# CONGENITAL ICTHYOSIS (HARLEQUIN FOETUS)

## Report of a case

BY

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Congenital icthyosis is a rare congenital abnormality. With the available literature in this place I am not able to find a reference about cases occurring in India. Cases have been reported in the British Journal of Obstetrics and Gynaecology.

Waring, in 1932, cited an early reference by Hart in 1750 to the disease. Caffier (1929) reported three cases occurring in a single family. Parer et al, in 1951, reviewed the literature and reported a case. Further cases were reported by Seitz in 1936, and Finlay and Bond in 1952. I. Kessel and S. N. Javet (1957) reported a typical case of "Harelquin foetus" where they did autopsy and found lower nephron nephrosis of the kidneys; other organs in the body were found to be normal. The mother subsequently gave birth to a normal infant.

**Case Report**

Mrs. A., a primipara, was admitted for labour pains on 13-9-57 at 8 A.M. Her general condition was good; and nothing abnormal was found on clinical examination. There was no albumen in urine, B.P. 120/80 and blood Kahn—negative. On palpation it was found to be a breech presentation. The pelvis was found to be adequate and she was put down for vaginal delivery. After ten hours of labour pains membranes ruptured and clear liquor amnii escaped. After another hour of strong bearing down pains it was found that the breech was not descending. A vaginal examination was done and it was found to be a full breech with cervix fully dilated and the breech at the brim. A breech extraction was done and a live male baby delivered.

The baby showed the typical appearance of a "Harlequin foetus" except that it did not have ectropion of the eyelids. The skin over the whole body was markedly thickened with cracks and fissures. The pinna of the ear was well formed but the edges were irregular and stuck on to the scalp. The external auditory meatus was occluded with thick whitish material which was at first thought to be vernix caseosa, but on trying to remove the material it was found to be the thickened skin of the external auditory meatus. Hair was well developed and the scalp also showed thickening and fissures. The external genital organs were normal. The limbs appeared oedematous with the skin showing same changes. The fingers and toes were not well formed (Fig. 1).

The baby cried well, opened its eyes, sucked well, passed urine and meconium. The baby lived for twenty-four hours.

The placenta membranes and cord appeared normal. The mother had an uneventful recovery. There was no similar abnormality in her or her husband's family.

**Biopsy of Skin:** Shows marked hyperkeratosis with thinning of stratum malphigii. Follicular keratotic plugs,

A plain skiagram of the baby after birth showed that the skeletal growth was normal (Fig. 4).

*Discussion*

The case reported by Kessel and Javet is a typical case of "Harlequin foetus" which is an extreme form of congenital icthyosis. The case reported by Finlay and Bond is of a milder type with collodion skin and lamellar icthyosis. Only the dorsum of hands, feet and toes was affected and the condition improved with desquamation of the abnormal areas leaving normal skin underneath.

According to R. W. B. Ellis a number of distinct conditions are included under the term icthyosis. In all there is dryness and scaling of the skin. Icthyosis vulgaris (simplex) is not usually manifest until later in infancy. There are congenital and foetal types, in which the infant is born with a hard covering which splits and desquamates shortly after birth. In true icthyosis congenita which is very rare the eyelids are everted and sucking may be interfered with. These cases are usually still-born or die within a few hours of birth.

Benign forms seen in the newborn are thought to be due either to an accumulation of vernix (icthyosis sebacea) or to persistance of a foetal membrane which subsequently splits and desquamates.

This case is not a typical "Harlequin foetus" as there was no ectropion, but all the other features were there. It is not mentioned whether there were any abnormalities of placenta, membranes and the cord in the other cases reported.

Thanks are due to my assistant Dr. Bangaru for her help and to student Jamal for the photograph of the baby. I thank the Superintendant for permitting me to publish this case.

*References*

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Fig. 1
Photograph of baby showing fissures in the skin and the whole body affected.

Fig. 2
Sections of skin showing hyperkeratosis and keratotic plugs.

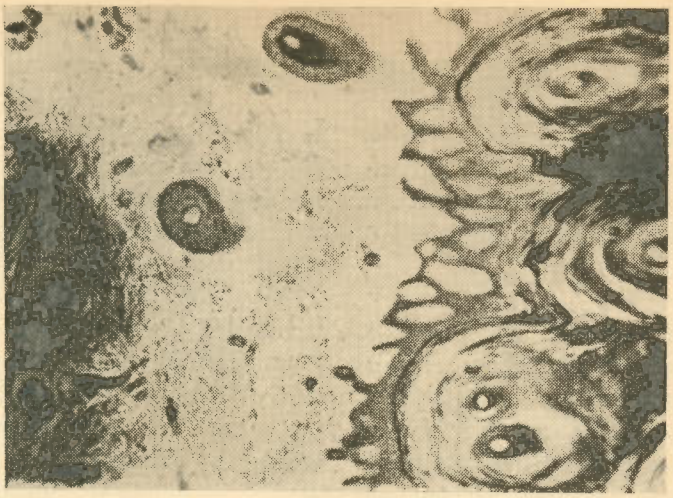

Fig. 3
Sections of skin showing hyperkeratosis and keratotic plugs

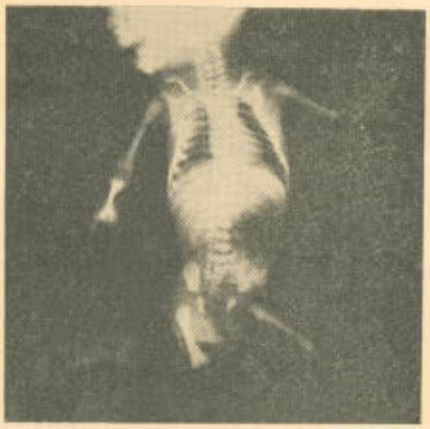

Fig. 4
Skiagram of baby showing normal skeletal growth.