USHER'S SYNDROME
AN ELECTROPHYSIOLOGICAL REVIEW
E. Burke, P. Rowland, P. Eustace.

In 1858 Von Graefe described a condition characterized by retinitis pigmentosa and profound deafness and in 1914 Usher clearly delineated the familial nature of this condition. Usher's syndrome is characterised by a progressive visual loss and profound deafness leaving the patient dependent on the sensory modality of touch.

It is inherited in an autosomal recessive fashion. There are two genetic types representing different clinical severity.
1. Retinitis pigmentosa and congenital deafness and vestibular disturbance.
2. R.P. and slowly progressive hearing impairment and normal vestibular function.

The incidence is 3/100,000 of general population and 3-4% of congenitally deaf population.

The sensory impairments associated with Usher's syndrome are believed to be caused by an inborn error in some metabolic process involving the pigment cells in the eye and ear which have a common origin during the embryonic stage of development.

In our review we screened children from a local deaf school where there are 670 pupils. In this group 30% have deafness associated with Rubella, 30% have a positive family history and 40% have no known aetiology. This last group is the group we are interested in screening for Usher's syndrome. We have also been alerted to the existence of misdiagnosed psychiatric patients who may be cases of Usher's syndrome.

We have screened 15 patients to date in this category and 8 of these have the characteristic features of Usher's syndrome.

In view of these findings we will screen those congenitally deaf children in the high risk group. These children should have an electroretinogram at five years old when even if asymptomatic they may well show changes in the electroretinogram, and appropriate educational measures can then be commenced.

THE MANAGEMENT OF AMAUROSIS FUGAX

This study on Amaurosis Fugax involved twenty patients over a two year period. We defined Amaurosis Fugax as "an episode of uniconal visual loss of less than four hours duration, which left no residual impairment of either visual field or visual acuity" (Wilson and Russell, 1977).

Analysis of the figures showed that 40% of attacks lasted less than ten minutes, while 80% lasted less than two hours. Males outnumbered females in a ratio of two to one. The age group was 45-80 years, with a mean of 65 years. Twenty per cent had hypercholesterolaemia, and ten per cent had hypertension, diabetes and angina respectively. Retinal emboli were recorded in two patients only.

Seventy per cent of patients gave a history of smoking. Amaurosis Fugax in the non-smoker was less common than in smokers and occurred in an older age group.

Twenty per cent of patients had an ipsilateral carotid bruit. Any patient with a bruit, subsequently was shown to have a stenosis greater than 50% on Doppler ultrasound scanning. Ultrasonography of the extracranial carotid system revealed that 70% showed plaque in the internal carotid artery, and 25% had a stenosis greater than 50%.

Sixty per cent of patients were treated with a combination of persantin and aspirin and 25% subsequently went on to have endarterectomy.

We concluded that Amaurosis Fugax is a significant early symptom of carotid disease, a condition which is treatable satisfactorily.

GHOST CELL GLAUCOMA

Ghost cell glaucoma is a form of secondary open angle glaucoma which results from trabecular meshwork obstruction caused by degenerate red blood cells and cellular debris in the anterior chamber.

Following vitreous haemorrhage, red cells degenerate into small, spherical, tan-coloured red cell ghost cells from which most intracellular haemoglobin has been lost, and whose cell membranes have become relatively rigid and cannot deform to the same extent as can fresh red cells.

Ghost cell glaucoma can develop where a defect in the anterior hyaloid face permits passage of the cells to the anterior chamber, where their relative rigidity makes passage through the trabecular meshwork more difficult, and causes aqueous outflow obstruction.

Three cases were presented with vitreous haemorrhages resulting from retinal neovascularization secondary to branch vein occlusion (2 cases) and proliferative diabetic retinopathy (1 case). Onset of ghost cell glaucoma occurred between 18 months and 4 years following initial vitreous haemorrhage. Intracocular pressures were controlled by medical treatment in one case and by trabeculectomy in 2. With adequate control of intraocular pressure, and in the absence of significant macular pathology the prognosis for vision is good, as the natural course of the condition is to resolve spontaneously when the ghost cell reservoir in the vitreous is exhausted, usually leaving an undamaged trabecular meshwork.

Despite experimental evidence that this condition cannot occur in the presence of an intact anterior hyoid face (haemolytic ghost cell glaucoma. Campbell, D. G. and Essigmann, E. M., Arch. Ophthalmol. 97, 2141-2146, 1979) none of these 3 patients had any history of, or clinical findings to suggest previous surgery or significant ocular trauma. Spontaneous dissolution of the anterior hyaloid face may therefore not be so uncommon as previously was thought.

JUXTAPAPILLARY SUBRETINAL NEOVASCULARIZATION
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Fourteen patients with juxtapapillary subretinal neovascular membranes were seen in the Eye and Ear Hospital between 1972 and 1982. The ages of the patients ranged from 13 to 75 years and were subdivided into two distinct groups: a
ACUTE ANGLE-CLOSURE GLAUCOMA AND CENTRAL RETINAL VEIN OCCLUSION

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The association of non rubeotic angle-closure glaucoma and central retinal vein occlusion is a very rare clinical situation.

In such cases, angle-closure glaucoma has usually been diagnosed within two to three weeks. The shallowing is reversible, but subsequent neovascular glaucoma may develop.

We present an elderly man with one month’s history of progressive, painless loss of vision followed by an episode of acute angle-closure glaucoma in that eye. Fundus examination revealed evidence of recent venous occlusion. A sustained reduction in intraocular pressure was achieved with Pilocarpine and Diamox. The angle remained 75% closed.

Two months after this initial presentation, neovascular glaucoma supervened.

This rare entity should not be confused with primary closed angle glaucoma. Unlike the latter, the treatment of choice is medical. Tridectomy is ineffective. There are two possible mechanisms involved. The first is antero-lateral displacement of a congested ciliary body. This results in relaxation of the zonule, which allows the lens to move forwards, causing a pupil block. The second possible mechanism is transudation of fluid from the retina. The pressure exerted by this additional fluid in the vitreous displaces the iris and lens anteriorly.

In both mechanisms as the excess fluid is absorbed the angle-closure process is reversed.

BILATERAL CENTRAL RETINAL VEIN OCCLUSION

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Bilateral central retinal vein occlusion is unusual, and is almost unknown in young patients.

We present a 30 year old female, who suffered from bilateral painless loss of vision. Visual acuity was reduced to counting fingers in both eyes. Funduscopy revealed bilateral central retinal vein occlusion.

She had a past history of hypertension, hypercholesterolemia and allergic asthma. Because of the history of atopy, a fluorescein angiogram was not performed, instead serial photographs were used as a means of follow-up.

Three months after the vascular accident, rubeotic glaucoma developed in the right eye. Examination of the left eye remains unchanged and there is no evidence of ischaemia.

Central retinal vein occlusion is a disease primarily of the elderly and this patient is the youngest we have seen reported in the literature. There are several well known risk factors, hypertension, hyperlipidaemia, blood dyscrasia, diabetes and primary open angle glaucoma.

Argon laser photocoagulation reduces the incidence of neovascularization in patients with an ischaemic type of central retinal vein occlusion, while patients with a hyperpermeable type of central retinal vein occlusion do not appear to benefit.

IRIS BURN

M. Hickey-Dwyer and D. Mooney.

A 4 year old child presented with an abnormal pupil. Two months previously she had been using a standard instamatic camera. As she did not know how to correctly use the camera she directed the lens towards her face and positioned the flashbulb in front of her eye. Consequently on firing the flashbulb the skin surrounding the eye was superficially burned, the eyelashes and eyebrow singed and the iris was burned in an area extending from 2 o’clock to 10 o’clock. No conjunctival, corneal or lens damage resulted and fundoscopy revealed a normal retina.

Follow-up has shown no changes in the original ocular findings. The pupil is now oval shaped and non-mobile.

The external burns resulted from a physical burn. The iris burn occurred secondary to transmission of light through the cornea, with resultant absorption by iris pigment and conversion to heat energy and subsequent denaturation of protein resulting in an iris burn.

We advocate that a judicious eye should be kept over flashbulbs in the hands of children.

LATE INFECTION AFTER POSTERIOR CHAMBER INTRAOCULAR LENS IMPLANT

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The case report of a 77 year old man was presented. He underwent a left extra capsular lens extraction with the insertion of posterior chamber intraocular lens. He made a satisfactory recovery.

Six weeks later he developed a severe anterior segment infective uveitis. Four weeks previously he had four teeth extracted by his dentist. We are convinced this caused a bacteraemia and a nidus of infection settled on the recent cataract operation.

Cardiologists accept any bacteraemia within an eight week period of a bacterial endocarditis as a cause of the infection.

This patient’s visual acuity recovered to 6/24 after intensive antibiotic and steroid therapy.

We propose all patients with a recently inserted intraocular lens should have prophylactic antibiotic prior to dental and other minor procedures.

A SURVEY OF INTRAOCULAR FOREIGN BODY INJURIES 1978 - 1985

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During the 8 years from 1978 to 1985, 79 patients with intraocular foreign body (IOFB) were treated at the Royal