Structural Hair Abnormalities in Hidrotic Ectodermal Dysplasia (HED)

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Significant chemical and physical alterations have been demonstrated in the hair of hidrotic ectodermal dysplasia (HED) patients as compared with normal subjects (Gold and Scriver, 1971). We found marked alterations of the hair surface in 2 patients—father and daughter—suffering from the autosomal dominant HED of the tricho-onychotic subgroup.

Striking deviations are already seen by low magnification. Hair specimens of all scalp regions show varying diameters, splitting of the cuticle, irregular margin contours, cracks and initial fractures without node formation. The hair tips are paintbrush-like (Fig. 1).

The cuticular surface structure is seriously altered as shown by an imprint technique. Partly there is a bizarre arrangement with dissolution of the scale pattern

Fig. 1a and b. Hair shafts in HED. a Diameter variations (microscopical magnification 22.5 ×). b Irregular contours, splitting of the cuticle, cracks and incipient fractures (microscopical magnification 100 ×)
(Fig. 2), partly remainings of the normal cuticular arrangement can still be seen. In other hair specimens, there is a complete loss of pattern. The shaft shows longitudinal fissures, fibre-like tear-offs and surface defects. When the cuticle is entirely lacking the cortex is denuded resembling a tree stem with longitudinal breaks (Fig. 3).

Up to now the hair abnormalities in most instances have been considered from a clinical viewpoint. Most authors have stressed the scanty hair growth and the short, brittle, lustreless hair. Salamon (1966) and Korting (1974) have described them as normal by light microscopy. However, demonstration of the hair surface by an imprint technique has shown, that there is a damage of the cuticle extending up to a complete “decuticulation” so that the cortex is exposed. It may be argued whether there exists a severe disorder of hair formation the cuticular damage being the main event, or whether the keratin structure and/or amorphous matrix is predominantly altered.

The observed hair changes are not identical to those described by Bottom et al. (1972) in the distal portion of the hair shaft.