DISEASE:
Hereditary pediatric Behçet-like disease

NAME:	Hereditary pediatric Behçet-like disease
DESCRIPTION:	A rare autosomal dominant autoinflammatory syndrome characterized by early onset systemic inflammation with autoimmune manifestations and more rarely, humoral immune deficiency and increased production of circulating proinflammatory cytokines, variably manifesting with recurrent oral aphthous ulcers, genital ulcers, arthralgia or arthritis, periodic fever, uveitis, and severe gastrointestinal involvement (pain, diarrhea, vomiting, rectal bleeding).
ORPHACODE:	476102
SYNONYMS:	Behçet-like disease due to HA20 Behçet-like disease due to haploinsufficiency of A20
XREF(S):	Orphanet OMIM ICD-10 OMIM
ANALYTE(S):	ELF4 TNFAIP3 RELA
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