Surgical implications in exudative retinal detachment

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Background

Surgeries for retinal reattachment form the mainstay of treatment for rhegmatogenous, tractional and combined retinal detachment. However, they do not form the first line of management in exudative retinal detachment. Massive exudative retinal detachment not responding to the conventional line of treatment or not permitting the performance of conventional line of treatment is chosen for surgical intervention. It is a mandatory prerequisite to identify the pre-existing factor that has been instrumental in the initiation of exudative retinal detachment (ERD).

ERD occurs due to breakdown of the complex interactions of forces that maintain the normal apposition of the retina, RPE, Bruch’s membrane and choroid (Table 1). Failure of these forces, due to various causes enlisted below, potentially could lead to formation of fluid under the neurosensory retina. Central serous chorioretinopathy (CSCR) known to be a very common cause for localized ERD either in stressed males or patients who are on steroid medication for various ailments are potentially at risk for CSCR. This variety of CSCR is described as typical and is treated with lifestyle modification, avoiding steroids in all forms; focal laser photocoagulation is done to leak identified on fundus fluorescein angiogram for extrafoveal leaks and photodynamic therapy (PDT)/micropulse diode laser for subfoveal leak.

When the fluid is non-resolving and retinal detachment is chronic and bullous, permanent damage to retinal pigment epithelium as well as outer retinal structures occurs and persistence of subretinal fibrin may lead to subretinal fibrotic scar formation. To avoid these complications, surgical intervention is planned after failure of conventional treatment. However, surgical treatment is never the first line of management in these conditions.

Surgical options

Drainage of subretinal fluid (SRF)

- External approach
- Internal approach

External approach

Methods of external drainage of SRF

1. Scleral cut down and choroidotomy
   - Needle opening of choroid
   - Laser opening of choroid

2. Needle aspiration

3. 26-G trocar and cannula drainage by retraction

Steve Charles technique¹ of needle drainage is a relatively safer procedure can be combined with Chandelier illumination offers the comfort of using BIOM with the hands free for drainage of SRF under direct visualization and also allows surgeon to withdraw needle as the retina flattens avoiding complications like retinal incarceration, subretinal haemorrhage and vitreous loss. Continuous monitoring² with indirect ophthalmoscopy during the

Table 1: Causes of exudative retinal detachment

<table>
<thead>
<tr>
<th>Choroidal causes:</th>
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<tbody>
<tr>
<td>a. Idiopathic choroidal vascular hyperpermeability *CSCR*</td>
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<td>b. Localized choroidal vascular hyperpermeability *Choroidal haemangioma*</td>
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<td>c. Tumours of choroid *Choroidal melanoma*</td>
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<td>d. Impaired outflow through sclera *Uveal effusion syndrome*</td>
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<td>e. Inflammatory conditions of choroid and sclera *VKH syndrome* *Sympathetic ophthalmitis* *Posterior scleritis*</td>
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<td>f. Ischaemia and altered choroidal hypperpermeability *Preeclampsia, malignant hypertension, disseminated intravascular coagulation, idiopathic polypoidal choroidal vasculopathy*</td>
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<th>Retinal causes:</th>
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<tr>
<td>a. Retinal angiomatosis</td>
<td></td>
</tr>
<tr>
<td>b. Coats’ disease</td>
<td></td>
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<tr>
<td>c. Retinal venous occlusive disease</td>
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<tr>
<td>d. Retinal artery macroaneurysm</td>
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<tr>
<td>e. Idiopathic frosted branch angiitis</td>
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<th>Miscellaneous causes:</th>
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<tr>
<td>a. Post-retinal reattachment surgery</td>
<td></td>
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<tr>
<td>b. Post-laser therapy</td>
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<td>c. Post-cryotherapy</td>
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drainage has also been reported to be free from complications.

Subretinal aspiration and injection device (SA-AID) introduced by Kang et al.\(^3\) to facilitate controlled external drainage of SRF. This device provides a safe approach to the subretinal space because it penetrates the eye wall obliquely and allows a changeable, predetermined length of the needle tip. The surgeon can also directly observe the retina with an indirect ophthalmoscope during the drainage.

**Internal approach**
- Pars plana vitrectomy with endodrainage through a drainage retinotomy, endolaser/cryopexy and tamponade with gas/silicon oil.
- Pars plana vitrectomy with liquid perfluorocarbon assisted external drainage.
- Pars plana approach endolaser without vitrectomy.

**Pearls for external drainage**
- **External drainage done for bullous RD in periphery**
- **If using needle, bevel down will avoid retinal aspiration.**
- **Choroidotomy opening and needle can get occluded by subretinal fibrin/cholesterol.**
- **AC maintainer may help for near complete drainage in selected situations.**

**Central serous chorioretinopathy**
Longstanding CSCR can cause severe vision loss due to significant photoreceptor damage and later on may lead to subretinal fibrotic scar formation and foveal scarring (Figure 1).

Various authors have claimed to have good results via both external and internal drainage approaches of SRF in non-responding chronic CSCR.

Bondalapati et al.\(^4\) had reported external drainage of SRF in six male patients with ERD following chronic CSCR with no recurrences. External drainage was performed with 26-G needle (Steve Charles technique). Chandelier-assisted external drainage gives the advantage of keeping the surgeon’s hands free to perform drainage. An external approach has been modified by various authors. Figueroa et al.\(^5\) performed sutureless sclerotomies beyond equator one in each inferior quadrant without SRF drainage followed by erect positioning. Retinal reattachment occurred within 24 h of surgery allowing focal laser photocoagulation to the leaking lesions on FFA. Good visual gain was recorded noted by the authors postoperatively with the interesting fact of nil surgical complications that were recorded.

Reasonable success rates were also claimed by authors\(^6\) who had done an internal approach by parsplana vitrectomy (PPV) with scleral buckle. Chen et al.\(^7\) suggested the use of liquid perfluorocarbon in a procedure which combined both PPV
All precipitating factors should be treated posteriorly. Liquid perfluorocarbon was used to attach the posterior retina while the SRF was drained via the external drainage and then applied diode laser to the area of leak. However, the visual outcome was poor which they attributed to long duration of macular detachment and subsequent subretinal fibrosis adjacent to the central retina. A similar observation was also made by Kang et al.⁸ who concluded that surgical treatment would be beneficial if performed early to prevent subretinal proliferation, which usually tends to have a negative influence on the final visual prognosis.

**CSCR**

- CSCR usually is treated well with photodynamic therapy/laser/ micro pulse diode laser.
- Exudative detachment usually occurs due to a RPE rip which may heal in few months.
- All precipitating factors should be treated before resorting to surgical treatment

**Diffuse choroidal haemangioma**

Diffuse choroidal haemangioma is a benign vascular tumour associated with cutaneous, ocular or central nervous system findings and may also be associated with Sturge-Weber syndrome. Diffuse red-orange thickening of the posterior choroid that has poorly defined borders and extends over a broad area.

Ultrasonography shows a diffusely thickened choroid on B scan, while A scan shows moderate to high internal reflectivity with an overlying SRF. Magnetic resonance imaging in $T_1$-weighted images are hyperintense while it is isointense to the vitreous in $T_2$-weighted images. Conventional therapy includes photodynamic therapy, external beam radiation therapy and oral propranolol. Shanmugam et al.⁹ in their unpublished data have treated a patient with diffuse choroidal haemangioma with bullous RD, resistant to conventional therapy with external drainage of SRF along with intraoperative transpupillary thermotherapy with good control of tumour and resolution of SRF.

**Uveal effusion syndrome**

Uveal effusion syndrome (UES) may be idiopathic or associated with abnormalities of choroid or sclera. It is basically a diagnosis of exclusion when all other causes are ruled out. Histological studies have demonstrated disruption of collagen fibers and filling of the intertrabecular spaces with amorphous glycosaminoglycan-like material obtained from the excised scleral tissue.

Gass¹⁰ hypothesized that abnormal thickness of the sclera could possibly compromise the choroidal vascularization, favouring the accumulation of extravascular proteins in the choroid and the ciliary body and thereby increasing fluid migration into the suprachoroidal space producing a ciliochoroidal detachment with secondary movement of water and proteins into the subretinal space. Eye being devoid of lymphatics intracocular extravascular proteins are removed via Schlemm’s canal and aqueous veins anteriorly while posteriorly by trans-scleral diffusion and through the emissary canals into the orbital tissues. Since increased thickness of the sclera was preventing the trans-scleral diffusion, partial thickness scleromas would help in successful reabsorption of subretinal fluid.

Posterior microphthalmos (PM) is an entity that has been very newly introduced to differentiate from nanophthalmos. Both have short axial length, except the fact that PM has normal corneal diameter while nanophthalmos has reduced corneal diameter. Clinical features reported in PM are reduced axial length with a normal-sized cornea and normal anterior segment and a disproportionately small size of the posterior segment. Other features reported are high hyperopia, uveal effusion, elevated papillomacular retinal fold, fine retinal striae, tapetoretinal degenerations similar to fundus flavimaculatus and fundus albipunctatus, retinoschisis, reduced foveal avascular zone, pseudodiplopiedema and macular holes.¹¹⁻¹⁵

Drainage of SRF is never successful as an independent procedure as it leads to higher chances of recurrences. The most common treatment performed is full-thickness sclerectomies to provide an exit for choroidal fluid, and this has been well supported in the literature by a large case series¹¹ which showed anatomical improvement in single procedure amounting to 83% and visual recovery of two or more lines in 56% of eyes.

Faulborn et al.¹⁶ described regression of SRF and resolution of choroidal detachment following pars plana full-thickness unsutured sclerotomies without sclerectomy in their series of five eyes of four patients with UES due to nanophthalmos.

**Coats’ disease**

The most popular and widely used classification for Coats’ disease is by Shields et al.¹⁷

- **Stage 1**: Telangiectasia
- **Stage 2**: Telangiectasia and exudation (2A extrafoveal exudation, 2B foveal exudation)
- **Stage 3**: ERD (3A subtotal and 3B total)
- **Stage 4**: Total retinal detachment and secondary glaucoma
- **Stage 5**: Advanced end-stage disease

Multiple modalities have been employed to treat including diathermy, laser photocoagulation, cryotherapy, SRF drainage, scleral buckling surgery, pars plana vitrectomy and intravitreal anti-vascular endothelial growth factor (VEGF) therapy. Treatment is aimed at destroying abnormal vasculature and aneurysmal dilations.
The surgical approach is contemplated at stages 3 and 4 as serous bullous retinal detachment that does not permit regular laser ablation or transconjunctival cryopexy of retinal vessel telangiectasias (Figure 2).

Interventional case series of 13 patients by Daniela et al.\textsuperscript{18} presented a modified surgical technique in the treatment of retinal detachment secondary to advanced Coats’ disease in children and reported on long-term anatomical and functional outcome. Their patients underwent PPV including exocryotherapy applied after fluid-air exchange in order to achieve complete treatment of the vascular changes, to reduce associated side-

\textbf{Figure 1:} CSCR with subretinal fibrosis.

\textbf{Figure 2:} Colour funds montage of successfully drained ERD following Coats’ disease.
effects and to avoid retinectomy and silicone oil tamponade. They described nil further therapy in 31%, additional therapy in 69% and revisional surgery with silicone oil tamponade in 46% of patients as well as visual improvement (27%), visual stability (36%) and visual deterioration (36%).

Cai et al.19 have described the effectiveness of endolaser photocoagulation by a two-port pars plana nonvitrectomy approach for treating Coats' disease with shallow ERD treatment. In their study of 25 eyes with treatment sessions varying from one to five, the retina reattachment rate was 96% and retinal redetachment rate was 4%. The proposed technique involves laser application directly to telangiectasia within the retina without targeting the RPE to create thermal injury in a non-vitrectomized eye with good peripheral retinal examination. Injection of viscoelastic was performed to minimize vitreous traction at the side ports.

Risk of vitreoretinal traction20 on usage of bevacizumab for Coats' disease with ERD was alerted by Ramasubramanian et al. in their analysis of eight patients with Coats' disease manifesting total or partial ERD where the retinal telangiectasia was treated with standard laser photocoagulation and/or cryotherapy plus additional intravitreal bevacizumab (1.25 mg/0.05 ml). Authors reported resolution of retinopathy (100%), SRF (100%), retinal exudation (75%) and vitreous fibrosis developed (50%).

Use of intravitreal bevacizumab is controversial. Victor et al.21 in their study of 24 patients used intravitreal bevacizumab followed by laser vascular ablation in the management of advanced Coats' disease presenting with ERD. None of the patients in their series developed vitreous fibrosis.

Rishi et al.22 have described treatment and outcome patterns in 307 Indian eyes with Coats' disease. Subretinal drainage was done in 21 (6.8%) cases and an attached retina was found in 12 and detached retina in 9 cases. Vitrectomy was done in 8(4.2%) cases and an attached retina was found in 7 and detached retina in a single case.

**Coats disease**

- **Laser is the main stay of treatment in Coats disease**
- **SRF drainage attempted in combination with cryotherapy for bullous peripheral detachments.**
- **The aim of SRF drainage is to prevent phthisis or painful blind eye and not visual gain.**
- **Retinotomy should be avoided and endodrainage should be performed in cases of combined retinal detachment with signs of chronicity.**
- **Retinoblastoma should be excluded in all children with bullous detachment and leukocoria.**

**Vogt–Koyanagi–Harada syndrome**

The Vogt–Koyanagi–Harada syndrome (VKH) is an autoimmune-mediated genetically influenced bilateral, diffuse granulomatous uveitis associated

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**Figure 3:** Polypoidal choroidal vasculopathy with ERD.
with poliosis, vitiligo, alopecia and central nervous system and auditory signs.

Tyrosinase or tyrosinase-related proteins have been proposed to be possible target antigens. Till date, there is no single diagnostic test which can pinpoint the diagnosis of VKH. We do not have any evidence in the literature regarding the occurrence of sensitization to melanocytic antigens.

VKH disease diagnosis is purely based on exclusion of other diseases. Ultrasonography is mandatory to demonstrate diffuse choroidal thickening, without a positive ‘T’ sign. FFA should show the presence of pinpoint areas of choroidal leakage.

The conventional treatment includes control of inflammation by regional, oral and intravenous corticosteroids, cyclosporine, antimetabolites and alkylating agents.

A successful surgical treatment (vitrectomy) for bilateral bullous retinal detachment in a patient with VKH disease is reported. Gaun et al. has described parsplana lensectomy and vitrectomy being done on a 78-year-old female with severe reduction of visual acuity in both eyes because of an extremely bullous non-rhegmatogenous retinal detachment accompanied by VKH disease. They had also supplemented with systemic and topical corticosteroid therapy. They reported no recurrence of retinal detachment even after tapering the dose of corticosteroid. They had concluded that vitrectomy may be an effective therapeutic option in the treatment for severe bullous retinal detachment associated with VKH disease.

Vogt-Koyanagi-Harada syndrome
- Chronic VKH can be differentiated from CSCR by a course of steroids and improvement with no worsening.
- Control of inflammation is the mainstay of treatment.
- SRF drainage is attempted in chronic, non-responsive bullous retinal detachments like in Coats disease.
- Recurrences are common.

Von Hippel–Lindau disease
Von Hippel–Lindau disease (VHL) is a multisystem familial tumour syndrome with autosomal dominant inheritance with high penetrance. Ocular lesions are characterized by retinal capillary haemangioma which is the most common presentation of VHL disease. Extraocular lesions include central nervous system haemangioma, endolymphatic sac tumour of the inner ear, pancreatic cysts, pancreatic adenoma, pancreatic islet cell tumours, pheochromocytoma, renal cyst, renal carcinoma, cystadenoma of the epididymis and the broad ligament. VHL gene is a tumour suppressor gene and is mapped to the short arm of chromosome 3p25. Genetic testing can be done with greater precision. Use of surveillance protocols is recommended for improving survival rate through early detection.

Conventional therapy to retinal capillary haemangioma is based on size of lesion, amount of exudation and the coexisting clinical condition. Treatment modalities are observation, laser photocoagulation, transpupillary thermotherapy, cryotherapy, photodynamic therapy, plaque radiotherapy and proton therapy. ERD in VHL can occur due to retinal capillary haemangioma itself or secondary to treatment of retinal capillary haemangioma.

Fang-Yi et al. have reported a rare case of VHL disease with multiple peripheral retinal capillary haemangioma (RCHs) treated by PDT combined with intravitreal bevacizumab injection, which showed significant tumour regression but with persistent exudative RD that required surgical intervention. The massive exudation which developed during the second day was probably due to the summation reaction of three large RCHs to PDT. The authors further recommended that fractionated PDT may be used in cases with multiple RCHs where one tumour is treated at a time to reduce the post-treatment reaction.

Kim et al. indicated that treatments for large RCHs in patients with VHL disease must be based on careful selection. Although PDT is considered one of the useful treatment options for RCH, PDT combined with an intravitreal injection of bevacizumab can lead to permanent and severe deterioration of vision because of massive ERD. Therefore, frequent fundus examinations should be performed after PDT. They could successfully reattach the retina with PPV, internal SRF drainage, retinotomy, endodiathermy, cryotherapy and endolaser therapy on the RCH, and injection of silicone oil and bevacizumab.

The two theories explain the increased exudation from retinal haemangioma following PDT. First, it could be increased leakage from the vascular lesion, a response known to occur with laser photocoagulation and cryotherapy. Alternatively, PDT can affect the choroidal vasculature, causing ischaemia and even shutdown of the choriocapillaris or retinal capillaries with breakdown of the vascular barrier and significant SRF exudation, a response seen in PDT treatment of choroidal neovascularization.

Von Hippel–Lindau disease
- Peripheral tractional detachment can occur following laser/cryotherapy and can be treated with scleral buckling.
• Occurrence of iatrogenic retinal breaks should be prevented while performing vitrectomy in bullous retinal detachments.
• Absence of posterior vitreous detachments and presence of thick fibrous sheets is challenging.
• Bleeding can be uncontrollable.
• Endocryotherapy can be used to treat vascular lesions

**ERD in retinopathy of prematurity**

Ittiara et al. had reported the rare complication of bilateral ERDs in a preterm infant (25 weeks of gestational age) treated with only intravitreal bevacizumab for ROP. The bilateral ERDs developed after 1 year. The right eye was treated with intravitreal bevacizumab, laser ablation and scleral buckling, resulting in resolution of the exudation and detachment. The left eye was treated with vitrectomy and lensectomy, but persistent exudation and detachment remained. This case study demonstrates the need for frequent and longer follow-up after bevacizumab injection for retinopathy of prematurity until the completion of retinal vascularization or until the completion of peripheral ablation with laser photocoagulation.

**Other conditions with ERD**

- Exudative detachments can occur with posterior scleritis which can be managed with anti-inflammatory agents or anti-infective therapy based on proper diagnosis.
- Exudative detachments can occur after endophthalmitis or choroidal abscess and control of infection is the mainstay of treatment. Therapeutic and diagnostic vitrectomies may be required for proper medical management.
- Exudative detachments can occur following idiopathic polyoidal choroidovasculopathy (Figure 3) and anti-VEGFs are the mainstay of treatment. Vitrectomy may be required to treat breakthrough vitreous haemorrhage in conjunction with anti-VEGF to settle the exudative element. Gas tamponade helps in displacement of subretinal bleeds in these patients with or without the use of tissue plasminogen activator.
- Exudative detachments can present with choroidal detachments following sudden decompression of globe in glaucoma filtration surgeries and can be managed medically similar to exudative RD following laser or cryotherapy.
- Masquerade syndrome can present with exudative detachments of retina, and choroid and malignancy (primary or secondary) should be excluded before resorting to any surgery.

**Conclusion**

Exudative detachments may show typical shifting fluid and the ethology should be ascertained. The primary cause should be treated and exudative detachments may resolve. Surgery is reserved for limited patients which do not respond to conventional therapy. Recurrences are common even after surgery and visual results may not be encouraging. Patients may require long-term follow-up to achieve a complete remission.

**References**


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