

pression owing to cerebral oedema; the foramina typically close later, because of further bony overgrowth.³ In allied conditions such as osteopetrosis, lacrimal surgery may be successful initially, but ossification of the rhinostomy may require further surgery.⁴ It is hoped that in the present case, modification of the disease process by medical therapy and the fashioning of a large rhinostomy might provide a longer lasting palliation of the symptoms of this rare, but distressing condition.

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An unusual form of blink reflex induced by pressing on a frontal burr hole

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In a patient with Arnold-Chiari type I malformation and drained hydrocephalus communicans, we observed the unique clinical pattern of a sustained contraction in the orbicularis oculi muscle when pressing on the burr hole. It can be speculated that digital pressure on the dura mater elicited an unusual form of blink reflex. It is also conceivable that this phenomenon was related to a slight increase in intracranial pressure.

Case report

A 60-year-old woman complained of intermittent contractions of the right orbicularis oculi muscle. She presented an Arnold-Chiari type I malformation, with closed Magendie foramen. She had successively undergone a ventriculoperitoneal shunt, numerous external ventricular drainages through a right frontal burr hole, suboccipital craniectomy with C1-C2 laminectomy, and opening of the Magendie foramen. Three years later, she noted narrowing of the right palpebral fissure when pressing with her finger on the scalp overlying the frontal burr hole. She also complained of vertical diplopia and difficulty in walking.

Neuro-ophthalmic examination showed that visual acuity and visual fields were normal. The patient showed signs of left ocular tilt reaction with tilting of the head to the left, left comitant hypotropia 6 dioptres in magnitude, and conjugate leftward cyclotorsion. Corneal sensitivity and resting position of the eyelids were unremarkable. However, when pressing with the finger on the scalp overlying the bone aperture previously made by trephination, a sustained contraction of the orbicularis oculi occurred.

Only the pretarsal fibres of the lower part of the orbicularis muscle were involved. This phenomenon was observed each time, and as long as, digital pressure was applied in the trephination area. Neurological examination further showed slight ataxia and instability without cerebellar syndrome.

Comment

This patient showed a most unusual phenomenon, consisting of a unilateral contraction of orbicularis oculi muscle which occurred when pressing on a burr hole in the frontal bone. The mechanisms which produce this phenomenon are poorly understood. The possibility of a voluntary contraction of the eyelid was discarded because of the sectorial involvement of fibres in the orbicularis oculi muscle. We suggest that the pressure applied on the frontal burr hole irritated an area of the dura mater innervated by the trigeminal nerve,¹ thereby inducing an unusual, segmental form of blink reflex.

A second hypothesis, although much less probable, may be considered to explain the occurrence of the lid phenomenon reported here. It is based on a possible increase in intracranial pressure caused by pressing on the burr hole. Indeed, it cannot be excluded that the blinking was a sign of increased intracranial pressure, causing irritation of the facial nerve, even though the rise in intracranial pressure induced by digital pressure was probably moderate. Cranial nerve function is occasionally altered following elevation of intracranial pressure. The most common changes involve the optic and the abducens nerves.² In this condition, single cases

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of oculomotor and trochlear nerve involvement have also been reported.³⁻⁶ With intracranial hypertension, changes in seventh nerve function are exceptional. Peripheral facial nerve palsy was observed in five individuals suffering from benign intracranial hypertension.^{7,8} In one additional patient facial myokymia has been observed, thus showing that, with intracranial hypertension, facial nerve involvement can result in axonal hyperactivity.

No clear explanation was offered as to the pathophysiology of the facial nerve involvement in intracranial hypertension, although it was felt that this dysfunction represented a non-specific pressure related phenomenon. In our patient, it is conceivable that the displacement of intracranial nervous structures related to the Arnold-

Chiari type I malformation played a role in the mechanism of this peculiar eyelid phenomenon, by allowing the seventh cranial nerve to be affected by elevation in intracranial pressure.

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Fuchs' heterochromic uveitis associated with retinitis pigmentosa in a father and son

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Fuchs' heterochromic uveitis (FHU) in patients with retinitis pigmentosa (RP) is a rare entity. Only five cases with this combination have been reported in literature.¹ In four of these cases FHU was associated with simplex RP. In one case the retinal degeneration was demonstrated to be of autosomal recessive inheritance.¹

We report on the coexistence of autosomal dominant RP and unilateral FHU in a father and son. RP as well as FHU are relatively infrequent diseases. The prevalence of RP is 25 in 100 000, while the dominant form accounts for less than 25%.² FHU is even more exceptional, since its prevalence is estimated at 1.8 in 100 000.³

tion was performed. Throughout the years the keratic precipitates have always been present. Intraocular pressures remained normal. On the most recent examination visual acuity was 0.125 in the right eye and light perception in the left aphakic eye. A small anterior and posterior subcapsular cataract was present in the right lens. Both vitreous cavities contained cells.

The proband's healthy 33-year-old son, case III-5, was noticed to be night blind from the age of 3. When he was 14 years of age the diagnosis RP was established. Five years later keratic precipitates were noticed in the left eye, together

Case reports

Case II-3 (Fig 1) was a healthy 56-year-old man with a history of night blindness from early childhood. At 29 years of age visual acuities were 0.25 in the right eye and hand movements in the left eye. Slit-lamp examination of the left eye revealed keratic precipitates, iris atrophy with a darker coloured iris than in the right eye (inversed heterochromia), and a dense cataract. Posterior synechiae were absent. The anterior segment of the right eye was unremarkable. On funduscopy he had pale optic discs, attenuated vessels, and peripheral bone corpuscle-shaped pigmentations in both eyes. The right visual field was constricted to 30 degrees central vision. There were no symptoms of redness, pain, or photophobia. Case II-3 fulfilled all criteria of unilateral FHU and bilateral RP. After this initial examination an intracapsular lens extrac-

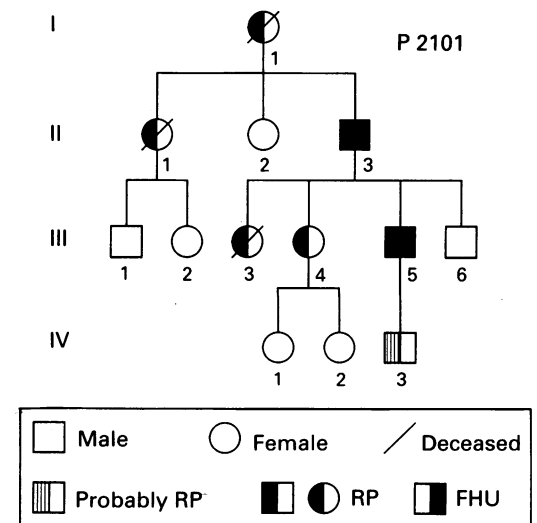


Figure 1 The family pedigree.

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