then perhaps one could use an anti diarrhoeal drug, such as Lomotil, though only with caution. This is because, in our experience, it is not possible to predict what dose will be toxic in children.

Both Dr Karan and Dr Limaye emphasise that in our cases severity of the symptoms was related to the dose ingested, and they also imply that symptoms only occur above a recommended therapeutic dose 0.25 mg/kg a day. But they fail to observe that on the contrary the figures for the lower limit of the ingested dose (mild, 0.62 mg/kg; moderate, 0.25 mg/kg; and severe, 0.77 mg/kg) did not correlate with the severity of symptoms. We suggest that if it is necessary to use Lomotil the prescribing doctor should take cognisance of the risks.

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Infantile cortical hyperostosis with raised immunoglobulins

Sir,
Ramchander and Ramkissoon (Archives, 1978, 53, 426) drew attention to the IgA and IgM levels in 2 cases of infantile cortical hyperostosis (Roske-de Toni-Caffey syndrome). They said that a virus infection during intrauterine or neonatal life might be the cause of this disorder.

I know 2 Italian families in which this disease was diagnosed in two generations: in the first family, in the mother and her son; in the other family, the father, the daughter, and the father’s first-cousin all had the disease (Duillo and Cerruti Mainardi, 1969). A few months ago another girl was born in the second family, and she shows the same disorder at age 8 weeks.

These cases, as do many other familial ones in the literature, support de Toni’s hypothesis (de Toni, 1943) that genetic factors may play a role in the aetiology of this mysterious disease.

References


Polyarthritis associated with Wilson’s disease

Sir,
A 14-year-old boy was admitted to hospital in August 1978 with an acute attack of polyarthritis which he had had for one day, and which affected both knees and elbows.

There was no history of sore throat, and the family history was unremarkable. On examination he was pale, and the affected joints showed effusion with tenderness and heat. Small spider telangiectases were present over the upper sternum, and both spleen and liver were slightly enlarged. Slit-lamp examination showed Kayser Fleischer rings in both corneae.

Full blood picture, ESR, urine analysis, liver function tests, ECG, serum urate, Hb electrophoresis, C-reactive protein, latex-fixation test, throat swab culture, lupus erythematosus phenomenon, were all negative. ASO titre, 333 IU/ml. Percutaneous liver needle biopsy showed fatty changes. Serum copper 77.96 μg/100 ml (12 μmol/l), serum caeruloplasmin 105 mg/l (10.5 mg/100 ml), urinary copper 261 μg/24 h (4.09 μmol/24 h).

The boy subsequenly developed two further attacks of polyarthritis during the next 2 months; the findings and investigations then were similar with ASO titre 166 and 250 IU/ml.

He responded well to paracetamol initially, and was then put on d-penicillamine, when he remained well until last seen in June 1979.

The association of polyarthritis with Wilson’s disease has not been previously reported, and may have been coincidental, but slit-lamp examination of atypical cases of arthritis in children would be prudent.

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Sodium valproate, pregnancy, and neonatal hyperglycinaemia

Sir,
It has been demonstrated that sodium valproate (SV) crosses the placenta freely and that maternal serum levels will reflect fetal serum levels (Archives, 1979, 54, 240). The SV concentration in the serum of a neonate was of the same order as that of the mother’s at delivery, but fell to insignificant levels by 5 days and was undetectable at 29 days. As this child appeared healthy, it was concluded that the use of SV in pregnancy was safe.

On the other hand, administration of SV is known to cause hyperglycinaemia and hyperglycinuria in patients suffering from epilepsy (Jaeken et al., 1977; Kamoun and Parvy, 1978; Similä et al., 1979). In experiments on animals it has been shown that the rate of influx of glycine and other amino-acids into the brain is higher in the newborn period than later in life (Seta et al., 1972; Banos et al., 1978). In addition, high concentrations of glycine, which are found in nonketotic hyperglycinaemia and many organic acidemias, are associated with severe impairment of neuronal function, and therefore a high plasma level of glycine during early postnatal life may be a risk factor (Tanaka, 1975; von Wendt et al., 1978). Therefore, we monitored the clinical state and the levels of plasma glycine and serum SV postnatally in 2 neonates; in both, the mothers had received SV treatment during pregnancy.
Infantile cortical hyperostosis with raised immunoglobulins.

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