Salicylate poisoning in dermatological treatment

Sir,—We read with interest the article on the successful treatment of a harlequin fetus.1 We note that the baby was initially treated with topical liquid paraffin containing 1% salicylic acid every three hours. This resulted in significant percutaneous salicylate absorption and systemic toxic manifestations. We have had similar problems with salicylate toxicity after the topical application of salicylate containing preparations for the management of skin disorders in two children.

One was an infant born at 38 weeks’ gestation and weighing 2500 g. His skin was covered by a collodion like membrane which within 24 hours started to crack and peel in large plaques. Topical 2% salicylic acid in aqueous cream was applied every three to four hours. On day 3 the baby vomited feeds and had a persistent metabolic acidosis. His salicylate concentration was 3 mmol/l. Topical salicylate treatment was stopped and a high fluid intake ensured by intravenous administration. He made a complete recovery.

The second patient was a 12 year old boy with severe ichthyosis. Treatment was started with topical 2% salicylic acid in simple cream applied to the whole body twice daily. The salicylate concentration increased to 5% on day 3 of treatment and 10% on day 5. On day 8 he developed symptoms of salicylate toxicity. His blood salicylate concentration was 3.5 mmol/l. Topical salicylate treatment was stopped. Intravenous fluids and bicarbonate were given and complete clinical and biochemical recovery was achieved after two days.

Topical salicylate acid in paraffin or simple cream is supported by several authors as treatment in a number of skin disorders.2,3 Our two cases and the one described by Ward and Jones1 show that significant percutaneous salicylate absorption can occur not only in newborn infants but also in older children, especially when salicylate preparations of increasing strength are used. We suggest that all topical salicylate treatment should be routine ly monitored with salicylate blood concentrations especially during the initial few days after onset or after any changes in treatment.

P GALEA
K M GOEL
Royal Hospital for Sick Children, Yorkhill,
Glasgow G3 8ST

Lichen sclerosus

Sir,—In response to the article on lichen sclerosus by Dr Berth-Jones et al, we would like to take this opportunity to support the link between lichen sclerosus and sexual abuse.1 I disagree that the chances of these two problems occurring together would seem very remote. I have had my care over 400 cases of lichen sclerosus and am well versed in the appearance of lichen sclerosus lesions at sites of trauma—for example, where wrist bands and bra straps cause friction. What is more, some patients report extra genital lesions occurring at sites of trauma—for example, oven burns.

It is my feeling that vulval trauma may precipitate vulval lichen sclerosus. I have seen two cases, a girl aged 4 and another aged 6, with definite lichen sclerosus of the vulva and they have also been sexually abused. Another 8 year old girl with a five year history of anogenital lichen sclerosus had also probably been sexually abused at the age of 3.

It is therefore important to accept that lichen sclerosus does occur at sites of trauma and that the trauma may be sexual abuse.

C J HARRINGTON
Royal Hallamshire Hospital,
Sheffield S10 2JF

1 Berth-Jones J, Graham-Brown RAC, Burns DA.

Fragile X mental retardation

Sir,—It is not only in the context of mental retardation without dysmorphic features that the general paediatrician should consider fragile X syndrome.1 It can present as a connective tissue dysfunction which mimics Ehlers-Danlos or Marfan’s syndrome with joint hyperextensibility so severe as to lead to dislocation, high arched palate, and cutis hyperelastica. There may also be mild dysmorphic features such as epicardiac folds and large malformed auricles. Cardiovascular features include mitral valve prolapse and dilatation of the ascending aorta.2 We agree that fragile X is an important diagnosis to exclude, and accept the need to be selective in deciding which patients should be screened, but would suggest that the screening criteria might usefully be broadened to include patients with connective tissue or cardiovascular abnormalities with no specific developmental delay. We also consider that full cardiac assessment should be performed in proved cases, because of the importance of diagnosing mitral valve prolapse for the need for appropriate prophylaxis against infective endocarditis in these patients.3

R FRANKLIN
A BUSH
Department of Paediatric Cardiology,
Brompton Hospital,
Fulham Road,
London SW3 6HP

Lichen sclerosis and sexual abuse

Sir,—Dr Berth-Jones et al refer to the potential for misdiagnosis of severe anogenital lichen sclerosus as sexual abuse.1 They state that ‘there is absolutely no evidence to support any link between lichen sclerosus and sexual abuse’ and the ‘chances of these two problems occurring together would seem very remote indeed’. We wish to report such a case.

We have treated 20 girls aged 4–11 years with lichen sclerosus over a two and a half year period. One girl, aged 7–5 years, presented with a four month history of intense diurnal frequency of micturition and wetting, and a sore vulva. The appearances were suggestive of early lichen sclerosis with patchy pallor and inflammation, plus minor fusion of the labia minora. Subsequently the changes became much more marked with characteristic thickened sclerotic white plaques, wrinkling, areas of acute inflammation, excoriation, and purpura. Substantial improvement was secured with topical clobetasone. There have, however, been psychological problems and concern about possible abuse has persisted.

The girl has a complex social background, her stepfather being a schedule 1 offender (physical injury in a different family). Careful social and psychological investigation initially failed to support a diagnosis of sexual abuse by the next year, coincident with worsening of her lichen sclerosus, she developed frequent encopresis, not associated with faecal retention. She eventually disclosed sexual abuse by her stepfather and much of her story was validated independently by her young brother.

A diagnosis of lichen sclerosis does not exclude sexual abuse, the diagnosis of which can, in some cases, be supported by non-specific statements by the child than upon physical findings. There is the additional possibility that chronic trauma in the presence of low grade skin infection may on occasion lead to the changes of lichen sclerosus.

B L PRIESTLEY
S BLEHLEN
The Children’s Hospital,
Western Bank,
Sheffield S10 2TH


Fetal ascites: an unusual presentation of Niemann-Pick disease type C

Sir,—We read with interest the paper of Maconochie et al as we have recently seen a case of Niemann-Pick disease type C with an
almost identical presentation. The baby was born to a 41 year old Pakistani woman, gravida 12 para 6. The parents were first cousins. The unfortunate obstetric history included four first trimester spontaneous abortions and a 29 week stillbirth. Another baby, an anencephalic, died in the neonatal period. Finally a girl, born at full term, died at 4 weeks of age after a three day illness characterised by fever, convulsions, and hepatosplenomegaly. Postmortem examination showed excessive deposition of sphingomyelin in the spleen.

In a subsequent pregnancy haemorrhage was noted at 26 weeks gestation. An ultrasound scan showed fetal ascites but no structural abnormalities. The baby was born at 34 weeks gestation weighing 2590 g. There was gross ascites and hepatosplenomegaly. Jaundice was evident on the first day of life (total bilirubin concentration 111 μmol/l), conjugated bilirubin 83 μmol/l). During the next two weeks there was clinical and laboratory evidence of continuing hepatic dysfunction, and the baby died from liver failure at 19 days.

Investigations excluded other causes of fetal ascites and neonatal liver disease. Peripherial leucocyte enzyme assays were normal. Bone marrow aspirate showed foamy histiocytes. A percutaneous postmortem splenic biopsy specimen showed accumulation of sphingomyelin. Partial deficiency of sphingomyelinase was found in cultured skin fibroblasts. The fibroblasts showed appreciably reduced intracellular esterification of exogenous lipid-protein derived cholesterol, consistent with Niemann-Pick disease type C.

Clearly this disease should be considered in the differential diagnosis of fetal ascites and conjugated hyperbilirubinaemia in the neonate. Whether, in our case, any of the previous abortions or the stillbirth was affected is speculative. Undoubtedly, increasing experience with ultrasound will assist in antenatal recognition of the disease. With regard to postnatal investigation our case emphasises the importance of skin fibroblast culture. In addition to studying cholesterol esterification, partial deficiency of fibroblast sphingomyelinase at a discrete chromosome level may be found, whereas peripheral leucocyte levels are typically normal.

D J MANNING
R G PEARSE
Neonatal Intensive Care Unit,
Jessop Hospital for Women,
Leegoe Grove Road,
Sheffield S7 7RE


The school health service through parents’ eyes

SIR,—We read with interest the personal paper on the school health service by Dr Perkins and would like to make some comments.

It is important to have a clearly defined function communicated to all users of the service. At the moment many teachers misunderstand and have inappropriate expectations of what we should achieve, which leads to disappointment and downgrading of the service in teachers’ eyes that is communicated to parents.

Inadequate facilities are provided in many schools, which must communicate lack of interest in the service to parents and children. Difficulties arise because of rules that have developed for historical reasons—for example, we have little access to simple investigation and also most things must be referred back or through general practitioners before action can be taken. This apparent lack of authority even in areas of special interest must convey low status.

Certainly communication could be improved by having a named doctor and nurse visiting the school regularly throughout the school year. In order to do the job efficiently we must have a clearly defined role about which all users have appropriate expectations. We should be able to fulfil that role in as direct a way as possible to be seen as having appropriate authority. Above all it must be seen as a useful service to parents and teachers and children. These are the personal views of the signatories.

P BLACKWELL,
G N DAVIES,
C RAVINDRANATH
Community Services (Child Health),
Grimby District General Hospital,
Grimby, South Humberside DN33 2BA


Chlamydia trachomatis in infants: a prospective study

SIR,—We were interested to read the paper by Preece et al dealing with chlamydial infection in children but feel that some important points may have been omitted. In infants, Chlamydia trachomatis infects multiple sites including the rectum and vulva. Screening from conjunctiva and pharynx alone may underestimate transmission from infected mothers. Where a more complete screening practice has been used, longer carrier status has been found. Failure to recognise the reality of genital carriage, and especially the declaration that a child is free from infection on the basis of limited investigation, carries serious social and legal implications, as some practitioners regard the presence of vaginal chlamydia as an indicator of sexual abuse. Topical treatment of chlamydial conjunctivitis does not clear the child of the pathogen at other sites and leaves it vulnerable to completely avoidable respiratory infection. Finally, we wish to challenge the assumption that prenatal screening for chlamydia is not cost effective. No account has been taken of an important part of the equation, namely the pregnant woman herself and of the morbidity she may avoid by receiving treatment earlier rather than later.

K POWER,
G M MIDDLETON,
G E FORSTER
The Whitchapel Clinic,
The London Hospital,
Whitechapel,
London E1 1B


Drs Preece, Anderson, and Thompson comment: Our recent paper explored the postnatal morbidity in infants associated with perinatal transmission of maternal C trachomatis infection. While it is interesting to document the persistence of rectal and vaginal carriage this was not our primary aim, and we agree that studies to document such carriage and persistence of infection are required to distinguish from later acquisition. We agree that in clinical practice, oral erythromycin is now the treatment of choice in all chlamydial infections in children.

Finally, our statement about cost effectiveness of screening for maternal infection was based on the subsequent perinatal and postnatal morbidity in infants. The practicality and effectiveness of such a screening programme is discussed in more detail in our companion paper. We feel that our data do not support such a programme to protect infants from chlamydial infection for the reasons outlined.

With reference to maternal infection, pregnancy may provide an opportunity to screen for C trachomatis infection in adult women. However, there may be other strategies that will be more logical and cost effective in the management of adult infection. As Drs Powell et al point out we do not address this issue, which needs more extensive and designed studies of the epidemiology of C trachomatis infection in adult women.


Egg and breast milk based nitrogen sources compared

SIR,—In their paper comparing the effects of intravenous Vamin 9 glucose and Vaminolact on the plasma amino acid profile, Punits et al discuss the possible neurotoxic effects of raised phenylalanine concentrations. They conclude that Vaminolact is preferable to Vamin 9 glucose because it was associated with less marked increases in plasma phenylalanine concentrations. In support of this view they refer to findings in early treated children with phenylketonuria showing that intellectual outcome is inversely associated with average phenylalanine control during treatment (as well as age at the start of treatment). I doubt whether it is appropriate to liken the effects of short term changes in phenylalanine in children receiving Vamin 9 glucose (which also need more specific therapeutic support such as taking tyrosine) to the effects of longterm phenylalanine control in phenylketonuria, which is associated with decreased rather than increased concentrations of tyrosine.

It may be useful to consider the likely effects of the raised phenylalanine concentrations on the uptake of amino acids into the brain. This will depend upon the plasma concentrations of the other large neutral amino