

Nonsyndromic congenital nail disorder 10

Description

Nonsyndromic congenital nail disorder 10 is a condition that affects the fingernails and toenails. Affected individuals have extremely thick nails (onychauxis) that separate from the underlying nail bed (onycholysis) and can appear claw-like. Some fingers and toes may be missing part of the nail (hyponychia).

In affected individuals, the nails are often abnormal from birth. However, the abnormalities may not be noticeable until later in childhood because the nails tend to grow more slowly than normal.

Individuals with nonsyndromic congenital nail disorder 10 do not have any other health problems related to the condition.

Frequency

Nonsyndromic congenital nail disorder 10 is likely a rare disorder. At least 14 affected individuals have been described in the scientific literature.

Causes

Nonsyndromic congenital nail disorder 10 is caused by mutations in the *FZD6* gene, which provides instructions for making a protein called frizzled-6. This protein is embedded in the outer membrane of many types of cells, where it is involved in transmitting chemical signals from outside the cell to the cell's nucleus. The frizzled-6 protein plays an especially critical role in the growth and development of nails, particularly the attachment of the nail to the nail bed.

FZD6 gene mutations that cause nonsyndromic congenital nail disorder 10 lead to the production of a frizzled-6 protein that cannot get to the cell membrane where it is needed or that cannot transmit signals into the cell. As a result, the growth and development of nails is poorly regulated, which leads to onycholysis and other abnormalities of the fingernails and toenails.

[Learn more about the gene associated with Nonsyndromic congenital nail disorder 10](#)

- FZD6

Inheritance

This condition is inherited in an autosomal recessive pattern, which means both copies of the gene in each cell have mutations. The parents of an individual with an autosomal recessive condition each carry one copy of the mutated gene, but they typically do not show signs and symptoms of the condition.

Other Names for This Condition

- Claw-shaped nails
- Nail disorder, nonsyndromic congenital, 10
- NDNC10
- Onychauxis, hyponychia, and onycholysis

Additional Information & Resources

Genetic Testing Information

- Genetic Testing Registry: Nonsyndromic congenital nail disorder 1 (<https://www.ncbi.nlm.nih.gov/gtr/conditions/C0406443/>)

Patient Support and Advocacy Resources

- National Organization for Rare Disorders (NORD) (<https://rarediseases.org/>)

Catalog of Genes and Diseases from OMIM

- NAIL DISORDER, NONSYNDROMIC CONGENITAL, 1; NDNC1 (<https://omim.org/entry/161050>)

Scientific Articles on PubMed

- PubMed (<https://pubmed.ncbi.nlm.nih.gov/?term=%28%28nonsyndromic+congenital+nail+disorder+1%29+OR+%28onychauxis,+hyponychia,+and+onycholysis%29%29+OR+%28%28autosomal+recessive+nail+dysplasia%29+AND+%28FZD6%29%29+AND+english%5Bla%5D>)

References

- Frojmark AS, Schuster J, Sobol M, Entesarian M, Kilander MBC, Gabrikova D, Nawaz S, Baig SM, Schulte G, Klar J, Dahl N. Mutations in Frizzled 6 cause isolated autosomal-recessive nail dysplasia. *Am J Hum Genet.* 2011 Jun 10;88(6):852-860. doi: 10.1016/j.ajhg.2011.05.013. Citation on PubMed (<https://pubmed.ncbi.nlm.nih.g>

ov/21665003) or Free article on PubMed Central (<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3113248/>)

- Naz G, Pasternack SM, Perrin C, Mattheisen M, Refke M, Khan S, Gul A, Simons M, Ahmad W, Betz RC. FZD6 encoding the Wnt receptor frizzled 6 is mutated in autosomal-recessive nail dysplasia. *Br J Dermatol*. 2012 May;166(5):1088-94. doi: 10.1111/j.1365-2133.2011.10800.x. Epub 2012 Apr 4. Citation on PubMed (<https://pubmed.ncbi.nlm.nih.gov/22211385>)
- Raza SI, Muhammad N, Khan S, Ahmad W. A novel missense mutation in the gene FZD6 underlies autosomal recessive nail dysplasia. *Br J Dermatol*. 2013 Feb;168(2):422-5. doi: 10.1111/j.1365-2133.2012.11203.x. Epub 2012 Oct 5. Citation on PubMed (<https://pubmed.ncbi.nlm.nih.gov/22861124>)

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