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