PAEDIATRIC DERMATOLOGY

NAIL SYNDROME PATELLA: NEED FOR THOROUGH EXPLORATION

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Objective: We report the observation of two different families

Observation: It is transmitted as an autosomal dominant trait with variable expression. Anomalies are located on the 9q34 genes LMXMB position are similar to those of ABO blood groups and adenylate cyclase and which encode a protein belonging to the family of LIM- homeobox domain, involved in limb morphogenesis and the anterior chamber of the eye, the maturation of renal podocytes increased collagen type I synthesis

Methods: In fact, 80 to 90 percent of patients have onycho - skeletal abnormalities from birth .In our patients, the nail pterygium was reported at birth and orthopedic symptoms, confirmed by the very rich radiological exploration in both observations. During the SNP renal disease is objectified in half of cases among both men and women. In the first family were not noted or renal abnormalities in the father or to his daughter when we have objectified proteinuria in the second family. Ocular involvement is absent in both cases. Our first observation is characterized by the predominance of onycho - skeletal involvement. Results: This reinforces the hypothesis not responsible for renal disease allelic mutation. Scarf and coll.ont mentioned the existence of a new type of pediatric renal impairment where the nail and bone is absent.

Conclusion: The development of means including immune histochemical diagnostic and genetic genital will in the near future to determine the risk of developing since renal impairment was observed that the collagen fibrils are a constant manifestation of the SNP and are also observed in patients with renal impairment is not clinically detectable.

Keywords: Nail; Nephropathy; Syndrome Patella