# REVIEW

# EPIDERMOLYSIS BULLOSA.

**(Review of literature and report of a case.)**

By

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Epidermolysis Bullosa is an uncommon disease of the skin characterised by an extraordinary tendency to the formation of blebs after friction or trauma. The disease occurs in all races and in all parts of the world. Recognition and knowledge of epidermolysis bullosa date back to just under hundred years. Goldscheider (1882) was credited with the earliest record of the simplex variety. Tilbury Fox (1879) described the dystrophic form which is more severe but less common. Elliot (1895) reported a case of simplex type in a male aged 30 years who has suffered from the disease since his 5th year. Luistgarten (1896) reported two such cases. Herlitz (1935) collected 14 cases of epidermolysis bullosa from the literature and added 8 of his own. Touraine 1942 classified this disease into three types:

1. Epidermolysis bullosa simplex (dominent)
2. Epidermolysis bullosa dystrophica.
   (a) hyperplastic type (dominent)
   (b) dysplastic type (recessive)
3. Epidermolysis bullosa lethalis (recessive).

Greenburg 1944 found an incidence of 0.2% among dermatologic patients seen in the Army Station Hospital during 2 year period. Since then periodical case reports have been published from all over the world. Literature pertaining to the disease from India is sparse and the following case of epidermolysis bullosa dystrophic form is being recorded for its rarity.

## CASE REPORT

A Hindu, female M. J. aged 7 years was admitted into the dermatological ward of the Government General Hospital, Guntur for a complaint of bullons eruptions since birth. The present complaint started as bullous eruptions after about 4 hours after birth confining mostly to the extremities and also on the trunk. These bullae were of varying sizes and ruptured leaving raw areas which subsequently healed. The discharge from the ulcerated areas is serous in nature. Of late the bullae are getting infected and turning into pustules. Lesions are more over the joints and painful. Iching is present. At birth she had nails which were healthy and subscquently nails became loose and dropped off.

* Received for publication on 7-5-1966.

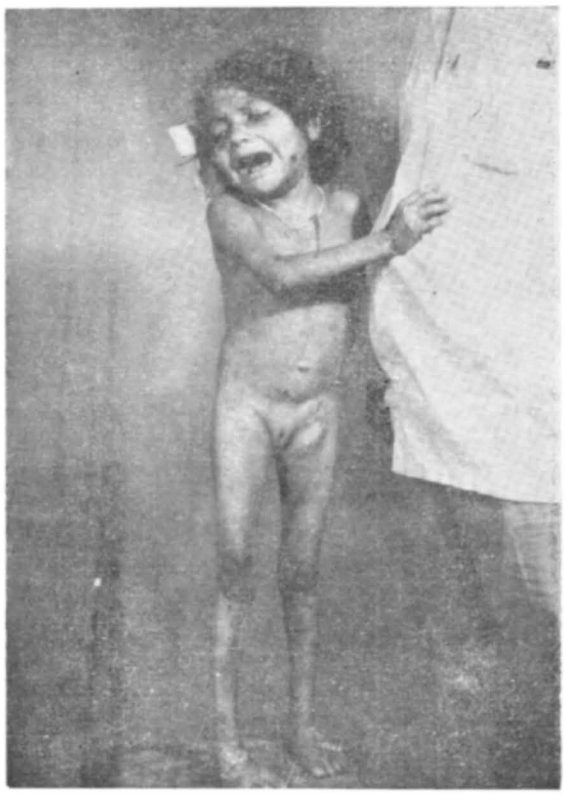


Fig. 1:—Clinical photograph showing the bullous eruptions of various sizes; some of them ulcerated.

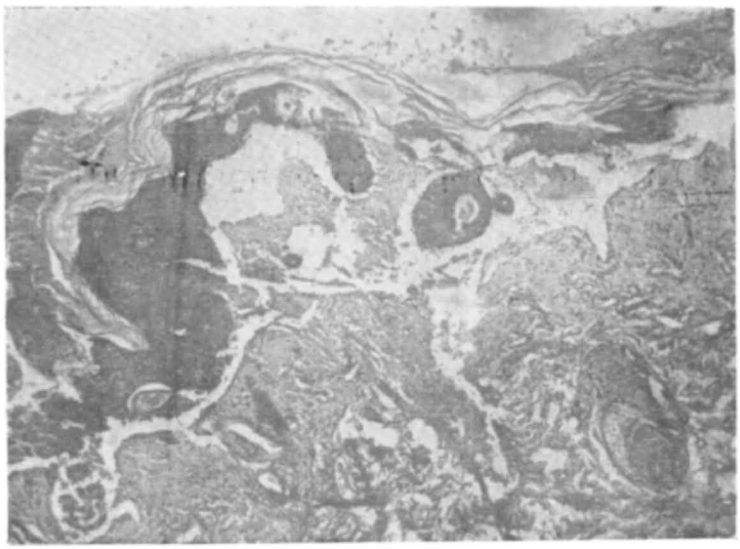
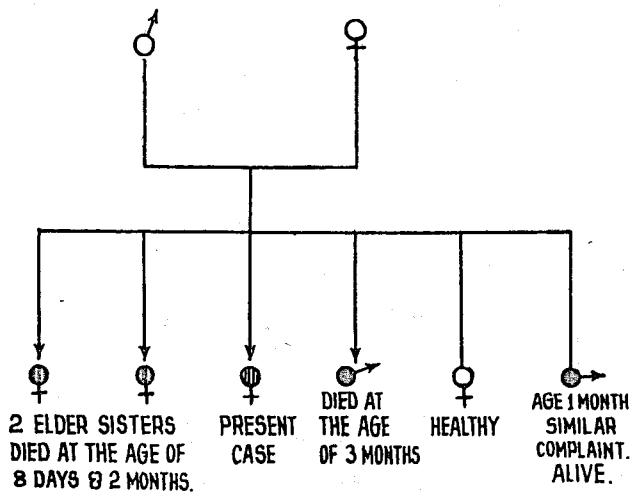


Fig. 2.—Photo micrograph illustrating sub-epidermal bullous formation with areas of haemorrhages H & E x 60.

## FAMILY HISTORY (Chart I)

Patient had two elder sisters and two younger sisters and one brother. Her two elder sisters suffered from similar complaint and expired at the age of 8 days and 2 months respectively. The patient is the third child. The fourth child (brother) had

CHART SHOWS THE FAMILY OF THE PATIENT WITH THE NO. OF CHILDREN WHO HAD SUFFERED FROM THE DISEASE.




a similar attack and died at the age of 3 months. The fifth child (female) aged 3 years is healthy. The sixth child aged 1 month is having similar complaint and is alive. No history of consanguinity between the parents. Patient comes from a middle class family. No history of any other illness.

## GENERAL AND SOCIAL CONDITION

Examination revealed an ill-nurished female child of about 7 years, anaemic, the nails of the fingers and toes are absent. Flaccid bullous eruptions over the right leg are seen. Nikolsky's sign is positive. Multiple raw areas of varying sizes and shapes are present over the extremities and trunk covered with dry crusts. Some of these healed areas are hypopigmated. Teeth, eyes, hair normal. Patient is able to stand and walk.

## INVESTIGATIONS

B. P:—100/70
Urine:— No albumin, No sugar.
Stool:— No ova or cysts.
Total W. B. C:— 9600/ cmm.
Difft. count: Polymorph: 72%, Lymphocyte: 22%, Eosinophile: 4%, Monocyte: 2%.
E. S. R: 15 mm 1st hour.
X-ray of bone: No abnormal changes seen in the bones.

## BIOPSY (No. 1304/66)

Shows sub-epidermal bullous formation with areas of haemorrhage and infection. Sections stained with elastic tissue stain did not reveal any change in the elastic tissue. Histological appearances are consistent with that of epidermolysis bullosa. (Fig. 2).

## TREATMENT

1. The patient was put on Predinisolone tablets.
   T. D. S. for 5 days.
   One tab. twice a day for 5 days.
   ½ tab. twice a day being continued.
2. Mullitivitamin drops.
3. Proteinix 1 tea spoon full twice a day.
4. Nebacortril ointment for local application.
5. Joints are protected from injury.

## COMMENT

Epidermolysis bullosa dystrophic type is the most severe and destructive form. It may be present at birth or at infancy. In the case recorded the disease appeared immediately after birth. Bullae follow slight trauma and often they seem to be without any obvious injury. Any part of the body may be affected though the sites of prediliction are, hands elbows and knees. The lesions heal with marked scarring atrophy, hyper-pigmentation and depigmentation. Bullae occasionally have been noticed in the mucous membranes of the lips, mouth, tongue, pharynx. But we have not come across such mucosal lesions.

In many of these cases a family history of similar complaint amongst the brothers or sisters is usually obtained and in the case recorded the patient's 3 sisters and one brother had similar complaint and they all died. This type is transmitted as a simple recessire factor. Consanguinity of the parents is frequently present but it is not seen in our case. The aetiological factors are not clearly understood.

Engman and Mook 1905 first suggested that the change may be due to inadequate development or even absence of elastic fibres in the upper corium. Leoni has supported this view. Special stains to demonstrate any change in the elastic tissue were employed but were negative in our case. Roberts et al suggested that perhaps the primary defect lies in the basal cells and not in the dermis. They have demonstrated vacuolar degeneration of the basal cells in clinically normal appearing areas. They are of the opinion that the changes in the elastic tissue may be secondary.

Various dystrophies of the nails are seen in these cases. They are frequently atrophic and sometimes completely destroyed. In fact in some infants the nails are absent at birth. In the case recorded, though the nails were present at birth and

healthy, as the disease progressed, the nails became loose, soft and have fallen off. The teeth are deficient, small and also decay early due to deficient enamel. No abnormalities in teeth are noticed in our case.

Prognosis must be guarded. Most patients die early in life but few live long enough to have children. This case is being treated with cortisone and supportive therapy and is being followed up periodically to judge the prognosis.

## SUMMARY

A case of epidermolysis bullosa-dytrophic type in a female child of 7 years is recorded.

## ACKNOWLEDGEMENT

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