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Perspective

Potential Developments in Primary Congenital Glaucoma Therapy

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DESCRIPTION

Primary Congenital Glaucoma (PCG), also known as infantile glaucoma, is a rare but potentially devastating condition characterized by elevated Intraocular Pressure (IOP) due to developmental abnormalities of the trabecular meshwork and aqueous outflow system. Early diagnosis and prompt intervention are essential to prevent irreversible optic nerve damage and preserve vision in affected infants and children. In recent years, significant advancements have been made in the management of PCG, offering improved treatment outcomes and quality of life for patients. This article explores the latest interventions for PCG, including surgical techniques, medical therapies, and emerging technologies.

PCG typically presents within the first few years of life, with onset usually occurring in the neonatal or infancy period. The classic triad of symptoms includes photophobia, epiphora (excessive tearing), and blepharospasm (eyelid squeezing). As the disease progresses, corneal edema, optic nerve cupping, and enlargement of the globe (buphthalmos) may become evident on clinical examination. The underlying pathophysiology of PCG involves impaired aqueous outflow due to developmental abnormalities of the trabecular meshwork, Schlemm's canal, and the anterior chamber angle. This results in increased resistance to aqueous humor drainage, leading to elevated IOP and subsequent optic nerve damage if left untreated. The management of PCG typically involves a combination of medical therapy and surgical intervention, adjust to the individual patient's clinical presentation, disease severity, and response to treatment. The primary goals of treatment are to lower IOP, preserve visual function, and prevent progression of optic nerve damage.

Medical therapy plays a limited role in the management of PCG and is primarily used as adjunctive treatment to lower IOP temporarily before surgical intervention. Topical medications such as prostaglandin analogs, beta-blockers, and carbonic anhydrase inhibitors may be prescribed to reduce aqueous production and increase outflow facility. However, their efficacy in infants and young children is limited by poor ocular penetration and systemic side effects. Trabeculotomy, either as an ab externo or ab interno procedure, involves the surgical opening of Schlemm's canal and the trabecular meshwork to improve aqueous outflow. This technique is particularly effective in infants and young children with PCG, offering high success rates in lowering IOP and preserving vision.

Trabeculectomy with or without antimetabolites (e.g., mitomycin-C) may be performed in cases of refractory or advanced PCG with extensive scarring of the trabecular meshwork. This procedure creates a filtering way to facilitate aqueous drainage from the anterior chamber to the subconjunctival space, effectively lowering IOP. Goniotomy involves the surgical incision of the trabecular meshwork to create a passage for aqueous outflow. This procedure is typically performed using a gonioprism under direct visualization with a surgical microscope and is particularly suitable for cases of isolated trabecular meshwork dysgenesis. Cycloablative procedures, including Diode LaserTransscleral Cyclophotocoagulation or Endoscopic(TSCP) Cyclophotocoagulation (ECP), may be considered as adjunctive therapy in cases of refractory PCG with inadequate IOP control. These procedures target the ciliary body to reduce aqueous production and lower IOP.

Minimally Invasive Glaucoma Surgery (MIGS) procedures, such as the iStent implant or the Kahook Dual Blade, offer a less invasive approach to lowering IOP by bypassing the trabecular meshwork and improving aqueous outflow. These procedures are particularly suitable for mild to moderate cases of PCG and may reduce the need for more invasive surgical interventions. Aqueous shunt devices, such as the Ahmed valve or the Baerveldt glaucoma implant, provide an alternative pathway for aqueous drainage in cases of refractory PCG with failed trabeculectomy or trabeculotomy. These devices are particularly effective in maintaining long-term IOP control and reducing the risk of surgical complications. The prognosis for PCG depends on the age at diagnosis, severity of disease, and response to treatment. Early diagnosis and prompt intervention are critical for optimizing visual outcomes and preventing irreversible vision loss. With timely and appropriate management, the majority of

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children with PCG can achieve stable IOP control and preservation of visual function. However, despite advances in treatment, some cases of PCG may be challenging to manage, particularly those with advanced disease, extensive optic nerve damage, or associated systemic abnormalities. These cases may require lifelong monitoring and multidisciplinary care to address both ocular and systemic comorbidities.