LETTERS

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Problems involved with the use of comforters

While I share many of the concerns expressed by Gill in his diatribe on dummies there are a number of concerns over the use of dummies 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 by 1857. Unfortunately by that 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 prolongeddummy sucking producing malocclusions in the primary dentition, these abnormalities are mainly self correct on cessation of the habit which is usually before 5 years of age.1

G B Winter
Emeritus Professor of Children’s Dentistry, 1 Hanfield Close, Elsine, Herts WD6 3JD, UK

References
1 Gill D. A diatribe on dummies. Arch Dis Child 2002; 87:222

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 of the limbs. 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 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,2 is not widely recognised. Suspected episodes of cataplexy have been reported in eight of 35,1 four of 25,3 and three of 17 patients with PWS. However, cataplectic manifestations are often “difficult to prove”, requiring a detailed history that is perhaps seldom available or elicited. We suggest that cataplexy may be relatively common in PWS and enquiries regarding its signs and symptoms 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; gcbv35@udcf.gla.ac.uk

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

Table 1 Conditions in which cataplexy is a recognised feature

<table>
<thead>
<tr>
<th>Familial isolated cataplexy</th>
<th>Narcolepsy–cataplexy syndrome</th>
<th>Hypnagogic/hypothalamic structural lesions</th>
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Kawasaki disease following meningoococcal sepsicaemia

We report a case of Kawasaki disease (KD) following meningoococcal sepsicaemia 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 maculopapular rash. Meningococcal sepsicaemia was diagnosed clinically and the boy was managed with fluid support and intravenous antibiotics. His recovery was complicated by developing respiratory syncytial virus infection 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 >15 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 but continued to 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 could 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; gcbv35@udcf.gla.ac.uk

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

References

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; avrramanan@hotmail.com

References
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 aminoacidaemia, 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 conjugated hyperbilirubinaemia, greatly in:

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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 caloric 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. 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.1 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 0QQ, UK; rachel_howells@hotmail.com

References

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 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 Subbedar
Liverpool Women’s Hospital, Crown Street, Liverpool L8 7SS, UK; nvsubahedr_lwh@yahoo.com

References

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.