

DISEASE:**Infantile-onset periodic fever-panniculitis-dermatosis syndrome**

NAME:	Infantile-onset periodic fever-panniculitis-dermatosis syndrome
DESCRIPTION:	A rare genetic autoinflammatory syndrome characterized by early-onset of repeated episodes of fever, nodular neutrophil-rich panniculitis, arthralgia, and lipodystrophy. Additional reported features include diarrhea, failure to thrive, lymphadenopathy, and vasculitis. Laboratory examination may reveal elevated serum C-reactive protein and leukocytosis with neutrophilia in the absence of infection.
ORPHACODE:	500062
SYNONYMS:	ORAS OTULIN deficiency OTULIN-related autoinflammatory syndrome Otulipenia
XREF(S):	Orphanet ICD-10 OMIM
ANALYTE(S):	OTULIN
CREATED:	13 May 2019 - 01:02
CHANGED:	22 Jun 2023 - 16:14

Source URL: <http://gentest.healthdata.be/disease/2626>

RELATED CONTENT

Related Genetic Tests

- [Periodic Fever \(88 genes\)](#)

Related Laboratories

- [Centre de Génétique Humaine - Erasme ULB](#)

Related Analytes

- [OTU deubiquitinase with linear linkage specificity](#)

Related Gene Panels

- [Periodic Fever \(88 genes\) - ULB](#)

Source URL: <http://gentest.healthdata.be/disease/2626>