LETTERS

If you have a burning desire to respond to a paper published in *ADC* or *F&N*, why not make use of our "rapid response" option?

Log on to our website (www.archdischild.com), find the paper that interests you, click on "full text" and send your response by email by clicking on "submit a response".

Providing it isn't libellous or obscene, it will be posted within seven days. You can retrieve it by clicking on "read eLetters" on our homepage.

The editors will decide, as before, whether to also publish it in a future paper issue.

Problems involved with the use of comforters

While I share many of the concerns expressed by Gill¹ in his diatribe on dummies there are a number of matters mentioned which require amplification or correction. The first patent on the India rubber nipple resembling the present day dummy was recorded in 1845 and was described in use in its present form in London in 1927. Unfortunately by this time the practice of dipping the dummy in a variety of sweetening agents to make it a more effective pacifier had become established and this habit was noted to be associated with the early onset of dental caries. No doubt the loss of primary incisors mentioned by Gill is due to their destruction by rampant dental caries associated with the persistent use of sweetened pacifiers and their subsequent extraction due to spreading infection, pain, and loss of sleep. The association of dummy sucking with malocclusion is more complex than stated. While there is a general agreement on the effect of prolonged dummy sucking producing malocclusions in the primary dentition, these abnormalities are mainly self corrective on cessation of the habit which is usually before 5 years of age.²

G B Winter

Emeritus Professor of Children's Dentistry,
1 Hartfield Close, Elstree, Herts WD6 3JD, UK

References

- 1 Gill D. A diatribe on dummies. *Arch Dis Child* 2002;86:222.
- 2 Winter GB. Problems involved with the use of comforters. *Int Dent J* 1980;30:28-38.

Cataplexy in the Prader-Willi syndrome

We report cataplexy, sudden atonic episodes provoked by emotion, in three patients with Prader-Willi syndrome (PWS) and suggest that cataplexy may be relatively common in this condition.

Detailed questioning of the mother of an 18 year old woman who had PWS elicited a history of recurrent attacks, apparently induced by laughter, with sudden loss of power in all the patient's limbs. If standing, she would slump to the floor but recover completely after a few seconds. She had no history of the sleep paralysis or hypnagogic hallucinations and there was no family history of cataplexy, narcolepsy, or epilepsy. Her EEG was unremarkable. Episodes of cataplexy and of narcolepsy, despite excellent weight control, have been reported by two other patients with

Table 1 Conditions in which cataplexy is a recognised feature

Familial isolated cataplexy
Norrie's disease
Niemann-Pick disease type C
Coffin-Lowry syndrome
Narcolepsy-cataplexy syndrome
Pontomedullary/hypothalamic structural lesions

PWS who attend this hospital, an 8 year old girl and a 10 year old boy. Only one of the three patients possesses the HLA DR15 (DR2) DQB1*0602 haplotype that is strongly associated with the narcolepsy-cataplexy syndrome.

Cataplexy is usually precipitated by emotion provoking laughter, anger, or joy. The affected individual often falls to the ground without losing consciousness and the phenomenon is often mistaken for an epileptic or cardiac event.¹ It can occur in isolation as a dominantly inherited trait or in association with a number of other conditions (table 1). The association between PWS and cataplexy, though described previously,¹⁻⁵ is not widely recognised. Suspected episodes of cataplexy have been reported in eight of 35,¹ four of 25,² and three of 17³ patients with PWS. However, cataplectic manifestations are often "difficult to prove",¹ requiring a detailed history⁵ that is perhaps seldom available or elicited. We suggest that cataplexy may be relatively common in PWS and enquiries regarding its signs should always be made, especially in any patient with a past diagnosis of paroxysmal events.

E S Tobias, J L Tolmie

Duncan Guthrie Institute of Medical Genetics,
Yorkhill Hospitals, Glasgow G3 8SJ, UK;
gbcv55@udcf.gla.ac.uk

J B P Stephenson

Fraser of Allander Neurosciences Unit, Royal
Hospital for Sick Children, Yorkhill, Glasgow
G3 8SJ, UK

References

- 1 Helbing-Zwanenburg B, Kamphuisen HAC, Mourtazaez MS. The origin of excessive daytime sleepiness in the Prader-Willi syndrome. *J Intellect Disabil Res* 1993;37:533-41.
- 2 Cassidy SB, McKillop JA, Morgan WJ. Sleep disorders in the Prader-Willi syndrome. 1989 David W Smith Workshop on Malformations and Morphogenesis, Madrid, Spain. *Proc Greenwood Genet Centre* 1990;9:74-5.
- 3 Clift S, Dahlitz M, Parkes JD. Sleep apnoea in the Prader-Willi syndrome. *J Sleep Res* 1994;3:121-6.
- 4 Parkes JD. Genetic factors in human sleep disorders with special reference to Norrie disease, Prader-Willi syndrome and Moebius syndrome. *J Sleep Res* 1999;8(suppl 1):14-22.
- 5 Stephenson JBP. *Fits and faints*. London: MacKeith Press, 1990:144.

Kawasaki disease following meningococcal septicaemia

We report a case of Kawasaki disease (KD) following meningococcal septicaemia which we believe has not been described before. A 14 month old boy presented to his local hospital with a four day history of being unwell, fever, and blanching maculopapular rash. Meningococcal septicaemia was diagnosed clinically

and the boy was managed with fluid support and intravenous antibiotics. His recovery was complicated by developing respiratory syncytial virus positive bronchiolitis and secondary surgical emphysema. Polymerase chain reaction) was positive for group B meningococcus on day 3. Blood and urine cultures were negative. He continued to spike high temperatures in the ward; a lumbar puncture performed on day 13 showed normal cerebrospinal fluid microscopy and biochemistry. Other investigations, including cranial computed tomography scan of his brain and abdominal ultrasound (including renal vessel Doppler studies) were all normal. He continued to spike high temperatures with pleomorphic erythematous rash, non-purulent conjunctivitis, red enlarged lips, red gums, red inflamed tongue, and axillary lymphadenopathy >1.5 cm. A clinical diagnosis of KD was made; he was treated with intravenous immunoglobulin and aspirin with good effect. Platelet count on day 14 was 933 (admission platelet count was 187). On day 18 he was noted to have mild peeling of his scrotum, hands, and feet. An echocardiogram showed left coronary artery ectasia. He was discharged on day 22 with follow up arrangements including repeat echocardiogram. He was, however, lost to follow up and no further data are available.

Discussion

A number of epidemiological and clinical observations suggest that KD may be caused by an infectious agent. These include geographic clustering of outbreaks, often with a seasonal predominance and the acute self limited nature of the illness. Many of the clinical features of KD are similar to those of other infectious diseases, for example, adenoviral infection and scarlet fever. Staphylococci, streptococci, and Epstein-Barr virus are some of the infectious agents implicated in KD.¹ An unusual degree of immune activation caused by bacterial and viral protein toxins acting as superantigens is currently considered to be the basis of pathology in KD.²⁻⁴ We believe that our case shows the possibility that a meningococcal toxin could act as a superantigen to cause KD. We were unable to find any published record of such an association in the literature. The currently proposed hypothesis to explain the pathogenesis is that a genetically susceptible host becomes colonised on the mucous membranes of the gastrointestinal tract by an organism that produces a toxin which behaves as a superantigen. We propose that a toxin producing meningococcus could cause KD in the same fashion as toxic shock syndrome toxin producing *Staphylococcus aureus*. It is possible that our patient coincidentally had both illnesses at around the same time. Understanding the aetiology of KD remains a major unresolved issue in paediatrics. Although there is no conclusive data to support the superantigen induced disease theory for KD, evidence suggesting that superantigens may mediate KD is growing.

A V Ramanan, E M Baildam

Department of Paediatric Rheumatology, Royal
Manchester Children's Hospital, Manchester, UK
Correspondence to: Dr A V Ramanan, 508, 77 Elm
Street, Toronto, Ontario M5G 1H4, Canada;
avramanan@hotmail.com

References

- 1 Leung DYM, Meissner HC, Fulton DR, et al. Toxic shock syndrome toxin-secreting *Staphylococcus aureus* in Kawasaki syndrome. *Lancet* 1993;342:1385-8.

- 2 **Curtis N**, Zheng R, Lamb JR, *et al*. Evidence for a superantigen mediated process in Kawasaki disease. *Arch Dis Child* 1995;**72**:308–11.
- 3 **Abe J**, Kotzin BL, Jujo K. Selective expansion of T cells expressing T-cell receptor variable regions V beta 2 and V beta 8 in Kawasaki disease. *Proc Natl Acad Sci U S A* 1992;**89**:4066–70.
- 4 **Leung DYM**, Giorgio RC, Kazemi LV, *et al*. Evidence for superantigen involvement in cardiovascular injury due to Kawasaki syndrome. *J Immunol* 1995;**155**:5018–21.

ARC syndrome: an expanding range of phenotypes

A female infant, born to consanguineous Pakistani parents, was noted shortly after birth to have dysmorphic features, including prominent occiput, beaked nose, high arched palate, and arthrogryposis with dislocated hips and rocker bottom feet. Ichthyosis was also present. Metabolic acidosis developed within a few hours of birth and severe conjugated hyperbilirubinaemia within two days.

Renal tubular acidosis was manifest by generalised aminoaciduria, phosphaturia, and an N-acetylglucosamine:creatinine ratio of >1000. Liver investigations revealed similar findings to those previously reported, with conjugated hyperbilirubinaemia, greatly increased alkaline phosphatase, but normal γ glutamyltransferase.¹ Plasma and urinary bile acids were normal. Histology of the patient's liver revealed the presence of normal numbers of bile duct and no lipofuscin deposition or inflammatory changes. No giant cells were present.

Recurrent episodes of necrotising enterocolitis occurred during the first two months of life (no organisms were identified in either the blood or faeces at the time of the original or recurrent episodes). Repeated episodes of septicaemia occurred later. Marked failure to thrive persisted despite high calorie enteral feeds and correction of acidosis. The patient died at the age of 10 months.

This patient differs in two ways from previous reported cases. Firstly, liver histology varies from that reported by Eastham and colleagues, in whose patients the liver biopsy specimens all showed giant cell transformation.¹ It may be possible that the histology did not show typical features due to early timing of the biopsy. It is however possible that our case represents a phenotypic variant of the same disorder.

Secondly, we believe our case to be the first reported to have necrotising enterocolitis. No immunodeficiency has been identified in our patient, unlike others in the literature.^{2–4} It was noteworthy that the patient was receiving hyperosmolar formula feeds at the time of the

first episode. The occurrence of necrotising enterocolitis should warn clinicians of the potential risk of hyperosmolar feeds in severely growth retarded infants with acidosis, even when born at or after term.

R Howells, U Ramaswami

University Department of Paediatrics, Box 116,
Level 8, Addenbrooke's Hospital, Cambridge
CB2 2QQ, UK; rachel_howells@hotmail.com

References

- 1 **Eastham KM**, McKeirnan PJ, Milford DV. ARC syndrome: an expanding range of phenotypes. *Arch Dis Child* 2001;**85**:415–20.
- 2 **Lutz-Richner AR**. Familiäre Gallenangmissbildungen mit tubuläre Niereninsuffizienz. *Helv Paediat Acta* 1973;**28**:1–12.
- 3 **Papadia F**. Biliary malformation with renal tubular insufficiency in two male infants: third family report. *Clin Genet* 1996;**49**:267–70.
- 4 **Horslen SP**, Quarrell OW. Liver histology in the arthrogryposis multiplex congenita, renal dysfunction and cholestasis (ARC) syndrome: report of three new cases and review. *J Med Genet* 1994;**31**:62–4.

Echocardiography on the neonatal unit

Two dimensional, M mode and Doppler echocardiography is widely used by paediatric cardiologists to evaluate cardiac structure and function in neonates, infants, and older children. Anecdotally, it is also being used increasingly by neonatologists in the early newborn period.^{1,2} We have recently undertaken a postal questionnaire survey of 38 neonatologists working in referral centres to review current UK practice.

Thirty seven neonatologists responded to the questionnaire. Nineteen units performed more than 15 echocardiograms per month, six performed 10–15/month, and 12 performed less than 10/month. Echocardiograms were usually performed by paediatric cardiologists and/or neonatologists, but also occasionally by echocardiographic technicians. Neonatologists performed echocardiograms in two thirds of responding units. The commonest indications for echocardiography were: diagnosis/exclusion of congenital heart disease, assessment of ductal patency and haemodynamics, assessment of myocardial function, and assessment of pulmonary hypertension.

Only 12 (32%) units had 24 hour access to paediatric cardiology service on site; of those who did not, 18 units usually had access to these services on an on-call basis. Babies were transferred out of the neonatal unit for echocardiography in 13 (35%) responding units. Indomethacin was used to treat a symptomatic persistent ductus arteriosus (PDA)

following a purely clinical diagnosis in 15 (41%) units.

This survey shows that echocardiography on the neonatal unit is often performed by a neonatologist rather than a cardiologist, presumably reflecting the (lack of) availability of 24 hour on-site paediatric cardiology services, even in neonatal referral centres. In a considerable number of units babies are either transferred out of the neonatal unit for echocardiographic assessment or receive treatment for PDA without prior echocardiographic confirmation. Such situations are undesirable and reflect the need for greater access to echocardiography on the neonatal unit, a service that is likely to be provided increasingly by neonatologists themselves in the future.

Although several paediatric echocardiography courses are available, currently there is no formal accreditation process for neonatologists. We believe there is a need to evaluate the reliability of echocardiography in the hands of neonatologists in a systematic way and are currently conducting such a study.

S Moss, N V Subhedar

Liverpool Women's Hospital, Crown Street,
Liverpool L8 7SS, UK;
nvsubhedar_lwh@yahoo.com

References

- 1 **Evans N**. Echocardiography on neonatal intensive care units in Australia and New Zealand. *J Paediatr Child Health* 2000;**36**:169–71.
- 2 **Skinner JR**. Echocardiography on the neonatal unit: a job for the neonatologist or the cardiologist? *Arch Dis Child* 1998;**78**:401–7.

CORRECTION

In July's *Archives* (*Arch Dis Child* 2002;**87**:85), the correction mentioned "the following table": this was incorrect. The sentence should have read "The corrected amounts are listed in the revised figures". No table was missing, and readers can view the revised figures at www.archdischild.com, as mentioned in the original correction. We apologise for the error.

Please see the Archives website (www.archdischild.com) to view the corrected figures.