

active tuberculosis. We have studied all active cases and all those receiving chemoprophylaxis for the subsequent five years after excluding recently arrived immigrants. Two important conclusions emerge.

First, although the total number of active cases was small, none had been immunised with BCG. Second, there were very few children from ethnic minorities with active disease and the rate for children whose families originated from the Indian subcontinent was similar to that for white children (table 2). The rate for Asian children is considerably lower than those reported in the 1983 national survey¹ and this is likely to be due partly to a decrease in the numbers of new immigrants, partly to the improved living standards of some Asian communities, but also to the protective effect of BCG immunisation of Asian infants. Neonatal BCG immunisation has been shown to give children an estimated protection against tuberculosis of between 64%³ and 75%.⁴

The results of this study require confirmation from other urban centres, whose ethnic minority communities may have different socioeconomic characteristics. In Blackburn, for example, 89% of notified childhood tuberculosis was among Asians.⁵ One possible source of bias in our study was that ethnic minority children may have been under-represented in the detected cases of tuberculosis due to less good use of the health services. Some support for this possibility is provided by the fact that four of the 11 more advanced cases came from an ethnic minority, but the numbers are too small to make definite conclusions.

Poverty, as shown by residence within a desig-

nated urban priority area, was a better indicator than ethnic group of the risk of tuberculosis (table 2). Selection of infants for BCG immunisation by ethnic group or by family history, while ensuring adequate protection for the immunised, has resulted in a small number of cases occurring in unimmunised children, usually from poorer sections of the city. Any form of selective policy has its problems. It may be wiser to immunise all babies on the postnatal wards, as is the practice in some hospitals,⁴ particularly because of the expected increase in the prevalence of adult tuberculosis associated with AIDS.⁶

We thank the clinicians involved for permission to study children under their care.

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Transient intracranial hypertension of infancy

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SUMMARY Four infants were seen on six separate occasions with febrile illnesses associated with a bulging anterior fontanelle and irritability. They had signs of a transient form of intracranial hypertension.

The irritable infant who presents with a bulging fontanelle and fever may have meningitis. We have recently seen four infants because of a bulging

fontanelle and irritability, associated with poor feeding and fever. None of them had otitis media, meningitis, or an intracranial space occupying lesion and their signs settled within a few days.

CASE 1

A previously healthy 8 month old girl presented with a 24 hour history of vomiting and fretfulness. Examination showed an irritable, febrile infant with a head circumference of 46.0 cm (75th centile), diastasis of cranial sutures, and a bulging anterior

fontanelle. There was no otitis media or focal signs of infection.

A computed tomogram showed mild communicating hydrocephalus, and lumbar puncture showed a raised opening pressure of 40 cm cerebrospinal fluid. She improved rapidly after lumbar puncture and was discharged home after 48 hours. Four days later she was readmitted with similar physical signs and a repeat computed tomogram was identical to the first one. Daily lumbar punctures were started and her symptoms resolved in two days.

CASE 2

A 5 month old boy was admitted to hospital with a three day history of fever, irritability, and vomiting. His parents had noticed his anterior fontanelle bulging for 24 hours. He had been born at 33 weeks' gestation and had had no neonatal problems. The head circumference at birth was on the 50th centile and his head growth was normal up to 3 months of age.

Examination showed an alert, mildly feverish infant whose head circumference was 45.0 cm (97th centile). The fontanelle was tense and bulging, nevertheless there was no suture separation. A cerebral ultrasound scan was normal.

He settled over two days and was discharged home, but readmitted three weeks later with a bulging fontanelle. His head circumference had increased to 46.1 cm. Because of concern about a possible space occupying lesion a computed tomogram was performed, which showed mild communicating hydrocephalus. His signs of raised intracranial pressure settled spontaneously over three days.

CASE 3

A 6 month old boy presented with a 12 hour history of fever and bulging of his fontanelle, both of which had been noticed by his parents. He had been born at term and had had birth asphyxia requiring intubation for seven minutes but he had made an uneventful recovery. Follow up over the next three months showed normal growth and development.

His temperature was 38.8°C and he was irritable on handling with episodes of screaming. His fontanelle was tense and bulging and his head circumference was 43.0 cm (25th centile). There were no focal signs of infection. Examination of his cerebrospinal fluid and a cerebral ultrasound scan were normal.

His fontanelle slackened promptly after lumbar puncture and his irritability rapidly resolved. He was discharged three days later.

CASE 4

A previously healthy 7 month old boy presented

with an acute illness of eight hours duration, during which he had been screaming and was reluctant to feed. He was found to be slightly irritable and had a temperature of 37.5°C. His anterior fontanelle was bulging at rest. The head circumference was on the 97th centile. A cranial ultrasound scan was normal. Because his general condition improved and there was concern about raised intracranial pressure an initial lumbar puncture was temporarily deferred. His fever and symptoms settled somewhat on treatment with regular paracetamol. Two days after admission he developed a left upper lobar pneumonia which responded to antibiotics; three days later his fontanelle was normotensive.

Discussion

These infants had six episodes of acutely raised intracranial pressure without evidence of central nervous system infection or a space occupying lesion. All had negative cerebrospinal fluid, blood, and urine cultures and normal plasma electrolyte and calcium concentrations. No specific cause could be found for the raised intracranial pressure but all episodes were associated with a febrile illness, in one case a chest infection. Two of the four cases had normal sized ventricles on cranial ultrasound scan and these are similar to benign intracranial hypertension.¹ The two other infants had enlarged ventricles and widened subarachnoid spaces on a computed tomogram: features of a mild communicating hydrocephalus. Follow up of all four children for several months showed normal developmental progress and normal head growth.

We suggest that a non-specific infective illness has interfered with absorption of the cerebrospinal fluid from the arachnoid villi. The widened cerebrospinal fluid spaces in two cases support this hypothesis. Benign intracranial hypertension has previously been described in children during roseola infantum,² upper respiratory tract infections,¹ and otitis media.³ Other conditions causing benign intracranial hypertension were excluded.⁴⁻⁶

The first three infants had rapid resolution of their symptoms after lumbar puncture. This procedure reduces intracranial pressure, an effect which may last for up to a day if cerebrospinal fluid continues to leak from the dural puncture site.

We wish to add a note of caution. The infant with signs suggestive of meningitis but who has a normal lumbar puncture may have transient intracranial hypertension. If there is no clinical improvement, however, the possibility of early meningitis that has failed to cause an initial pleocytosis must be considered—antibiotics should be commenced and the lumbar puncture repeated.

This group of infants developed a transient rise in intracranial pressure without evidence of central nervous system infection. Discussion with colleagues leads us to believe that this condition is much more common than has previously been recognised.

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Cerebral systemic lupus erythematosus

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SUMMARY Cerebral systemic lupus erythematosus is uncommon in childhood. Three further cases are presented to draw attention to the unusual and varied clinical manifestations of this condition.

The diagnosis of cerebral systemic lupus erythematosus (SLE), even in adults, is easily overlooked in the absence of more widespread systemic involvement by the disease. In children there is an even greater likelihood of misdiagnosis because of the rarity of the disorder in this age group.¹ The present report of three children who presented with cerebral manifestations of SLE emphasises not only the varied features of such neurological involvement but also the need to investigate any obscure neurological illness in children by immunological tests.

Case reports

Case 1. A boy aged 6 years presented with writhing movements of the left arm followed several weeks later by sudden flexion and extension movements of the left leg. These worsened over the next three months and were accompanied by the evolution of dystonia of the left eyelid and episodic deviation of the eyes to the left. Six months later he developed focal tonic clonic seizures affecting the left side which were difficult to control, despite the use of phenytoin, sodium valproate, clonazepam, and phenobarbitone.

Over the next two years he followed a fluctuating course, with variation in his dystonia and mobility. Attention span, behaviour, and both reading and writing all deteriorated. Investigations, including biochemical screening of plasma and urine for inherited or acquired metabolic disorders, a computed tomogram of the brain and nuclear magnetic resonance brain scan, carotid angiography, and brain biopsy all failed to give a diagnosis. An area of atrophy in the head of the left caudate nucleus was the only finding of note. At the age of 9 years he developed a 'lupus syndrome' characterised by arthralgia, lymphadenopathy, and a morbilliform rash, which was initially thought to due to his anticonvulsant treatment. The presence of changes pathognomonic for SLE on immunohistology of a skin biopsy specimen, together with the presence of characteristically high titre antibodies to double stranded DNA (table), however, suggested that the whole illness, including the presenting neurological features, was the result of SLE. Treatment with prednisolone (40 mg/1.73 m²/day) was introduced with a dramatic improvement not only in the lupus syndrome but also in behaviour and intellectual function. Gradual withdrawal of the prednisolone over a period of a month was followed by a deterioration which, however, responded once more to the reintroduction of steroids and a small supplement of azathioprine. These measures were followed by an appreciable improvement in behaviour, speech, and in school performance. Control