ARCHIVES OF DISEASE IN CHILDHOOD

The Journal of the British Paediatric Association

NOTES TO CONTRIBUTORS Papers for publication should be sent to the Editors, *Archives of Disease in Childhood,* BMA House, Tavistock Square, London WC1H 9JR (fax 0171-383-6668) and should be prepared in accordance with the 'Instructions to authors' published in the January and July issues of the journal. Copies of these instructions may be obtained from the editorial office on receipt of a stamped addressed envelope.

NOTICE TO ADVERTISERS Applications for advertising space and rates should be made to the Advertisement Manager, *Archives of Disease in Childhood,* BMA House, Tavistock Square, London WC1H 9JR.

NOTICE TO SUBSCRIBERS *Archives of Disease in Childhood* is published monthly. The annual subscription rates are £204 ($322). Orders should be sent to the Subscription Manager, *Archives of Disease in Childhood,* BMA House, Tavistock Square, London WC1H 9JR. Orders can also be placed with any leading subscription agent or bookseller. (For the convenience of readers in the USA subscription orders, with or without payment may also be sent to *British Medical Journal,* PO Box 408, Franklin, MA 02038. All inquiries however must be addressed to the publisher in London.)

Subscribers may pay for their subscriptions by Access, Visa, or American Express by quoting on their order the credit or charge card preferred together with the appropriate personal account number and the expiry date of the card.

All inquiries regarding air mail rates and single copies already published should be addressed to the publisher in London.

The British Paediatric Association is a registered charity.

Published by the BMJ Publishing Group
Typeset by Bedford Typesetters Ltd.
Printed by The Devonshire Press Ltd.

Second class postage paid, Rahway NJ. Postmaster: send address changes to: Archives of Disease in Childhood, c/o Mercury Airfreight International Ltd Inc, 2323 Randolph Avenue, Avenel, NH 07001, USA.

ISSN 0003-9888

LUCINA

Orthopaedic problems are common in children with prune belly syndrome (*Journal of Bone and Joint Surgery* 1995; **77A:** 251–7). Twenty five of 40 children with the syndrome had musculoskeletal problems, including 13 with significant hip disease and 11 with spinal problems. Three children had renal osteodystrophy.

*In assessing the benefits of anticonvulsant drug treatment or epilepsy surgery it is clearly easiest to count seizures. Equally clearly, other factors must be just as important and work on adult patients in the United States (*Annals of Neurology *1995; 37: 158–66) has attempted to incorporate health related quality of life measures into assessments of the results of epilepsy surgery. What really matters is whether the patients think they have benefited or not.*

Work in Italy gives hope of producing varieties of wheat which can be tolerated by people with coeliac disease (*Gut* 1995; **36:** 375–8). Wheats lacking several gliadin components have been tested in vitro using coeliac intestinal mucosa and found to be much less toxic. Further, more gliadin deficient, varieties are being developed and clinical trials can't be far off.

*It becomes important to recognise rare diseases when they are treatable. One such condition is Segawa's disease (hereditary progressive dystonia with marked diurnal fluctuation). This disabling condition responds dramatically to treatment with levodopa and some cases do not show the characteristic diurnal fluctuation. It has now been shown that dopa responsive dystonia, with or without diurnal variation, is caused by mutation of a gene on the long arm of chromosome 14 (*Annals of Neurology *1995; 37: 405–8).*

Three French doctors were asked by the British Audiology Society to find out how neonates and infants were being tested for hearing loss across Europe (*International Journal of Pediatric Otorhinolaryngology* 1995; **31:** 175–82). They found that all newborns were regularly tested only in Germany and Hungary and behavioural tests are most often used. They recommend objective tests such as auditory brain stem responses and transient evoked otoacoustic emissions and prefer the latter because of ease and rapidity of use. Ideally all newborn babies should be tested. A selective testing policy will result in many hearing impaired children being missed but could be dictated by financial considerations. Children with a significant sensorineural hearing loss should be discovered as soon as possible after birth.

*Doctors in Ohio argued that suppressing the inflammatory response within the lungs in patients with cystic fibrosis might help to preserve lung function. In a double blind trial 85 patients with mild lung disease were given either ibuprofen or placebo for four years (*New England Journal of Medicine *1995; 332: 848–54). Those assigned to ibuprofen maintained both lung function and weight better than the placebo group. Adverse reactions were no more common in the treatment group.*

Much thought has been given to the problem of delivering drugs specifically to target cells, especially to liver cells. A new approach is to deliver the drug within an artificially created chylomicron (*Nature Medicine* 1995; **1:** 221–5). Work on rats suggests that by doing this it may be possible to deliver antiviral drugs directly to hepatocytes in sufficient concentration to be effective without risk of toxicity to other organs and provides hope for the treatment of chronic hepatitis B virus carriers.

*Children with recurrent breath holding spells (BHS) deserve to have their haemoglobins checked. An association between BHS and iron deficiency anaemia has been known about for many years and the point is underlined by recent case reports (*Journal of Pediatrics *1995; **126:** 395–7) describing two children whose BHS resolved rapidly with correction of their anaemia.*

Malaria remains an important cause of early childhood morbidity and mortality in sub-Saharan Africa. A study of 9774 infants born in Cameroon in 1978 (*International Journal of Epidemiology* 1995; **24:** 204–17) shows that malaria contributed to about 9% of deaths under the age of two years. Factors associated with malaria mortality included lack of antenatal care, overcrowding, bottle feeding, lack of immunisation, other infections, and being firstborn.

*Hydroxyurea stimulates production of haemoglobin F and might, therefore, be of benefit to patients with sickle cell disease. An American study of adult patients (*New England Journal of Medicine *1995; 332: 1317–22) has shown a reduction in rate of sickle cell crises from a median of 4·5 per year in those given placebo to a median of 2·5 per year in those taking capsules of hydroxyurea. The treatment group had very significant ($p<0·001$) reductions in transfusion requirement and in the chest syndrome. Although there were no important adverse effects in this trial long term safety remains uncertain. Trials in children are in progress.*

Recent work in Canada on cell biology in the lesions of fibrous dysplasia of bone may lead to an understanding of the pathogenesis of osteosarcoma as well as of fibrous dysplasia (*New England Journal of Medicine* 1995; **332:** 1546–51). The fibroblasts in the lesions of fibrous dysplasia were shown to have increased expression of the c-*fos* proto-oncogene and this is thought to be a consequence of increased adenylate cyclase activity. Such changes are important for intracellular signalling pathways and fundamental to cell growth and differentiation. Osteosarcoma tissue also shows increased c-*fos* expression and it is suggested that this could be a primary step in the genesis of the tumour.