almost identical presentation.¹ 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 hydrops 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 $\mu\text{mol/l}$, conjugated bilirubin 83 $\mu\text{mol/l}$). 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. Peripheral 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 lipoprotein 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 may be found, whereas peripheral leucocyte levels are typically normal.

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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.¹

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.

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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.¹ In infants, *Chlamydia trachomatis* infects multiple sites including the rectum and vulva.² 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 shown.³ Failure to recognise the reality of genital carriage, and especially the declaration that a child is free from infection on the basis of limited sampling, carries serious social and legal implications, as some practitioners regard the presence of vaginal chlamydia as an indicator of sexual abuse.⁴ 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 antenatal 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.

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2 Schacter J, Grossman M, Holt J, *et al*. Infection with Chlamydia trachomatis: involvement of multiple anatomic sites in neonates. *J Infect Dis* 1979;139:232-4.

3 Bell TA, Stamm WE, Kuo C-C, *et al*. Chronic Chlamydia trachomatis infections in infants. In: Oriel D, Ridgway G, Schacter J. *Chlamydial infections*. Cambridge: Cambridge University Press, 1986:305-8.

4 Ingram DL, White ST, Occhiuti AR, *et al*. Childhood vaginal infections: association of chlamydia trachomatis with sexual contact. *Pediatr Infect Dis* 1986;5:226-9.

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.¹

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.² 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 specifically designed studies of the epidemiology of *C trachomatis* infection in adult women.

1 Rettig PJ. Chlamydial infections in paediatrics: diagnostic and therapeutic considerations. *Pediatr Infect Dis* 1986;5:158-62.

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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, Puntis *et al* discuss the possible neurotoxic effects of raised phenylalanine concentrations.¹ 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).² I doubt whether it is appropriate to liken the effects of short term changes in phenylalanine in children receiving Vamin 9 glucose (which also caused rises in other amino acids including tyrosine) to the effects of longterm phenylalanine control in phenylketonuria, which is associated with decreased rather than increased concentrations of tyrosine.

It may be more useful to consider the likely effects of the raised phenylalanine concentrations on the uptake of amino acids into the brain.³ This will depend upon the plasma concentrations of the other large neutral amino

acids, as well as the phenylalanine concentrations and the transport characteristics of the blood-brain barrier. Although we know rather little about the latter in sick, human infants, comparison of the ratios of phenylalanine to the sum of the concentrations of tyrosine, tryptophan, and the branched chain amino acids in infants receiving breast feeds, Vamin 9 glucose, and Vaminolact provides a comparative measure of phenylalanine uptake in the three groups of infants.

Using the average plasma amino acid values in breast fed infants⁴ and in those given Vamin 9 glucose and Vaminolact¹ the ratios were 0.1, 0.35, and 0.27 respectively. The data suggest that phenylalanine uptakes into the brain in infants receiving Vamin 9 glucose and Vaminolact were similar, and that both products are likely to result in higher uptake of phenylalanine than breast feeding. In those infants receiving Vamin 9 glucose who had particularly high phenylalanine concentrations the ratio would be higher only if the concentrations of tyrosine and other amino acids failed to rise with phenylalanine. The increased ratio in infants receiving Vaminolact was largely accounted for by their lower tyrosine concentrations, and the ratios of tyrosine to the other large neutral amino acids (0.2, 0.2, and 0.06 respectively) suggest that tyrosine uptake was lower in infants receiving Vaminolact than in breast fed infants; in infants receiving Vamin 9 glucose it was similar. However, if tryptophan is considered the reverse situation pertains (ratios 0.13, 0.04, and 0.12 respectively).

These simplified ratios cannot be expected to give an accurate prediction of brain amino acid uptake but do provide useful comparisons. In the present state of knowledge it seems wisest in infants receiving intravenous amino acids to aim at plasma amino acid profiles similar to the averages observed in well nourished breast fed neonates. By this criterion Vaminolact requires the addition of tyrosine (and a reduction of threonine) before it can be said to be 'better' than Vamin 9 glucose.

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- 2 Smith I, Beasley M. Intelligence and behaviour in children with early treated phenylketonuria. A report from the MRC/DHSS Phenylketonuria Register. *European Journal of Clinical Nutrition* 1989;43:1-5.
- 3 Pardridge WM. Brain metabolism: a perspective from the blood brain barrier. *Physiol Rev* 1983; 63:1481-535.
- 4 Wu PYK, Edwards N, Storm MC. Plasma amino acid pattern in normal breast fed infants. *J Pediatr* 1986;109:347-9.

Drs Puntis, Ball, Preece, et al comment:

We thank Dr Smith both for her remarks on our paper and her interesting speculations. While we do not disagree with her comments, there are several points which should be raised in reply. We do feel that when examining available amino acid sources it is reasonable to focus attention on those amino acids such as phenylalanine that under certain circumstances are known to have deleterious clinical effects at high concentrations. Our data allow for certain generalisations to be made when comparing the two amino acid solutions, but we feel that Dr Smith is rather brave to make inferences about phenylalanine uptake by the brain in our patients. We did not conclude that Vaminolact is preferable to Vamin 9 glucose simply because it was associated with less marked increases in plasma phenylalanine, but because, in general, the plasma concentrations of the various amino acids (including phenylalanine and tyrosine) in Vaminolact fed infants tended to be closer to the mean described in breast fed infants than those in the Vamin 9 glucose fed group. By this criterion Vaminolact appears 'better' than Vamin 9 glucose. Whether or not we should be attempting to reproduce the amino acid profiles found in breast fed infants is a moot point and the ideal composition for amino acid sources remains a subject for debate.

Aneurysms of the vien of Galen

SIR,—We would like to add a small but

important point to the discussion of aneurysms of the vein of Galen by O'Donnabhain and Duff.¹ We would agree that surgery for the aneurysm itself is both difficult and seldom successful but may be a necessary part of the management of the infant with high output cardiac failure. Infants with hydrocephalus comprise a different clinical group who should undergo shunting as the initial surgical procedure. Having stabilised these patients with hydrocephalus it is important to be aware that further surgery may not be necessary as there are a number of cases in the literature in which the aneurysm has subsequently undergone spontaneous thrombosis.

All the reported cases of a thrombosed aneurysm of the vein of Galen presented initially with hydrocephalus, although one case presented with subarachnoid haemorrhage in addition to hydrocephalus. Spontaneous thrombosis does facilitate the successful surgical removal of the aneurysm, the first such case being a 5 month old boy described by Heinz *et al.*² Calcification of the aneurysm occurs in 14% of patients and the aneurysm may calcify completely. We have recently described a 3 month old boy with hydrocephalus whose aneurysm of the vein of Galen was completely thrombosed and calcified by 3 years.³ We now consider him 'cured' after ventriculoperitoneal shunting only and we would advocate that early surgery, if possible, be confined to the treatment of hydrocephalus.

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- 3 Chapman S, Hockley AD. Calcification of an aneurysm of the vein of Galen. *Pediatr Radiol* 1989;19:541-2.