
Toledo type brachyglymania

Editor,—I read with interest the paper by Grain et al dealing with the first UK case of a type of brachyglymania (short trunk) which is associated with both peripheral corneal punctate opacities only seen by slit lamp and a qualitative abnormality of glycosaminoglycans (chondroitin sulphate).1 These data confirm our previous findings in four siblings with this autosomal recessive condition.2 We agree with the authors' statement that these cases represent a distinct type of spondyly dysplasia. Natural history, physical examination, and ophthalmological, radiographic, and biochemical findings in the case reported by Grain et al coincide with those of our cases, except for two points. First, some of our cases had irregular chondrocostal fusion. Second, advanced bone age was not present in our cases and this may explain why final adult height in our male cases (3-10th centile) was not as short as the one predicted for the case reported by Grain et al (3rd centile).

As stated by the authors, this disease may be a currently unrecognised cause of short stature. We have suggested for the diagnosis of this brachyglymania, type I, that a slit lamp examination as well as detailed glycosaminoglycan studies should be performed as routine procedures.3 The latter test is currently available only in some laboratories, but it is of crucial importance for the diagnosis of brachyglymania type I. As autosomal recessive and autosomal dominant patterns of inheritance are involved in the four types of brachyglymania,1 the distinction among them will require an adequate clinical management of the patients and will give further support for adequate genetic counselling.

Present efforts in this type of brachyglymania should be directed to DNA studies and among the candidate genes one should include those involved in glycosaminoglycan metabolism. Sequencing and cloning of the gene for brachyglymania would allow a more precise diagnosis and genetic counselling for this condition.

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Acyclovir in chickenpox

Editor,—Virological evidence for the reactionivation of chickenpox contracted in infancy has recently become available and is related to the immune status of the host.1 Secondary attacks of chickenpox and early reactivation as zoster have been reported after the treatment of normal children with chickenpox suggesting that the immune response may be impaired after acyclovir treatment.2 We report the case of severe primary varicella infection in an infant who should have been protected by passive maternal antibody. His mother had been treated with acyclovir for chickenpox before delivery.

A 25 year old woman presented at 38 weeks' gestation with a vesicular rash. The diagnosis of chickenpox was confirmed by the detection of specific IgM antibodies to varicella zoster virus and she was treated with acyclovir 800 mg five times daily for seven days. Nine days after the development of the rash she delivered a healthy boy. Six days after delivery he developed a vesicular rash and fever, and varicella zoster virus was detected in vesicular fluid. He was successfully treated with a five day course of acyclovir (100 mg/kg daily).

This infant was born nine days after his mother developed chickenpox and, in accordance with current guidelines for the UK, he did not receive active immumunisation globulin.3 We postulate that the use of acyclovir to treat the mother's infection may have affected her immune response to the virus leading to reduced passive transfer of immunity to her fetus. When he was born he was at increased risk of varicella infection, which he subsequently developed. This case highlights concerns over the effect of acyclovir on the immune response to chickenpox and also suggests that the present guidelines for passive immunisation against varicella zoster virus may leave a proportion of infants born to mothers treated with acyclovir at unnecessary risk.

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Expulsion of ventriculoperitoneal shunt tubing

Editor,—A baby girl of 18 months was admitted to our unit on 3 September 1982 with a two week history of irritability, vomiting, and refusal to sleep. She had a fever of 38-5°C and a lumbar puncture showed cerebrospinal fluid protein of 3-6 g/l, 750 polymorphonuclear leucocytes/mm³, protein of cerebrospinal fluid, and Gram positive cocci on staining.

A diagnosis of pneumococcal meningitis was made and the child was treated with triple chemotherapy: penicillin, sulphadimidine, and chloramphenicol as was routine in 1982. She remained clinically unwell and developed a third and sixth nerve palsy. Computed tomography of the head showed marked dilatation of the lateral and third ventricles and she was referred to the neurosurgeons who subsequently inserted a ventriculoperitoneal shunt. The child recovered from her meningitis but remained globally retarded in her development with regular seizures and unable to speak.

At the age of 14 years, she was rehydrated with gente of some group. was well and her bowels was normal. Urine culture and analysis was negative and a plain abdominal radiograph failed to reveal any abnormality. The apparent abdominal pain that the child was suffering persisted intermittently for several weeks and she was reviewed and examined on several occasions. No clinical evidence of organic disease was elicited. After approximately eight weeks of intermittent symptoms the child was referred to the local neurosurgeon. The child recovered and was eventually discharged after being reviewed and assessed. There was no relapse of meningitis but she remained severely disabled.

A rare example of nodular lymphoid hyperplasia causing a ventriculoperitoneal shunt failure is described. The finding is relevant to the practical aspects of shunting in children with congenital hydrocephalus.

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Infant length measurements

Editor,—Like Professor Frank Falkner1 I was interested to read Dr Doull's article on the reliability of infant length measurement,2 even though I am a little disappointed to find no reference to the Neonatometer—an instrument for measuring crown-heel length in infancy designed and written up by Bob Holding (from Holtain Ltd) and myself 24 years ago3 in the Archives. This design allowed the child to lie comfortably on a flat platform as was given to the technique in the training of observers with the neonatometer,4 95% of all observations of crown-heel length were likely to lie between plus and minus 3·4 mm of the true value. These represented acceptable, length and reliable measurements. The constant pressure pad fitted to the number counter, allowing it to automatically lock, added particular precision. We also showed that mothers were well able to hold the head.

But a good reliable instrument is one thing, it is quite another to convince people of the value of length measurement in infants. Along with weight and head circumference, length is important — not only for the more immediate assessment of growth status but also to help evaluate a problem of growth in an older child by looking back at earlier measurements.

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Toledo type brachyolmia.

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