

Lucina

The prevalence of alpha-thalassaemias is up to 5% in Hong Kong and up to 40% in some parts of South East Asia. Haemoglobin H consists of four beta-globin chains. Instead of the usual complement of four normal alpha-globin genes (two on each chromosome 16), people with haemoglobin H disease have only one. At the other three alpha-globin gene sites, they have either three deletions or two deletions and one mutation. Now, work in Hong Kong (*New England Journal of Medicine* 2000;343:544–50) has shown that those with a mutation have more severe disease but iron overload is equally a problem in both types.

Eight of 13 American and four of five French boys with cerebral X-linked adrenoleukodystrophy survived bone marrow transplantation at ages 5 to 11 years and were followed for five to ten years (*Lancet* 2000;356:713–18). Verbal intelligence remained normal in 11 and non-verbal abilities remained stable or improved in seven. Eight showed continuing demyelination on magnetic resonance imaging (MRI) but later stabilised, one showed no change, one improved, and two showed complete reversal of MRI abnormalities.

Is laparoscopic surgery safe in children with ventriculoperitoneal shunts? A small study in St Louis, Missouri (*Journal of Pediatric Surgery* 2000;35:1104–5) suggests that it is. Three of ten children had postoperative complications but none was related to the shunt. Six of the laparoscopies were for fundoplication and the complication rate in these six children was the same as in 17 without a shunt who had the same operation.

In Finland (*Journal of the American Medical Association* 2000;284:993–1000), the parents of 1062 healthy 7 month old infants were randomised to receive counselling about a children's diet low in saturated fat and cholesterol or standard health education; 496 of the children also had neurodevelopmental testing when aged 5. The results of these tests were essentially the same in the two groups. At 5 years old, mean serum cholesterol concentration was 4.27 mmol/L in the diet group and 4.41 mmol/L in the controls. The diet reduced both high and low density lipoprotein cholesterol concentrations but did not alter the ratio of high density lipoprotein cholesterol to total cholesterol.

The quality of the evidence that abused children become abusing adults has been questioned. A systematic review of 10 studies (*Lancet* 2000;356:814–19) showed that only two met more than five of eight

methodological standards. Four studies (including the one which met all eight standards) showed a significant increase in risk and four (including the one which met six standards) did not. Even the authors of the "eight star" study pointed out that intergenerational continuity is not the rule and many abused children become competent parents. The positive results in the study were confined to primiparous mothers of low socioeconomic status who had been severely abused.

It has been estimated (*Lancet* 2000;356:795–801) that some 6% of deaths in Austria, France, and Switzerland are caused by air pollution and that pollution from motor traffic is responsible for about half the damage. Traffic generated air pollution causes around 295 000 episodes of "bronchitis" and over 160 000 attacks of asthma in children in these countries each year. An increase in inhalable particulate matter of 10 $\mu\text{g}/\text{m}^3$ would cause a 4.4% increase in asthma attacks in children.

Analysis of samples taken from children with *Haemophilus influenzae* type B (Hib) meningitis in Boston, USA in the early 1970s (*Paediatric Infectious Disease Journal* 2000;19:589–91) has shown that some (and almost all of those over two years old) developed a rapid antibody response after the onset of disease though initial antibody levels were low. They must have been naturally immunologically primed, probably by cross-reacting flora. The implication is that immunological priming (as induced by Hib vaccine) may not prevent invasive disease, but may reduce its severity. Circulating antibody to capsular polysaccharide at the time of encounter with the organism is important for complete protection.

At the Hospital for Sick Children in Toronto (*New England Journal of Medicine* 2000;343:689–94) 100 children aged over 5 years old, with severe asthma, were randomly assigned to a single dose of either oral prednisone (2 mg/kg) or inhaled fluticasone (2 mg) along with repeated doses of inhaled salbutamol and ipratropium. Mean forced expiratory volume in one second (FEV₁) increased over four hours from 44% to 63% in the prednisone group and from 46% to 56% in the fluticasone group. FEV₁ fell over this time in a quarter of those given fluticasone but in none of those given prednisone. These authors conclude that inhaled steroid should not be used in acute asthma.

A study in a New York neonatal intensive care unit (*New England Journal of Medicine*

2000;343:695–700) has supported previous suggestions that one factor which might predispose to pseudomonas colonisation is the wearing of artificial fingernails by staff. Increasing age also increased the risk of pseudomonas carriage on the hands of unit staff.

Animal and human evidence has suggested that zidovudine given perinatally might damage cardiac muscle mitochondria. Now an American serial ultrasound study (*New England Journal of Medicine* 2000;343:759–66) of 440 infants born to HIV positive mothers has not shown any effect of perinatal zidovudine treatment on left ventricular structure or function. A 1999 French report, though, suggested that mitochondrial damage from zidovudine (shown in eight of 1754 exposed children) is most likely to present with neurological symptoms. Nevertheless, the benefits of zidovudine in preventing vertical transmission of HIV far outweigh the possible harm.

Bactericidal/permeability-increasing protein (BPI) binds endotoxin and has antibacterial activity. A UK/US trial of a recombinant BPI fragment (rBPI₂₁) given to children with severe meningococcal sepsis (*Lancet* 2000;356:961–7) showed no reduction in mortality but trends towards reduced need for severe amputations and better outcome. The average time to administration of rBPI₂₁ was six hours and it is suggested that giving it much earlier (with the first antibiotic dose) would give better results.

There are many good reasons for advocating breast feeding but new evidence suggests that the prevention of schizophrenia is not one of them. Data from 1946 and 1958 British birth cohorts (*British Journal of Psychiatry* 2000;177:218–21) showed no significant difference in breast feeding rates between those who developed schizophrenia and those who did not. Previous case control studies had suggested that breast feeding might be protective.

Psychologists in the West Midlands (*British Journal of Medical Psychology* 2000;73:327–38) used the technique of discourse analysis after interviewing three sets of parents of children being assessed for an autistic spectrum disorder. The parents were thought to have an "ideological dilemma" in that they regarded themselves as the experts about their own children but also acknowledged their need of the expertise of the professionals. Balancing the two viewpoints is, of course, an important part of clinical practice.