3. APPROPRIATE CLINICAL MANAGEMENT OF WOMEN WITH HERLYN-WERNER-WUNDERLICH SYNDROME

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Introduction. Uterine didelphys with obstructed hemivagina and ipsilateral renal agenesis are assigned to the Herlyn-Werner-Wunderlich (HWW) syndrome. It generally reveals in adult women or a post-pubertal adolescent in whom hematometrocolpos generates a noticeable pain on the flank of the blind hemivagina.

Case presentation. A 14-year-old adolescent was admitted with right pelvic pain and dysmenorrhea. The symptoms have become more intense over the past two weeks. Menarche occurred at 13-years-old and the patient had a history of regular menses with cyclic pelvic pain. On first clinical examination, tenderness in the right lower abdomen and normal vulva, anus and hymen were noted. The ultrasound examination revealed a right pelvic mass (90/74 mm), double endometrial echoes, and hematocolpos. Uterine didelphys with stasis signs in the right corn, renal agenesis on the right, doubled kidney on the left were found on CT with contrast. After receiving the informed accord, the surgical team accomplished a vaginal septum incision and identified a hematocolpos, which was drained. The next follow-up visit was planned for one month later, when the patient revealed the absence of the symptoms.

Discussion. The explicit causation and pathogenesis of HWW syndrome are still not known, but are possibly associated to the role of Wolffian ducts in the evolution of internal genital organs and kidneys, and müllerian ducts fusions. For this reason, an embryologic anomaly of one Wolffian ducts may cause unilateral renal agenesis associated with obstructed hemivagina. It is not initially diagnosed, because of the regular menstruations from the blind hemivagina. The literature mentions a right-sided prevalence of the obstructed system. Despite such an advance in diagnostic technology, the significant heterogeneity of its clinical manifestation suggests it hinders quick recognition. The absolute treatment of HWW syndrome is the incision of as much of the obstructing vaginal septum as possible. Fertility is maintained with this surgery, which is not remarkably decreased in women with uterine didelphys.

Conclusion. The immediate and correct diagnosis of female genital organ disorders, including HWW syndrome, is obligatory to avert complications and maintain future fertility.