Ahmed Glaucoma Valve Implant for Childhood Glaucoma in Sturge-Weber Syndrome with Choroidal Hemangioma

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We present our experience with implantation of an Ahmed glaucoma valve in a 9-year-old girl with Sturge-Weber syndrome with choroidal hemangioma. In this case of childhood glaucoma, the choice of surgical procedure should be based on the efficacy and elimination of intra-and post-operative complications. An Ahmed glaucoma valve offered safety and efficacy in controlling glaucoma. This technique avoided sudden drops in intraocular pressure and decreases the risk of intraoperative and postoperative choroidal effusion and hemorrhage. The surgical procedure was not complicated: a one-staged implant was satisfactory and a prophylactic sclerostomy was not required. *(Chang Gung Med J 2006;29:528-31)*

Key words: Ahmed glaucoma valve, Sturge-Weber syndrome, choroidal hemangioma.

The best surgical procedure for childhood onset glaucoma in Sturge-Weber syndrome (SWS) combined with choroidal hemangioma is very challenging. The presence of a choroidal hemangioma, with a thin-walled endothelium, might increase the risk for expulsive hemorrhage when sudden intraoperative hypotony develops.[1] Goniotomy and trabeculotomy are less effective in childhood SWS.[2] The results of trabeculectomy without anti-scarring medicine are poor,[3,4] but adjunctive anti-metabolites to modulate wound healing can cause postoperative hypotony and other risks.[5,6] The choice of surgery should be based on the efficacy and elimination of intra- and post-operative complications.

This article describes primary implantation of an Ahmed glaucoma valve (AGV), an aqueous shunting device with a unidirectional valve design to avoid postoperative hypotony, in a 9-year-old girl with SWS with choroidal hemangioma. Our experience suggests that AGV minimizes potential complications and offers good control of intraocular pressure (IOP).

CASE REPORT

A 9-year-old girl with a right side facial nevus flammeus (Fig. 1) was referred in May 2001 with a history of glaucoma in the right eye, which had been under treatment with a β-blocker (Bunolgan), a topical carbonic anhydrate inhibitor (Trusopt) and 2% pilocarpine. Her best-corrected visual acuity (BCVA) was right eye (RE) 20/30 and left eye (LE) 20/20. The IOP was RE 25 mmHg and LE 12 mmHg. The optic nerve heads were cupped with cup-disc ratios of RE 0.8 and LE 0.3. Fundoscopy and transverse

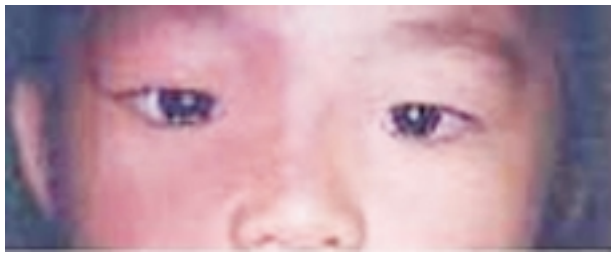

Fig. 1 A girl with a right side facial nevus flammus.

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ultrasound B-scan showed a large choroidal hemangioma over the posterior pole. (Fig. 2) In addition, the MRI revealed a vascular malformation at the right temporal lobe.

Latanoprost (Xalatan) was given to her and the IOP was controlled around 18-22 mmHg in the right eye during the following two years. In July 2003, IOP elevated up to 31 mmHg (RE) and visual acuity started to deteriorate (BCVA 20/50). We decided to place the Ahmed Glaucoma Valve to control IOP. An uneventful surgical procedure without posterior sclerostomy was undertaken under general anesthesia in August 2003. Ahmed glaucoma valve was placed over the superior temporal quadrant in the right eye. (Fig. 3) Shallow anterior chamber occurred on the first postoperative day and resolved spontaneously thereafter. Neither severe choroidal effusion nor intraocular hemorrhage was observed during and after the surgery. Choroidal hemangioma remained stable without any choroidal expansion postoperatively. Postoperative IOP was 7 mmHg (1 day), 11 mmHg (1 week), 16 mmHg (1 month), 21 mmHg (3 months), 18 mmHg (6 months) and 19 mmHg (1 year), respectively, without antiglaucoma medication. The visual acuity returned to preoperative BCVA 20/50 in one month postoperatively.

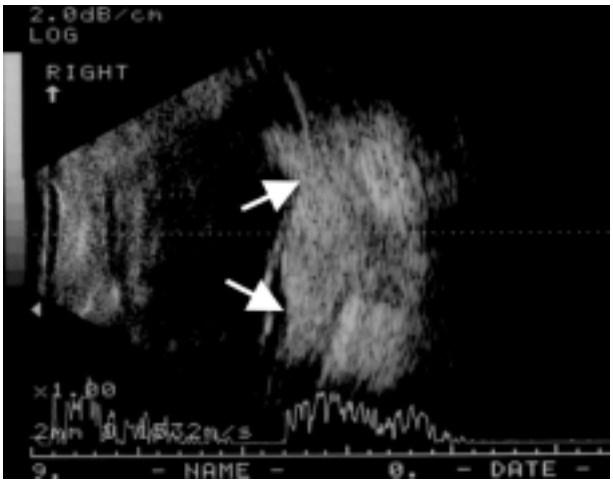

Fig. 2 A transverse ultrasound B-scan showed a large choroidal hemangioma over the posterior pole (arrows).

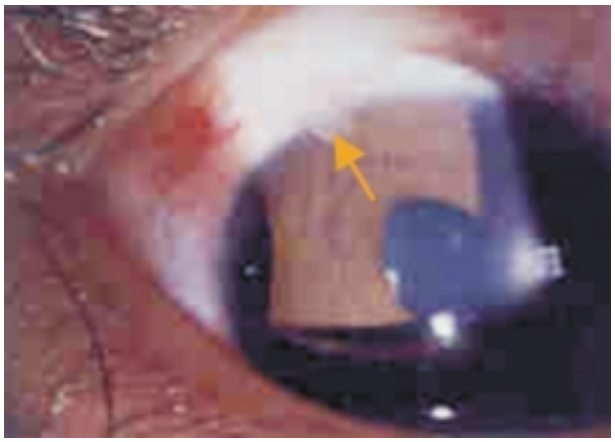

Fig. 3 An Ahmed glaucoma valve was placed over the superior temporal quadrant in the right eye (arrow).

DISCUSSION

The clinical manifestation of glaucoma associated with SWS is usually present when the facial hemangioma involves the lids or conjunctiva. The onset of glaucoma may present from infancy to early adulthood.[7] The mechanism of IOP elevation could be related to developmental anomalies of the anterior segment, increased episcleral venous pressure, or both.[8] Although medications may be sufficient to control glaucoma that occurs in later life, the early onset glaucoma usually requires surgical intervention.[2] Several literatures suggested that goniotomy and trabeculotomy offer good results when the glaucoma occurred in infancy, in which the developmental angle anomaly was supposed to be present.[2,9] But goniotomy and trabeculotomy have lower success rates when done for SWS than when used for primary congenital glaucoma. This has led to the opinion that an additional filtering surgery to bypass the episcleral veins is needed to achieve long-term effectiveness.[10,11] A primary procedure of combined trabeculotomy-trabeculectomy was hence used in SWS with glaucoma and better results were achieved.[12-14] Several studies have reported on the use of an aqueous shunt insertion for the management of glaucoma in SWS.[15-18]

The major surgical concerns in this case were the age of the patient and the presence of a large choroidal hemangioma. For a 9-year-old patient, a goniotomy or trabeculotomy would not be suggested because it would be less successful.[2,10,11] With a fragile choroidal hemangioma, any intraocular surgery carries a higher risk of massive hemorrhage. The best surgical modality would be one that produces the least hypotony during the procedure. Patients with SWS receiving a trabeculectomy still have the potential sight-threatening complications of choroidal

effusion and hemorrhage when IOP suddenly decreases,[1,2,10,11] and a prophylactic sclerostomy is usually required.[10-14] The Ahmed glaucoma valve implant has a unidirectional valve which prevents postoperative hypotony. It offers potential advantages over valveless implants, in which internal occlusion or external ligature is usually needed to avoid postoperative hypotony.[18,19] It was introduced in Taiwan in 2000 and is the only officially approved aqueous shunt in Taiwan. Under the health insurance system in this country, it is assumed to be a last resort in complicated glaucoma when a conventional trabeculectomy is contraindicated or not available. In our patient, a prophylactic sclerostomy was not needed during the surgical procedure. Postoperative hypotony was transient and the IOP returned to normal within a week. Furthermore, the choroidal hemangioma did not change significantly after surgery, and exudative retinal detachment, a common situation associated with an underlying hemangioma, did not occur.[1] Although there are several studies reporting surgical results for childhood glaucoma in Sturge-Weber syndrome,[2,10-18] to our knowledge, this is the first case report to address the concomitant presence of choroidal hemangioma in Sturge-Weber syndrome.

In summary, an Ahmed glaucoma valve offers safety and efficacy in controlling glaucoma in pediatric Sturge-Weber syndrome combined with choroidal hemangioma. The surgical procedure is not complicated: a one-stage implant is satisfactory and a prophylactic sclerostomy is not required. This technique avoids sudden drops in IOP and decreases the risk of intraoperative and postoperative choroidal effusion and hemorrhage.

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Sturge-Weber 症候群合併脈絡膜血管瘤之幼年型青光眼：使用 Ahmed 青光眼瓣膜手術成功之病例

吳秀琛 林耕國

本文報告一位九歲 Sturge-Weber 症候群合併脈絡膜血管瘤之女童接受 Ahmed 青光眼瓣膜手術成功控制眼壓之病例。臨床上面對這類極特殊之幼年型青光眼的手術方法除了必須考慮成功率以外，很重要的也需防止術中與術後可能發生的併發症。本案例的經驗証實 Ahmed 青光眼瓣膜確實能夠有效控制眼壓並提供足夠的安全性。這種技術主要可以避免術中與術後眼壓急遽下降導致嚴重的脈絡膜滲出和出血；此外手術步驟亦不是太複雜：單次手術植入即可完成而無需預先鞏膜切開術之執行。(長庚醫誌 2006;29:528-31)

關鍵字：Ahmed 青光眼瓣膜，Sturge-Weber 症候群，脈絡膜血管瘤。

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