tympanotomy was performed and a fistula was found in the foot plate of the stapes. The stapes was removed and the oval window was plugged with temporal fascia. Since then (18 months) no further attacks of meningitis have occurred.

The second case was a girl, born following a normal pregnancy, and delivery, to healthy unrelated parents. Her development was normal except for deafness discovered at 12 months of age. She was fitted with a hearing aid in both ears. She has had three attacks of pneumococcal meningitis at the age of 21 months, 23 months, and 27 months, and was referred to the Hospital for Sick Children. Examination revealed a normal healthy child with a hearing aid in both ears. Throat, nose and right ear were normal; the left ear showed air bubbles and fluid in the middle ear. The tympanic membrane was dull. There was no other abnormality. Her investigations also were not diagnostic: haemoglobin and white cell count were normal. Serum immunoglobulins IgG and IgA were normal, IgM slightly raised. Nitroblue tetrazolium test was normal; cerebrospinal fluid (CSF) showed no cells and no organisms were cultured. Protein was 0.1 g/l (normal); skull radiographs and tomograms of the mastoids were normal. Metrizamide cisternogram did not demonstrate a leak into sinuses, and was normal. CSF flowed through the subarachnoid space. A technetium brain scan also showed normal CSF distribution and no abnormal accumulation was demonstrated. Hearing test confirmed severe bilateral sensorineural hearing loss. She also had an exploratory tympanotomy, and this showed a leak of perilymph from the central part of the footplate of the stapes. The stapes was removed from the oval window and the cavity was plugged with tragal perichondrium and post-auricular muscle. She has remained free from attacks of meningitis for the past 42 months.

Recurrent meningitis in the presence of other systemic infections may indicate an immunological disorder. However, when it occurs without systemic infections, an anatomical defect is likely. When confronted with a child with the triad recurrent meningitis, hearing loss (especially sensori-neural) and evidence of middle ear pathology even without abnormality in the tomographic study of temporal bones, then labyrinthine fistula should be suspected. Detailed investigations, as in both our cases, may fail to show a site of CSF leak. Therefore in such a child exploratory tympanotomy should be undertaken.

We thank Mr John Evans and Mr Robert

Pracy for allowing us to report these children.

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Errata

In the October issue of the *Journal of Neurology, Neurosurgery and Psychiatry* the letters by Quinn *et al* and by Plant were printed without their figures. We apologise to these authors and publish below the letters in full.

Insulin-induced hypoglycaemia does not abolish chorea

Sir: Pathological changes occur in the hypothalamus in Huntington's disease.¹ Insulin tolerance tests have been used to examine hypothalamic function in such patients, and mild abnormalities of growth hormone secretion have been described.²⁻⁴ In the course of such an investigation, Keogh *et al*² noted that chorea ceased some 30 min after the insulin injection and was not evident for the next 60 to 75 min in all of the twelve patients studied. They did not think that this dramatic change was due to an altered level of consciousness, for "all patients were awake throughout the investigations and were checked repeatedly to see that they were capable of verbal communication". Subsequently, Lavin *et al*³ described similar observations in another group of eight patients with Huntington's disease, in all of whom chorea disappeared for at least an hour within about half-an-

hour of the insulin injection. Such a dramatic effect on chorea might provide some clue as to the pathophysiology of that movement disorder, so we have repeated the study concentrating on the effect of insulin-induced hypoglycaemia on the chorea.

Five patients with Huntington's disease (four males and one female; aged 30 to 70 years; with disease duration from 2 to 13 years; four on no drugs and one on tetrabenazine 25 mg three times daily) with obvious chorea were studied. After an overnight fast, blood was withdrawn for glucose estimation, and insulin (0.1 mg/kg) was injected into the opposite arm. Blood sugar and clinical response were measured every 10 min for 60 min, and then every 20 min for a further 60 min. The severity of chorea was rated using a specially designed scale described in detail elsewhere.⁵ In addition the number of choreic movements occurring at rest in one selected region, such as an eye, finger or toe depending on the individual patient, was counted over a 60 sec period. Blood sugar fell below 2.0 mmol/l and symptoms and/or signs of hypoglycaemia developed in all subjects. However, the intensity of chorea did not alter. Three subjects fell asleep during the test, and chorea disappeared in two, but on arousal their chorea was of the same severity as before insulin. Unfortunately, insulin-induced hypoglycaemia had no effect on chorea in our patients.

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Fig Chorea severity scores (above) and blood glucose levels (below) before and after injection of insulin 0.1 ul/kg iv. On the left are the mean values ($\pm$ 1SD) for three patients observed for 120 minutes after injection; on the right are the individual values for two other patients who required intravenous glucose.

Periodic alternating nystagmus in a case of hereditary ataxia and its treatment with baclofen

Sir: I describe below a patient with a dominantly inherited cerebellar degeneration who presented with oscillopsia, and was found to have periodic alternating nystagmus. His symptoms were virtually abolished following treatment with baclofen, which confirms a recent finding in two out of three cases of periodic alternating nystagmus similarly treated.¹ Periodic alternating nystagmus itself is a rare and extraordinary form of spontaneous nystagmus. It is a horizontal, usually jerk nystagmus which changes direction periodically. Less than a hundred cases are to be found in the literature in association with a considerable variety of disorders including multiple sclerosis, posterior fossa malformations and tumours, phenytoin intoxication and neurosyphilis. One previous case has been reported in a hereditary ataxia (Friedreich's ataxia),² but the veracity of the diagnosis in that case has been questioned.³

A 34-year-old salesman presented with a four year history of increasingly troublesome oscillopsia. His father and grandfather had both developed ataxia in middle

life. The former is living, has been investigated elsewhere, and a diagnosis of hereditary ataxia made; he has nystagmus, but details are not known. No other family members are as yet affected. On examination cavoid feet and alternating nystagmus were noted, but no other abnormalities. Routine blood count and biochemistry were normal. Acanthocytes were looked for but none seen. Serological tests for syphilis were negative. Skull radiographs were normal but a CT scan showed marked brain stem and cerebellar atrophy. Nerve conduction studies and visual evoked responses were within normal limits. The upper trace in the figure is a continuous record of the patient's periodic alternating nystagmus with eyes in the primary position. The cycle length is 182 seconds with a left beating phase of 80 seconds, a right beating phase of 76 seconds and two null phases of 12 and 14 seconds respectively. The second trace shows a series of saccades 30° to either side of the primary position, recorded at a lower gain. At the start of the trace the null position is at 30° left, as it shifts to the primary position first degree nystagmus to the left appears and increases in amplitude. It has been commented previously that the nystagmus can be conceptualised as resulting from periodic shifts of

the null zone.⁴ Other features were also similar to most previously reported cases (for example the three analysed in detail by Baloh *et al.*).⁵ The spontaneous nystagmus was superimposed on saccades and smooth pursuit. Optokinetic nystagmus could be produced only during null phase or when the stimulus was moving in the same direction as the slow phase of the nystagmus.

Traces three and four were recorded in a similar manner whilst the patient was taking baclofen 10 mg thrice daily. Spontaneous nystagmus in the primary position has been abolished, first degree nystagmus still occurs during saccadic eye movements to right and left but there is no longer any shift of the null position.

Oscillopsia is common in periodic alternating nystagmus.⁶ It is particularly troublesome because, for most of the cycle, nystagmus occurs in the primary position. This patient was rendered asymptomatic for most activities whilst taking baclofen because this feature was abolished. Periodic alternating nystagmus is uncommon and easily missed. The present case illustrates that whatever the underlying disorder, it is worthwhile looking specifically for the condition in any patient complaining of oscillopsia as it appears to be one of the few such eye movement dis-