Hypohidrotic ectodermal dysplasia (HED) is a rare inherited multisystem disorder that belongs to the group of diseases known as ectodermal dysplasias. Ectodermal dysplasias typically affect the hair, teeth, nails, sweat glands, and/or skin. HED is primarily characterized by partial or complete absence of certain sweat glands (eccrine glands), causing lack of or diminished sweating (anhidrosis or hypohidrosis), heat intolerance, and fever; abnormally sparse hair (hypotrichosis), and absence (hypodontia) and/or malformation of certain teeth. Many individuals with HED also have characteristic facial abnormalities including a prominent forehead, a sunken nasal bridge (so-called "saddle nose"), unusually thick lips, and/or a large chin. The skin on most of the body may be abnormally thin, dry, and soft with an abnormal lack of pigmentation (hypopigmentation). However, the skin around the eyes (periorbital) may be darkly pigmented (hyperpigmentation) and finely wrinkled, appearing prematurely aged. In many cases, affected infants and children may also exhibit underdevelopment (hypoplasia) or absence (aplasia) of mucous glands within the respiratory and gastrointestinal (GI) tracts and, in some cases, decreased function of certain components of the immune system (e.g., depressed lymphocyte function, and rarely cellular immune hypofunction), potentially causing an increased susceptibility to certain infections and/or allergic conditions. Many affected infants and children experience recurrent attacks of wheezing and breathlessness (asthma), respiratory infections; chronic inflammation of the nasal passages (atrophic rhinitis), scaling, itchy (pruritic) skin rashes (eczema), and/or other findings.