Correspondence

Placental steroid sulphatase deficiency

Sir,

I would like to comment on the paper on placental steroid sulphatase deficiency by Attenburrow *et al.*¹ The authors state that this condition is associated with delayed onset of labour, poor progress during labour, and failure to breast feed. In a recent paper dealing with 23 cases of placental steroid sulphatase deficiency, that is approximately one fourth of all published cases, neither prolonged gestation nor failure of cervical dilatation during labour was conspicuous.² Furthermore, we have been able to show that women whose pregnancies had been complicated with placental steroid sulphatase deficiency were generally as successful in establishing breast feeding as other women.³ In this context it is notable that two of the children from family B reported by Attenburrow *et al* were in fact breast fed.

A few additional points should be of interest to paediatricians. It is by now generally accepted that placental steroid sulphatase deficiency is the antenatal forerunner of recessive X-linked ichthyosis. Our 23 women gave birth to 25 boys who all have acquired this type of ichthyosis. Typically, at the age of 1 to 3 weeks a general peeling of the skin with rather large, light, and loosely attached scales over the entire integument was noticed. During the next weeks, this immature scaling was replaced by the 2 to 5 mm polygonal, brownish, and more firmly attached scales characteristic of recessive X-linked ichthyosis. A high proportion of boys with steroid sulphatase deficiency and recessive X-linked ichthyosis experience testicular maldescent. In our series of 80 patients, nine had testicular maldescent, mainly in the form of uni- or bilateral inguinal cryptorchidism.

Attenburrow *et al* suggest that parents with placental steroid sulphatase deficient boys seek appropriate advice on the skin changes; I wonder what that might entail. In our experience, topical treatment of recessive X-linked ichthyosis has been found cumbersome and only slightly beneficial. Recently, however, a therapeutic trial with a cream containing 10% cholesterol has yielded promising results in respect of efficacy and acceptability.⁴

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Dr Attenburrow comments:

I was interested to read Dr Lykkesfeldt's comments. There are obviously conflicting views as to whether there are problems with labour and breast feeding in association with placental steroid sulphatase deficiency. In fact reported patients, on the whole, will be a rather selected

group who are investigated further because of problems during pregnancy. In a review by Taylor⁵ of 48 patients, however, 46% of primigravidas started labour spontaneously as did 39% of multigravidas. Of those in whom labour was established, 39% required caesarean section, of which 54% were induced and 28% were of spontaneous onset. A high incidence of intervention! I must acknowledge that two out of our four children reported were breast fed.

The point about cryptorchidism is noted. Theoretically, one might expect problems with sexual maturation. None of our patients had testicular maldescent.

As regards treatment of ichthyosis, we have tended to advise urea cream (BNF) in association with emulsifying ointment. I would agree that this has had a modest effect only. Certainly a cholesterol cream has theoretical appeal, as the underlying problem is thought to be accumulation of cholesterol sulphate in the stratum corneum.⁶

References

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- 3 Lykkesfeldt G, Bock JE. Lactation in placental steroid sulphatase deficiency. *Br J Obstet Gynaecol* 1985;**92**:179-80.
- 4 Lykkesfeldt G, Høyer H. Topical cholesterol treatment of recessive X-linked ichthyosis. *Lancet* 1983;**ii**:1337-8.
- 5 Taylor NF. Review: placental sulphatase deficiency. *Journal of Inherited Metabolic Disease* 1982;**5**:164-76.
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Welfare of children in hospital

Sir,

We at the National Association for the Welfare of Children in Hospital (NAWCH) were gratified to read your editorial¹ which raised many issues pertinent to our work in England today.

Although it is now 25 years since the sound, humanitarian recommendations of the Platt Committee's report on the Welfare of Children in Hospital were adopted as official Department of Health policy, we are constantly reminded that for many children and their families, a hospital stay can be a nightmare experience of enforced separation and conflict. Our survey of parental access and family facilities on wards admitting children in 1982² showed that less than half those wards admitting children had abandoned visiting rules for parents.

We believe that our association, with a membership