

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 Wenera

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

[*Źródło](#)

orphonet