication ranges from 0% to 11% in children with various devices [12, 34, 37, 38, 40, 43, 51, 56, 87]. Permanent motility disorders represent a late complication and may be due to mechanical restriction of the extraocular muscles secondary to adhesion or scarring or to a large bleb or episcleral plate.

Options After Failed Surgery

Elevated IOP after GDD insertion may be due to either tube obstruction at either the tube tip or in the valve mechanism or GDD failure due to bleb encapsulation. A number of treatment options are available once GDDs fail to control IOP effectively either through plate fibrosis, scarring, or bleb encapsulation. Recommencement of topical glaucoma medications is the usual first-line option. Failing this, of the pediatric glaucoma experts surveyed [46], 26% would proceed with cyclodestruction, 26% would revise the GDD (capsule excision with or without anti-scarring agent), and 23% would insert a second GDD. A nonrandomized chart review of 17 eyes suggests that secondary cyclodestruction versus secondary GDD has equivocal results at 2 years [64]. The amount of cyclodestructive treatment required to achieve the desired degree of IOP reduction may be difficult to titrate and may be associated with high rates of retinal detachment, phthisis bulbi, and other vision-threatening complications [74]. Shah et al. demonstrated that after failed GDD, an additional GDD offered better IOP control than revision by excision of an encapsulated bleb [88]. A further retrospective case series of 22 eyes did not demonstrate higher-than-expected rates of complications associated with GDDs [89]; however, both these studies were in an adult population.

Conclusion

Glaucoma drainage devices are increasingly useful in the management of childhood glaucoma. They can be used as a primary procedure in certain situations or as a secondary procedure where more conventional surgery (e.g., angle surgery) has been carried out and failed. We hope that this chapter has provided a comprehensive review of why, when, and how GDDs should be used in the treatment of childhood glaucoma.

The reader may be excused for coming away from this chapter with some pessimism, especially after the main conclusion above that these devices have a 50% failure rate at 5 years. We believe it is important to put this in perspective, however. The eyes in which GDDs are generally used are those same eyes that would have almost certainly gone blind prior to the introduction of these devices and are best implanted sooner than later to optimize long-term visual prognosis. The authors have patients who maintain useful and sometimes excellent vision many years (even decades) after implantation in early childhood. We encourage those caring for children with glaucoma to add the use of GDDs to their surgical portfolio. This recommendation is particularly important in resource-constrained settings now that low-cost GDDs are available worldwide. Successful GDD cases make all the hard work of managing these children among the most rewarding long-term aspect of childhood glaucoma care.

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