

cryptorchid testes. In: Fonkalsrud ED, Mengel W, eds. *The undescended testis*. Chicago: Year Book Publishers, 1981:57-74.

7 Hadziseimovic F. *Cryptorchidism—management and implications*. Berlin: Springer-Verlag, 1983.

Drs Rao, Wilkinson, and Benton comment:

We are grateful to Dr Goh and Dr Hutson for their comments. We agree that testicular screening should start at the neonatal examination. Testicular screening in our district does begin with the neonatal examination—as mentioned in our article. What we discovered as a result of our audit was that a major proportion of cases of undescended testes were not detected at the neonatal stage but at the school entry examination at 5 years of age. We are therefore compelled to recommend that a 'final' screen at around 5 years is continued at present. Detection at 5 years may not be ideal but it is better than no detection at all.

A programme of systematic and uniform developmental checks for all boys under 5 years is being introduced throughout the district, in keeping with the national directive on child health surveillance.¹ If the new programme achieves the desired effect of early detection, testicular screening at school entry may become unnecessary in the future.

The purpose of our audit was to assess the effectiveness of local screening for undescended testes. To achieve this it was necessary and appropriate to examine the involvement of all the professional groups who contributed to the developmental surveillance of those aged 0-5, including health visitors. General practitioners were unaware that health visitors were not required to check testes, as indeed were two out of the three authors of this study. This fact only became generally recognised as a result of the audit.

The ineffectiveness of a programme is so often caused by problems which seem completely obvious—in retrospect. The purpose of audit is to reveal these problems, however self evident they may appear, as frequently this is the only way to motivate change in an organisation.

1 Department of Health and Social Security. *Child health surveillance: implementation of the new GP contract*. (EL (90)P/36.) London: Department of Health, 1990.

Factors associated with chronic lung disease in preterm infants

SIR,—Professor Cooke's assertion that . . . 'parenteral lipid emulsions should be restricted to older preterm infants, or to those without respiratory symptoms'¹ is not warranted by the original data he presents or the published work he cites.

I do not dispute that his analysis demonstrates *association* between chronic lung disease and parenteral lipid therapy but correlation is a notoriously weak proof of *cause*. In this case why cannot one contend that the longer a baby is ventilated, the more likely he is to receive parenteral lipid? The conclusions of such an analysis also depend on which variables one includes or, more importantly, ignores. The vitamin A intake (lowest in the infants receiving most parenteral nutrition) could exemplify the latter category.

In support of his hypothesis, Professor Cooke cites a small randomised study² as demonstrating that lipid-treated infants ' . . . showed a significant excess of chronic lung disease . . .', though the difference in incidence

of bronchopulmonary dysplasia (BPD) between the lipid-treated and control groups was not significant by conventional criteria (lipid group 14/20, control group 10/22, $p < 0.1$). Admittedly the duration of respiratory support and supplemental oxygen treatment was greater in the lipid-treated group ($p < 0.05$), but the alveolar-arterial oxygen tension difference was somewhat greater in these infants *before* randomisation. It is true that a randomised study of *vitamin A supplementation* demonstrated a reduced incidence of both BPD and retinopathy,³ but avoidance of lipid emulsion could only exacerbate fat soluble vitamin deficiency.

Fat is a major nutrient which provides about half the non-protein energy intake in most parenteral nutrition regimens and it could not be omitted without constraining amino acid intake too. There is a temptation to increase glucose intake to overcome this problem but this increases both oxygen consumption and carbon dioxide production, further stressing a ventilated immature baby. Lipid emulsions also supply essential fatty acids, important for growth of the nervous system and the synthesis of prostaglandins and leukotrienes. An infant of 26 weeks' gestation (1% fat by weight) would rapidly develop deficiency.

I do not dismiss the possibility that some relationship between chronic lung disease and lipid infusion exists, but would argue that randomised studies of dosage and solution composition are the way forward, not draconian proscription. Neonatal intensive care certainly is a 'continuing experiment'¹ but let us make sure it is adequately controlled.

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- 1 Cooke RWI. Factors associated with chronic lung disease in preterm infants. *Arch Dis Child* 1991;66:776-9.
- 2 Hammerman C, Aramburo MJ. Decreased lipid intake reduces morbidity in sick premature neonates. *J Pediatr* 1988;113:1083-8.
- 3 Shenai JP, Kennedy KA, Chytil F, Stahlman MT. Clinical trial of vitamin A supplementation in infants susceptible to bronchopulmonary dysplasia. *J Pediatr* 1987;111:269-77.

SIR,—We read with interest Professor Cooke's paper on factors associated with chronic lung disease (bronchopulmonary dysplasia, BPD) in preterm infants.¹ His retrospective study shows an association between lipid use in the first 21 days of life and the incidence of chronic lung disease. Details of lipid use are scanty—there is no information on total duration of lipid emulsion, time of start of emulsion in other than 10 day bands, whether lipid emulsion was reduced as enteral feeds were increased, how often hypercholesterolaemia or hypertriglyceridaemia occurred and if lipid emulsion was reduced accordingly, how many babies received 4 g/kg/day of lipid emulsion (20 ml/kg/day of 20% Intralipid) and so on. An association was also noted between BPD and both gestational age and septicaemia. However, association does not imply causation, and can only generate a hypothesis to be tested.

The association noted from this retrospective study should be investigated by a prospective randomised controlled study, comparing the current nutritional package in use in the Mersey unit for babies of 24 to 30 weeks' gestation with an intervention in which lipid is

used only in babies of this gestational age range who have no respiratory disease. With the quoted (1988/9) rate of BPD in the unit of 60%, an intervention to reduce the rate to 40% would require a sample size of 126 babies in each group for the study to have a power of 90% and significance level of 0.05. As 40% of babies die or are breathing air by 28 days, the total sample size would need to be 420 babies. In a single centre study, this would require 10 years to complete at current rates of preterm birth in the Mersey region. If the intervention reduced the rate of BPD to 25%, a total sample size of 123 babies would be required.

Without such a study, we believe that the last paragraph of this paper is invalid. Professor Cooke recommends that parenteral lipid should be restricted to older preterm infants, or those without respiratory symptoms in the belief that the risks of an increased incidence of BPD outweigh 'theoretical gains made from early lipid infusion'. We believe that the balance of risk and benefit should be reversed, and that the harm caused by lipid deprivation may greatly outweigh any theoretical reduction in the incidence of BPD. Undernutrition is a major problem in babies who develop BPD, with late use of lipid emulsion and frequent periods of lipid-free alimentation being among the main reasons.² Lipid emulsion is energy rich and thus helps avoid the risk of fluid overload while achieving the desired energy goal. Moreover, essential fatty acid deficiency quickly develops in preterm babies who are deprived of lipid.³ Fat soluble vitamins are usually supplied as an additive to parenteral lipid emulsion. There is increasing speculation that vitamin A deficiency affects the repair of injured squamous epithelium in preterm lungs, and thus contributes to the development of BPD.⁴ We would welcome a multicentre study to test Professor Cooke's hypothesis that lipid emulsion use causes an increased incidence of BPD.

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- 1 Cooke RWI. Factors associated with chronic lung disease in preterm infants. *Arch Dis Child* 1991;66:776-9.
- 2 Wilson DC, McClure G, Halliday HL, Reid MMcC, Dodge JA. Nutrition and bronchopulmonary dysplasia. *Arch Dis Child* 1991;66:37-8.
- 3 Cooke RJ, Zee P, Yeh Y-Y. Essential fatty acid status of the premature infant during short-term fat-free parenteral nutrition. *J Pediatr Gastroenterol Nutr* 1984;3:446-9.
- 4 Shenai JP, Chytil F, Stahlman MT. Vitamin A status of neonates with bronchopulmonary dysplasia. *Pediatr Res* 1985;19:185-9.

Professor Cooke comments:

Chronic lung disease (CLD) in preterm infants results from lung damage and abnormal healing caused by a wide variety of factors. My study is aimed to discover the cause for a step change in the incidence of CLD in our unit in 1987/8. The only variable examined that satisfactorily explained such a change was the early use of parenteral lipid, introduced at this time because of concern about poor postnatal weight gain in patients with CLD. It would of course be possible to explore this variable further in more detail and it is quite possible that only some infants, probably the sickest, are adversely affected by very early lipid infusion.

Dr Williams suggests that length of ventilation could account for the association. When duration of ventilation during the first month of life is entered in the regression, it does not account for the correlation seen. This variable was not included in the final paper because, as explained, it both may cause and be caused by CLD. Vitamin A at 800 U per day was used through the period of the study either in oral or intravenous supplements. Dr Williams is somewhat selective in his quotations from the paper of Hammerman and Aramburo, in omitting to mention that 7/20 lipid-treated infants went home on oxygen compared with 0/22 of the controls.

I agree with Dr Wilson and his senior colleagues when they state that association does not necessarily imply causation, and that my cohort study can only generate a hypothesis to be tested. However, they robustly counter my proposition that the 'theoretical gains made from early lipid infusion may be outweighed by an increase in CLD' with their own small case control study showing an association between low energy intake and CLD. Surely what is sauce for the goose is good for the gander? Also the two sets of findings are not incompatible.

Dr Wilson and colleagues offer me advice on the size of randomised controlled trial which they now feel should be conducted. Trial size calculations are based on prior assumptions about the size of the effect of the intervention and the incidence of the target condition in the population. Their calculations are correct for their assumptions, although they give no reason for choosing a reduction of CLD from 60% to 40% as the effect sought. The odds ratio for lipid use in the first 21 days was 8.1 in the study reported, suggesting that a considerably smaller study may be needed. I certainly support randomised trials and further studies are currently underway.

While there is good reason to believe that better nutrition both before and after birth may make an important contribution to improved recovery from lung disease in the preterm infant, there is also good reason to believe that early use of lipid infusions may not be the best way of achieving this.

The firmness with which this was expressed in my concluding remarks is at least partly due to editorial changes, so perhaps they should share the reputation for draconian proscription!

Aetiology of malocclusion of the teeth

SIR,—In his excellent article about the aetiology of malocclusion of the teeth,¹ Professor Leighton does not mention the most common cause of malocclusion we see on this side of the Atlantic, at least in an inner city population of New York City: prolonged bottle feeding and/or pacifier use.

We see many children over the age of 1 year who already show the typical V shape of the incisors in the maxilla, as compared with the U shape of the mandible, indicating the above, or to a much lesser extent, thumbsucking. We, therefore, encourage our housestaff very strongly to promote discontinuation of bottle feeding or pacifier use after 1 year of age. The American Association of Pediatric Orthodontists is in full agreement with this.

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1 Leighton BC. Aetiology of malocclusion of the teeth. *Arch Dis Child* 1991;66:1011-2.

Professor Leighton comments:

Many surveys have shown a relationship between sucking habits and malocclusion of the teeth in the deciduous dentition, and even in the mixed dentition. However, it has been shown in a longitudinal study extending from birth to maturity (more than 17 years of age), that the effects of the sucking habit, an increased overjet and an open bite, are both resolved eventually when the habit ceases before the permanent dentition is established.^{1 2} Crossbites are usually corrected spontaneously when the deciduous canines are shed. There were a few cases where a large overjet remained, but these occurred just as frequently where no sucking habit had been indulged. The use of a comforter or prolonged use of a feeding bottle might be justifiably condemned for hygienic reasons, but not because of any effect it may have on the teeth, which is transitory.

- 1 Leighton BC. Symposium on aspects of the dental development of the child. 2. The early development of cross-bites. *The Dental Practitioner* 1966;17:145-52.
- 2 Leighton BC. Early recognition of normal occlusion. In: McNamara, J. ed. *The biology of occlusal development*. Ann Arbor: The Center for Human Growth and Development, University of Michigan, 1977:147-67.

Vocal cord paralysis as a presenting sign of acute spinal muscular atrophy (SMA type 1)

SIR,—Acute diaphragmatic dysfunction can be the first sign of spinal muscular atrophy (SMA) type 1 (Werdnig-Hoffmann syndrome),¹ but early respiratory distress due to vocal cord paralysis has to our knowledge not been reported.

Case report

A girl aged 5 weeks was admitted for a progressive inspiratory stridor of recent onset. Pregnancy, birth, and the first month of life were uneventful. Tendon reflexes were brisk. Moderate axial hypotonia without limb weakness was attributed to her respiratory problem. Direct laryngoscopy showed a complete paralysis of both vocal cords in paramedian position. Lungs and diaphragm were normal on the chest radiograph. Tracheostomy was required at 6 weeks. Extensive work-up to look for a known cause of vocal cord paralysis in infancy remained negative.²

At 4 months, swallowing difficulties were reported. Three weeks later a massive hypotonia with severe weakness of all extremities, areflexia, and fasciculations of the tongue were found. Facial and ocular movements were spared.

Electromyography showed denervation, with fibrillation potentials in proximal and distal muscles of the left leg and pectoralis major. Sensory and motor conduction velocities were normal, with a reduced compound muscular activity amplitude. A biopsy specimen of the quadriceps showed group atrophy with increased connective and adipous tissue. No inflammatory reaction, abnormal material, or significant ultrastructural changes were found. Serum creatine kinase was normal.

The child died at 5 months. A necropsy was refused.

Clinical evolution and results of the electromyography and muscle biopsy were typical of SMA type 1, but vocal cord paralysis was a

very puzzling presentation. Tracheostomy was required before the occurrence of peripheral signs of the disease. Classically, bulbar dysfunction appears late in the course of SMA type 1, respiratory failure being a consequence of progressive involvement of intercostal muscles.

Isolated vocal cord dysfunction has been described in SMA, but in a variant of the juvenile form (type 3) with distal weakness.³ The diagnosis of SMA type 1 should be considered when the clinician is confronted by unexplained vocal cord paralysis in infants.

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- 1 McWilliam RC, Gardner-Medwin D, Doyle D, Stephenson JBP. Diaphragmatic paralysis due to spinal muscular atrophy. An unrecognized cause of respiratory failure in infancy. *Arch Dis Child* 1985;60:145-9.
- 2 Cohen SR, Birns JW, Geller KA, Thompson JW. Laryngeal paralysis in children, a long term retrospective study. *Ann Otol Rhinol Laryngol* 1982;91:417-24.
- 3 Young ID, Harper PS. Hereditary distal spinal muscular atrophy with vocal cord paralysis. *J Neurol Neurosurg Psychiatry* 1980;43:413-8.

Trends in preterm survival and cerebral haemorrhage

SIR,—The observation by Cooke that the survival of preterm infants over a 10 year period improved while cerebral haemorrhage diagnosed by ultrasound showed no significant trend remarkably makes no comment on the incidence of this pathology among deaths as opposed to survivors.¹ If the author wishes to demonstrate a relationship, or the absence of a relationship, between a potentially lethal condition and the numbers of deaths (or survivors as practitioners in child health tend to view the matter) then knowing the incidence of the condition among patients who die is invaluable.

It cannot be assumed that the overall incidence of cerebral haemorrhage as shown by ultrasound during life is identical to its incidence at death. If cerebral haemorrhage is more common in patients who die its lethal potential is to some degree demonstrated. If cerebral haemorrhage is more common in survivors, a beneficial effect or association has to be considered. If the incidence is the same then one has to regard cerebral bleeding as a phenomenon which is unrelated to death or survival and which is there by chance.

Alas the author cannot resolve this matter simply by re-examining the ultrasound reports and apportioning them to deaths or survivors. This is because it would be open to the criticism, as indeed the paper as it stands is open to the criticism, that cerebral haemorrhage occurred after the last cerebral scan and was in fact the major cause of death. This criticism can only be satisfied by postmortem ultrasonography or what is far better, necropsy. The advantage of the latter is that the age and extent of the haemorrhage and its relevance to the outcome is more accurately assessed.

As a pathologist I cannot help being amused by the irony of this situation. Pathologists are always open to the criticism that knowing the incidence of a disease in patients who die does not in itself indicate the incidence in those who are alive. The argument still carries