Uveal effusion, also referred to as choroidal detachment, can occur secondary to a number of ocular conditions that result in inflammatory or hydrostatic changes, including scleritis, posterior uveitis, and hypotony after glaucoma surgery. When no cause can be identified, this entity is termed uveal effusion syndrome (UES). It is by definition idiopathic but can be associated with nanophthalmia or hypermetropia in some cases.

The disease course is typically relapsing and remitting. Gradual superior visual field loss is the most common initial complaint, but patients may also report blurred vision or metamorphopsia. The magnitude of choroidal elevation in UES is highly variable, but it typically begins in the periphery. Simultaneous exudative retinal detachment that can involve the macula may occur (Figure 1A). Deep retinal or subretinal exudates may appear prior to serous detachment, and optic nerve edema may be present. There may be mild to moderate vitreous cell. Hyperpigmented changes in the retinal pigment epithelium (RPE), termed leopard spots, and permanently reduced VA can occur with chronic disease (Figure 1B). Spontaneous resolution occurs in most cases but may take months or several years.

**Epidemiology**

UES is extremely rare. Patients are typically middle-aged men. Approximately 65% of cases are bilateral. Approximately half of those requiring surgical intervention to reverse the condition in one eye will require surgery in the fellow eye. Nanophthalmos itself is always bilateral and may be sporadic or familial with either dominant or recessive patterns of inheritance.

**Theories of Pathogenesis**

Analyses of imaging with ultrasound biomicroscopy and MRI suggest that the sclera may be abnormally thickened in patients with UES. Histologic specimens of excised sclera in nanophthalmic eyes and eyes with UES demonstrate disruption of collagen fibers and the presence of glycosaminoglycan-like material within the interfibrillary spaces. The subretinal fluid in these eyes contains two to three times greater concentration of albumin and other proteins than in healthy eyes. In 50% of cases, cerebrospinal fluid protein is also elevated without pleocytosis. These findings suggest that transscleral exit of albumin may be impaired, leading to a hyperosmotic gradient into the choroid.

Another hypothesis is that compression of the scleral veins by swollen or abnormal scleral tissue results in choroidal fluid retention. Gass et al also reported hypoplastic or missing vortex veins in eyes with UES of both nanophthalmic and normal axial lengths. In nanophthalmic eyes with chronic angle closure, dilated episcleral veins and blood in Schlemm canal can be seen. These findings are indicative of

**Management of Uveal Effusion Syndrome**

Several surgical methods have been proposed to manage this rare condition.

**BY ZACHARY BODNAR, MD; AND PRITHVI MRUTHYUNJAYA, MD, MHS**
increased resistance to venous drainage from the eye. These proposed mechanisms are not mutually exclusive, and the end result is believed to be transudative exit of fluid from the choriocapillaris into the surrounding potential space. This leads to choroidal swelling and detachment. Fluid may also accumulate in the subretinal space, resulting in concurrent exudative retinal detachment.

Not all eyes with UES have been shown to have abnormalities of the sclera. Uyama et al performed lamellar sclerectomy in 19 eyes of 16 patients with UES. Histologic analysis of the excised sclera revealed disorganized collagen and deposition of proteoglycans in the extracellular matrix of the nanophthalmic eyes and eyes with thickened sclera but not in normal-sized eyes with normal scleral thickness. This may have implications for treatment, as the normal-sized, normal thickness eyes did not respond to sclerectomy.

**EVALUATION**

UES is a diagnosis of exclusion. The clinician should first rule out other causes of choroidal effusion and serous retinal detachment, such as posterior scleritis and uveitis, choroidal neoplasia such as uveal lymphoma, paraneoplastic syndromes, metastatic tumors of the choroid or retina, hypertensive choroidopathy, postoperative inflammation from cryotherapy or photocoagulation, and persistent hypotony.

IOP is usually normal in UES, and abnormal tonometry should prompt a search for other causes, as should signs of ocular inflammation. Central serous chorioretinopathy with multiple detachments of RPE can have a similar appearance to UES, but choroidal thickening and choroidal detachment will be absent. Dark-appearing choroidal detachments can be mistaken for suprachoroidal hemorrhage, peripheral exudative hemorrhagic chorioretinopathy, or choroidal melanoma.

**IMAGING**

B-scan ultrasonography can be used to document the presence of retinal and/or choroidal detachment. Often, the choroid and/or sclera will be thickened. It is not uncommon to see annular thickening of the anterior choroid, often captured best with ultrasound biomicroscopy. Ultrasound is helpful in identifying...
secondary causes of observed fundus changes, including choroidal tumors or the T-sign seen in posterior scleritis (a hyporeflective layer of fluid in the Tenon space surrounding the nerve).\textsuperscript{3,15} In ruling out other conditions such as scleritis, standardized A-scan ultrasound provides accurate axial length measurements to identify nanophthalmos.

Fluorescein angiography in UES typically shows areas of hypofluorescence corresponding to leopard-spot pigmentation, with angiographic leakage rarely identified.\textsuperscript{2,4} In the acute setting, the leopard spots will appear as hyperfluorescent on fundus autofluorescence (FAF), but over time this may change to a mix of hyper- and hypo-autofluorescence on FAF (Figure 2). Corresponding OCT through the spots will reveal focal thickening of the RPE. Both of these findings may diminish with resolution of subretinal fluid, but typically the FAF changes will persist.\textsuperscript{16}

Enhanced-depth OCT demonstrates increased choroidal thickness compared with normal eyes and the presence of large areas of hyporeflectivity that may correspond to engorged choroidal veins or expansion of the suprachoroidal space.\textsuperscript{17} Indocyanine green angiography appears to confirm the presence of dilated choroidal vessels.\textsuperscript{18} ERG may be abnormal but is nonspecific.\textsuperscript{2}

**Nonsurgical Management**

Much of the literature suggests that UES is refractory to medical management. Several reports have indicated that systemic corticosteroids and immunomodulating agents produce neither a dramatic nor lasting response.\textsuperscript{2,4,18} However, some more recent studies provide evidence that a subset of eyes with UES may respond well to oral, periocular, or topical steroids and poorly to surgical treatment.\textsuperscript{13} A large retrospective study by Shields et al analyzed the response of eyes with UES and normal axial length without evidence of scleral thickening to oral, periocular, or topical corticosteroids or a combination thereof. Of 104 eyes, 95% had resolution of effusions after a median of 3 months of steroid therapy, and only 5% required scleral window surgery.\textsuperscript{19}

Other proposed nonsurgical therapies include laser photocoagulation and topical application of a prostaglandin analogue. Kumar et al reported improvement in chronic exudative retinal detachment and modest improvement in vision in two patients with UES after treatment with long-term slow-release indomethacin and scatter laser photocoagulation, but the authors acknowledged that the improvement may have been spontaneous.\textsuperscript{18} Several other reports found no lasting benefit of laser photocoagulation to areas of angiographic leakage.\textsuperscript{2,4}

In principle, prostaglandin analogues could be helpful by increasing uveoscleral outflow.\textsuperscript{11,20} Derk et al reported resolution of effusions after 3 months in three patients with bilateral disease who were treated with a combination of topical latanoprost and oral acetazolamide.\textsuperscript{21} Prostaglandin analogues have not been evaluated for the treatment of UES in any large clinical studies.

**Surgical Management**

Several surgical treatments have been described, with the surgical goals being (1) direct (internal or external) drainage of the subretinal fluid, (2) reduction in scleral resistance to allow fluid evacuation from the suprachoroidal space, (3) unroofing of the vortex veins, or (4) a combination of approaches.

A common approach is to create full-thickness sclerectomies to facilitate the egress of suprachoroidal fluid. In a series of 23 eyes with UES and normal axial length, Johnson and Gass reported anatomic improvement within 6 months in 83% of eyes after a single procedure to create sclerectomies. After one or two procedures, 96% of eyes demonstrated anatomic improvement. The average time to reattachment was 2 to 3 months, with recurrence in 23% of eyes. Final VA improved by 2 or more lines in 56% of eyes, remained stable in 35% of eyes, and worsened in 9%. The median final VA improved from 20/100 preoperatively to 20/60.\textsuperscript{1} Jackson et al reported full resolution of choroidal effusions in seven of 14 eyes using this same technique.\textsuperscript{22} Two case reports describe a modification of this technique using mitomycin C applied to the sclerectomy bed to reduce scarring.\textsuperscript{23,24}

Sabrosa et al reported a successful revision of a failed deep sclerectomy surgery using a scleral punch and mitomycin C.\textsuperscript{25} Yepez and Aravalo used Ex-Press Glaucoma Filtration Device (Alcon) shunts to maintain patency of the sclerotomy.\textsuperscript{26} Schneiderman and Johnson described a case in which pars plana vitrectomy and internal drainage of subretinal fluid with C\textsubscript{3}F\textsubscript{8} gas tamponade was combined with partial thickness sclerectomies in a 73-year-old patient with UES.\textsuperscript{27}

Uyama et al described a technique they called sub scleral sclerectomy in which two-thirds—thickness scleral flaps measuring 4 mm by 5 mm are made in the inferotemporal and inferonasal quadrants. Beneath each flap, the remaining sclera is excised to expose the choroid. The edges of the scleral wounds are then cauterized, and the flap is loosely sutured. If a second procedure is required, additional sclerectomies can be performed in the upper quadrants. The authors also administered mannitol
and acetazolamide postoperatively to attempt to hasten the resolution of fluid. They reported that their surgical approach resulted in resolution of subretinal fluid and choroidal thickening in nanopthalmic eyes and normal-sized eyes with scleral thickening but not in eyes with normal axial length and scleral thickness.13

Ghazi et al described a modified technique wherein preoperative or intraoperative B-scan ultrasound is used to identify areas of maximal scleral choroidal thickening in each involved quadrant. These sites are then chosen for sclerostomy placement using a Kelly punch. For sites located anterior to the equator, a single sclerostomy punch within a 4-mm circumferential incision is made. At posterior sites, a 4-by-4-mm, two-thirds-thickness sclerectomy is also performed. These authors reported complete resolution of choroidal thickening and subretinal fluid in five of six eyes using this technique, and the majority resolved within 3 to 4 months.15

Faulborn and Kölli reported rapid resolution of choroidal effusions and serous retinal detachment in five eyes of four patients using another modified technique. They created 2-by-8-mm full-thickness scleral flaps without sclerectomy, 4 mm posterior to the limbus in each quadrant. The flaps were not secured with sutures.1 As there is a risk of hemorrhage associated with the creation of scleral windows, Bausili et al reported use of a CO₂ laser to perform simultaneous tissue dissection and cautery.28

Surgical decompression of the vortex veins where they exit the sclera has also been described. This procedure may be combined with drainage of choroidal or subretinal fluid with intravitreal injection of air.12 A technique described by Brockhurst combines scleral resection to unroof the vortex veins with sclerostomies to drain the suprachoroidal fluid. The sclerectomy begins at the insertion of the rectus muscles and extends to the exit of the vortex veins in each quadrant. Separate, smaller full-thickness sclerostomies are made in each scleral dissection bed for fluid drainage. In Brockhurst’s original report, retinal reattachment was achieved in eight of 10 eyes.12 Yue et al subsequently reported successful treatment of two nanopthalmic eyes with UES using this technique.9

Ozgonul et al suggested using a staged approach beginning with partial thickness sclerectomies. If a second surgery is required, revision of the flap using mitomycin C can be considered. Full-thickness sclerostomy, as per the Ghazi method, or unroofing of the vortex veins may be considered in eyes that are refractory to revision with mitomycin C.29

Each of the surgical approaches described above is an attempt to create a durable, low-resistance pathway for choroidal fluid to exit. Matlach et al proposed that it may be sufficient to drain the fluid intraoperatively using a penetrating diathermy probe to access the suprachoroidal space, with anterior chamber infusion placed to create a pressure differential. Complete flattening of the choroid can be achieved by the end of surgery using this technique, the authors asserted, but one of two eyes in the small series they reported required a repeat procedure for late recurrence.30

Our own preference is to perform...
Concomitant chronic serous retinal detachment involving the macula can lead to permanent vision loss. Multimodal imaging can be helpful to confirm UES and differentiate it from other causes of choroidal effusion. Although some eyes with normal axial length and no scleral thickening may respond to treatment with oral, topical, or periocular corticosteroids, many will require surgical intervention to facilitate the flow of fluid from the choroid through the sclera. However, a delay in diagnosis of this condition prior to definitive surgery may result in chronic visual compromise. The review provided above may be helpful for clinicians who encounter patients with this rare but potentially serious condition.

It pays to know about UES
UES is a rare idiopathic cause of choroidal detachment that may be associated with nanophthalmos and increased scleral thickness.

Figure 5. Creation of sclerotomy using Kelly punch.

four-quadrant scleral windows. After the conjunctiva is exposed, the four rectus muscles are isolated and the oblique quadrants revealed. A mark is made to outline a 5- by 5-mm scleral flap in mid-quadrant, about 4 mm from the rectus muscle insertion (Figure 3). Using a No. 57 scleral blade (Alcon), an 80% scleral thickness lamellar flap is outlined and dissected (Figure 4). Then a smaller, inner scleral 3-mm incision is made down to the glistening fibers of the outer choroid but clearly into the suprachoroidal space. It is important not to have any thin lamella of tissue covering the uveal tissue. Next, using a Kelly punch, the inner lip of the remaining scleral lamellar fibers are removed to create a round circle of exposed uveal tissue (Figure 5). The outer scleral flap is then amputated. (Alternatively, it can be sutured back without significant tension.) This is repeated in the other three quadrants if there is adequate exposure. We do not purposely try to unroof or disturb the vortex veins. Rarely, if there is a highly bulous exudative retinal detachment, secondary drainage of the fluid can be added, but this is often not necessary.

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Financial disclosure: None

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