Opis choroby *

Definicja

A rare congenital urogenital anomaly characterized by the presence of double uterus (didelphys, bicornuate or septum-complete or partial), unilateral cervico-vaginal obstruction (obstructed hemivagina-communicant, not communicant or septate and unilateral cervical atresia) and ipsilateral renal anomalies (renal agenesis and/or other urinary tract anomalies). Patients are usually diagnosed at puberty after menarche due to recurrent severe dysmenorrhea, chronic pelvic pain, excessive foul smelling mucopurulent discharge, spotting and intermenstrual bleeding (depending on the existence of uterine or vaginal communications). Fever, dyspareunia, and a palpable abdominal, pelvic or vaginal mass (mucocolpos or pyocolpos) may also be present.

Dane

Klasyfikacja
Zespół wad wrodzonych

Synonimy
Double uterus and obstructed hemivagina syndrome
Niedrożna szczątkowa pochwa - anomalia nerki po tej samej stronie
Zespół Herlyn i Wernera
Zespół OHVIRA
Zespół podwójna macica - niedrożna szczątkowa pochwa
Zespół Wunderlicha
Herlyn-Werner syndrome
OHVIRA syndrome
Obstructed hemivagina and ipsilateral renal anomaly
Wunderlich syndrome

Kod ORPHA
3411

Kod OMIM
192050

Kod ICD10
Q51.2

Kod ICD11
LD2F.1Y

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*Źródło

orphanet