PARTIAL THICKNESS SCLERECTOMY FOR UVEAL EFFUSION SYNDROME IN NANOPHTHALMIC EYES.

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ABSTRACT

Introduction: Nanophthalmos characterized by short axial length, high lens-to-eye ratio and thick sclera, is more prone to develop uveal effusion syndrome (UES). This rare entity can result in idiopathic exudative detachment of the choroid, ciliary body and retina. Abnormality in the scleral thickness with resultant obstruction of the vortex veins and reduced trans-scleral drainage of fluid is responsible for exudative retinal detachment (ERD).

Methods: A retrospective study of UES in nanophthalmic patients treated with partial thickness sclerectomy in tertiary eye care centre from January 2015 to June 2019. Five eyes of five patients (four males and one female) with a diagnosis of nanophthalmos suffered from angle closure glaucoma associated with ERD. Raised intra-ocular pressure (IOP) not amenable to conservative medical management were subjected to surgery. Lamellar sclerectomy was performed in two or more quadrants without drainage which was judged on the basis of maximum amount of exudative fluid present in the subsequent quadrants.

Result: The average age at surgery was 39.2 years and the mean follow-up duration was 9.2 months (6 to 18 months). Revision sclerectomy was required in 2/5 (60%) eyes. The serous fluid gradually resolved and retina remained reattached at the end of final follow up. The useful vision was preserved and IOP was normalized.

Conclusion: Nanophthalmic UES remains a challenging clinical entity. Partial thickness sclerectomy may be an effective option in the treatment of nanophthalmic UES, not amenable to the conventional medical management in a low resource setup.

Keywords: angle closure glaucoma, exudative retinal detachment, nanophthalmos, uveal effusion syndrome, sclerectomy.


INTRODUCTION

Nanophthalmos is a rare disorder characterized by small eyes with short axial length (AL) and a thickened sclera. The corneal diameter may reduce marginally and the size of the lens is normal or slightly increased. Usually, nanophthalmic patients suffer from hypermetropia between +10 to +20 dioptres (D). It usually occurs bilaterally and sporadically, but can also occur as an inherited form in either autosomal dominant or recessive pattern. The condition is thought to be caused by a developmental arrest of ocular growth. Ultrastructural and histochemical studies have demonstrated abnormal scleral collagen fibers and an alteration in the production of glycosaminoglycans in scleral cells.
The examination of nanophthalmic eyes frequently demonstrate crowding and shallowing of the anterior chamber structures mainly due to a high lens-to-eye volume ratio. This factor leads to high predilection to angle-closure glaucoma and uveal effusion in these susceptible eyes.5

Uveal effusion syndrome (UES) is a rare clinical entity resulting in idiopathic exudative detachments of the choroid, ciliary body and retina. Impaired vascular drainage associated with scleral thickening is assumed to be the primary etiology.6 Possible obstruction of the vortex veins and reduced transscleral drainage of protein rich fluid from the suprachoroidal space due to the thickened sclera is believed to be the major coverted cause.3, 7 The association between UES and nanophthalmic eyes was first established by Brockhurst.1

Gass emphasized the possible congenital scleral anomaly in majority of idiopathic UES while abnormal vortex veins might be another plausible explanation. UES could arise due to various ocular pathological states like postoperative hypotony, post scleral buckling surgery or posterior scleritis.6 Medical management have been tried with oral acetazolamide and oral/topical prostaglandin analogues but the success is limited, and yet to be fully validated.8 The management of these type of small eye phenotype represents a major surgical challenge to regulate the intraocular pressure (IOP) and to preserve the vision.

In this series, we describe the technique and results of sclerectomies in the treatment of UES in five patients with nanophthalmos.

METHODS

We reviewed the medical records of five patients who were referred to our department of vitreoretina from general ophthalmology clinic and glaucoma clinic for uncontrolled IOP despite maximum tolerated anti glaucoma medications from January 2015 to June 2019. Informed and written consent was obtained from all the patients. The identity of the patient has been anonymized throughout the text. Institutional approval was not required to publish the retrospective series. The study adhered to the tenets of the Declaration of Helsinki.

A detailed medical and ocular history was taken in each case. Each patient underwent comprehensive ophthalmic evaluation including measurement of visual acuity (VA), slitlamp biomicroscopy examination, dilated fundus evaluation by indirect ophthalmoscopy, applanation tonometry, and ocular biometry. Ultrasongraphic A-scans for AL measurement was obtained in all cases and B-scans were performed whenever indicated.

Surgical technique

All the cases were managed by surgical approach. The surgery was performed under peribulbar anesthesia under strict aseptic technique. After 360° conjunctival peritomy, blunt dissection of the tenon’s capsule was done with tenotomy scissors. Muscle hook was used to isolate all the recti and bridle suture was passed underneath those muscle. The margin of sclerectomies were located just behind the insertion of recti anteriorly and at the equator posteriorly. Partial thickness approximately 2/3rd (60%) thickness of sclerectomy (6 x 5 mm flap) was excised in two or more quadrants viz: superonasal (SN), inferonasal (IN), superotemporal (ST) and inferotemporal (IT). The site and number of sclerectomy was decided based on maximal convexity of exudative fluid evident upon pre-operative examination findings. No attempt was made to drain the subretinal fluid. The excised area was left bare. The subtenon and conjunctiva was closed with vicryl 8-0 suture (Figure 1). Post-operatively oral anti-microbial agent (tab. Ciprofloxacin 500 mg BD), oral steroid 1mg/kg body weight, oral acetazolamide, oral analgesics in as per need basis and oral proton pump inhibitors was initiated and continued for next five days. Oral steroid were tapered off in a weekly interval. Topical antibiotics, steroid drops in a tapering fashion, cycloplegic drops and
steroid antibiotic combination ointment were administered following from the day post surgery and after removal of an eye pad. The patients were followed up for more than six months at conclusion. Revision surgery was performed as similar to the procedure described earlier. The remaining quadrants where previous sclerectomy was not performed was selected. Post-operative management with respect to medications in the revised cases were also similar as described previously.

RESULTS
Case 1
A 48-year-old male presented with complaints of diminution of vision in both eyes (BE) since 4 months. He was previously managed with maximum tolerated antiglaucoma medications (gtt. Timolol maleate 0.5%, gtt. Brimonidine tartrate 0.2%, gtt. Dorzolamide hydrochloride 2%, gtt. Bimatoprost 0.01%) and also subsequently managed with Neodymium Yttrium Aluminium Garnett laser peripheral iridotomy (LPI) for high IOP. His visual acuity (VA) in right eye (RE) was hand movement (HM) and left eye (LE) 1/60 with no further improvement in refraction. There was no significant family history and no known co-morbidities. The IOP at presentation was RE 31 mmHg and LE 16 mmHg respectively. Anterior segment examination revealed shallow anterior chamber (AC) of van Herick (VH) grade II and patent LPI in BE. Dilated fundus examination showed ERD in RE (Figure 2). Ocular biometric readings was consistent with nanophthalmos with AL of 16.7 mm RE and 16.5 mm LE. Similarly BE were phakic with lens thickness (LT) of 4.2 mm RE and 4.1 mm LE. Ultrasonographic (USG) B scan for estimation of retina-choroid-sclera (RCS) complex thickness showed 2.5 mm RE and 2.35 LE. The choroidal and exudative detachment was more prominent in the IN and IT quadrants. Partial thickness sclerectomy in the two quadrants (IN, IT) without external fluid drainage was performed in the right eye.
First post-operative VA was HM but the IOP was lower at 21 mmHg, the retina was not completely attached and there was presence of ERD but decreased than pre-operative status. One month following the surgery the best corrected visual acuity (BCVA) improved to 1/60 and the IOP was stable. Three months post surgery the BCVA was 3/60 and the retina was re-attached completely. At the end of eight months, the IOP remained stable at 13 mmHg, the retina attached and the BCVA documented was 5/60. The recovery of the patient was uneventful.

Her VA was HM RE and she denied perception of light (DPL) LE. She was previously diagnosed as nanophthalmos and treated for secondary angle closure glaucoma with maximally tolerated anti glaucoma medications as well as LPI in her BE.

However, she was referred for persistently high IOP despite medications and deteriorating vision. Her IOP at presentation was 29 mmHg RE and 50 mmHg LE. Anterior segment examination showed shallow AC of VH I in RE and neovascularization of the iris and white pupillary reflex in LE. The AL reading was 14.2 mm RE and excluded in LE. The LT was 4.3 mm RE and 4.2 mm respectively. Fundus finding was consistent with ERD in RE with maximum amount of fluid in the inferior quadrants.

**Case 2**

A 62-year-old female was referred for progressive diminution of vision in her right eye since 2 months. Her VA was HM RE and she denied perception of light (DPL) LE. She was previously diagnosed as nanophthalmos and treated for secondary angle closure glaucoma with maximally tolerated anti glaucoma medications as well as LPI in her BE.

![Image](image1.png)

**Figure 2:** Preoperative findings of exudative retinal detachment with choroidal effusion of patient 1. (A) Fundus photo. (B) Ultrasonographic B scan of the same eye showing exudative retinal detachment.

![Image](image2.png)

**Figure 3:** Postoperative findings of patient 1. (A) Fundus photo of first postoperative day with resolving exudative fluid. (B) Echography of the same patient at three months follow-up which shows complete resolution of the fluid and attached retina. (C) Corresponding fundus photo at three months following surgery.
USG B scan revealed thickened RCS complex of 2.4 mm RE and closed funnel retinal detachment in LE. Conservative topical management with antiglaucoma medications, steroids and cycloplegic agent was continued in her left eye. Whereas, surgical option with two quadrants (IN and IT) partial thickness sclerectomy was opted for RE. She was in her course of recovery with VA 1/60 and IOP 19 mmHg at the end of second post-operative month, but the retina re detached and exudative fluid became prominent inferiorly and nasally during the third month post surgery.

Following a detailed discussion with the patient, she agreed to proceed with the surgery and similar procedure with lamellar sclerectomy was performed in the SN quadrant. There was no complications documented during the intraoperative as well as post-operative period. We had the record of the patient for the next three months where her BCVA was 1/60 and IOP was 17 mmHg, then he patient was lost for follow-up.

Case 3
A 46-year-old male with no other family history and systemic co-morbidities was referred for the complaint of diminution of vision in RE and uncontrolled IOP despite maximum tolerated antiglaucoma medications. The documented VA was HM RE and DPL LE. The recorded IOP was 27 mmHg in RE. The AL and LT measured in RE was 18.7 mm and 3.8 mm respectively. The AL and LT reading was not obtained in the fellow phthisical eye. The refractive status of the affected eye was +12 D. The surgery was performed in the inferior quadrants (IT, IN), where the exudative fluid was more prominent. Following surgery, the post-operative event was uneventful till six months. Then the patient had re-detachment and decision to proceed with second surgery was discussed with the patient and his family members. Further partial thickness sclerectomy was carried out in the remaining superior two quadrants (ST, SN) accounting for a total of four quadrant sclerectomy after the revision surgery. The patient recovered without any complications. The IOP was 16 mmHg and the retina remained attached with BCVA of 3/60 at the end of his last follow-up.

Case 4
A 41-year-old male who was previously prescribed and compliant with maximum tolerated anti-glaucoma medications and a patent LPI was referred with gradual diminution of vision and high IOP in right. VA was HM RE and counting fingers at 1 feet LE. The AL was 15.7 mm and 15.1 mm respectively and LT was 3.5 mm and 3.9 mm. The IOP in affected eye was 30 mmHg, similarly, the eye was hyperopic with +14 D on retinoscopy and the VA was not improved with refraction. Two quadrants (IN, IT) lamellar sclerectomy was performed. The patient was followed up for a duration of 11 months where the BCVA was 4/60 and the retina remained attached (figure 4) and the IOP was within a normal range of 12 mmHg.

Case 5
A 19-year-old male with progressive diminution of vision in both eye for one and half month duration. The VA was HM RE with no improvement in refraction and 4/60 LE which improved upto 6/36 with +16 D. IOP was 28 mmHg and 16 with maximum tolerated antiglaucoma medicines. The AL readings were 15.8 mm RE and 15.9 mm LE. The LT was 3.8 mm RE and 3.6 mm LE. The AC was shallow in RE with VH grade I RE and VH grade II LE. There was a marked ERD with choroidal

Figure 4: Fundus photo of patient no 4 at 9 months follow-up. Leopard spot appearance of the fundus is seen and the retina is attached.
detachment in right and surgery was proceeded accordingly in the most dependable portion of IN and IT quadrants. The exudative fluid resolved gradually over a period of two months. The patient recovered without any complications and he followed up with us for a period of 12 months post surgery. The findings of his last follow-up revealed a BCVA of 2/60 RE with an IOP of 17 mmHg and attached retina.

The average age at surgery was 39.2 years (range 19 – 48 years) and there were four males and one female patient in our series. Pre-operative findings of the patients revealed profound vision loss and exclusive right eye affection in all the cases. Fundus examination revealed ERD with smooth convexity and dome shaped configuration. IOP were high in all the cases pre-operatively despite maximally tolerated anti glaucoma medications and peripheral iridotomy. AL was below 19 mm in all cases, with a mean AL of 16.2 mm (14.2 mm to 18.7 mm) and average LT of 3.9 mm (3.5 mm to 4.2 mm) in the affected RE. Lens to eye volume in our series was 24.1%. Refraction showed high hyperopia with an average of +14.1 D (+12 D to +16D) of the affected RE and +14 D (+13 to +15.5 D) in the fellow eye. The details of the patient characteristics were depicted in Table 1.

Table 1. Patient Characteristics

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age(years)/Gender</th>
<th>VA at presentation</th>
<th>Previous Management</th>
<th>AL measurement (mm)</th>
<th>Lens Thickness (mm)</th>
<th>Pre-Operative IOP</th>
<th>Refractive Status</th>
</tr>
</thead>
<tbody>
<tr>
<td>RE</td>
<td>LE</td>
<td>RE</td>
<td>LE</td>
<td>RE</td>
<td>LE</td>
<td>RE</td>
<td>LE</td>
</tr>
<tr>
<td>1 48/M</td>
<td>HM 1/60</td>
<td>Anti-glaucoma meds + LPI</td>
<td>16.7</td>
<td>16.5</td>
<td>4.2</td>
<td>4.1</td>
<td>31</td>
</tr>
<tr>
<td>2 42/F</td>
<td>HM DPL</td>
<td>Anti-glaucoma meds + LPI</td>
<td>14.2</td>
<td>-</td>
<td>4.3</td>
<td>4.2</td>
<td>29</td>
</tr>
<tr>
<td>3 46/M</td>
<td>HM DPL</td>
<td>Anti-glaucoma meds</td>
<td>18.7</td>
<td>-</td>
<td>3.8</td>
<td>-</td>
<td>27</td>
</tr>
<tr>
<td>4 41/M</td>
<td>HM FC 1 feet</td>
<td>Anti-glaucoma meds + LPI</td>
<td>15.7</td>
<td>15.1</td>
<td>3.5</td>
<td>3.9</td>
<td>30</td>
</tr>
<tr>
<td>5 19/M</td>
<td>HM 4/60</td>
<td>Anti-glaucoma meds</td>
<td>15.8</td>
<td>15.9</td>
<td>3.8</td>
<td>3.6</td>
<td>28</td>
</tr>
</tbody>
</table>


On table finding showed tortuous episcleral vessels and abnormally dilated vortex veins. The
resected flap demonstrated a thickened sclera. Unfortunately, the details of histopathological examination of resected lamellar scleral flap could not be incorporated. Following from the day of surgery, the ERD gradually resolved and the retina remained attached in all the patients. However, revision surgery was required in two patients due to re-detachment and repeat accumulation of exudative fluid at third and sixth postoperative months respectively. Revision was carried out at one more quadrant in second patient and remaining two quadrants in third patient. Deliberate decision was taken in these two cases owing to their one eyed status and after patient was uneventful. The mean follow-up duration was of 9.2 months (6 to 18 months). The summary of the surgical management and final outcomes is presented in Table 2.

**CONCLUSION**

The term nanophthalmos indicates that the eye is small and in contrast to ordinary microphthalmos, not associated with other ocular malformations.10 This rare entity of nanophthalmic eyes are prone to develop glaucoma. The reason for increased IOP is postulated secondary to disparity in the ocular dimensions.11 The lens-to-eye volume which is approximately 4% in normal population is

<table>
<thead>
<tr>
<th>Patient</th>
<th>Sclerectomy Quadrants</th>
<th>BCVA</th>
<th>IOP (mmHg)</th>
<th>Status of retina</th>
<th>F/U duration (months)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Two quadrants IN, IT</td>
<td>5/60</td>
<td>13</td>
<td>Attached</td>
<td>8</td>
</tr>
<tr>
<td>2</td>
<td>Three quadrants IN, IT, SN</td>
<td>1/60</td>
<td>17</td>
<td>Attached</td>
<td>6</td>
</tr>
<tr>
<td>3</td>
<td>Four quadrants IN, IT, ST, SN</td>
<td>3/60</td>
<td>16</td>
<td>Attached</td>
<td>9</td>
</tr>
<tr>
<td>4</td>
<td>Two quadrants IN, IT</td>
<td>4/60</td>
<td>12</td>
<td>Attached</td>
<td>11</td>
</tr>
<tr>
<td>5</td>
<td>Two quadrants IN, IT</td>
<td>2/60</td>
<td>17</td>
<td>Attached</td>
<td>12</td>
</tr>
</tbody>
</table>


explanation of all the possible complications. The retina was attached and IOP remained stable at the final follow-up. The post-operative recovery of the deranged and increased to 10% - 30% in nanophthalmic eyes.12 Another possible mechanism suggested for angle closure glaucoma
is the development of uveal effusion. The serous choroidal detachment and secondary retinal detachment cause forward rotation of the ciliary body, relaxation of lenticular zonules, forward movement of the lens increasing the iridolenticular apposition and pupillary block resulting in angle closure glaucoma.

Glaucoma in nanophthalmic eyes has been treated with various modalities like laser iridoplasty, surgical iridectomy, filtration surgery, cataract extractions and vortex vein decompression. Unfortunately, these procedures are accompanied by an extremely high complication rate of failure to control the IOP and even loss of vision.

Vortex vein decompression for UES in nanophthalmic eyes was initiated by Brockhurst. In the original procedure, scleral beds were dissected at the equator, to decompress the vortex veins and relieve the uveal effusion. Gass used the same procedure to treat a case of uveal effusion successfully, as he believed that the abnormally thick sclera was the cause for the effusion. Wax et al suggested that the thickened sclera in nanophthalmos, may have mostly an anterior component. The improved transcleral fluid outflow by thinning it resulted in functional decompression of the anterior vascular bed, thus preventing the dreaded postoperative complications.

In a report for management of familial nanophthalmos, the scleral flap was not extended posteriorly to the equator and no attempt was made to unroof or transect the vortex veins. However, we managed our cases surgically by resecting the scleral flap anteriorly at the boundary of spiral of Tillaux contributed by the insertion of recti muscles and extending posteriorly upto the equator. We attempted the lamellar sclerectomy in the inferior quadrants as much as possible so that the superior quadrants can be preserved for the purpose of glaucoma surgery in future. No attempt was made to drain the subretinal fluid and vortex vein was not transected in our procedure. Similarly two quadrant vein decompression with subretinal fluid drainage at the same setting also revealed promising result but the cases have short follow up period.

This condition can be overlooked easily and attempted to be managed with laser peripheral iridotomies for high IOP and shallow anterior chamber. Nonetheless, the sudden drop in IOP after a peripheral iridotomy, results in a relative increase in the choroidal venous pressure and an aggravation of the uveal effusion. Medical therapy might be a safer alternative than surgery, but the results of medical therapy is variable and not validated.

The effectiveness of an unsutured sclerotomy or sclerectomy in curing or preventing uveal effusion supports the concept that inadequate transscleral outflow contributes to the production of uveal effusion. The benefit of dissection around vortex veins, may not result from decompressing the venous drainage, but from opening the channels for transscleral drainage. Transscleral channels made remote from vortex veins seem just as effective and with less risk of haemorrhage. The failed sclerectomy procedure was repeated by deep sclerostomy and adjunctive application of topical mitomycin. However we did not apply antifibrotic agent in the revision surgery for the fear of scleral melting as the long term reports of adverse effects is not yet available.

The success rate of these procedures in achieving an anatomic improvement was reported approximately 83% after a single procedure and about 96% after repeat surgery. Anatomical attachment achieved in our series after single procedure was 60% which improved 100% at the end of mean follow up of 9.2 months (6 to 12 months). The anatomical achievement did not translate into functional success. We were able to preserve of at least ambulatory vision in three
patients similarly gradual improvement in VA by two lines was achieved in remaining two patients.

The retrospective nature and short follow-up duration are the limitations of this series. This precludes us to propose a definitive statement with regards to long term anatomical success and stable VA. The management of nanophthalmic UES in our series is not a novel technique. However, nanophthalmic UES are generally rare, we believe that the findings from this study will contribute to the pool of knowledge in the management of this condition especially in the low resource setup.

Here, we have presented five cases of UES in nanophthalmic eyes who were successfully managed with partial thickness/lamellar sclerectomy. The surgery was well tolerated by the patients and no significant complications were noted at the end of the follow up period respectively. Though the management of this type of condition pose a challenge in itself, the method of partial thickness scleral sclerectomy could be a safe surgical modality in nanophthalmic UES.

REFERENCES


