

Surgical Approaches to Serous Retinal Detachment With Retina–Lens Touch in Eyes With Nanophthalmos

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Abstract

Purpose: To describe the visual outcomes and problems that resulted from surgical treatment of nanophthalmic complete retinal detachment (RD) with retina–lens contact. **Methods:** A multicenter retrospective case series with deep sclerectomy as a treatment was performed. **Results:** Five cases had extensive deep sclerectomies, 3 with intended drainage of subretinal fluid (SRF). The RD resolved 1 week postoperatively in 4 cases and within 1 month in 1 case. The visual acuity improved from light perception to a median of 20/100. Three cases had longstanding lens touch beyond 1 year and improved VA to 20/100, 20/150, and hand motions, respectively. Complications included focal lens dialysis in 2 cases (passive drainage of SRF) and lens or intraocular lens dislocation in 1 case each (active drainage of SRF). Ultrasound biomicroscopy and anterior optical coherence tomography showed a very narrow angle and short zonules. **Conclusions:** Deep sclerectomy results in good anatomic and functional improvements in advanced cases of nanophthalmos exudative detachment, which is often considered to be incurable.

Keywords

nanophthalmos, serous retinal detachment, sclerectomy, ultrasound biomicroscopy, uveal effusion, zonules, lens dislocation

Introduction

Massive retinal detachment (RD) with lens contact is an ominous sign and can be found in end-stage retinopathy of prematurity (ROP) or Coats disease.^{1–4} Scleral buckling or vitrectomy, subretinal fluid (SRF) drainage, transscleral cryotherapy, or endolaser treatment are frequently needed for eyes with severe Coats disease and RD.

Therapeutic problems occur as a result of massive RD with lens contact in the context of nanophthalmos. Intraocular surgery in eyes with nanophthalmos is risky because of the anatomic features related to a developmental arrest of ocular growth. These eyes have very anterior insertion of the retina, a large lens, short zonules, a shallow anterior chamber (AC) (with a propensity for angle closure), and a noticeably thickened sclera and choroid (Figure 1).^{5–18} Vitrectomy is dangerous because it can result in peripheral holes (anterior retinal insertion),¹² which can cause an exudative RD to become more complex and require silicone oil tamponade or multiple surgical procedures.^{12,13}

In this case series, we describe the anatomic and visual results of surgical procedures for serous RD with retina–lens touch in eyes with nanophthalmos.

Methods

The clinical data of all patients of all ages diagnosed with nanophthalmos (axial length [AL] <20.5 mm) who had massive exudative RD with lens contact were retrospectively analyzed. The study adhered to the tenets of the Declaration of Helsinki and was approved by the Institutional Review Board, Rafic

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Anatomic Features

- Deep-set orbit
- Narrow palpebral fissure
- Thick Tenon capsule
- Vascular episclera
- Small cornea
- Shallow anterior chamber
- Narrow angle
- Boomerang-shaped iris
- Poor dilation of pupil
- Anteriorly positioned ciliary processes
- Lack of ciliary ring expansion
- Thick lens
- Short zonules
- Strong hyaloideocapsular ligament
- Non-liquefied compact vitreous
- Small optic disc
- Optic disc drusen
- Thickened sclera
- Thickened choroid
- Fovea plana
- Attenuation of foveal avascular zone
- Tortuous retinal vessels

Figure 1. Special anatomic features of nanophthalmos.⁵⁻¹⁸

Hari University Hospital. Patients provided informed consent for their information and images to be published.

Eyes with congenital malformations, anterior or posterior microphthalmos, ROP, or Coats disease were excluded from the study. Patients taking topiramate or with a history of posterior scleritis or uveitis were also excluded.

The patients received a comprehensive ocular examination that included the best-corrected visual acuity (BCVA) (with spectacles) measured with the Snellen chart, intraocular pressure (IOP) measured with a Goldmann applanation tonometer or by puff tonometry, anterior segment assessment with slitlamp microscopy, ocular fundus assessment with 20.0 D and 90.0 D lenses and spectral-domain optical coherence tomography (OCT) with or without angiography, and intraocular lens (IOL) power and AL measured with an IOLMaster 700 swept-source OCT biometer (Carl Zeiss Meditec AG). In addition, previous glaucoma procedures or antiglaucoma medications were recorded. All cases previously had several thorough retinal examinations, and no retinal holes were noted.

Surgical Technique⁷

In Cases 1 to 3, lateral canthotomy was performed to allow easy access to the posterior sclera. This was followed by a 360-degree conjunctival peritomy with isolation of the rectus muscles. Bare

sclera was fully exposed, and vortex veins were identified in all quadrants. At least 90% of the scleral thickness was excised, extending from the recti insertions to the vortex veins for 3/4 quadrants, sparing the superior oblique muscle insertion. The initial groove was made with a 57 blade (BVI) to reach the desired depth. The scleral flap to be excised was pulled up with a forceps using the nondominant hand, while the plane of incision was extended using a crescent blade with the dominant hand. Cautery was avoided to enable a histologic study of the sclera.

In 2 cases (Cases 1 and 2), nonpenetrating deep sclerectomy was followed by passive leakage of SRF. The right scleral depth was confirmed by allowing a cotton applicator to come in contact with the remaining scleral wall and becoming moist or by directly watching liquid flowing intraoperatively. If not, a deeper sclerectomy was needed. In case of globe hypotony, very small intracameral injections of hyaluronic acid, a balanced salt solution, or gas (air, sulfur hexafluoride [SF₆], perfluoropropane) were given at the corneal limbus. In Cases 4 and 5, a 4-quadrant two-thirds 5 mm × 5 mm sclerectomy was created followed by a more central 2 mm × 2 mm complete sclerectomy to expose the choroid. In Cases 3 to 5, transscleral drainage of SRF was also performed using diathermy from 1 quadrant, and the eye was repressurized through a limbal incision with a balanced salt solution.

Postoperative Care

An examination was performed daily for the first week to monitor healing of the RD and treat ocular hypotony. When there was hypotony, intracameral injections at the limbus of air, SF₆, or hyaluronic acid were administered using 30- or 27-gauge needles.

Results

Five patients had light perception (LP) VA and unilateral retrolental RD (Figures 2 to 4). Despite the use of oral corticosteroids, uveal effusion developed over a few months to a few years from a shallow RD to retina–lens touch. Of the patients, 4 were men and 1 was a woman; the median age was 44.4 years. The sclera thickness was 2 to 3 mm. The retina reattached within 1 week of the sclerectomy in Cases 1 to 4 and within 1 month in Case 5 (Table 1).

The BCVA (with spectacles) improved from LP to a median of 20/100 at the latest follow-up (mean, 32.2 months; range, 6–90). The illness was first thought to be incurable in Cases 4, 5, and 2, with long-lasting RD of 24, 36, and 48 months, respectively, and VA recovery to 20/100, hand motions, and 20/150, respectively. In Case 5, the ellipsoid zone was severely disrupted, which was associated with poor vision (Figure 4). In Cases 1 and 2, the lens zonules were damaged to some extent (without subsequent sequelae), and in 2 other cases, the lens (Case 4, Figure 3) or the IOL (Case 5, Figure 4) had dislocated into the vitreous cavity. To control hypotony, the AC was filled, at the limbus, with hyaluronic acid (Case 1), air (Case 2), or a balanced salt solution (Cases 3, 4, and 5). In Cases 1 and 2,

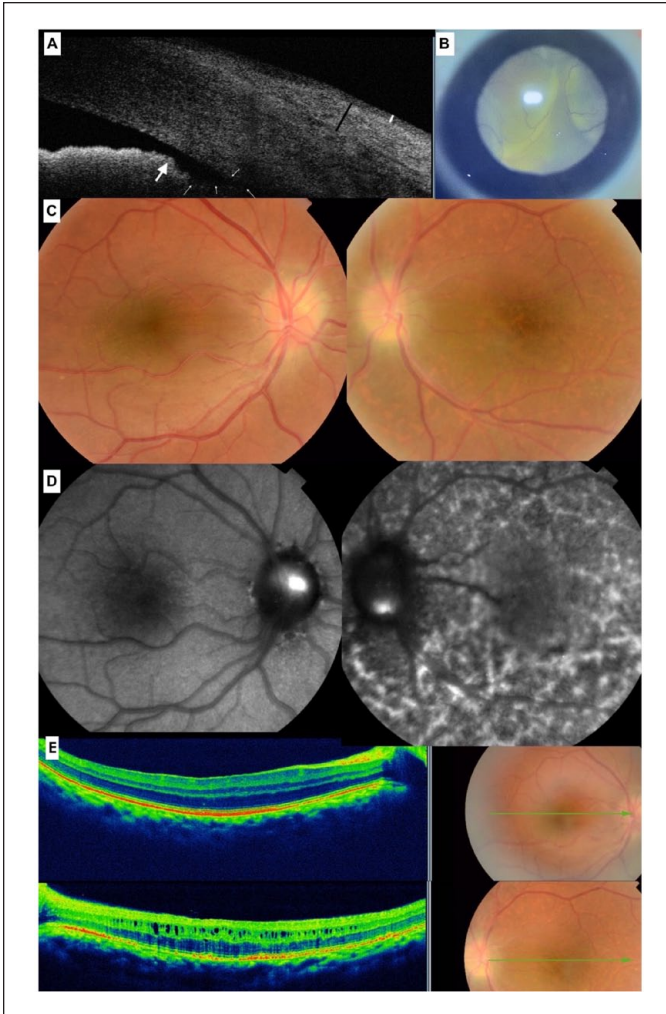


Figure 2. (Case 1) A 38-year-old White woman was referred for the treatment of an inferior serous RD in the right eye not responsive to an oral corticosteroid. (A) Anterior segment OCT of the fellow eye showed a narrow angle (small arrows), boomerang-shaped iris (large arrow), and thick episclera (black caliper) underlying bulbar conjunctiva (white caliper). On the day of surgery, the patient tested positive for COVID-19. Surgery was cleared 2 months later. Intraoperatively, total RD was noted (B). A nondrainage, 300-degree, 90% depth sclerectomy with passive spontaneous egress of SRF was performed. To avoid hypotony, hyaluronic acid (0.05 mL) was injected intracamerally with difficulty (great resistance noted), after which zonular dialysis occurred nasally. The retina flattened 1 week postoperatively. One month and 19 months postoperatively, the VA improved from light perception to 20/50. At the last follow-up, fundus photography (C), fundus autofluorescence (D), and SD-OCT (E) showed bilateral small optic discs, bilateral fovea plana, inner macular cystic spaces, and hyperfluorescent linear tracks in the unoperated left eye only. Abbreviations: OCT, optical coherence tomography; RD, retinal detachment; SD-OCT, spectral-domain OCT; SRF, subretinal fluid; VA, visual acuity.

zonular dialysis occurred after a cotton applicator was applied to the injection site in an effort to control the reflux of hyaluronic acid or air. The lens dislocated into the vitreous cavity in Cases 4 and 5 after the eye was reinflated with a balanced salt solution injected through a limbal incision.

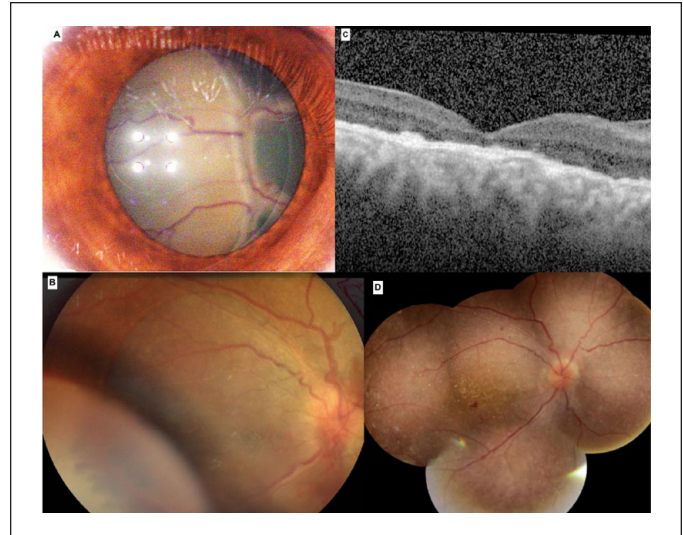


Figure 3. (Case 4) A 32-year-old White man presented reporting reduced vision in the right eye for 1.5 years. His VA was light perception in the right eye, which used to be the better eye. The axial length was 17 mm. He had leukocoria from the retina contacting the lens (A). Initially, the right eye appeared to be inoperable. A 4-quadrant, partial-thickness sclerectomy and sclerotomy surgery were performed in the left fellow eye. Twenty months after surgery in the left eye, a 4-quadrant partial-thickness sclerectomy with central 2 mm complete unroofing of the sclera was performed in the right eye. After SRF drainage, the globe was pressurized with intracameral balanced salt solution, with immediate dislocation of the crystalline lens into the vitreous cavity (B). The retina reattached on the first postoperative day (C). Subsequently, pars plana vitrectomy and lensectomy were performed with scleral fixation of the intraocular lens using the Yamane technique. At the last follow-up at 29 months, the retina was totally reattached and the corrected VA was 20/100 in the right eye (D). Abbreviations: SRF, subretinal fluid; VA, visual acuity.

Conclusions

To our knowledge, this is the largest series of massive RDs in nanophthalmic eyes with retina–lens touch in the literature. Deep sclerectomy with or without active drainage of SRF may result in surprisingly good anatomic and functional improvements in these advanced cases of nanophthalmos-related exudative detachment, which is often considered to be incurable. Conventional surgical interventions in exudative RD have included vortex vein decompression, sclerotomies, and small partial-thickness sclerectomies.^{19,20} In one case, Johnson and Gass¹⁹ performed a small rectangular equatorial quadratic one-half to two-thirds thickness sclerectomy, while in another case, Brockhurst²⁰ performed vortex vein decompression. Such interventions are very effective in the control of most uveal effusions. In young patients with nanophthalmos, exuberant fibrosis would originate from a very vascular episclera and active thick Tenon capsule and would cover the sclerotomies or the partial sclerectomy, mitigating the initial surgical success. More definitive and radical surgery was needed in nanophthalmos, as previously described.⁷ The sclerectomies involved 300 degrees from muscle insertion to the vortices,

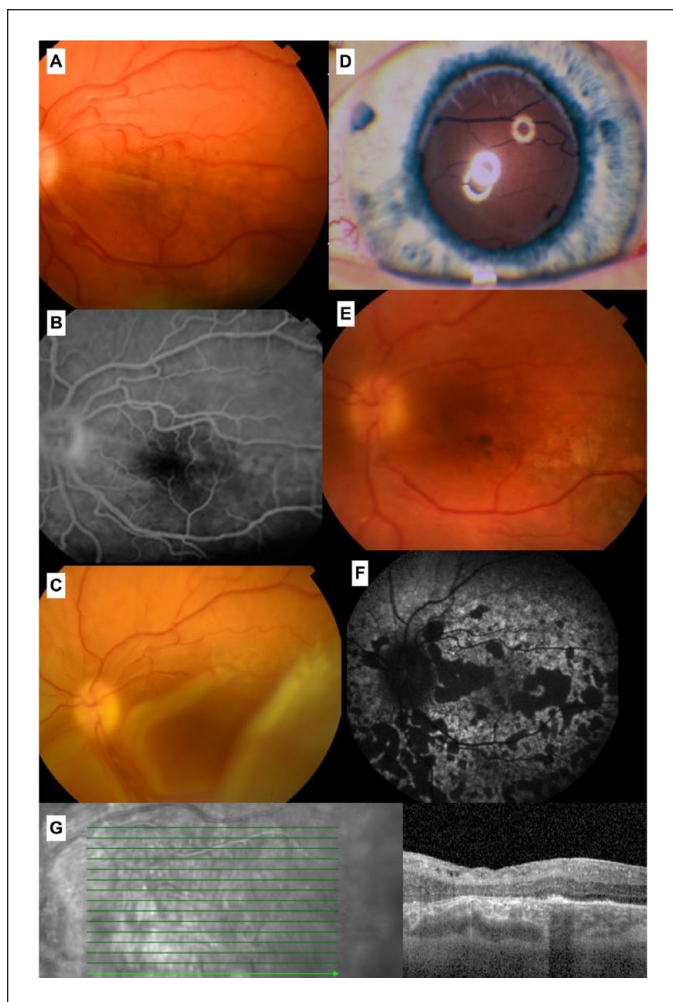


Figure 4. (Case 5) A 44-year-old White man had central visual loss of 7 years from exudative RD that was poorly responsive to years of intermittent pulse corticosteroid therapy. Funduscopy showed a shallow inferior RD with macular edema (A). The macula had hyperfluorescent spots with pinpoint leakage from the early stages of fluorescein angiography (B). The bullous detachment involved the macula 2 years later (C). One year later, the VA decreased to HM and then to LP. The retina was in close contact with the IOL (D). One year after lens–retina contact, a 4-quadrant, half-thickness sclerectomy with central complete sclerectomy and 1-quadrant drainage of SRF were performed. The IOL fell into the vitreous cavity immediately after injection of a balanced salt solution into the anterior chamber. After 1 month, the retina was completely attached. The retina flattened at the 1-month follow-up, and the VA improved to HM. Severe damage to the inner retina was seen on fundus photography (E), autofluorescence (F), and SD-OCT (G) 40 months postoperatively.

Abbreviations: HM, hand motions; IOL, intraocular lens; LP, light perception; RD, retinal detachment; SD-OCT, spectral domain optical coherence tomography; SRF, subretinal fluid; VA, visual acuity.

sparing the superior oblique muscle insertion and 90% depth. Another approach is to use mitomycin-C with small sclerectomies to prevent fibrosis at the surgical site.⁶

Nanophthalmic uveal effusion with total RD and retina–lens touch is an exceptionally rare condition. It begins as a localized uveal effusion and progresses to total detachment over a span of weeks to months. Early therapy in mild effusion or even prophylactic therapy has been advised.⁷ Historically in nanophthalmos, the extent and depth of the sclerectomy varied from 1 to 4 quadrants, 60% to 80% depth, with or without central complete sclerectomy, with or without linear 100% deep sclerotomy, and with or without SRF drainage.^{18–20}

Exudative RD in nanophthalmos is thought to result from a markedly thickened sclera or megasclera that blocks transscleral fluid outflow or compresses the vortex veins, leading to passive fluid accumulation in the suprachoroid and resulting in ciliochoroidal detachment. The fast resolution time of the uveal effusion and persistence of an attached retina during a long postoperative follow-up period favor transscleral fluid outflow obstruction in nanophthalmos. A second theory has implicated a primary collagen anomaly in nanophthalmos, such as disordered thickened scleral fiber arrangement with fraying.^{5,16,17}

The major complication encountered in 4 of 5 cases with the current surgical technique was zonular dialysis or lens dislocation (Table 1). The AC in nanophthalmos is very shallow and crowded due to a combination of a relatively big lens, thickened iris, narrow ciliary ring, and often peripheral anterior synechiae (Figure 1). The vitreous is also condensed, with a strong attachment to the posterior lens surface. Any minimal injection into the AC may cause a tremendous rise in IOP, resulting in swift reflux of the injected material or zonular dialysis. Moreover, placing an anterior maintainer runs the risk of lens touch. The findings of short zonules on ultrasound biomicroscopy in 1 case and inelasticity of the zonular fibers during surgery dictate a cautious, minimal injection of gas, a balanced salt solution, or a viscoelastic agent.

Based on the current small case series, it is difficult to compare drainage and nondrainage deep sclerectomy in the setting of total RD. On one hand, drainage sclerectomy leads to intraoperative flattening of the retina at the expense of total disruption of the zonules. On the other hand, nondrainage sclerectomy allows slower, yet more controlled, resolution of SRF, with more focal damage to the zonules because of the smaller intracameral volumes injected. Patients with nanophthalmos who have intraocular surgery often present with abnormal ciliary zonules.^{21–28} A mutation in myelin regulatory factor (MYRF) has been implicated in the pathogenesis of many cases of familial nanophthalmos in humans.²⁸ A nanophthalmos frameshift mutation of MYRF in mice²⁵ showed ocular phenotypes similar to their human counterpart,^{26–28} including a shallow AC and reduced zonular fiber density and structural dehiscence of zonular fibers. Zonular dialysis was noted in several case series (Table 2).^{21–26} Embryonic function of the *MFRP* gene is necessary for the eye to achieve normal dimensions by the third trimester, and a mutation in this or related genes would halt ocular development, leading to a lack of ciliary ring expansion, a thick crystalline lens, and dwarf zonules.^{26–28}

Table 1. Clinical Characteristics of 4 Nanophthalmic Eyes of 4 Patients With Total Retinal Detachment.

Characteristic	Case 1	Case 2	Case 3	Case 4	Case 5	Summary/Mean
Age (y)	38	49	59	32	44	44.4
Sex	F	M	M	M	M	I F; 4 M
Race	White	White	White	White	White	White 100%
Laterality	Left	Right	Left	Right	Left	Right 2; left 3
Baseline refraction (D)	+14	+12	+18	+15	+15	+14.8
IOL power (D) (Hoffer or Barrett Universal II)	+50	+45	+59	+33	NA	+46.8
Axial length (mm)	17.6	17.6	15.5	17.2	14.3	16.4
RD extent	Total lens touch	Total lens touch	Total lens touch	Total lens touch	Total lens touch	Total 100%
Evidence of progression from shallow RD to total RD	Yes	Yes	Yes	Yes	Yes	All progressive RD
Duration of RD at time of surgery (mo)	2	48	1	24	36	19
Deemed inoperable	No	Yes	No	Yes	Yes	Yes 60%
Previous laser peripheral iridotomy	Yes	Yes	No	No	Yes	Yes 60%
Previous topical glaucoma medication	No	Yes	No	No	No	20% glaucoma drops
Lens status	Phakia	Phakia	Pseudophakia	Phakia	Pseudophakia	60% phakia
Previous ocular surgery	No	No	Phaco w/ IOL implant	No	Phaco w/ IOL implant	No 60%
BCVA initial	LP	LP	LP	LP	LP	LP 100%
Follow eye RD	No	No	No	Yes	Yes	Yes 40%
UBM findings	No	Short thick zonules	No	No	No	Yes 20%
Trial of oral corticosteroid on presentation	Yes	No	Yes	Yes	Yes	Yes 80%
Trial (steroid) failed to decrease SRF	Yes	NA	Yes	Yes	Yes	Yes 80%
Thickened sclera	Yes	Yes	Yes	Yes	Yes	Yes 100%
Scleral excision	4-quadrant 90% depth 280°–300°	4-quadrant 90% depth 280°–300°	4-quadrant initial 60% with partial response then redo 90% depth 280°–300°	4-quadrant 90% depth 260°	4-quadrant half-thickness 5 mm × 7 mm sclerectomy with 100% 2 mm × 3 mm central sclerectomy	100% 4-quadrant 90% 280° mean depth
Drainage of SRF using diathermy	No	No	Yes	Yes	Yes	No 40%
Material used to reform AC intraoperatively	Hyaluronic acid	Air	BSS	BSS	BSS	BSS 60%
Lens status after AC reformation	Sector zonular dialysis	Sector zonular dialysis	Unchanged	Total dislocation of lens into vitreous cavity	Total dislocation of IOL into vitreous cavity	Sector dialysis 40%; total dislocation 40%
Time for SRF resorption (wk)	1	1	1 after reoperation 5 mo apart (only SRF superiorly resolved w/ 1 st superficial sclerectomy)	1	4	1
Preoperative or postoperative NSAID	No	No	No	No	No	No 100%
Postoperative topical corticosteroid	3 wk	3 wk	2 wk	4 wk	No	3 wk
Postoperative oral or sub-Tenon corticosteroid	20 mg prednisone with taper over 1 mo	Sub-Tenon triamcinolone	No	No	No	No 60%
Ancillary perioperative management	No	Inject SF ₆ and C ₃ F ₈ in AC to raise IOP	No	No	No	No 80%
Additional surgeries	No	No	Initial surgery was shallow sclerectomy w/ slow SRF resorption; repeat deep sclerectomy fastened SRF resorption tremendously	Lensectomy; vitrectomy; IOL fixation (Yamane technique) 1.5 mm from limbus	No	No 60%
Final CMT	447	303	338	362	328	356
BCVA postoperative (final if different)	20/50	20/150 (CF after cataract)	20/70	20/100	HM	Median 20/100
Follow-up (mo)	21	15	6	29	90	32.2
Absence of foveal pit	Yes	Yes	Yes	Yes	Yes	Yes 100%
Disruption of the ellipsoid zone	No	No	No	No	Yes	Yes 20%

Abbreviations: AC, anterior chamber; BCVA, best-corrected visual acuity; BSS, balanced salt solution; C₃F₈, perfluoropropane; CF, counting fingers; CMT, central macular thickness; IOL, intraocular lens; IOP, intraocular pressure; LP, light perception; Phaco, phacoemulsification; NSAID, nonsteroidal anti-inflammatory drug; RD, retinal detachment; SF₆, sulfur hexafluoride; SRF, subretinal fluid; UBM, ultrasound biomicroscopy.

Table 2. Zonular Deficiencies in the Nanophthalmos Literature.^{5,21–24}

Author ^a	Publication Year	Total Eyes (N)	Eyes With Zonular Defect, n (%) ^b	Hospital, Country
Day ²¹	2013	12	3 (25.0)	Moorfields Eye Hospital, UK
Zheng ²²	2017	11	2 (18.2)	EYE and ENT Hospital of Fudan University, China
Yosar ²³	2021	71	1 (1.4)	University of Sydney, Australia
Rajendrababu ⁵	2021	29	2 (6.9)	Aravind Eye Hospital, India
Ucar ²⁴	2022	61	7 (11.5)	Konyagoz Eye Hospital, Turkey
Total	2013–2022	184	15 (8.2)	5 countries in 3 continents

^aFirst author.

^bPercentage of total eyes.

It is critical to avoid a sudden drop in IOP during any intraocular surgery in nanophthalmos because it will exacerbate suprachoroidal fluid accumulation. Slow release of choroidal fluid via a nondrainage technique (very deep sclerotomy) allows for more controlled passive decompression of the choroidal detachment. Monitoring the IOP after sclerectomy is advised to regulate the IOP intraoperatively or 1 week postoperatively (at the slitlamp) via repeated, very small intracameral injections of air, expansile gas, or viscoelastic agents.

There are anecdotal reports of the use of medical therapy for exudative RD in nanophthalmos, such as corticosteroids,²⁹ nonsteroidal anti-inflammatory drugs,^{6,30} and intravitreal vascular endothelial growth factor.³¹ Medical therapy with oral corticosteroids in uveal effusion syndrome is well established but was deemed ineffective in nanophthalmos.^{10,11} Elgin et al⁹ described a patient with nanophthalmos and mild exudative RD that resolved rapidly after administration of the topical corticosteroid alone. The authors suggested that the corticosteroid reduced inflammation and stabilized the blood–ocular barrier, thereby improving regression. Topical and systemic corticosteroids were clinically ineffective in our cases and in the literature.¹² Exudative RD may result from the use of antiglaucoma medications (beta-blocker and acetazolamide). Changing the glaucoma therapy and reversing the angle closure by scleral excision with or without phacoemulsification of a clear lens should be considered.^{29,32,33}

Exudative RD accompanies a variety of diseases, including idiopathic uveal effusion, retinal or choroidal vascular disorders (chronic bullous central serous chorioretinopathy,³⁴ arteriovenous malformation³⁵), inflammatory disease (Vogt-Koyanagi-Harada, posterior scleritis),³⁶ and neoplastic processes; it also occurs after glaucoma filtering surgery and the use of medications (eg, sulfonamides and especially topiramate and acetazolamide). Control of IOP and inflammation are appropriate nonsurgical approaches.¹¹ Unlike rhegmatogenous RD, many cases of exudative RD, even if chronic, have a relatively acceptable visual prognosis, similar to the cases in the current series.^{34,37} Two cases (similar to those in the current series) with chronic central serous retinopathy–related grand bullous RD and poor vision recovered rapidly with long-lasting VA recoveries to 20/200 and 20/63, respectively.^{38,39} It is possible that in our first 4 cases, the nanophthalmic detached

outer retina was relatively protected from early and late atrophic degeneration for unknown reasons. Hypothetical explanations for the good visual recovery in our cases include a highly oxygenated thickened choroid, the short distance between the swollen choroid and the detached retina and, finally, decreased retinal metabolism in the setting of uveal effusion and a partially mature retina.

In conclusion, deep sclerectomies resulted in good visual improvement, even in longstanding serous RD with lens contact in eyes with nanophthalmos. Nondrainage sclerectomy may provide slower, but controlled, resolution of the uveal effusion with fewer lens dislocation problems. Additional studies of drainage and zonular preservation are needed.

Ethical Approval

Approval for this study was granted by the Institutional Review Board, Rafik Hariri University Hospital.

Statement of Informed Consent

Informed consent was obtained.

Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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