

# Quadrantic Partial Thickness Sclerectomy for Treatment of Uveal Effusion Syndrome

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**Purpose:** To report our experience with partial thickness sclerectomy in patients diagnosed as idiopathic uveal effusion syndrome.

**Study design:** Prospective case series

**Place and Duration of Study:** Layton Rehmatullah Benevolent Trust (LRBT) eye hospital, Lahore January 2010 to August 2013.

**Material and Methods:** Four eyes of two patients (one male and one female) with bilateral idiopathic uveal syndrome were included in study. The diagnosis was clinical.

**Results:** Both patients showed improvement clinically in visual acuity and in funduscopy in term of retinal reattachment.

**Conclusion:** Patient with uveal effusion syndrome responded better to surgical procedures like sclerectomy and didn't respond to medical treatment.

**Key words:** Uveal effusion syndrome, quadrantic sclerectomy, exudative retinal detachment, intraocular pressure (IOP).

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It was 1963<sup>1</sup> when Schepens and Brockhurst coined the term "uveal effusion". They reported choroidal and ciliary body detachment with exudation. In 1982, Gass and Jallow introduced term "idiopathic uveal effusion syndrome" to describe serous detachment of choroid, ciliary body and retina of unknown origin<sup>2</sup>. It is a rare ocular disorder affecting predominantly healthy young males and involvement is commonly bilateral. It is a diagnosis of

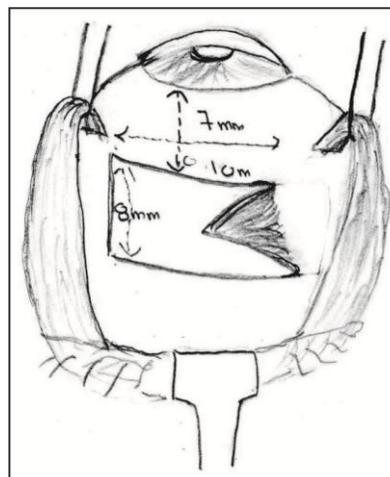
exclusion. The exudative retinal detachment follows a typical pattern which begins inferiorly. The anterior fibers of sclera attaching the choroid are long and tangentially oriented than posterior fibers so more fluid accumulation is seen anteriorly<sup>3</sup>. Other features are dilated episcleral vessels, blood in canal of schlemm, normal intraocular pressure (IOP), elevation of sub retinal and cerebrospinal fluid (CCF) proteins<sup>4,5</sup>. Marked anatomical alterations may be evident at the

level of retinal pigment epithelium with so called "leopard spot" hyper pigmentation<sup>1</sup>. There is no evident intraocular inflammation<sup>1</sup>. The natural history of disease shows activation and remission pattern and if patient is not treated timely, there may be permanent visual loss due to structural damage to photoreceptors and intra-retinal fibrosis. The milder forms usually resolve but visual prognosis for eyes presenting with bullous retinal detachment is poor<sup>6</sup>.

It responds poorly to medical treatment like corticosteroids and anti metabolites<sup>6</sup> and non-surgical treatment. Response to surgical options like scleral buckling and pars plana vitrectomy (PPV) is also not convincing. The successful retinal re attachment can be obtained by scleral thinning procedures like quadrant partial thickness sclerectomy.

### MATERIALS AND METHODS

Four eyes of two patients (one male and one female) with bilateral idiopathic uveal syndrome were included in study. The diagnosis was clinical. The patients with posterior scleritis, inflammatory orbital disease, arteriovenous fistula, pan retinal laser, recent retinal surgery, ocular trauma and ocular neoplasm were excluded. There was no history of taking drugs like sulfonamide and acetazolamide. The technique of sclerectomy involved the following steps. 360 degree peritomy around limbus is done. Four recti muscles are secured with bridle sutures as for retinal buckling procedure. One quadrant is exposed at a time. After securing homeostasis a rectangular area is marked with scleral marker 7 mm from the limbus measuring 10mm horizontally and 8 mm vertically. With 15 no blade a partial thickness scleral flap, almost 80% in thickness is raised and removed. The scleral thickness at this place is now 20%. The procedure is repeated in all four quadrants. Subretinal fluid may or may not be drained. The author recommends the drainage for early rehabilitation Fig 1.

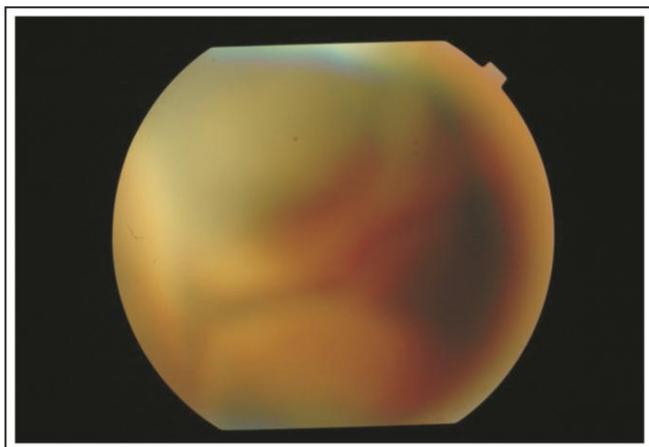


**Fig. 1:** Diagramtic representation of the procedure.

### Case 1

A young male presented with gradual decrease in vision in both eyes for the last six months. His vision

was perception of light (PL) in both eyes with exudative retinal detachment seen on fundus examination, fig. 2. His intraocular pressure (IOP) was 10 mm Hg both eyes. Quadrantic sclerectomy was done in both eyes with 1 week interval, fig. 3. Subretinal fluid (SRF) was drained in left eye but not in right eye. Hypotony was managed in left eye with intravitreal C<sub>3</sub>F<sub>8</sub> injection after SRF drainage.



**Fig. 2:** Preoperative fundus picture.

**Case 2**

A young female presented with gradual decrease in vision for the last 9 months. The vision was light perception (PL) in both eyes with normal IOP (8 mm Hg in right 6 mm Hg in left). She was hypermetropic of +7 diopter sphere in both eyes. Her axial length was 19 mm in both eyes. Her right eye was operated and quadrantic sclerectomy but she didn't report back for left eye surgery after 2 follow up visits.

**Table 1:** Patient data showing pre and post-operative vision and IOP.

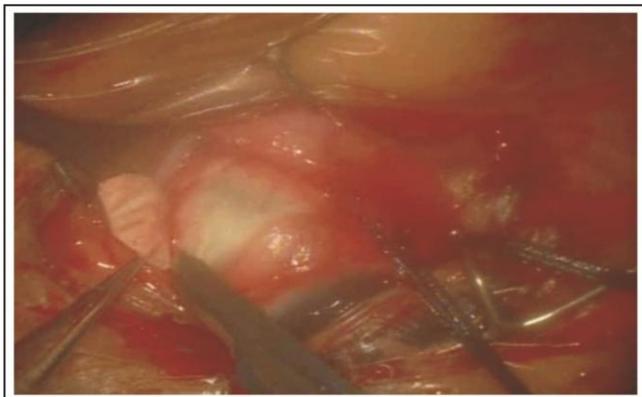
Serial No.	Eye	Pre-Operative		Post-Operative					
		Visual Acuity	IOP (mm Hg)	Visual Acuity			IOP (mm Hg)		
				1 wk	3 wks	12 wks	1 wk	3 wks	12 wks
Case 01 Male	Rt	PL+	10	6/36	6/24	6/24	10	10	10
	Lt	PL+	10	6/36	6/24	6/24	12	10	10
Case 02 Female	Rt	PL+	8	6/36	6/36	6/36	10	12	10
	Lt	PL+	10	-surgery not done	-	-	-	-	-

**RESULTS**

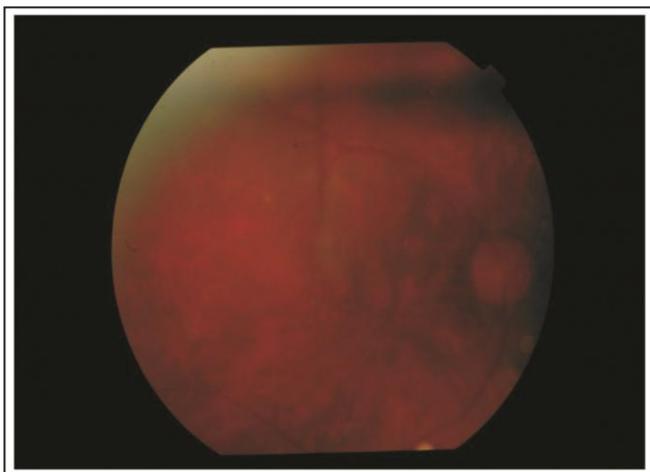
Three eyes were operated and quadrantic sclerectomy was performed in these. Male patient showed improvement in vision which was faster in left eye than right eye, in which sclerectomy was accompanied by SRF drainage and intravitreal C<sub>3</sub>F<sub>8</sub>gas injection. Patient's vision improved from PL to 6/24 in four weeks postoperatively in left eye. In the right eye it improved to 6/24 in six weeks. This was the eye in which SRF was not drained. His retina was attached in both eyes, fig. 4. He was examined at his last follow up visit on September 2014 and he had bilateral best corrected vision of 6/18 with normal IOP.

Female was examined one and three weeks postoperatively. Her vision improved from PL to 6/36 with glasses in her right eye. Her retina was attached. She was lost in follow-up.

Both patients were followed one week, three weeks and twelve weeks postoperatively.



**Fig. 3:** Quadrantic Sclerectomy.



**Fig. 4:** Fundus Picture - 1 month postoperatively.

**DISCUSSION**

Two patients with idiopathic uveal effusion syndrome were operated. It is a type of scleropathy affecting choroidal fluid dynamics. Abnormally thick sclera and short axial length were common in both eyes. Quadrantic sclerectomy in 3 eyes was successful in attaining retinal re attachment. Following scleral resection all 3 eyes showed retinal re attachment and improvement in visual acuity.

Theories regarding pathogenesis of uveal effusion syndrome suggest that it is more common in nanophthalmic eyes<sup>7</sup> where scleropathy is congenital and abnormally thick sclera<sup>8</sup> compresses vortex veins leading to impeded drainage. Similarly thick sclera is also seen in patients suffering from glycogen storage disease, mucopolysaccharidosis (Hunter syndrome)<sup>9</sup> where sclera is thick due to increased deposition of mucopolysaccharides. Decompression of vortex veins was attempted by Gass and he found that full thickness sclera incisions were effective in obtaining

reattachment of retina, supporting the hypothesis that it was primarily scleral thickening causing uveal effusion by obstructing protein diffusion (mainly albumin) out of sclera and obstruction of vortex veins. The thick sclera hinders trans-scleral protein diffusion which results in protein accumulation in choroidal extra vascular space. In the absence of any known systemic disorder, abnormal deposition of different materials is the cause of scleral thickening<sup>10,11,12</sup>. The intraocular pressure is one of the factors which determines trans-scleral flow of proteins in the eye<sup>13,14,15</sup>. Quadrantic sclerectomy was successful in treating uveal effusion syndrome by relieving choroidal effusion and non rhegmatogenous retinal detachment<sup>16,17</sup>. It supports the hypothesis that thick sclera prevents protein diffusion and decreasing scleral thickness can improve fluid outflow<sup>18</sup>.

Vortex vein decompression was first suggested by Schaffer in 1975<sup>6</sup>. Success of scleral thinning procedure was also reported by Brockhurst in 1980<sup>19,20</sup>.

**CONCLUSION**

Patient with uveal effusion syndrome responded better to surgical procedures like sclerectomy and didn't respond to medical treatment.

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