

Opis choroby *

Definicja

Constriction rings syndrome is a congenital limb malformation disorder with an extremely variable clinical presentation characterized by the presence of partial to complete, congenital, fibrous, circumferential, constriction bands/rings on any part of the body, although a particular predilection for the upper or lower extremities is seen. Phenotypes range from only a mild skin indentation to complete amputation of parts of the fetus (e.g. digits, distal limb). Compression from the rings may lead to edema, skeletal anomalies (e.g. fractures, foot deformities) and, infrequently, neural compromise.

Dane

Klasyfikacja

Zespół wad wrodzonych

Synonimy

Amniotic band sequence
Amniotic band syndrome
Congenital ring constrictions
Constriction band syndrome
Streeter dysplasia
Amniotic band sequence
Amniotic band syndrome
Congenital ring constrictions
Constriction band syndrome
Streeter dysplasia

Kod ORPHA

295000

Kod OMIM

217100

Kod ICD10

Q79.8

Kod ICD11

LD26.5

*Źródło

orphanet