LETTERS

If you have a burning desire to respond to a paper published in ADC or FeN, why not make use of our “rapid response” option?
Log on to our website (www.archdischild.com), find the paper that interests you, click on “full text” and send your response by email by clicking on “submit a response”.
Providing it isn’t libellous or obscene, it will be posted within seven days. You can retrieve it by clicking on “read eLetters” on our homepage.
The editors will decide, as before, whether to also publish it in a future paper issue.

C difficile induced pneumatosis intestinalis in a neutropenic child

A 4 year old boy presented with a 24 hour history of fever, cramping central abdominal pain with distension, and bloody diarrhoeal stools. He had developed acute myeloblastic leukaemia at the age of 1. He eventually required a matched unrelated graft which engrafted poorly, and subsequently developed graft versus host disease (GVHD). He was currently neutropenic.

Temperature was 38.5°C, pulse 150/min; he had abdominal distension and tenderness but no ascites.

An abdominal film (fig 1) revealed dilatation of the colon with gaseous linear tramlining of the bowel wall consistent with pneumatosis intestinalis (PI). Stools were positive for Clostridium difficile toxin A. PI resolved with bowel rest, intravenous fluids, meropenem, and metronidazole but he later died of relapsed leukaemia.

PI is described in children in Crohn’s disease,1 ulcerative colitis,2 leukaemia,3 trauma,4 HIV,5 and GVHD after BMT.6 C difficile, pseudomembranous colitis, and PI are reported in an immunocompetent adult patient.7 Conservative management is usually successful but acidosis and portal gas are associated with a poor outcome.8

Figure 1 Abdominal film showing dilatation of the colon and PI.

References
Table 1 Conditions in which cataplexy is a recognised feature

<table>
<thead>
<tr>
<th>Condition</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Familial isolated cataplexy</td>
<td></td>
</tr>
<tr>
<td>Norrie’s disease</td>
<td></td>
</tr>
<tr>
<td>Niemann-Pick disease type C</td>
<td></td>
</tr>
<tr>
<td>Caffin-Lowry syndrome</td>
<td></td>
</tr>
<tr>
<td>Narcolepsy-cataplexy syndrome</td>
<td></td>
</tr>
<tr>
<td>Fontanomeditullary/hypothalamic structural lesions</td>
<td></td>
</tr>
</tbody>
</table>

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 he makes that require amplification or correction. The first patent on the 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 d typography 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 shared with those of other infectious diseases, for example, adenoviral infection and scarlet fever. Staphylococcus, 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. 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 of KD 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 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 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.

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, hooked 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 with normal alkaline phosphatase, but normal conjugated hyperbilirubinaemia, greatly findings to those previously reported, with an N-acetylglucosamine:creatinine ratio of 1:1. Renal tubular acidosis was manifest by generalised aminoaciduria, phosphaturia, and 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 hyperbilirubinemia, greatly 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 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 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 hypomolar 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.

R Howells, U Ramaswami
University Department of Paediatrics, Box 116, Level 8, Aldenbrooke’s Hospital, Cambridge CB2 2QQ, UK; rachel_howells@hotmail.com

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.1,2 We have recently undertaken 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. Neonatologists performed echocardiograms in two thirds of responding units. The commonest indications for echocardiography were: diagnosis/exclusion of congenital heart disease, 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 symptomatic persistent ductus arteriosus (PDA) following a purely clinical diagnosis in 15 (41%) units.

This survey shows 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 echocardiographic 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 echocardiography 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.

S Moss, N V Subhedar
Liverpool Women’s Hospital, Crown Street, Liverpool L8 7SS, UK; nvsuhbedar_lwh@yahoo.com

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.