Dr El-Radhi comments: I accept the first criticism that about half of the 154 patients in our study were studied retrospectively. Currently we are carrying out a new study in which patients are only prospectively studied.

I do not accept the second comment that we are perhaps misguided in believing that lowering the fever in a child is good practice. Accumulated data from the past 20 years suggest that fever may be beneficial to the infected host. In febrile convulsions, infection with virus plays a major part. Could the child then become ‘neuro-immune’ around the age of 5 years rather than the currently held notion of cerebral maturation? More research is needed to determine the role of interleukin 1 in astrocytes.2

We, too, believe firmly that each child has his own threshold for eliciting a convulsion and that the higher the threshold the greater the fever required to trigger a convulsion. In other words, the child’s threshold can be determined by the degree of temperature at the time of the attack.

The group of children with the lowest fever (38–38.9°C) who experience more recurrences, should be of interest for follow up for a longer period in this group we may find the reported increased incidence of epilepsy after a febrile convulsion. Although it may be appropriate to consider anticonvulsant prophylaxis here, such prophylaxis appears to be more controversial now than a decade ago.


Growth hormone deficiency in children with chromosomal abnormalities

Sir,—We were intrigued by a recent report of Abusrewil et al.1 They described a boy with a ring 18 chromosome karyotype and complete growth hormone deficiency responsive to treatment. We are following up two patients with chromosomal deletions who have also responded favourably to growth hormone treatment.

Case 1, a boy, was born at 36 weeks' gestation weighing 1960 g. He had microcephaly and multiple malformations, including dysplastic right kidney, thoracic hemicardia, pulmonary stenosis, hernias, and hypospadias. Karyotype was male with partial ring 21 chromosome and partial monosomy 21. Holoprosencephaly was seen on a computed tomogram of the brain. Similar cases with a broad phenotypic spectrum have been described previously.2 3 At 20 years of age he was severely retarded, his length was 83-4 cm (~4-5 SD score), weight 107-2 kg, and bone age 18 months. Maximum stimulated growth hormone concentrations were 5-6 mIU/l and 1-4 mIU/l in the levidopa and clonidine test, respectively. Thyroxine concentration was normal. Growth hormone treatment with somotrem (Pentopin) at a dose of 1-25 mg subcutaneously three times a week was started and has been continued for 16 months now. Growth velocity increased from 3-4 cm/year pretreatment to 9-5 cm/year during treatment. Case 2, a girl, was born at term weighing 2700 g. She had microcephaly, downward slanted palpebral fissures, long hands with a proximal thumb, cleft soft palate and bilateral sixth and auricular canals. Primary hypothyroidism was diagnosed at age 7 years, but the growth rate did not improve much on treatment. At 11-4 years, she was only mildly mentally retarded with height -2.1 SD (~ 3-23 SD score), weight 21-3 kg, and bone age 8-8 years. Maximum growth hormone concentrations were 15-2 mIU/l in a clonidine test and 21-4 mIU/l in an insulin stimulation test, and 8-4 mIU/l in a 12 hour overnight study. Chromosome analysis showed deletion of the long arm of chromosome 18.4 A computed tomogram of the brain was normal. On treatment with somatrem, 1 mg subcutaneously three times a week for one full year height velocity improved from 3-6 cm/year to 6-7 cm/year. Growth hormone treatment was then stopped for eight months with a decrease in height velocity to 5-0 cm/year. For the past six months, the patient has received somatrem 1-25 mg subcutaneously daily and has grown at a rate of 8-25 cm/year.

It is remarkable that our patient with ring 21 chromosome abnormality also has growth hormone deficiency, like the patient of Abusrewil et al and two further patients they cite from the literature who all had a ring 18 chromosome karyotype.1 We agree that poor growth and short stature is a hallmark of many chromosomal disorders, but may occasionally be associated with growth hormone deficiency and needs to be diagnosed.

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Generalised lymphangiomatosis with chylothorax

Sir,—We read with interest the article by Dunkelman et al.,1 and would like to add a description of an infant with lymphangiomatous scalp lesions currently under our care. A 25 year old pregnant woman with two previous first trimester miscarriages was noted at 27 weeks' gestation to have polyhydramnios. Ultrasound scan of the fetus showed pleural and pericardial effusions, ascites, and multi-loculated fluid filled scalp lesions. The mother was blood group O negative and had irregular antibodies, although these were considered unlikely to cause haemolysis. At 33 weeks' gestation spontaneous labour resulted in the delivery of an hydridic infant boy weighing 2325 g. The scalp lesions were bilateral, symmetrical, and localised to the parietal region. They were up to 1.5 cm in diameter and similar in appearance to a bunch of grapes (figure). There was no obvious communication to other structures, aspiration yielding clear fluid. A biopsy specimen showed lymphangiomatous circumscription. The baby had bilateral pleural effusions aspirated and was initially ventilated for four days. Investigation failed to demonstrate any cause for the hydroms. Full skeletal survey showed no limb reduction or lytic lesions, only asymmetry of the thoracic cage. Aspiration of a small left pleural effusion at 1 week of age yielded clear fluid with numerous lymphocytes on microscopy. Subsequent chest radiography was consistent with pulmonary lymphangiecstasia and computed tomography of the chest at 1 month revealed interstitial thickening consistent with this diagnosis. Lung biopsy has not been performed. Chronic ventilator and oxygen dependence ensued before final successful extubation after 10 weeks. Enteral feeding was commenced at 1 month with small volumes of Pregestemil (Bristol-Myers).

Subsequently a symptomatric left pleural effusion has required thoracocentesis on five occasions, with the most recent specimen yielding a turbid fluid identifiable as chyle (protein 41 g/l and triglyceride 4-14 mmol/l) with abundant white cells, all lymphocytes. The infant is now 4 months of age and while remaining oxygen dependent is completely enterally fed with no symptoms attributable to lymphangiomatosus bowel involvement. The scalp lesions have not increased in size and are now epithelialising.

Generalised lymphatic abnormalities are recognised causes of hydroms fetalis.2 We are not, however, aware of any previous reports of an association between lymphangiomatous scalp lesions and chylothorax, which in this case presented antenatally.


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