LETTERS

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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 Gill in his diatribe on dummies there is a number of conditions (table 1) 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 1857. Unfortunately by the time the practice of dishing 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.

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References
1 Gill D. A diatribe on dummies. Arch Dis Child 2002; 86:222

Cataplexy in the Prader–Willi syndrome

We report cataplexy, sudden atomic episodes provoked by emotion, in three patients with Prader–Willi syndrome (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. 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, is not widely recognised. Suspected episodes of cataplexy have been reported in eight of 35; four of 25; 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 enquires regarding its signs and symptoms could always be made, especially in any patient with a past diagnosis of paroxysmal events.

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Table 1: Conditions in which cataplexy is a recognised feature

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<th>Condition</th>
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<td>Familial isolated cataplexy</td>
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<td>Narcolepsy–cataplexy syndrome</td>
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<td>Narcolepsy–sleep paralysis syndrome</td>
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Kawasaki disease following meningoococal septicaemia

We report a case of Kawasaki disease (KD) following meningoococal 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. Meningooococal 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 meningoccocus 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 caused by an infectious agent. These include geo-trophic superantigens may mediate KD is growing.

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References

www.archdischild.com
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, hooved 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. Plasma and urinai 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 caloric enteral feeds and correction of acidosis. The patient died at the age of 10 months. This patient differs in two ways from previous reports of ARC syndrome: an expanding range of phenotypes.

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 neonatalogists in the early newborn period. Two thirds of responding units perform echocardiograms in two thirds of responding units. The commonest indications for echocardiography were: diagnosis/exclusion of congenital heart disease, assessment of ductal patency and classification of septal defects. The commonest indications for echocardiography were: diagnosis/exclusion of congenital heart disease, assessment of ductal patency and classification of septal defects. The commonest indications for echocardiography were: diagnosis/exclusion of congenital heart disease, assessment of ductal patency and classification of septal defects.

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.