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Steroid dehydrogenase deficiency-dental anomalies syndrome

INSERM

Source

INSERM. (1999). Orphanet: an online rare disease and orphan drug data base. Steroid dehydrogenase deficiency-dental anomalies syndrome. ORPHA:3196

Steroid dehydrogenase deficiency-dental anomalies syndrome is an autosomal recessive liver disease which was associated with numerical dental aberrations in a consanguineous Arabi Saudi family. This association suggests that the same gene is involved in both defects. General hypomineralisation and enamel hypoplasia found in this family is thought to be secondary to malabsorption due to liver disease.