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 other conditions 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 1892. 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 correct on cessation of the habit which is usually before 5 years of age.1

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References

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 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,2 is not widely recognised. Suspected episodes of cataplexy have been reported in eight of 35,3 four of 25,4 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 may prove useful in the management of any patient with a past diagnosis of paroxysmal events.

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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. 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.5 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 mucosal 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 possibility that a meningococcal toxin could cause KD in the same fashion as toxic shock syndrome toxin producing Staphylococcus aureus is possible. If 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.

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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 arthrogyrosis 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 hyperosmolar formula feeds at the time of the original life (no organisms were identified in either faeces or blood at the time of the original admission). It may be possible that the 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 sepsis 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 previously reported cases. Firstly, liver 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.12 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.

**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 We have recently undertaken a postal questionnaire survey of 38 neonatologists working in referral centres in the UK currently conducting such a study.

In July’s Archives (Arch Dis Child 2002;87:85), the correction mentioned “the following table” as 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.
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Arch Dis Child 2002 87: 170
doi: 10.1136/adc.87.2.170

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