HERLYN-WERNER-WUNDERLICH SYNDROME AND DISTAL VAGINAL ATRESIA. TWO PARTICULAR CASES

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Background and aims Müllerian anomalies are rare, causing malformations of the female reproductive system as the Herlyn-Werner-Wunderlich Syndrome (HWWS) and distal vaginal atresia. These patients are usually asymptomatic until menarche. There may be associated with renal and urinary tract anomalies. We describe two cases of young girls with occluded vagina associated with urethrovaginal fistula.

Cases presentation The first patient is a 3-years-old asymptomatic girl with prenatal diagnosis of left renal agenesis. Ultrasoundography, performed 6 months prior to consultation, identified an anechoic cyst in hypogastrium. On genital examination, dismorphic girl with prenatal diagnosis of left renal agenesis. Ultrasoundography performed reveals urethrovaginal fistula. The second patient is a 4-years-old girl with occluded vagina suggesting distal atresia. Saline injected through the membrane eliminates by urethra. Cystoureterography confirms urethrovaginal fistula.

Conclusions The diagnoses of HWWS (uterus didelphys, unilateral low vaginal obstruction and ipsilateral renal agenesis) also known by OHVIRA and distal vaginal atresia in infancy and early childhood are unusual. The reported cases represent examples of premenarche diagnosis. In both patients, we detected urethrovaginal fistula, association not reported before in literature. Early diagnosis allows appropriate therapeutic management and prevents subsequent complications.

Methods We were studied retrospectively the medical records from these patients.

Results Out of the 46 cases, 60.8% were younger than 7 years, 41.3% were originated from urban environment; 27 cases were bilateral inguinal hernia; 14 presented various preoperative status/comorbidities (occlusive intestinal syndrome, acute dehydration, visceral adherence), 8 various surgical complications (bleeding, anaemia, scrotal swelling); the average length of stay was 3.69 days. Laparoscopic repair was significantly more frequent in patient younger than 7 years from urban environment (p = 0.001), but also with significantly more frequent comorbidities (p = 0.04) and post surgical complications (p = 0.003). Unilateral inguinal hernia develop fewer complications (p = 0.03) and require a shorter length of stay (p = 0.008). The patients without comorbidities presented a significant shorter length of stay (p = 0.05). The absence of surgical complications shortened significantly the length of stay (p = 0.01).

Conclusions Laparoscopic inguinal hernia repair may be considered a safer procedure, with fewer complications and shorter length of stay.

DISTAL VAGINAL ATRESIA. TWO PARTICULAR CASES

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