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 difficulties with his argument 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 the 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 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, Elsene, Herts WD6 3JD, UK

References

Cataplexy in the Prader–Willi syndrome

We report cataplexy, sudden atomic 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 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) DOB1*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,2,3 is not widely recognised. Suspected episodes of cataplexy have been reported in eight of 35,4 four of 25,5 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 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;
gbev55@udcf.gla.ac.uk

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

References

Kawasaki disease following meningoococcal septicaemia

We report a case of Kawasaki disease (KD) following meningoococcal 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. Meningoococal 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 meningoococcus 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 also seen in 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 We believe that our case shows the possible role of a meningoococal toxin acting 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 meningoococcus 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 Baldwin
Department of Paediatric Rheumatology, Royal Manchester Children’s Hospital, Manchester M8 8UX. Correspondence to: A V Ramanan, 508, 77 Elm Street, Toronto, Ontario M5G 1H4, Canada; avramanan@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 aminoaciduria, phosphaturia, and metabolic acidosis developed within a few hours of birth and severe conjugated hyperbilirubinaemia within two days. Liver investigations revealed similar findings to those previously reported, with a 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 function in neonates, infants, and older children. It is also being used function in neonates, infants, and older children. It is also being used function in neonates, infants, and older children. It is also being used function in neonates, infants, and older children.

In July’s Archives (Arch Dis Child 2002;87:85), the correction mentioned “the following table” 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.
ARC syndrome: an expanding range of phenotypes

R Howells and U Ramaswami

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