

## Bubble Wrap Retinopathy

An adolescent patient with a history of high myopia and prior retinal detachment has an acute retinal detachment with macrocysts in the fellow eye.

## BY GWEN COUSINS, MD; STANISLAV ZHUK, MD; AND ROY AROGYASAMI, MD

In this issue of Retina Today, Gwen Cousins, MD; Stanislav Zhuk, MD; and Roy Arogyasami, MD, discuss pearls for performing surgery for bubble wrap retinopathy.

We extend an invitation to readers to submit pearls for publication in Retina Today. Please send submissions for consideration to Dean Eliott, MD (dean\_eliott@meei.harvard.edu); or Ingrid U. Scott, MD, MPH (iscott@hmc.psu.edu). We look forward to hearing from you.





- Dean Eliott, MD; and Ingrid U. Scott, MD, MPH

15-year-old boy presented complaining of long-standing poor vision in his right eye. He had a history of high myopia with spectacle correction of -16.0 D and -15.5 D in the right eye and left eye, respectively. Three years prior, he was treated at an outside hospital for a retinal detachment in his right eye, which had a complicated course and eventually required silicone oil placement. He had a significant cataract in this eye, and the visual acuity was 20/400 (Figure 1). During attempted phacoemulsification by a local cataract surgeon, it was noted that oil came forward into the anterior chamber before the capsulorrhexis was initiated, and the case was aborted. After the procedure, the surgeon suggested that the patient seek vitreoretinal follow-up.

On B-scan ultrasound, the right eye showed no mass or hemorrhage, but the image was limited due to silicone oil artifact. Visual acuity in the left eye was 20/30, and no pathology such as lattice, tears, or holes was found on peripheral scleral depressed examination. The family was interested in having cataract surgery attempted again in order to improve vision in the right eye, and they understood the questionable visual potential of the eye due to prior retinal detachment and possible amblyopia.

There was no history of trauma, and the etiology of the retinal detachment was unknown to the referring hospital's retinal surgeon. When we saw the patient, he did not exhibit abnormal body habitus, joint extensibility, or skin elasticity, and the family told us there was no



Figure 1. White cataract in the right, silicone-filled eye.

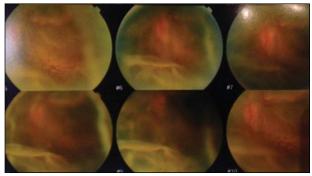


Figure 2. Series of images showing retinal detachment and dialvsis.

history of Marfan syndrome, Ehlers-Danlos syndrome, or Stickler syndrome. Aside from the retinal detachment, there was no other ocular history and family history of

TABLE 1: DIFFERENTIAL DIAGNOSIS	
Atypical Coats disease	No evidence of lipid and exudative retinal detachment
X-linked juvenile retinoschisis	Normally microcysts, not macrocysts
Atypical Vogt-Koyanagi-Harada syndrome	Not exudative retinal detachment, no skin, or central nervous system changes
Traumatic retinal detachment	No history of trauma
Retinal detachment with associated disease (ie, Marfan/Ehlers-Danlos/Stickler)	No history of these diseases or symptoms
Myopic retinal detachment	Most likely etiology

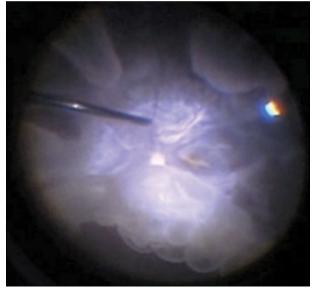


Figure 3. View upon entry showing multiple macrocysts in the retina.

retinal detachment.

Unfortunately, 7 months after the initial visit, the patient experienced a decrease in vision in the left eye and visual acuity was hand motion. They postponed cataract surgery in the right eye and vision in that eye was still 20/400. Examination of the left eye demonstrated a total retinal detachment with multiple macrocysts and inferior retinal dialysis. Grade B proliferative vitreoretinopathy was noted, with rolled edges of the dialysis and marked vitreous haze (Figure 2). The patient reported that the vision in his left eye had been decreased for only 1 day, which was surprising, as the retinal macrocysts implied chronic retinal detachment. Due to the patient's functionally monocular status with long-standing poor vision in the right eye, we believe his history to be reliable.

Thorough research did not yield any similar cases of acute retinal detachment with macrocysts in either adults or children. We thought the patient may have slowly detached his retina outside the macula and had a



Figure 4. Vitrector utilized to open macrocyst for retinal reapposition.

long-standing chronic retinal detachment, thus leading to the grade B proliferative vitreoretinopathy. A list of other potential differential diagnoses that may have led to the retinal detachment is provided in Table 1.

Of these possible diagnoses, we believed that myopia was the most probable predisposing etiology. The patient was not likely to have Coats disease or Vogt-Koyanagi-Harada syndrome because of the nonexudative nature of the retinal detachment. Further, the patient did not have a connective tissue disease such as Marfan syndrome, but the possibility of an unrecognized and rare genetic abnormality predisposing toward high myopia, retinal detachment, and macrocysts was not ruled out.

Regarding x-linked juvenile retinoschisis, we felt this was unlikely to be an atypical presentation of the disease because the patient did not have any foveal or peripheral retinoschisis on initial exam and initially had fairly good vision. Still, patients with x-linked juvenile retinoschisis

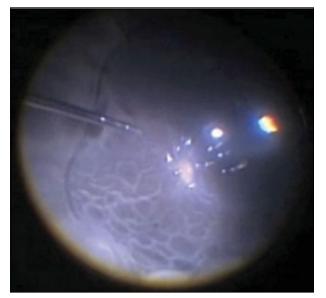


Figure 5. Perfluoro-n-octane liquid used. Note the macrocysts found even centrally.

do have an increased risk of retinal detachment, and early macular and peripheral changes may have been subtle and difficult to detect.

We explained the severe nature of the pathology and guarded prognosis in an extensive discussion with the family before proceeding to surgery. We felt that using general anesthesia in a hospital setting was the safest choice for this 15-year-old boy. Our surgical approach began with placing a scleral buckle to support the vitreous base. The large inferior dialysis and total retinal detachment necessitated the use of an encircling element. A 240 band and 70 sleeve were used to buckle. We used 7-0 polyethylene terephthalate sutures to hold secure the 240 band and placed it 3 mm posterior to the ora serrata with the loops extending from 10 mm to 13 mm posterior to the corneal limbus.

For vitrectomy, trocars were placed 3.5 mm posterior to the limbus. We used a dual bevel approach with initial scleral entry at 45° and subsequent perpendicular entry into the vitreous. Next, we performed 23-gauge pars plana vitrectomy with an Alcon Accurus system (the procedure took place in May 2010). We prefer the use of the BIOM noncontact viewing systems rather than a contact lens. We used 2% hydroxypropyl methylcellulose (Ocucoat, Bausch + Lomb) to preserve the clarity of the cornea.

Upon entry, we noted that the retinal dialysis extended over 270°, leaving only the superonasal retina attached. We also noted multiple macrocysts throughout the retina. The posterior hyaloid was already detached. Because there was significant vitreous haze, triamcinolone acetonide (Kenalog, Bristol-Myers Squibb) was not required

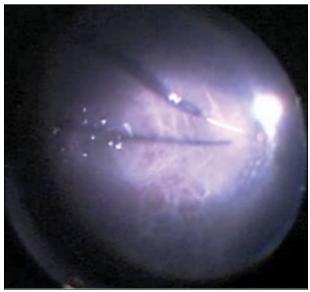


Figure 6. Endolaser used on areas of retinal reapposition.



Figure 7. Marked epiretinal membrane in the left eye.

to stain the vitreous. Vitreous traction was removed with the help of scleral depression, but the macrocysts were particularly problematic at the edge of the dialysis because they prevented retinal apposition to the underlying retinal pigment epithelium. Multiple cysts at the edge of the dialysis had to be opened with the vitreous cutter so that the retina would reattach. Many of the cysts were noted to be bigger than the size of the optic nerve and surprisingly large when viewed in profile at the edge of the dialysis (Figures 3-6).

Perfluoro-n-octane (Perfluoron, Alcon) was used to flatten the retina, and extensive endolaser was applied 360° in the retinal periphery; approximately 3000 applications were placed. Fluid-air exchange was performed, and we



widened the superotemporal sclerotomy for placement of 1000 centistoke silicone oil for long-term tamponade. We sutured all sclerotomies and closed the sclera with polyglactin. We recommended face-down positioning and saw the patient the following day.

Postoperatively, the retina remained flat and the patient's vision improved dramatically. By 1 month, the patient had a visual acuity of 20/25 in the left eye. This may seem surprising, but it is important to remember that the index of refraction of the lens (n = 1.5) and vitreous (n = 1.336) create an interface that is greatly altered after vitreous substitution. Smith and colleagues noted that when the vitreous is substituted with silicone oil (n = 1.40) in a phakic patient, there can be a 10 D to 14 D hyperopic shift. Obviously, this shift occurs at the lens plane and not at the spectacle plane. Our patient, who was a -15.5 D myope at the spectacle plane, was likely near -13 D at the lens plane, thus making the silicone oil shift push him to near refractive emmetropia.

Four months postoperatively, the patient developed an epiretinal membrane (ERM) with an associated decrease in vision to 20/100 (Figure 7). Fortunately, the retina remained attached under silicone oil. After another 2 months, when we felt he was stable, we elected to remove the silicone oil and peel the ERM.

Upon removal of the silicone oil, the retina promptly detached into a funnel configuration. Surprisingly, even previously lasered tissue did not remain adhered and there was no evidence of traction from PVR. The attempt to peel the ERM was aborted, and the silicone oil was replaced. The patient continued to have a difficult postoperative course in the subsequent year with the development of cataract, which was removed, followed by posterior capsular opacification, which was treated with YAG laser.

After the YAG capsulotomy, the patient had a mild anterior chamber reaction that may have been secondary to the silicone oil or repeated surgical manipulation. Steroids reduced the inflammation. Eventually the patient was tapered off steroids and is stable with 20/100 visual acuity. Currently, he is working and understandably hesitant to attempt silicone oil removal with an ERM peel again.

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