LETTERS

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Orbital varices and orbital wall defects

Orbital varices are a vascular hamartoma typified by a plexus of low pressure, low flow, thin walled and distensible vessels that intermingle with the normal orbital vessels.1–4 If freely communicating with the orbital circulation, engorgement of varices can occur by increasing venous pressure through the Valsalva manoeuvre,5 bending posture,6 coughing or straining and these, in turn, lead to the clinical characteristics of variable proptosis, intermittent pain, and orbital haemorrhage.7,8

Observation is usually warranted for small lesions, but surgical intervention may be necessary in advanced cases: indications for surgical intervention include non-resolving episodes of thrombosis, severe disfiguring proptosis or displacement of the globe, and optic nerve compression.9,10 Surgery can be extremely difficult, as varices are very friable and intimately intermixed with normal orbital structures; there is also a significant risk of visual loss as a result of haemorrhage or optic nerve damage, the latter being generally caused by vascular compromise.11,12 The association of orbital venous anomalies with orbital wall defects provides a further source of surgical difficulty because of the close proximity of intracranial structures and the continuity with extraorbital or intracranial venous anomalies.

Case series

The orbital database, at Moorfields Eye Hospital, was used to identify patients with a clinical diagnosis of low pressure orbital varices and their orbital imaging (computed tomography and/or magnetic resonance image) was reviewed. Images were examined for evidence of orbital expansion, osseous defects of the orbit, nose or sinuses, and anomalies of the frontal lobes. Patients who had either orbital or intracranial surgery before the date of imaging were excluded from the investigation.

The clinical diagnosis of orbital varices was identified in 310 patients, and imaging was available for 223 patients (72%). Six patients with previous orbital or intracranial surgery were excluded and nine cases (4%) had associated anomalies of the neighbouring orbital walls (table 1).

Four cases (patients 1–4) were associated with “pitting” of the orbital wall secondary to orbital varices (fig 1A). Another three cases (patients 6–8) were associated with enlarged superior orbital fissure and two cases (patients 5 and 9) with multiple orbital roof “defects” (fig 1B). Orbital varices were present up to the dural space in two cases (patients 4 and 5), and involved the frontal lobe parenchyma in one case (patient 6; fig 1C, D).
Table 1 Characteristics of nine patients with orbital wall defects in association with orbital varices

<table>
<thead>
<tr>
<th>No</th>
<th>Side</th>
<th>Age (years) at referral</th>
<th>Sex</th>
<th>Main location of orbital varix</th>
<th>Expansion of orbit</th>
<th>Absent walls</th>
<th>Ethmoid</th>
<th>Cribriform</th>
<th>Frontal</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Left</td>
<td>6</td>
<td>M</td>
<td>Medial and extensive superomedial</td>
<td>Present</td>
<td>Small roof defect</td>
<td>Pitted bone and smaller ethmoid</td>
<td>L-low R-normal</td>
<td>Dips low at cribriform</td>
</tr>
<tr>
<td>2</td>
<td>Right</td>
<td>21</td>
<td>F</td>
<td>Extraconal-medial</td>
<td>Tiny thin area SNQ</td>
<td>Pitted bone and smaller ethmoid</td>
<td>Compressed</td>
<td>R-low L-mild</td>
<td>Low frontal lobe over cribriform</td>
</tr>
<tr>
<td>3</td>
<td>Left</td>
<td>62</td>
<td>M</td>
<td>Superomedial</td>
<td>Present</td>
<td>Pitted roof and small defects of veins</td>
<td>Normal</td>
<td>Normal</td>
<td>Hint of varix but otherwise normal</td>
</tr>
<tr>
<td>4</td>
<td>Right</td>
<td>58</td>
<td>F</td>
<td>Panorbit intraconal and extracranal</td>
<td>Present</td>
<td>Post superior wall and pitted bone</td>
<td>Normal</td>
<td>Normal</td>
<td>Varices up to dural space</td>
</tr>
<tr>
<td>5</td>
<td>Right</td>
<td>47</td>
<td>M</td>
<td>Panorbit intraconal and extracranal</td>
<td>Absent</td>
<td>Posterior orbital roof</td>
<td>Normal</td>
<td>Normal</td>
<td>Varices up to dural space</td>
</tr>
<tr>
<td>6</td>
<td>Right</td>
<td>14</td>
<td>F</td>
<td>Posterior intraconal, superior ethmoid</td>
<td>Present</td>
<td>Enlarged SOF</td>
<td>Normal</td>
<td>Normal</td>
<td>Varices into frontal lobe</td>
</tr>
<tr>
<td>7</td>
<td>Left</td>
<td>40</td>
<td>M</td>
<td>Posterior intraconal</td>
<td>Present</td>
<td>Enlarged SOF and small lateral wall</td>
<td>Slightly smaller</td>
<td>Unknown</td>
<td>Normal</td>
</tr>
<tr>
<td>8</td>
<td>Left</td>
<td>37</td>
<td>F</td>
<td>Posterior intraconal and extracranal</td>
<td>Present</td>
<td>Very enlarged SOF, patchy SNQ defects posteriorly</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>9</td>
<td>Left</td>
<td>66</td>
<td>M</td>
<td>Extracranal–superior (large)</td>
<td>Present</td>
<td>Posterior orbital roof</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
</tbody>
</table>

SNQ = suprasellar quadrant; SOF = superior orbit fissure.

One patient (case 2) had thinning of the suprasellar quadrant of the orbit, nasal orbital wall pitting, and a low ipsilateral cribriform plate, when first seen at age 21 in 1981 (fig 1E, F). On repeat imaging 20 years later (2001), this patient was noted to have developed proptosis, a defect in the superonasal wall of the orbit, and a new mid-line nasal encephalocele (fig 1J, I).

Comment

Fine cut (3 mm) orbital CT scans easily delineate varices and diagnostic phleboliths, which occur from thrombus formation,2 and provide an excellent natural contrast between brain, bone, and varix. The typical findings for varices include an ill defined multiloculated mass, with some patchy contrast enhancement, in communication with the neighbouring orbital circulation3; diffusion expansion of the orbital walls is well recognised in some cases, especially in childhood lesions.

Several factors may have biased the study population: many are symptomatic patients, having been referred from other ophthalmic units in consideration for surgical intervention. The apparent incidence of orbital wall defects (4%) in our series may, therefore, be a slight overestimate. In a minority of patients, orbital varices may be associated with orbital wall defects, and such defects may, eventually, lead to an encephalocele formation. Clinicians should be aware of these, apparently unreported, associations before embarking on surgical intervention for orbital varices.

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References


4 Rubin PAD, Ramula HD. Orbital venous anomalies demonstrated by spiral computed tomography. Ophthalmology 1997;104:1463–70


Exenteration of invasive conjunctival squamous cell carcinoma

Ocular surface squamous neoplasia (OSSN) includes conjunctival intraepithelial neoplasia with dysplasia, carcinoma in situ and conjunctival squamous cell carcinoma (SCC). Besides ultraviolet B irradiation identified as an risk factor, OSSN is associated with human papillomavirus type 16 and 18 (HPV-16, HPV-18).1,4 The exact role and possible prognostic value of p53 overexpression is unclear and little is known about its expression during the development of conjunctival SCC.

Case reports

Patient 1

A 75 year old man was referred with a 2 year history of an extensive conjunctival papillomatous tumour of the left eye covering three quarters of the cornea with visual acuity of hand perception. A full thickness biopsy was performed.

Both patients underwent orbital exenteration including removal of the eyelids. Histopathologically the focal invasive, completely removed tumour of patient 1 grew in a papillomatous manner. The tumour cells of the conjunctival neoplasm showed strongly enlarged nuclei with prominent nucleoli, and formed cohesive units with intercellular bridges (fig 1B).

The exophytic tumour of patient 2 was predominantly intraepithelial with foci of subepithelial invasion. Focal tumour anaplasia was observed in the otherwise moderately differentiated tumour with squamous cell differentiation.

Immunostaining of both specimens revealed strong p53 (monoclonal mouse anti-human p53-protein DNA), overexpression (>26% of epithelium cells) and low expression of p21 (<6% of epithelium cells) of the invasive region of the tumour indicating an inactivating p53 mutation (fig 1C). While in patient 1 expression for p53 was found in all epithelial layers, in patient 2 it was expressed suprabasally. In contrast, both p53 and p21 showed moderate reactivity in the dysplastic region up to the middle layer of the tumour (fig 1D). In the apical layer epithelium cells were occasionally p21 positive.

Immunostaining for HPV (HPV screening antibody, Virofem Diagnostica, Germany) was positive in patient 1.

Comment

The high recurrence rate of OSSN of 9–64% after resection seems to depend on the histopathological grade and status of surgical margins.5 HPV-16 and HPV-18 are considered
to be possible cofactors involved in initiation and early progression of OSSN. Therefore both of the presented tumours were clinically papillomatous, immunostaining for HPV was positive only in patient 1. The tumour suppressor gene p53 has been found to be inactivated in over 50% of human cancers. In OSSN, overexpression of p53 has been previously reported in some SCC of conjunctiva. In the SCC of our patients, p53 overexpression indicating inactivating p53 mutations were observed only in the invasive part of the tumour, but not in the carcinoma in situ. While Dushku and coworkers assumed that p53 mutations could be an early event in tumour development consistent with ultraviolet radiation, our findings clearly indicate that mutations of p53 are a late event that occurs with disease progression, as observed with other solid tumours.

Our results of two exenterated advanced stages of SCC emphasise the necessity to remove dysplastic OSSN completely to prevent progression to invasive carcinomas. Identification of inactivating p53 mutations may indicate an increased risk for invasiveness. Therefore immunohistochemical analysis of biopsy specimen may help in the management of these tumours.

Familial pseudotumoral sclerochoroidal calcification associated with chondrocalcinosis

Sclerochoroidal calcification is the deposition of calcium at the level of the sclera and choroid. Two entities have been described: metastatic calcifications resulting from deposition of calcium in normal tissues caused by phosphocalcic metabolism abnormality such as primary and secondary hyperparathyroidism, pseudohypoparathyroidism, hypervitaminosis D, vitamin D intoxication, hypophosphataemia, sarcoidosis, Barter syndrome, and Gietelman syndrome; and dystrophic calcifications caused by secondary deposition of calcium in abnormal tissues despite normal serum levels of calcium and phosphate.

Sclerochoroidal calcifications can also be idiopathic. We describe the first case of hereditary form of sclerochoroidal calcifications associated with familial articular chondrocalcinosis.

Case report

A 69 year old man was admitted to the department of ophthalmology in November 1999 with gradual deterioration of vision in both eyes. He had a medical history of familial articular chondrocalcinosis. His father, brother, and son were treated for the same disease. On examination, best corrected visual acuity was 20/120 in the right eye and finger counting in the left eye. Slit lamp examination and ocular tension were normal. The funduscopy revealed multiple bilateral pseudotumoral white choroidal masses in both eyes (fig 1). Ultrasound examination of the eyes confirmed the calcific nature of the lesions.

On fluorescein angiography in March 1979 the pseudotumoral lesions were smaller and did not involve the macular area in left eye (fig 1, left). He had an extensive metabolic evaluation with familial chondrocalcinosis. His father, brother, and son were treated for the same disease.

References


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The brother’s funduscopy revealed multiple bilateral, extrafoveal pseudotumoral white choroidal masses (fig 2, top). The son’s funduscopy revealed plaque-like and only slightly elevated lesions seen in the midperiphery (fig 2, bottom). Ultrasonograms confirmed the calcific nature of these lesions.

Comment
In 1997, Shields et al described a case of sclerochoroidal calcifications in a normocalcaemic patient who had chondrocalcinosis. We first describe a familial case of sclerochoroidal calcifications associated with calcium pyrophosphate dihydrate (CPPD). In this family, autosomal dominant inheritance is highly likely because there are affected individuals in each generation, there is male to male transmission, and every affected member has an affected parent.

Inheritance in sclerochoroidal calcifications has never been described; however, hereditary forms of chondrocalcinosis have already been described.

In our report, a patient had a 24 year follow up showing a progressive involvement of the macular area, suggesting a growth of the calcifications. Two types of calcifications have been described previously, the plaque-like and the pseudotumoral type.

To our knowledge, it has never been determined if the plaque-like lesions evolve into tumour-like lesions. In 1992, Schachat and associates reported 10 cases with follow up ranging from 7 months to 10 years, for whom no change in the lesion was seen. This is the first observation with 24 years of follow up suggesting a possible evolution of plaque-like lesions to pseudotumoral lesions.

We suggest that every patient affected by familial chondrocalcinosis should have an ophthalmic examination to detect sclerochoroidal calcification. These lesions seem to be evolving in time with possible involvement of the macula. Choroidal neovascularisation is also a vision threatening complication of sclerochoroidal calcifications. Our case suggests the need to perform ophthalmological examination in patients and family members of patients affected by chondrocalcinosis.

Whole body PET/CT imaging for detection of metastatic choroidal melanoma
Metastatic choroidal melanoma typically presents in the liver. Therefore, liver enzyme assays are the most common haematological evaluation performed after treatment.

In 1985, The Collaborative Ocular Melanoma Study required periodic medical evaluations including a physical examination, liver functions studies, a complete blood count, and a chest x ray. If liver enzymes exceeded 1.5 times normal, computed tomography (CT) of the abdomen was required. If low attenuation hepatic nodules suggested metastatic disease, fine needle aspiration biopsy of the liver tumours provided cytopathological confirmation.

Positron emission tomography (PET) is a molecular imaging technique that uses radiolabelled molecules to image metabolic activity in vivo. When whole body PET was combined with computed radiographic tomography (CT), PET/CT put anatomy and function on the same page making practical a new era of physiological imaging.

This study examines the ability of positron emission tomography combined with computed tomography (PET/CT) to allow for detection of previously occult metastatic melanoma.

Case report
A 77 year old woman presented with a 15.4 x 15 mm width and 13.2 mm high collar button shaped choroidal melanoma with a large secondary retinal detachment in her right eye. Her preoperative medical evaluation

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References

proved negative. She was treated by enucleation.

Two years later a follow up medical evaluation revealed elevated liver function studies (table 1) and a chest x ray showed a pleural effusion. CT of the abdomen with contrast revealed multiple low attenuation hepatic foci consistent with metastatic melanoma.

A PET/CT was requested. Fifty minutes after intravenous administration of 15.2 mCi of fluordeoxyglucose, whole body PET/CT imaging revealed enlarged para-aortic lymph nodes and a subcutaneous nodule in the abdominal wall (fig 1). The CT portion of the PET/CT also revealed two 3 mm nodules in the right upper lobe. PET imaging was able to reveal multiple bony metastasis that were not seen on the CT portion (fig 1). Both CT and PET showed a large liver metastasis. Since it is a physiological assay, PET also demonstrated the metabolic activity of the metastatic tumours (fig 1).

Comment
In this case, whole body PET/CT was found to be capable of uncovering metastases not seen with abdominal CT alone. This led us away from considering regional perfusion of the liver, hepatic resection, and towards systemic treatment.

Therefore, when PET/CT identifies extrahepatic involvement, it can have a significant impact on the management of patients with metastatic choroidal melanoma.

PET/CT could also be used for initial staging. Early detection of occult metastases

Table 1  Patient characteristics

<table>
<thead>
<tr>
<th>Physical</th>
<th>Examination</th>
<th>Blood examination</th>
<th>X ray CT scan abdomen with contrast</th>
<th>Whole body PET/CT</th>
</tr>
</thead>
<tbody>
<tr>
<td>General</td>
<td>Icterus</td>
<td>No lesions noted</td>
<td>1.4 cm subcutaneous nodule in the anterior abdominal wall</td>
<td>Hypermetabolic focus in the subcutaneous tissue of the anterior abdominal wall</td>
</tr>
<tr>
<td>Subcutaneous tissue</td>
<td>2 nodules in the anterior abdominal wall</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Nodes</td>
<td>None</td>
<td>No lesions noted</td>
<td>Enlarged para-aortic lymph nodes on the left side</td>
<td>Hypermetabolic focus in the upper abdomen</td>
</tr>
<tr>
<td>Lungs</td>
<td>No abnormalities noted</td>
<td>Pleural effusion right lung base and calcified hilar nodes</td>
<td>Two small 3 mm nodules in the right upper lobe. Right pleural effusion</td>
<td>No foci noted</td>
</tr>
<tr>
<td>Liver</td>
<td>Enlarged</td>
<td>High bilirubin, AST, ALT, alkaline phosphatase, GGT</td>
<td>Low attenuation lesion &gt;20 cm in greatest diameter in the right lobe with calcification seen posteriorly. Numerous low attenuation lesions throughout both lobes of the liver</td>
<td>Enlarged. Large hypermetabolic focus in the right lobe with mass effect. Numerous hypermetabolic foci throughout remainder of liver</td>
</tr>
<tr>
<td>Kidney</td>
<td>8 mm cyst midpole and 2 cm cyst upper pole of left kidney</td>
<td>Large right renal cyst. Additional smaller cysts</td>
<td>Photopenic defect due to large right renal cyst</td>
<td></td>
</tr>
<tr>
<td>Bone</td>
<td>No lesions noted</td>
<td>No lesions noted</td>
<td>Hypermetabolic foci in the right skull base, left scapula, left humerus, sternum, multiple bilateral ribs, thoracic and lumbar spine, pelvis, and bilateral femurs</td>
<td></td>
</tr>
</tbody>
</table>

Figure 1  On the left, the CT demonstrates the anatomy; on the right the PET shows areas of hypermetabolism (glucose uptake); in the middle the two images are fused. PET/CT revealed enlarged para-aortic lymph nodes and a subcutaneous nodule in the anterior abdominal wall. The PET imaging portion of the PET/CT was able to reveal multiple bony metastases that were not seen on the CT portion of the examination. Both CT and PET showed a large liver metastasis.
offers the potential to avoid ineffective and expensive enucleations, radioactive plaques, proton irradiation, eye wall resections, or laser treatment. Local therapies would be abandoned in favour of systemic treatments.

Another issue related to PET/CT is cost. Up to five times more than CT of the abdomen, PET/CT is only covered (Medicare) for melanoma staging/restaging when the stage of the cancer remains in doubt after completion of conventional imaging (or if the clinical management would differ depending on the PET findings). Since PET/CT revealed extrahepatic loci in this case, it changed our clinical approach. There is little doubt about the improved ability of PET/CT to detect lesions; the real issue is cost and if the results will change outcomes.

This study goes one step further, CT, MRI, or PET alone. By combining whole body PET and CT, this examination joins anatomy and function in one examination (fig 1). The relative efficacy of PET/CT to locate metastases should be evaluated within the framework of a prospective study.

Trans-Tenon’s retrobulbar triamcinolone infusion for small choroidal neovascularisation

Intravitreal and sub-Tenon’s corticosteroids are being evaluated for the treatment of choroidal neovascularisation (CNV). 6-8 We reported on the efficacy of triamcinolone acetonide administered as a trans-Tenon’s retrobulbar infusion (triamcinolone infusion) in reducing inflammation in uveitic eyes. 6 Here we evaluated the same treatment in eyes with small subfoveal CNV.

Case reports

Triaemcinolone infusion was performed in 22 eyes of 22 patients with subfoveal CNV of greatest diameter less than or equal to 1 disc diameter (DD). The diagnoses were age related macular degeneration (AMD) in 14 eyes, idiopathic CNV in four eyes, polypoidal choroidal vasculopathy (PCV) in three eyes, and punctate inner choroidopathy (PIC) in one eye. One AMD eye had undergone ablative argon laser photococoagulation for CNV previously, but no other eyes had received previous treatment. The median post-triamcinolone infusion follow up was 7.5 months (range 4–27 months). Pretreatment fluorescein angiography (FA) revealed predominantly classic CNV in 12 eyes and predominantly occult CNV in 10 eyes. Records were reviewed retrospectively and did not require institutional review board approval. Informed consent was obtained before each procedure.

The patient’s eye was prepared with povidone-iodine and sterile drapes applied. After topical anaesthesia, conjunctiva and Tenon’s capsule were incised in the inferotemporal quadrant. A 23 gauge curved blunt cannula approximately 2.1 cm in length (#HS-2764, Handaya Co, Ltd, Tokyo, Japan) was inserted to the hub into sub-Tenon’s space and 20 mg (0.5 ml) triamcinolone acetonide (Bristol Pharmaceutical, KK, Tokyo, Japan) was infused. The wound was left unsutured and 0.5% levofloxacin was instilled topically three times a day for 1 week.

Onset of CNV fibrosis was observed in 14 eyes (64%) by 3 months post-triamcinolone infusion (fig 1). Rates of fibrosis were 50% (7/14 eyes) for AMD, 100% (4/4 eyes) for idiopathic CNV, 67% (2/3 eyes) for PCV, and 100% (1/1 eye) for PIC. Fibrosis did not correlate with CNV size or lesion composition. FA performed at 3 months showed decreased lesion leakage in 12 eyes (53%), no change in five eyes (23%), and increased leakage in five eyes (23%). Best corrected visual acuity (VA) at 3 months for all eyes improved by ≥0.2 logarithm of the minimum angle of resolution (logMAR) in four eyes (18%), no change in five eyes (23%), and increased leakage in five eyes (23%).

The median VA for all eyes was 0.30 before treatment (range 0.08–1.0) and 0.24 at 3 months after treatment (range 0.05–1.2). Of the 14 eyes with AMD, the VA at 3 months improved by ≥0.2 logMAR in one eye (7%), remained

![Figure 1](http://www.bjophthalmol.com)

**Figure 1** Colour fundus photographs and fluorescein angiography late images of an eye in a 65 year old patient with age related macular degeneration and small subfoveal choroidal neovascularisation before (A, B; best corrected visual acuity 0.3) and 3 months after triamcinolone infusion (C, D; best corrected visual acuity 0.7).
unchanged in 10 eyes (71%), and worsened by >0.2 logMAR in three eyes (21%). In these AMD eyes, the median decimal VA was 0.30 before treatment (range 0.08–1.0) and 0.20 at 3 months after treatment (range 0.05–0.7). Complications such as intraocular pressure elevation, infection, or cataract progression were not noted in any eyes.

### Comment
This interventional case series shows that trans-Tenon's retrobulbar infusion of triamcinolone acetonide resulted in less fibrosis in the majority of eyes with small CNV. This effect of corticosteroids in inhibiting CNV growth probably involves several pathways. The effect of corticosteroids in inhibiting neovascular cells that participate in the neovascular response probably has a prominent role. Triamcinolone acetonide has specifically been shown to inhibit basic fibroblast growth factor induced migration and tube formation of choroidal microvascular endothelial cells. In a rat model, triamcinolone acetonide inhibits choroidal neovascularization in a laser-treated rat model. Arch Ophthalmol 2001;119:399–404.

### Table 1
Anomalies and cancers reported in offspring of IVF

<table>
<thead>
<tr>
<th>Condition</th>
<th>Reference</th>
</tr>
</thead>
<tbody>
<tr>
<td>Neuroectodermal</td>
<td>White et al.²</td>
</tr>
<tr>
<td>tumours</td>
<td>Kobayashi et al.³</td>
</tr>
<tr>
<td>Neuroblastoma</td>
<td>Kramer et al.²</td>
</tr>
<tr>
<td>(Note: associated</td>
<td>Michalek et al.³</td>
</tr>
<tr>
<td>with the use of</td>
<td></td>
</tr>
<tr>
<td>fertility drugs)</td>
<td></td>
</tr>
<tr>
<td>Retinoblastoma</td>
<td>Moll et al.³</td>
</tr>
<tr>
<td></td>
<td>Crusberg et al.³</td>
</tr>
<tr>
<td></td>
<td>Artero et al.³</td>
</tr>
<tr>
<td>Hepatoblastoma</td>
<td>Melamed et al.³</td>
</tr>
<tr>
<td></td>
<td>Toren et al.³</td>
</tr>
<tr>
<td>Clear cell kidney</td>
<td>Toren et al.³</td>
</tr>
<tr>
<td>sarcoma</td>
<td></td>
</tr>
<tr>
<td>Lymphoma</td>
<td>Kobayashi et al.³</td>
</tr>
<tr>
<td>Transposition of</td>
<td></td>
</tr>
<tr>
<td>the great arteries</td>
<td>Lancaster³</td>
</tr>
<tr>
<td></td>
<td></td>
</tr>
<tr>
<td>Neural tube defects</td>
<td>Berg et al.³</td>
</tr>
</tbody>
</table>

Several types of congenital anomalies and cancers have been reported in the literature, primarily in the form of case reports. The relation between IVF and these conditions is not definitively established.

### Table 2
Retinoblastoma in children born through IVF

<table>
<thead>
<tr>
<th>Reference</th>
<th>Sex and age at diagnosis</th>
<th>Eye</th>
<th>Causes of subfertility</th>
<th>DNA 13q14 analysis</th>
<th>Assisted reproductive technique</th>
</tr>
</thead>
<tbody>
<tr>
<td>Artero et al.³</td>
<td>NR, 30</td>
<td>Unilateral</td>
<td>NR</td>
<td>NR</td>
<td>IVF with donor sperm</td>
</tr>
<tr>
<td>Mall et al.³</td>
<td>F, 38</td>
<td>Bath</td>
<td>Unexplained</td>
<td>Exon 8</td>
<td>IVF with 6 courses of clomid</td>
</tr>
<tr>
<td>Mall et al.³</td>
<td>M, 15</td>
<td>Left</td>
<td>Maternal cause</td>
<td>Normal</td>
<td>IVF</td>
</tr>
<tr>
<td>Mall et al.³</td>
<td>F, 24</td>
<td>Right</td>
<td>Unexplained</td>
<td>Intron 3</td>
<td>IVF with 8 AI attempts</td>
</tr>
<tr>
<td>Mall et al.³</td>
<td>M, 8.5</td>
<td>Bath</td>
<td>Unexplained</td>
<td>mutated</td>
<td>IVF with 8 AI attempts</td>
</tr>
<tr>
<td>This report</td>
<td>F, 32</td>
<td>Left</td>
<td>Paternal cause</td>
<td>Normal</td>
<td>IVF with ICSI</td>
</tr>
</tbody>
</table>

NR, not reported; AI, artificial insemination; ICSI, intracytoplasmic sperm injection.

### References
PostScript

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References


Angle closure glaucoma after laser photoagulation for retinopathy of prematurity

Infantile angle closure glaucoma (AGC) is a rare consequence of retinopathy of prematurity (ROP) and usually occurs a few years after laser treatment for ROP. A Medline search for AGC following laser photoagulation extracted only one case. In the case, AGC occurred in 2 weeks after laser photoagulation and although occurrence of iris bombe in both eyes was described, the mechanism for the AGC was not fully clarified.4

We present a case of bilateral AGC that occurred within a several weeks after the laser photoagulation for ROP. We shall discuss the importance of ultrasound biomicroscopy (UBM) in the diagnosis.

Case report

A baby girl, born at 25 weeks gestation weighing 796 g, was diagnosed with stage 2 plus, zone 2 ROP bilaterally at 33 weeks. Diode laser photoagulation, 986 applications right eye and 629 left eye, with 200–240 mW, 0.4 second duration, was performed by a paediatric ophthalmologist. On the following day, severe hyphaema was observed bilaterally but there was no evidence of choroidal detachment by B-mode ultrasound sonography. Topical atropine and corticosteroid were started and she was...
endothelium. As a result, the anterior capsule
strated iris bombe bilaterally, and the entire
branes and iridohyaloid vessels were
17 mm Hg left eye. Persistent pupillary mem-
chamber became shallow.
formed in the both eyes and the anterior
photocoagulation induced posterior synechia,
the lens was displaced into the anterior
the left eye as measured with a caliper installed

dip, peripheral iridectomy was performed bilat-
erally (fig 1). Postoperatively, her peripheral
anterior chamber deepened bilaterally although
the lens in the right eye was still
endothelium. Indirect
ophthalmoscopy revealed normal cup to disc
ratio. The IOP fell to normal levels bilaterally.

Figure 2 Composite images of the anterior
segment scanned with UBM (top) and schematic
representations (bottom). (A) Left eye. Posterior
synechia and iris bombe can be seen. C, cornea; L, lens; AC, anterior chamber; S, posterior synechia; PC, posterior chamber. (B) Right eye. The high echo lines (arrowheads) along the corneal endothelium represent the iris adherent to the corneal endothelium and the iris bombe. The lens is displaced into the anterior chamber and the anterior chamber is completely obliterated. The arrow represents the posterior capsule of the lens. Note that the cornea is distended and is thin as a result of the high intracocular pressure. The thickness of the cornea at the centre is 394 μm, and 613 μm in the left eye as measured with a caliper installed in the UBM model 840 Humphrey.

followed up conservatively. During the follow
up period, total posterior synechia was
formed in both eyes and the anterior
chamber became shallow.
At 39 weeks, the corneal diameter had
increased, and the anterior chamber was
extremely shallow bilaterally. The intraocular
pressure (IOP) in the right eye was elevated
to 28 mm Hg, and she was referred to our
hospital.

Our examination showed that the corneal
diameter was increased to 11 mm bilaterally.
Slit lamp examination showed corneal
oedema and shallow anterior chamber depth
bilaterally, especially in the right eye. The
corneal oedema made the funduscopy
difficult bilaterally. The IOP under the general
anaesthesia was 33 mm Hg right eye and
17 mm Hg left eye. Persistent pupillary mem-
branes and iridohyaloid vessels were
observed but ruberosis iridis was not observed
(fig 1).

UBM images of anterior segments dem-
strated iris bombe bilaterally, and the entire
right iris surface was adherent to the corneal
endothelium. As a result, the anterior capsule of the lens was also attached to the corneal endothelium (fig 2). Choroidal detachment and a retrolental mass were not observed by B-mode ultrasound sonography (fig 2).

Peripheral iridectomy was performed bilat-
erally (fig 1). Postoperatively, her peripheral
anterior chamber deepened bilaterally although
the lens in the right eye was still
endothelium. Indirect
ophthalmoscopy revealed normal cup to disc
ratio. The IOP fell to normal levels bilaterally.

Comment
Shallow anterior chambers in ROP patients are
known to be caused by various factors—
for example, choroidal detachment after
excessive photocoagulation, development of
retrolental mass, or relative increase in lens
thickness, but usually the cause of shallow
anterior chamber cannot be determined. In
our case, the development of hyphaema after
photocoagulation induced posterior synechia,
and the iris bombe followed. The displace-
ment of the anterior chamber structures was
induced by the forward movement of the iris-
brain diaphragm in the right eye, and the
ocular fragility in premature baby may
explain this deformity.

Vitreous haemorrhage is known to occur in
7.9% of ROP cases after photocoagulation. In
our case, there is a possibility that the
hyphaema was derived from vitreous haem-
morrhage. Another possibility is an acciden-
tal photocoagulation of persistent pupillary
membranes and/or iridocorneal vessels
caused the hyphaema. We are not aware of
such morphological changes after photo-
coa
gulation for ROP.

ACG that occurs immediately after retinal
photocoagulation in ROP patients is rare, but
is still an important complication. In ROP
patients, the lens and its ligament are weak,
and therefore not only ACG but also lens
displacement occurred. It is important that
we be aware of the possible development of
ACG following retinal photocoagulation for
ROP.

Sequential treatment of central retinal vein occlusion with intravitreal tissue plasminogen activator and intravitreal triamcinolone

Treatment for central retinal vein occlusion (CRVO) remains disappointing despite recently proposed intravitreal surgical tech-
niques. We previously introduced the use of intravitreal tissue plasminogen activator (TPA) for acute central retinal vein occlusion in 1999. Numerous investigators have con-
firmed its safety and suggested that it may
have a beneficial role in the treatment of
acute central retinal vein occlusion.

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Figure 1 (A) Fundus photograph shows an
acute CRVO with dilated retinal veins,retinal
haemorrhages, and retinal oedema. (B) Fundus appearance 1 month after intravitreal injection
of tissue plasminogen activator. (C) Photograph
of the same eye 5 months later.
investigators found the porcine retina was, in fact, permeable to TPA. Our clinical experience with intravitreal TPA in humans with CRVO and large submacular haemorrhages strongly suggests that intravitreal TPA does cross the human retina. Greenberg et al were the first to report the possible beneficial effect of intravitreal steroids on patients with chronic CRVO.[8] This report describes a sequential treatment strategy for patients with CRVO who present early in the course of their disease and can be performed in the office while avoiding the risks of vitrectomy. It utilizes intravitreal TPA in the acute phase of the vein occlusion to attempt clot lysis, and then treats any remaining vascular leakage with intravitreal triamcinolone.

Case report
A 59 year old obese, hypertensive flight instructor presented with a sudden decrease in vision for 7 days in the right eye. Vision was 20/400 right eye and 20/20 left eye. The patient was diagnosed with an acute CRVO in the right eye (fig 1A). The left eye was normal. After being advised of the risks and benefits, the patient elected to undergo intravitreal injection of TPA (75 μg). Thirteen days later, the patient noted marked improvement in vision with 20/200 vision. Thirty four days after the injection, the patient’s vision was 20/60 (fig 1B).

Six months after intravitreal TPA injection, the vision remained 20/30, but the patient still complained of metamorphopsia and blurry vision despite resolution of other findings of CRVO (fig 1C). Fluorescein angiogram (FA) revealed persistent macular oedema (fig 2A). Optical coherence tomography (OCT) showed the foveal thickness to be 331 μm with mild intraretinal oedema. After being advised of the risks and the benefits, the patient then underwent injection of intravitreal triamcinolone (4 mg).

Six weeks after intravitreal triamcinolone, the FA returned to normal and OCT showed decreased foveal thickness from 331 μm to 291 μm. The patient reported a significant improvement in vision with decreased metamorphopsia. Vision was 20/25 with no late leakage on the fluorescein angiogram (fig 2B).

Comment
To our knowledge, this represents the first published case of CRVO treated sequentially with intravitreal TPA for the acute phase and intravitreal triamcinolone for the chronic phase. TPA is a drug that must be used early in the course of thrombus formation to be effective. We do not recommend its use for patients with chronic symptoms. Intravitreal steroids appear to decrease the blood-retinal barrier breakdown and macular oedema, but recurrent oedema may occur since the steroids do not appear to affect the thrombus itself.

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Severe post-laser suprachoroidal haemorrhaging in a diabetic patient receiving anticoagulants
Although the aetiology is not well understood, expulsive suprachoroidal haemorrhaging (ESH) is the most severe complication associated with intraocular surgery. Anticoagulants are considered a risk factor for spontaneous suprachoroidal haemorrhaging in cases with high myopia, age related macular degeneration, and diabetic retinopathy.[9] However, ESH post photoacoagulation is extremely rare regardless of anticoagulant therapy. We have experienced a severe case of post-laser ESH correlated with anticoagulant therapy, which resulted in irreversible visual disturbance.

Case report
A 70 year old woman was diagnosed with pre-proliferative diabetic retinopathy based on fluorescein angiographic examinations. Two months before diagnosis, she underwent right eye cataract surgery. During the past 6 years, the patient received warfarin (4 mg/day) and aspirin (81 mg/day) because of atrial fibrillation after myocardial infarction. Laser photoacoagulation was performed in her right eye with a Nidek MC-7000, yellow-green laser. Operating conditions were 200–280 burns per session with a spot size 200 μm, exposure 0.2 seconds, power 100–120 mW using a Quadrальной contact lens (Volk, Tokyo, Japan). Treatment was separated into three partitions with a minimum 2 week interval between sessions. Three days after final photoacoagulation, the patient had a sudden visual loss to hand movements in slit lamp examinations, the retina seemed to be detached from the posterior surface of the implanted intraocular lens. Severe choroidal detachment was found by fundus examination (fig 1). The B-mode ultrasonography showed massive haemorrhaging in the choroidal space (fig 2). In systemic examinations, multiple purple spots were observed in both her arms. Microhaematuria was also noted. Blood examination revealed blood sugar 167 mg/dl; platelet number 179 000 ×10³/μl; PT% 19% (control 70–120); PTs 28.7 seconds; PT INR 4.72 (control 1); PT on June 5, 2004.

Severe post-laser suprachoroidal haemorrhaging in a diabetic patient receiving anticoagulants

Comment
We have described a case of ESH after laser photoacoagulation in a patient receiving anticoagulant therapy. Laser photoacoagulation is known as an effective treatment for various oculic disorders and is a widely used, non-incision surgical procedure. However, a number of complications have been reported, with some citing an irreversible visual disturbance. On the other hand, anticoagulant therapy is prevalent after cardiac/brain infarctions, which necessitates a long photoacoagulation system management. In the present case, the PT INR was extremely prolonged (a respected value of 2–3 is

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appropriate for post-cardiac infarction). Presumably, choroidal microbleeding initiated by photoocoagulation persisted because of an overly suppressed coagulation system; blood pooled in the choroidal space, which assumed an ESH. To our knowledge there is only one other similar case reported by Khairallah et al that showed post-laser choroidal haematoma in a diabetic patient treated with anticoagulant. Even though ESH incidence is low, extreme caution must be exercised when performing laser therapy in patients using anticoagulants, because of potentially serious outcomes. An age of 65 years or more, history of stroke, history of gastrointestinal bleeding, a serious morbid condition (recent myocardial infarction, renal insufficiency, or severe anaemia), and atrial fibrillation are five high risk factors for major bleeding in outpatients treated with warfarin. If possible, preoperative coagulation system examinations are recommended for high risk patients receiving anticoagulant treatments.

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regimens with reported efficacy including a 6 month regimen of rifampicin (with streptomycin also given in the initial phase only) for tuberculous uveitis. Furthermore, the WHO recommendations are for active extraocular tuberculosis that has been diagnosed by specimen examination or strong clinical evidence, and give no recommendations for latent infection. As we have previously reported in a series on intraocular tuberculosis, systemic examination failed to identify a focus of active tuberculosis in the majority of our patients, and we have come to suspect that the uveitis we observed may be an immune response to latent tuberculosis antigen sequestered elsewhere. Therefore, the patients we described were given a diagnosis of presumed intraocular tuberculosis, that is with uveitis presumed to be related to the Mycobacterium tuberculosis organism. Furthermore, we would like to clarify that in the cases of presumed ocular tuberculosis that received trans-Tenon's retrobulbar triamcinolone infusion, this treatment was judged to be effective in two of three eyes. Regardless, since the focus of active or latent tuberculosis was never identified in our patients, a two drug regimen of isoniazid and rifampicin was used as a therapeutic trial for antituberculosis therapy. A similar therapeutic trial for ocular tuberculosis, albeit with isoniazid alone, has been previously advocated in Japan by Ishihara and Ohno. With regard to the second comment, among the 16 patients who were receiving some form of systemic immunosuppressive therapy, we did not notice any difference in outcome when compared to patients who were not on immunosuppressive therapy. In other words, the efficacy of trans-Tenon's retrobulbar triamcinolone infusion was the same. However, we suspect that the recurrence rate after triamcinolone infusion may be different, and we are currently investigating this possibility.

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Owls' eyes move

"Double crossed," the cover illustration and article by Schwab on the barn owl refers to the alleged immobility of the owl's eyes. This is a myth which should not be perpetuated in the BJO. The owl's eyes do in fact move, and while the amount is not large, it is just enough for two papers on the subject. The phrase "nearly immobile" is preferable.

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NOTICES

Low vision care

The latest issue of Community Eye Health (No 49) deals with the problems and management of low vision. For further information please contact: Journal of Community Eye Health, International Resource Centre, International Centre for Eye Health, Department of Infectious and Tropical Diseases, London School of Hygiene and Tropical Medicine, Keppel Street, London WC1E 7HT, UK; tel: +44 (0)20 7612 7964; email: Anita.Shah@lshtm.ac.uk; online edition: www.jceh.co.uk. Annual subscription (4 issues) UK £28/US$45. Free to developing country applicants.

Elimination of avoidable blindness

The 56th World Health Assembly (WHA) considered the report on the elimination of avoidable blindness (doc A56/26) and urged Member States to: (1) Commit themselves to supporting the Global Initiative for the Elimination of Avoidable Blindness by setting up a national Vision 2020 plan by 2005; (2) Establish a national coordinating committee for Vision 2020, or a national blindness prevention committee to help implement the plan; (3) Implement the plan by 2007; (4) Include effective monitoring and evaluation of the plan with the aim of showing a reduction in the magnitude of avoidable blindness by 2010; (5) To support the mobilisation of resources for eliminating avoidable blindness. The WHO also urged the Director-General to maintain and strengthen WHO's collaboration with Member States and the partners of the Global Initiative for the Elimination of Avoidable Blindness as well as aid in the coordination and support of national capability.

4th International Congress on Autoimmunity

The 4th International Congress on Autoimmunity will take place 3–7 November 2004 in Budapest, Hungary. The deadline for the receipt of abstracts is 20 June 2004. Further details: Krones International Global Congress Organisers and Association Management Services, 17 Rue du Cendrier, PO Box 1726, CH-1211 Geneva 1, Switzerland (tel: +41 22 908 0488; fax: +41 22 732 2850; email: autoim04@krones.com; website: www.krones.com/autoim04).