

DISEASE:**Sterile multifocal osteomyelitis with periostitis and pustulosis**

NAME:	Sterile multifocal osteomyelitis with periostitis and pustulosis
DESCRIPTION:	Sterile multifocal osteomyelitis with periostitis and pustulosis is a rare, severe, genetic autoinflammatory syndrome characterized by usually neonatal onset of generalized neutrophilic cutaneous pustulosis and severe, recurrent, multifocal, aseptic osteomyelitis with marked periostitis, typically affecting distal ribs, long bones and vertebral bodies. High levels of acute-phase reactants (with no fever associated) and onychosis are frequently observed additional features.
ORPHACODE:	210115
SYNONYMS:	Autoinflammatory disease due to interleukin-1 receptor antagonist deficiency DIRA Interleukin-1 receptor antagonist deficiency OMPP
XREF(S):	Orphanet OMIM ICD-10
ANALYTE(S):	IL1RN
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