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 other issues (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 1927. Unfortunately by this time the practice of dishing the dummy in a variety of sweetening agents to make it a more efective 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 efect 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.2

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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) who have acquired the condition. There appears to be no family history of cataplexy in any of these families.

Three patients from two families have been studied. The boys, aged 2, 10 and 13 years, all have psychomotor retardation, hyperphagia, and obesity, and each has a deletion showing the presence of the insulin-like growth factor 2 (IGF2) gene. There is no other evidence for the presence of Carney complex (1).

The association of these patients with PWS and cataplexy, though described previously,3,4 is not widely recognised. Suspected episodes of cataplexy have been reported in eight of 35,5 four of 25,6 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.

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References


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. 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 efect. 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.2 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.3,4 We believe that our case shows the possibility that a meningoococal 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 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.

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References

conjugated hyperbilirubinaemia, greatly in
findings to those previously reported, with
>1000. Liver investigations revealed similar
generalised aminoaciduria, phosphaturia, and
gated hyperbilirubinaemia within two days.
also present. Metabolic acidosis developed
hips and rocker bottom feet. Icthyosis was
prominent occiput, beaked nose, high arched
Pakistani parents, was noted shortly after
A female infant, born to consanguineous
range of phenotypes
ARC syndrome: an expanding
range of phenotypes
A female infant, born to consanguineous
features, including prominent occiput, boaked nose, high arched
and arthrogryposis with dislocated hips and rocker bottom feet. Icthyosis was
also present. Metabolic acidosis developed
R wells, Philadelphia.
N-acyetylglucosamine:creatinine ratio of
Liver investigations revealed similar
findings to those previously reported, with
conjugated hyperbilirubinaemia, greatly in
increased 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 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 Septicemia occurred later. Masked 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 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. 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 severe
ly 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.1 We have recently under-
taken 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. Neonatol-
gists performed echocardiograms in two
thirds of responding units. The commonest
indications for echocardiography were:
diagnosis/exclusion of congenital heart dis-
ease, 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 symp-
tomatic persistent ductus arteriosus (PDA)

following a purely clinical diagnosis in 15
(41%) units.
This survey showed 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 echocardi-
ographic 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 themselves in
the future.
Although several paediatric echocardi-
ography courses are available, currently there is
no formal accreditation process for neonolo-
gists. 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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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.archdischild.com) to view the
corrected figures.

www.archdischild.com
ARC syndrome: an expanding range of phenotypes

R Howells and U Ramaswami

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Updated information and services can be found at:
http://adc.bmj.com/content/87/2/170.4

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References

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