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Case report

Retinal pigment epithelium changes in pediatric patients with glaucoma drainage devices

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ABSTRACT

Purpose: Retinal changes secondary to hypotony are usually described as wrinkling or folding of the inner portion of the choroid, the retinal pigment epithelium (RPE), and the outer retinal layers in the macular area due to scleral wall collapse. We describe a new retinal finding in children with suspected hypotony after implantation of Baerveldt Glaucoma Implant (BGI).

Observations: Four patients in our series developed significant RPE defects after BGI implant. The RPE defects appeared as elongated white lines observed solely in the posterior pole, in no particular pattern, and seemed to be worse in infants with anterior segment dysgenesis and with collagen disorders.

Conclusion and importance: Children have thinner and more elastic scleral walls than adults. This characteristic may cause the inward scleral wall to collapse when the eye is hypotonic. The resulting redundancy of the retina leads to wrinkling and RPE defects characterized by hypopigmented lines predominantly in the macular area. Such findings, to our knowledge, have not been previously reported in pediatric patients.

1. Introduction

Glaucoma drainage devices such as the Baerveldt Glaucoma Implant (BGI) (Abbott Medical Optics, Santa Ana, CA) are used in pediatric glaucoma patients when control of the intraocular pressure (IOP) is not achieved with medical therapy or with angle surgeries. Success rates after BGI implantation have been reported to be 80–94% in several studies.[1–7] However, postoperative complication rates associated with shunt surgeries remain high, with most early complications occurring secondary to post-operative hypotony, including choroidal effusions and/or hemorrhages, shallow or flat anterior chambers (AC) with or without induced aqueous misdirection, or retinal changes.[8] Post-operative hypotony is a significant occurrence following pediatric BGI implantation, with greater than one-third of patients experiencing hypotony within the first 6 months following surgery.[9]

There is a paucity of information regarding the specific, chronic retinal changes that can occur in pediatric patients following this complication, as well as their pathologic mechanism. Our purpose is to describe a new retinal finding in children after implantation of Baerveldt Glaucoma Implant. We suspect these changes occurred during a period of hypotony after implantation of the BGI.

2. Findings

2.1. Case 1

A seven-month old male with infantile glaucoma underwent trabeculotomies in both eyes within the first month of life. Due to poorly controlled intraocular pressure of 28 mm Hg, a BGI was implanted in the left eye. Five months later, an exam under anesthesia revealed an IOP of 11 mm Hg (Fig. 1) and hypopigmented RPE changes involving the temporal half of the posterior pole of the left eye (Fig. 2).

2.2. Case 2

A four-month old infant with SHOX gene mutation, and Peters anomaly in both eyes presented with congenital glaucoma and uncontrolled IOP of 40 mm Hg in the right eye and 30 mm Hg in the left eye. The patient underwent corneal transplants in both eyes and BGI placement on the right eye, followed by the left eye two weeks later.

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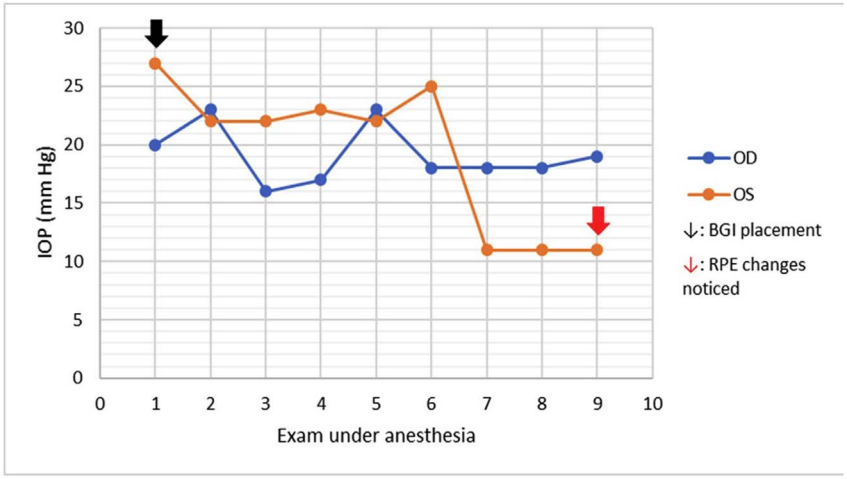

Fig. 1. Case 1 IOP measurements at time of BGI placement and of retinal findings.

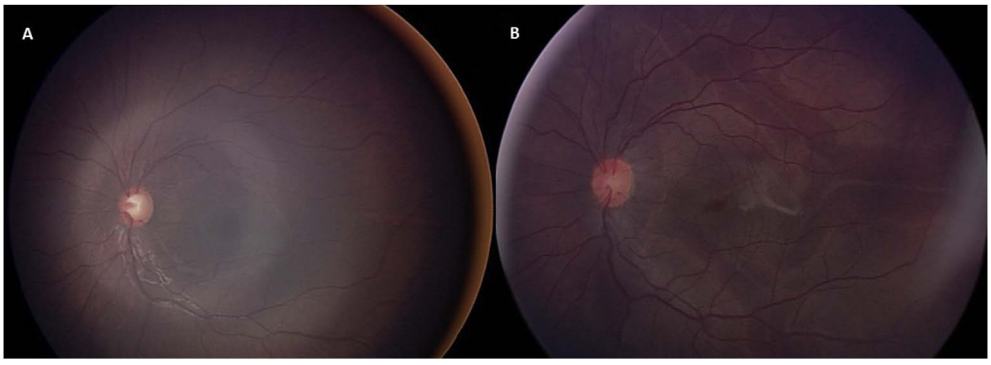

Fig. 2. A. Fundus picture before Baerveldt Glaucoma Implant placement. B. Retinal pigment epithelium changes noticed after BGI placement.

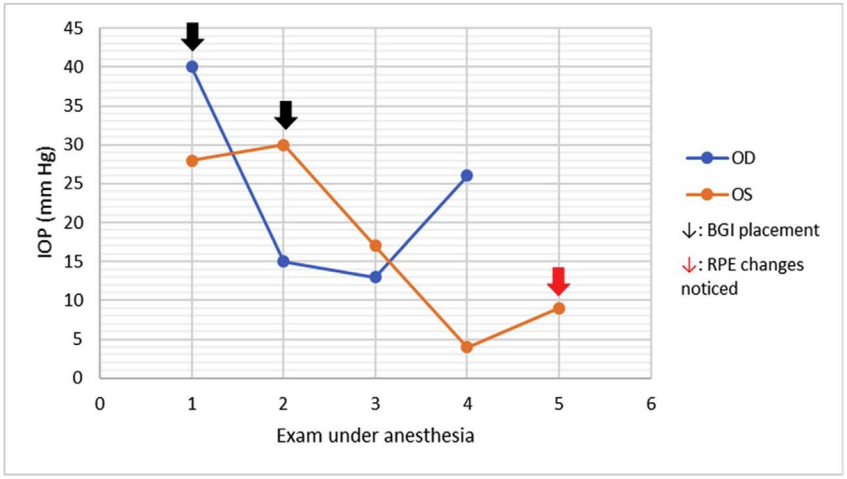

Fig. 3. Case 2 IOP measurements at time of BGI placement and of retinal findings.

Thirteen months after surgery, an exam under anesthesia revealed an IOP of 9 mm Hg in the left eye (Fig. 3), as well as RPE changes in the posterior pole extending from the optic disc to the foveal area (Fig. 4).

2.3. Case 3

A one-year old female with an undefined collagen vascular disorder, hip dysplasia and club feet was referred for lens opacity in the left eye and bilateral corneal opacity and congenital glaucoma associated with Peter's Anomaly and Axenfeld Rieger's Syndrome. Due to uncontrolled IOP in both eyes of approximately 40 mm Hg, a pars plana vitrectomy and lensectomy with BGI placement was performed in the left eye, followed by a pars plana vitrectomy, penetrating keratoplasty and BGI placement on the right eye 4 weeks later. An exam under anesthesia 3 months later showed IOP on the left eye of 6 mm Hg (Fig. 5) and fundus RPE changes compromising the macular area (Fig. 6).

2.4. Case 4

A four-month old female presented with endogenous candida endophthalmitis on the right eye at 34 weeks gestational age. She underwent pars plana vitrectomy and lensectomy. Subsequently, she developed aphakic closed angle glaucoma with IOP 35 mm Hg and underwent BGI placement. On follow-up exams, IOP fluctuated between 21 and 32 mm Hg. Nine months later, an exam under anesthesia revealed IOP of 29 mm Hg (Fig. 7) and RPE changes in the macular area