ectodermal dysplasia society

EPIDERMOLYSIS BULLOSA
AND
ECTODERMAL DYSPLASIA

Epidermolysis Bullosa (also called EB) is the general term for a group of disorders characterized by increased fragility and blistering of the skin. There are many such conditions, and more than 25 separate forms have been described to date. Like the EDs, the EBs are genetically transmitted with different types having different modes of inheritance. Usually EB is apparent from birth or early infancy; occasionally, individuals with certain forms may experience their first blisters later in life. In severe types of EB, there can be involvement of the eyes, the linings of the digestive and genitourinary tracts and the mucous membranes of the respiratory tract.

Blistering in EB is induced by friction and by exposure to high environmental temperatures. Therefore, although individuals with EB have no difficulty with sweating or heat regulation, they may be heat intolerant because of the marked increase in blistering that occurs in hot weather or in overly warm indoor environments. Avoidance of overdressing and provision of air-conditioning in hot weather are good rules for EBs as well as EDs.

Hair growth and structure is normal in EB. However, if there is extensive blistering and scarring of the scalp, particularly in the forms of the disease that heal poorly, scalp hair may be sparse or even absent. The nails are often affected in EB, especially in certain forms of the disease. The nail plates may be lost temporarily or permanently or may be markedly abnormal in appearance. These changes are usually due to trauma to the nails or blistering around the nail fold. The teeth are usually normal in structure and number, but may be subject to decay because of mucous membrane problems. Blistering in the mouth, often causing severe discomfort and may limit attempts at brushing and caring for the teeth. The junctional form of EB is also associated with a defect in tooth enamel. It is very important for the EB patient, like the EDs, to obtain regular dental care from dentists knowledgeable about this condition.

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