Single Central Incisor Syndrome

National Cancer Institute

Source

National Cancer Institute. Single Central Incisor Syndrome. NCI Thesaurus. Code C131003.

An autosomal dominant condition usually caused by mutation(s) in the SHH gene, encoding sonic hedgehog, a secreted protein involved in the organization and morphology of the developing embryo. This condition is characterized by multiple, mainly midline, developmental variations, including the presence of a tooth in the center of the maxillary dental arch in both primary and permanent dentition, and any combination of the following: holoprosencephaly, congenital nasal malformation (choanal atresia, midnasal stenosis, or congenital pyriform aperture stenosis), cleft lip and/or palate, hypopituitarism (including hypothyroidism and hypogonadism, resulting in variations in genital development including small penis or incomplete masculinization in male infants), congenital heart anomalies, and developmental delay.