Editorial

Childhood Glaucoma

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Childhood glaucoma is a heterogeneous cluster of disorders occurring in early years of life. According to the American Academy of Ophthalmology[1], congenital glaucoma is present at birth or diagnosed up to 12 months of life. Infantile glaucoma is present from 1 year to 3 years and after 3 years onwards, it is termed as juvenile glaucoma. This division however can be arbitrary in our country where child is brought often too late for clinical advice.

Most of the pediatric glaucoma have no specific identifiable cause and are considered as primary glaucoma. However, when glaucoma is associated with some specific disease, it is called as secondary glaucoma. Some of the disorders associated with childhood glaucoma include Axenfeld – Rieger syndrome, Sturge-Weber syndrome, Aniridia and Neurofibromatosis. The chronic use of topical steroids, trauma and cataract surgery remain the other associated factors.

About 10% of primary congenital/infantile glaucoma are inherited with specific gene mutation. The secondary glaucoma associated with neurofibromatosis and Aniridia are inherited by autosomal gene, which can be passed on to 50% of affected children[2].

The prevalence of primary congenital glaucoma (PCG) is not known in this country but it occurs in about 1:10,000 live births in USA[3]. In Saudi Arabia PCG is estimated to have a prevalence up to 10 times higher than in USA occurring in 1:2500 to 3000 live births[4].

The condition is usually managed surgically. The surgical techniques are designed to eliminate the resistance of aqueous outflow created by the structural abnormalities in the anterior chamber angle. These congenital changes include presence of non-permeable Barkan's membrane covering the trabecular meshwork and anterior insertion of ciliary body and Iris, overlapping the trabecular meshwork[5,6].

Childhood glaucoma poses a huge diagnostic challenge, as young children are uncooperative and difficult to examine in routine setup. The Intraocular pressure (IOP) reading can be difficult to obtain even under sedation as these sedating agents can influence the readings of IOP so these children should be examined and referred to a special unit equipped with handheld tonometer, portable slit lamp and anesthetic facilities.

Managing childhood glaucoma is one of difficult tasks for glaucoma specialist. The goal of treatment should be better control of IOP and preservation of vision. Glaucoma surgery remains the eventual treatment. Anti-glaucoma drops are used to stabilize the IOP until surgery is scheduled. Because of serious side effects of medical therapy in the young age, topical drops are carefully chosen. Usually Beta-blockers and Alpha agonists are avoided because of their potential harmful side effects.

Broadly, the surgical treatment is divided into 3 categories.

1. Angle surgeries like Goniotomy and Trabeculotomy are meant to enhance the aqueous outflow pathway. Goniotomy is usually preferred if cornea is clear. In case of hazy cornea when angle structures cannot be visualized, trabeculotomy is the procedure of choice. It is generally accepted that success rate of these 2 procedures is similar with same degree of disease severety[7,8,9]. However, advocates of Goniotomy argue in its favor when considering the long-term effect especially for future glaucoma surgery such

as trabeculectomy as conjunctiva and sclera remains untouched, increasing the future success of drainage surgery. With the availability of anti-fibrotic Mitomycin C (MMC), this may not be the case[10]. Recently 360 degrees canaloplasty is taking momentum, which can be performed from outside or inside the eye. The success rate of circumferential 360 degrees trabeculotomy in eyes with PCG varies from 77% – 92% after 1 – 4 years[11,12,13].

2. The second set of surgical category is to create external outflow like trabeculectomy or placement of glaucoma drainage devices (GDD). The result of trabeculectomy in children has been poor but with the advent of adjunctive use of MMC, surgical outcome has improved. Our results with MMC augmented trabeculectomy in PCG has shown that 58% of children up to 3 years of age maintained the IOP of less than 15 mm Hg at the end of 1 year follow-up[14]. Molteno first published his results of use of GDD in children in 1973[15]. Since then several researchers have used Ahmed Glaucoma Valve and Baerveldt implant in children. At 1 to 2 years follow-up, many workers have reported a success rate of 80% but this has reduced to 50% in long-term follow-up[16,17]. If one looks at the literature, there is no superiority of one device over other. However, Baerveldt implant may provide slight better long-term IOP control but Ahmed implant has shown lesser complications[18,19].
3. The third category is procedures causing reduction of aqueous production. This includes cyclo-destructive procedures usually carried out with trans-scleral diode laser of 810 nm. This procedure is traditionally reserved for refractory cases where routine surgery has failed. However, accurate laser application in big eyeball with distorted landmarks can be extremely difficult. Endoscopic diode laser can be used in these cases but its use in phakic eyes remains controversial.

The best surgical procedures in childhood glaucoma should consider age of the patient, underlying cause of glaucoma, associated ocular factors, any previous ocular surgery, and extent of visual damage and above all surgical expertise of treating specialist and facilities available locally.

The diagnosis of childhood glaucoma not only affects the child and parents emotionally but also can hinder child's education in long term and also can be burden on the family to finance multiple surgeries and hospital costs. In these cases, it may be prudent for primary physician to refer such child to an institute where these factors can be addressed.

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