nonsyndromic congenital nail disorder 10

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.

Genetic Changes

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.

Inheritance Pattern

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

Diagnosis & Management

Genetic Testing

- Genetic Testing Registry: Nail disorder, nonsyndromic congenital, 10
  https://www.ncbi.nlm.nih.gov/gtr/conditions/C3279974/

Other Diagnosis and Management Resources

- American Osteopathic College of Dermatology: Onycholysis
  http://www.aocd.org/?page=Onycholysis

General Information from MedlinePlus

- Diagnostic Tests
  https://medlineplus.gov/diagnostictests.html
- Drug Therapy
  https://medlineplus.gov/drugtherapy.html
- Genetic Counseling
  https://medlineplus.gov/geneticcounseling.html
- Palliative Care
  https://medlineplus.gov/palliativecare.html
- Surgery and Rehabilitation
  https://medlineplus.gov/surgeryandrehabilitation.html

Additional Information & Resources

MedlinePlus

- Encyclopedia: Nail Abnormalities
  https://medlineplus.gov/ency/article/003247.htm
- Encyclopedia: Onycholysis (image)
  https://medlineplus.gov/ency/imagepages/2010.htm
- Health Topic: Nail Diseases
  https://medlineplus.gov/naildiseases.html

Educational Resources

- Disease InfoSearch: Nail disorder, nonsyndromic congenital, 10
  http://www.diseaseinfosearch.org/Nail+disorder%2C+nonsyndromic+congenital%2C+10/8967
- KidsHealth from Nemours: Your Nails
  http://kidshealth.org/en/kids/your-nails.html
- MalaCards: nail disorder, nonsyndromic congenital, 10,
  http://www.malacards.org/card/nail_disorder_nonsyndromic_congenital_10
- Merck Manual Consumer Version: Deformities, Dystrophies, and Discoloration of the Nails
  http://www.merckmanuals.com/home/skin-disorders/nail-disorders/deformities,-dystrophies,-and-discoloration-of-the-nails
- Orphanet: Autosomal recessive nail dysplasia
  http://www.orpha.net/consor/cgi-bin/OC_Exp.php?Lng=EN&Expert=280654

Patient Support and Advocacy Resources

- British Association of Dermatologists: Skin Support
  http://skinsupport.org.uk/

ClinicalTrials.gov

- ClinicalTrials.gov
  https://clinicaltrials.gov/ct2/results?cond=%22nonsyndromic+congenital+nail+disorder+10%22+OR+%22Onycholysis%22

Scientific Articles on PubMed

- PubMed
  https://www.ncbi.nlm.nih.gov/pubmed?term=%28%28nonsyndromic+congenital+nail+disorder+10%29+OR+%28onychauxis,+hyponychia,+and+onycholysis%29%29+OR+%28%28autosomal+recessive+nail+dysplasia%29+AND+%28FZD6%29%29+AND+english%5Bla%5D

OMIM

- NAIL DISORDER, NONSYNDROMIC CONGENITAL, 10
  http://omim.org/entry/614157

MedGen

- Nail disorder, nonsyndromic congenital, 10
  https://www.ncbi.nlm.nih.gov/medgen/481604

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