LEEPERS

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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 are a number of points which I would like to make. To begin with, there is no scientific data to suggest that milk aids in the future 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 dummying 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.

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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 frequent 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 within 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, 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 enquiries regarding its signs and symptoms should always be made, especially in any patient with a past diagnosis of paroxysmal events.

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References

Kawasaki disease following meningococcal septicemia

We report a case of Kawasaki disease (KD) following meningococcal septicemia 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 septicemia 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 (PCR) 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 25 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 could always be made, especially in any patient with a past diagnosis of paroxysmal events.

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conjugated hyperbilirubinaemia, greatly in-
an N-acetylglucosamine:creatinine ratio of
generalised aminoaciduria, phosphaturia, and
also present. Metabolic acidosis developed
Pakistani parents, was noted shortly after
A female infant, born to consanguineous
range of phenotypes
ARC syndrome: an expanding
formation.
specimens all showed giant cell trans-
varies from that reported by Eastham and
died at the age of 10 months.
markedly underweight despite high calorie enteral
or recurrent episodes). Repeated episodes of
the blood or faeces at the time of the original
colitis occurred during the first two months of
present.
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 in-
creased alkaline phosphatase, but normal γ
 glutamyltransferase. 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 enterocoi-
litis 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 calorie enteral
feeds and correction of acidosis. The patient
died at the age of 10 months.
This patient differs in two ways from previ-
ous cases. Firstly, liver histology varied from
that reported by Eastham and
colleagues, in whose patients the liver biopsy
specimens all showed giant cell trans-
formation. It may be possible that the histol-
yogy 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. 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 sever-
ely growth retarded infants with acidosis,
even when born at or after term.

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Echocardiography on the
neonatal unit
Two dimensional, M mode and Doppler echo-
cardiography is widely used by paediatric car-
diologists 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.

Two dimensional, M mode and Doppler echo-
cardiography 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.

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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 fig-
ures at www.archdischild.com, as mentioned in
the original correction. We apologise for the error.

Please see the Archives website (www.archdis-
child.com) to view the corrected figures.