Read Item - The Ectodermal Dysplasias

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Abstract: Doctor's resource on the Ectodermal Dysplasias

The Ectodermal Dysplasias

Ectodermal dysplasia defines a group of inherited developmental syndromes with disorders in more than one ectoderm derived tissue. The defect presumably manifests during early embryological development some time after the third week post-conception, when ectoderm can first be distinguished from mesoderm and endoderm, but before the end of the third month when ectodermal cells have become committed to differentiate into specific derivative structures.

Ectodermal cells during this period (between 3 weeks and 3 months) have the potential to develop into either neuroectoderm or surface ectoderm.

Any developmental abnormality that damages ectodermal cells in this window period prior to committed differentiation may result in abnormalities in the infant in a large number of organs. In general, those foetuses that survive to conception often manifest abnormalities of hair, nails, epidermis, teeth and eccrine glands. These abnormalities form the basis of classification for ectodermal dysplasias.

Other abnormalities that may be occur include deafness, mental retardation, breast hypoplasia, cleft lip and palate, syndactyly, ectrodactyly, skeletal abnormalities and distinctive facies. Bony abnormalities, although reflecting an abnormality of a mesenchyme derived tissues, do not exclude a diagnosis of ectodermal dysplasia.

In order to fulfil the criteria for an ectodermal dysplasia, there must be a congenital, non-progressive abnormality in at least two ectodermal derived tissues.