Wrinkly Skin Syndrome

Alternative Names
WSS

WHO International Classification of Diseases
Diseases of the musculoskeletal system and connective tissue

OMIM Number
278250

Mode of Inheritance
Autosomal recessive

Gene Map Locus
2q32

Description
The wrinkly skin syndrome is an autosomal recessive syndrome in which there is wrinkling of the skin on the dorsum of the hands, feet, and abdomen; increased number of creases on the palm and soles; hypermobility of the small joints of the hands associated with intrauterine growth retardation (IUGR); poor postnatal growth; and developmental delay.

Molecular Genetics
In a report on two children with wrinkly skin syndrome scientists noted a del 2q32. This observation is suggestive that the collagen genes COL3A1 and COL5A2 could be candidate genes for gerodermia osteodysplastica and wrinkled skin syndrome. Mutation in COL3A1 causes Ehlers-Danlos IV (EDS IV), while mutation in COL5A2 gene produce EDS type I and II phenotype.

Epidemiology in the Arab World

Palestine
Al-Gazali et al. 2001 reported three patients for a Palestinian consanguineous couple, residents of the United Arab Emirates, who also have one normal child. The parents were symptomatically normal and had a history of miscarriages at five months and two months. All children had similar dysmorphic facial features consisting of broad and prominent forehead, hypotelorism with epicanthal folds, prominent bulbous nose, flat malar region, and large protruding ears. All had wrinkling of the skin more marked on the dorsum of the hands, feet, and abdomen; hyperextensibility of the joints, particularly of the hands; and aged appearance. Intrauterine growth retardation, subsequent failure to thrive, developmental delay, and variable degree of osteoporosis was also present in all of them. The older children developed progressive prognathism. In view of the similarities and overlap between geroderma osteodysplastica, wrinkly skin syndrome, and cutis laxa and developmental delay, Al-Gazali et al. (2001) suggested that all these syndromes represent variable manifestation of the same disorder.

Saudi Arabia
Karrar et al. (1983) reported a brother and sister born to a Saudi couple who showed aging appearance, wrinkled skin over the hands and feet, inelastic skin, prominent veins over the hands, and other musculoskeletal and connective tissue manifestations. Both children were small for their age and had congenital dislocation of the hips.

Syria
Al-Gazali et al. 2001 reported two children for a Syrian consanguineous couple, residents of the United Arab Emirates, with no family history of any significant problems. Both children had similar dysmorphic facial features consisting of broad and prominent forehead, hypotelorism with epicanthal folds, prominent bulbous nose, flat malar region, and large protruding ears. They had wrinkling of the skin more marked on the dorsum of the hands, feet, and abdomen; hyperextensibility of the joints, particularly of the hands; and aged appearance. Intrauterine
growth retardation, subsequent failure to thrive, developmental delay, and variable degree of osteoporosis was also present in both of them. In view of the similarities and overlap between geroderma osteodysplastica, wrinkly skin syndrome, and cutis laxa and developmental delay, Al-Gazali et al. (2001) suggested that all these syndromes represent variable manifestation of the same disorder.

**United Arab Emirates**
[See also: Palestine > Al-Gazali et al., 2001; Syria > Al-Gazali et al., 2001].

**References**


** Contributors**

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