LETTERS TO THE EDITOR

Model to identify potentially preventable cerebral palsy of intrapartum origin

EDITOR,—It is highly desirable to identify potentially preventable intrapartum cerebral palsy, even retrospectively, and I congratulate Gaffney *et al*¹ on attempting to create such a model for identification. They propose a stepwise process in which: (i) cases with recognised postneonatal and prenatal causes are excluded; (ii) signs compatible with intrapartum brain damage are sought, and if present; (iii) intrapartum care is evaluated according to predefined standards. I have no authority to evaluate standards of intrapartum care, but am concerned that their study suggests a degree of certainty concerning the possibility of retrospectively identifying an intrapartum cause in individual cases which current knowledge cannot support, and consequently, that they calculate that 12.4% of cerebral palsy cases are *potentially preventable* in the intrapartum period.

Assignment of a postneonatal cause for cerebral palsy is straightforward, requiring recognition of a potentially brain damaging event or illness after observable postnatal neurological normality. In contrast, there are cogent reasons why antepartum causes may frequently be missed, and in a proportion of these, the cause is likely to be assigned to the intrapartum period.

The data supplied by Gaffney *et al* confirm the difficulty of assigning the timing of cause. For 45% ((87 + 8/210)) of cases, no postneonatal, intrapartum, or prepartum cause was recognised. These cases are most likely to be the result of an unidentified prepartum cause. A proportion of those cases with an unrecognised prepartum cause may be less tolerant of the stress of labour and therefore likely to have abnormal cardiotocography, depressed Apgar scores, and abnormal neurological function. Unfortunately, no intrapartum or neonatal signs specifically indicate that brain damage has occurred during delivery.

Ideally, the same criteria required for assigning a postneonatal cause should be required for an intrapartum one—that is, the presence of a potentially brain damaging event or illness following neurological normality. Prenatally, neurological normality is difficult to ascertain, but we could apply the first criterion. In Gaffney *et al*'s paper only eight of the 35 babies included in step 5 (with evidence of fetal distress or an obstetric emergency) had detailed evidence of an adverse intrapartum event likely to initiate brain damage. Most of the remaining 27 infants seemed only to have had signs of fetal distress, particularly bradycardia, without an intrapartum event to explain the fetal distress. Bradycardia is not defined in the paper, and is widely reported to infer any fetal heart rate measurement of < 120 beats per minute. It has been suggested that a normal healthy fetus must experience a bradycardia of < 60 beats per minute for 10 minutes or more before permanent brain damage occurs. Therefore, until such bradycardia occurs, it does not constitute a brain damaging event,

but, in addition to cardiotocograms, it only provides evidence of fetal intolerance to labour without identifying the cause.²

The model therefore has a grey area: cases without a recognised cause for their cerebral palsy, for whom the first sign of a problem is intolerance to labour. For this reason the maximum likelihood we assigned to birth asphyxia as the cause of spastic cerebral palsy was "very likely".³ Events justified this caution, when, in a subsequent study we found that one of our "very likely" cases had two other similarly affected family members (one of whom had no evidence of intrapartum asphyxia), making familial cerebral palsy more likely.⁴

If cause cannot be unequivocally assigned to the intrapartum period, the precise proportion of cases potentially preventable by intrapartum care cannot be determined. A lower limit might be obtained by requiring a potentially brain damaging intrapartum event unrelated to the prior neurological status of the fetus, followed by neurological encephalopathy. This assumes that it is statistically unlikely for an independent emergency to occur in already severely compromised fetus. The data given by Gaffney *et al* show that the lower limit is between 0 and 8 cases (0-3.8%), depending on the proportion of the eight cases who received suboptimal care for their obstetric emergency. An upper limit could be obtained from the proportion deemed to receive suboptimal care for their signs of fetal distress, assuming that these cases all had normal brains prior to labour in the absence of a recognised prenatal cause. I feel that this upper limit will underestimate it, but until we can determine the neurological status immediately before and after labour, we cannot be more precise.

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- 1 Gaffney G, Flavell V, Johnson A, Squier MV, Sellers S. Model to identify potentially preventable cerebral palsy of intrapartum origin. *Arch Dis Child* 1995;73:F106-8.
- 2 Blair E. A research definition for birth asphyxia? *Dev Med Child Neurol* 1993; 449-5.
- 3 Blair E, Stanley F. Intrapartum asphyxia: a rare cause of cerebral palsy. *J Pediatr* 1988;112: 515-19.
- 4 Blair E, Stanley F, Hockey A. Intrapartum asphyxia and cerebral palsy. *J Pediatr* 1992; 121: 170-1.

Drs Gaffney and Johnson comment:

Dr Blair highlights the continuing uncertainty about the time, or times, at which damage to the developing brain occurs. As we clearly point out in our paper, only a proportion of children with cerebral palsy that is prenatal in origin can be identified at present. This reflects an inability of clinicians or epidemiologists to recognise abnormal fetal brain development before the onset of labour. Many clinicians and lawyers continue to regard most cerebral palsy as being of intrapartum origin.

We are suggesting that if the principles which we outline in our paper are applied, it will be clear that most children do not have an intrapartum origin to their cerebral palsy. Further work is urgently needed in the area of antenatal cardiotocograms, the interpretation of patterns of fetal movement, and other research aimed at identifying, antenatally, babies with abnormal neurological function.

Neonatal otoacoustic emission screening (OAE) for deafness: psychological costs

EDITOR,—The paper by Watkin carefully addressed many of the issues involved in establishing universal neonatal hearing screening.¹ He found that a universal screen could produce a good yield (2/1000) at a cost that was not prohibitive. He acknowledged that the priority for universal screening programmes is to increase specificity without losing sensitivity. An earlier study by Watkin *et al* showed that most parents of deaf children would have welcomed neonatal identification.² How do the parents of false positive cases feel after a fairly long sequence of screening and investigation involving OAE and auditory brainstem response (ABR) testing? It would be wise to consider the experience of established universal screens such as phenylketonuria and congenital hypothyroidism. Marteau noted that the parents of neonates falsely diagnosed on routine screening as having hypothyroidism, reported great strain on their marriages and difficulties in their relationships.³ The psychosocial effects of universal hearing screening may be similar. Such problems may arise for parents between an initial OAE fail and subsequent confirmation of normality.

Watkin provided every mother with an explanatory leaflet before the test. Parents may require more than this. Edwards, with reference to neonatal screening in general, emphasised the need for pretest counselling and informed consent.⁴

A prospective study to measure levels of parental stress in false positive cases as against pass cases and cases not screened would usefully contribute to the debate on the use of targeted or universal neonatal screening.

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- 1 Watkin P M. Neonatal otoacoustic emission screening and the identification of deafness. *Arch Dis Child* 1996; 74: F16-F25.
- 2 Watkin P M, Beckman A, Baldwin M. The views of parents of hearing impaired children on the need for neonatal screening. *Br J Audiol* 1995; 29: 259-62.
- 3 Marteau T M. Psychological costs of screening. *BMJ* 1989; 299: 527.
- 4 Edwards A. Pretest counselling and informed consent should be prerequisites. *BMJ* 1996; 312: 183.

Capillary blood sampling: should the heel be warmed?

EDITOR,—From their study Dr Barker and colleagues¹ concluded that neonatal heel skin temperature was not an important factor in capillary blood sampling. While this conclusion may be valid in respect to their outcome measures (time taken to collect a standard volume of blood, the number of repeat procedures, and the infant's behavioural response), it ignores the influence that heel capillary circulation may have on the characteristics of the sampled blood.

In 1963 I studied 28 healthy term infants aged 0-72 hours in relation to their capillary blood pH and haematocrit. Blood was sampled from both heels of each infant either without prior preparation, or after either immersion of the lower leg in water at 40°C for 2 minutes, or alternatively, after persistent gentle massage for 2 minutes. These two

forms of preparation seemed to be equally effective in improving the capillary circulation in that their pH and haematocrit findings were very comparable. Blood taken from unprepared heels, however, tended to have a significantly lower pH. On occasions, especially soon after birth and when peripheral cyanosis was present, the difference might be considerable (pH 7.3 vs pH 7.1). Haematocrit values seemed to be less affected, though in individual infants differences of 5% or more (plus or minus) were encountered, especially when polycythaemia was present. Not surprisingly I found that other parameters might also be influenced by heel preparation. For example, the capillary blood glucose concentration might be profoundly depressed when capillary circulation was poor.

May I therefore caution readers of Dr Barker *et al.*'s interesting study to reflect carefully before omitting measures designed to ensure a good local circulation prior to heel sampling.

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- 1 Barker DP, Willetts B, Cappendijk VC, Rutter N. Capillary blood sampling: Should the heel be warmed? *Arch Dis Child* 1996; 74: F139-F40.

Dr Barker and Rutter comment:

Professor Dunn's comments on the value of heel warming in improving capillary pH are not supported by the study of McLain *et al.* cited in our paper.² In this study 158 paired arterial and capillary samples were collected from the heels of preterm infants, with heel warming performed to 40 °C for five minutes before collection of half the capillary samples. No significant difference in the discrepancy between arterial and capillary pH was found in relation to warming, and, overall, only 18% of the paired samples were discrepant by more than 0.05 pH units.²

However, the sampling method used may be an important factor. Manual sampling to an uncontrolled depth may reach vessels in which flow is more responsive to local temperature, at the expense of increased distress to the infant.³

- 2 McLain BI, Evans J, Dear PRF. Comparison of capillary and arterial blood gas measurements in neonates. *Arch Dis Child* 1988;63:743-7.

- 3 Harpin VA, Rutter N. Making heel pricks less painful. *Arch Dis Child* 1983;58:226-8.

X linked immune dysregulation, neonatal insulin dependent diabetes, and intractable diarrhoea

EDITOR,—We report an Italian family with a similar clinical picture to that described by Peake *et al.*¹

At birth, the proband (III 4) had petechiae and a platelet count of 14000/mm³ (fig 1). A bone marrow examination showed that the megakaryocytes were severely reduced in number. The child also had widespread eczema from the first few days of life. The child had first been admitted at the age of 2 months: his platelet count has been 12000/mm³, with an eosinophil count of 800/mm³, an IgG of 95 mg/dl, an IgM of 200 ml/dl, an IgA of 144 mg/dl, and an IgE of 2132 U/ml. Lymphocyte numbers, CD4:CD8 ratio, glucose tolerance, urinary acids and karyotypes were all normal.

Soon after admission he developed diarrhoea which did not respond to total parenteral nutrition. At the age of 3 months, biochemical signs of parenchymal renal disease were detected. The parents refused further treatment and the child died shortly afterwards.

III 1 died at the age of 6 months as a result of intractable diarrhoea. Total parenteral nutrition led to glucose intolerance. At the age of 1 month III 3 developed insulin dependent diabetes mellitus and diarrhoea; an intestinal biopsy specimen showed severe jejunal mucosal inflammation associated with infiltration of the lamina propria by lymphocytes and plasma cells. Autoimmune enteropathy was subsequently diagnosed. Despite treatment with steroids, cyclosporin, and total parenteral nutrition, he died at the age of 7 months as a result of severe hepatic damage and septic complications.

III 7 had had diarrhoea from the age of 45 days and normal glucose tolerance. Serum enterocyte autoantibodies and thyroid microsomal autoantibodies were detected and he was treated with cyclosporin, steroids, and total parenteral nutrition. He died at the age of 12 months.

III 8 had intractable diarrhoea and died at the age of 2 months.

III 9 had diabetes and died at the age of 4 months.

III 11 developed coeliac disease and is following a gluten free diet at the time of writing. He remains well at the age of 7 years.

The thrombocytopenia is a feature of another X linked disease—Wiskott-Aldrich syndrome. Thrombocytopenia, liver and renal diseases are rare in the families with neonatal diabetes and diarrhoea reported to date.

The case reported by Peake had agenesis of the islet of Langerhans; our patient had neonatal amegakaryocytic thrombocytopenia. These two features are not associated with immune dysregulation, which is probably not the primary defect.

Our family is interesting because of the different associations in each child, a picture which is not dissimilar to mitochondrial disease, but we have no other evidence in support of this hypothesis. It is our belief, however, that all the patients with X linked

neonatal diabetes and diarrhoea reported to date, have, in fact, one disease only.

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- 1 Peake JE, McCrossin RB, Byrne G, Shepherd R. X linked immune dysregulation, neonatal insulin dependent diabetes and intractable diarrhoea. *Arch Dis Child* 1996; 74: F195-9.

Pain response in neonates

EDITOR,—Dr Ramenghi and colleagues have elegantly shown that intraoral administration of sweet tasting solutions reduces response to pain in the newborn.^{1,2} Although young infants have been the focus of study so far, the findings may be applicable to older children. According to the following extract, this already seems to have been recognised some time ago. Charlotte Bronte published her novel *Villette* in 1853, based on her experiences of studying and working in Brussels. The character of Lucy Snowe relates what happened when the child of her employer broke an arm and was attended by the doctor, the bones being successfully reset once the child had been calmed with sugar water:

"I, at least, was taken up with endeavouring to soothe Fifine; whose cries (for she had good lungs) were appalling to hear....So now, when we get a little calmer, we must commence business; and we will soon have that unlucky little arm bandaged and in right order. Hereupon he called for a glass of eau sucrée, fed her with some teaspoonful of the sweet liquid...promised her more when the operation should be over, and promptly went to work.; much as he had hurt her, she held out her hand to bid him a friendly good-night."³

Lucy, the shrewd observer of this scene, considered that "the little gourmand's heart had been won through her palate." This seems a more poetic way of saying that the antinociceptive effect of sucrose is mediated through endogenous opiate release.

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- 1 Ramenghi LA, Wood CM, Griffith GC, Levene MI. Reduction of pain response in premature infants using intraoral sucrose. *Arch Dis Child* 1996;74:F126-8.

- 2 Ramenghi LA, Griffith GC, Wood CM, Levene MI. Effect of non-sucrose sweet tasting solution on neonatal heel prick responses. *Arch Dis Child* 1996;74:F129-31.

- 3 Bronte C. *Villette*. Everyman edition. London: JM Dent, 1993: 92-3.

Erratum

Fetal growth velocity: kinetic, clinical, and biological aspects.

Arch Dis Child 1996;74:F10-15.

For "anoxia" on page F14, lines 12 and 15, please substitute "damage". Interestingly, this error arose because the original manuscript from Italy used the expression "a noxa" (an injury or damage).

Figure 1 Family M