

## RAPP-HODGKIN SYNDROME (RHS)

The Rapp-Hodgkin syndrome (RHS) is the name given to an Ectodermal Dysplasia syndrome that is associated with cleft lip and palate. There are other ED syndromes with cleft lip and palate (EEC and Hay-Wells), but RHS does not share the hand defects of the former or eyelid defects of the latter. The sweat dysfunction in RHS is not as severe as it is in the Christ-Siemens-Touraine ED (X-linked recessive hypohidrotic ED) and some affected people perspire through their scalp. The hair in RHS grows slowly and is coarse. Like patients with Hay-Wells syndrome, they may have inflammatory scalp dermatitis leading to scarring alopecia in adulthood. As a rule, individuals with RHS have more teeth than those with Christ-Siemens-Touraine ED. Because of the associated cleft lip and palate, people with RHS may also have speech problems and repeated ear infections. Additional features occasionally present include narrowed ear canals, abnormalities of the tear ducts, drooping eyelids, cleft uvula and hypospadias.

RHS is inherited as an autosomal dominant trait. This means that either sex may be affected, that males and females are affected equally severely and that the features of the syndrome may be passed from one generation to the next. Not everyone with RHS shows the exact pattern of features; some people may be very mildly affected or not even have one or another of the features that constitute the syndrome. It has been suggested that RHS and the Hay-Wells syndrome are different forms of the same entity.

As with most of the EDs, general health, intelligence and life-span are within normal limits.

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### Supporting a normal lifestyle

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