Progress of a harlequin fetus treated with etretinate

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'Harlequin fetus' is the descriptive term for the most severe form of congenital ichthyosis. This condition has also been known as ichthyosis fetalis and has previously been fatal either *in utero* or shortly after birth (Wells & Kerr 1965). In this brief report the progress and treatment of a harlequin fetus to 43 weeks is described.

Case report

A baby with clinical features of harlequin fetus of gestational age 34 weeks and weight 1.75 kg was seen at birth. The skin was hard and thickened and appeared composed of hexagonal plates which split to reveal moist erythematous areas. There was severe ectropion and eclabium. The nose was flat and the ears were rudimentary (Figure 1). The baby was immobile with limbs held in fixed flexion and had deformed hands and feet.

Treatment was with humidified temperature control, standard infant feeding via a nasogastric tube aiming at 200 ml/kg/day, baths and frequently applied emollients. Etretinate was given at a dose of 1 mg twice daily, then reduced to 1 mg daily.

At 10 months the baby weighed 4.4 kg and had dry erythematous ichthyotic skin. The nose had developed normally, there was no eclabium and the shape of the ears had improved (Figure 2). She moved all joints and could transfer objects from hand to hand. Her ectropion had improved spontaneously, although plastic surgery was contemplated on her eyelids to further

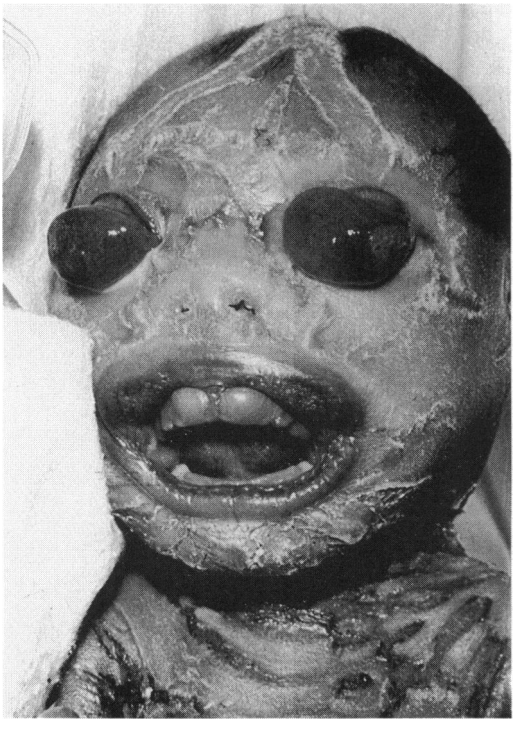

Figure 1

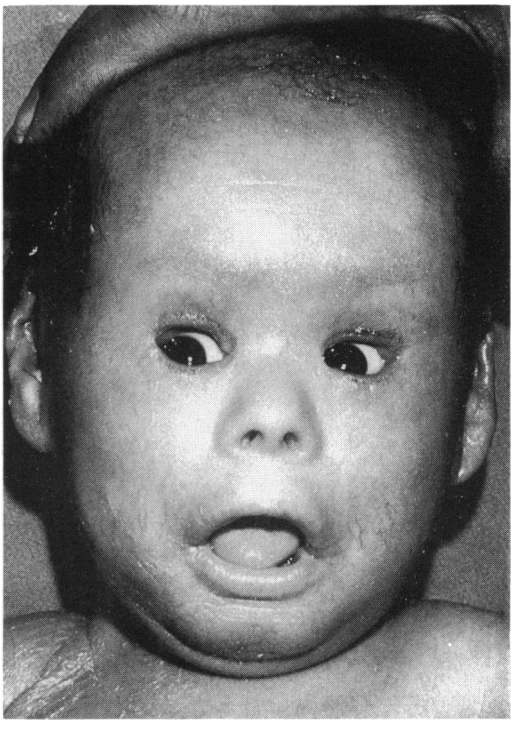

Figure 2.

protect her sclera. This has subsequently been carried out.

Discussion

Harlequin fetus is a syndrome which should be differentiated from collodion baby from which it is clinically distinct. The picture of a harlequin fetus is that of a baby deformed by the thickness and contraction of its own skin and while a collodion baby may have some similar features, namely ectropion, eclabium and transient deformities of hands and feet, the thin shining layer which covers the baby bears little resemblance to the rigid keratin of a harlequin fetus. The collodion baby may be considered to be the phenotypic expression of several genotypes, but since our patient is the only documented harlequin fetus surviving to 1 year at the time of this report, it is not possible to classify this entity, although the literature suggests that it may progress to ichthyosiform erythroderma (Lawlor & Peiris 1985).

Acknowledgements

We wish to thank Dr J Burton for allowing us to submit this short report. A full case report is published in the *British Journal of Dermatology* (Lawlor & Peiris 1985).

References

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