

## CASE REPORT

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# Neonatal hydrometrocolpos: a clinical case of difficult diagnosis

## Hidrometrocolpos neonatal: a propósito de un caso de difícil diagnóstico clínico

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### ABSTRACT

The diagnosis of fetal malformations continues to be a challenge for the obstetrician-gynecologist, despite advances in fetal medicine and ultrasonographic technology. Hydrometrocolpos is a dilatation of the uterus and vagina due to accumulation of secretions, product of genital obstruction usually associated with congenital malformations. We present the case of a female newborn with congenital hydrometrocolpos, whose diagnostic suspicion in pelvic studies was ovarian cysts, but it was due to the increase of bilateral hydronephrosis. At birth, it was decided to surgically decompress the urinary tract, at which time hydrometrocolpos associated with imperforate hymen was found.

**Key words:** Uterus, Hymen.

### RESUMEN

El diagnóstico de las malformaciones fetales sigue siendo un reto para el gineco-obstetra, a pesar de los avances en medicina fetal y en tecnología ultrasonográfica. El hidrometrocolpos es una dilatación del útero y vagina por el acúmulo de secreciones, producto de una obstrucción genital usualmente asociada a malformaciones congénitas. Presentamos el caso de una recién nacida con hidrometrocolpos congénito, cuya sospecha diagnóstica en los estudios pélvicos fue de quistes de ovario, pero se debía al incremento de una hidronefrosis bilateral. Al nacimiento, se decidió descomprimir quirúrgicamente la vía urinaria, momento en el que se encontró el hidrometrocolpos asociado a himen imperforado.

**Palabras clave:** Útero, Himen.

### INTRODUCCIÓN

The development of the female genital apparatus begins in the fifth week after fertilization from the Müllerian ducts, in a cranio-caudal direction, with the fusion of what will constitute the uterus and keeping separate the portions that will give rise to the fallopian tubes<sup>(1)</sup>. The hymen represents the union of the bulbs of the vaginal sinus with the urogenital sinus, which is usually perforated in embryonic life and, when it does not occur, can lead to vaginal malformations<sup>(2)</sup>.

Congenital malformations of the uterus occur in 0.5% of the population and vaginal malformations in 0.025%. Hydrometrocolpos or hydrocolpos is a rare condition, with an incidence of 1/16.000 female births. It consists of dilatation of the uterus and vagina due to accumulation of secretions resulting from glandular hypersecretion stimulated by maternal hormones, which is associated with obstructive congenital malformations, such as imperforate hymen, septum or vaginal atresia, due to anomalies of the urogenital sinus or cloaca<sup>(1,2)</sup>. Many cases are newly discovered in adolescence<sup>(1)</sup>. We present the case of a newborn with congenital hydrometrocolpos.

### CLINICAL CASE

We present the case of a female newborn born of normal vaginal delivery at 39 weeks, weighing 3,575 grams, Apgar 9 at one minute and 9 at 5 minutes. The clinical course of the pregnancy was uneventful, with a single umbilical artery and mild left pyelectasis of 9 mm at 24 weeks;

no additional pelvic lesion was described. In a subsequent ultrasound at 34 weeks, a 36 mm intrapelvic echolucent image was described, with an inner papilla and scarce peripheral vascularization on Doppler, which was classified as an ovarian cyst. In an ultrasound control by another observer one week later, the image of the same size and bilateral moderate pyelectasis of 11 and 12 mm was described.

At 36 weeks, a third evaluator mentioned similar characteristics and reported it as ovarian cyst versus renal cyst. It was assumed that the pyelectasia could be secondary to tumor compression and, as the fetus remained stable, it was decided to wait for delivery. On further evaluation, vaginal delivery was considered due to previous normal vaginal delivery and good fetal health conditions.

After birth, an ultrasound of the newborn's abdomen correlated with intrauterine findings of an ovarian cyst with irregular borders and intracystic papilla (Figure 1). However, CA 125 and alpha-fetoprotein levels were elevated: CA 125 112 U/mL (reference value less than 35) and AFP 17 928 ng/mL (reference value 0 to 7). Renal evaluation determined moderate bilateral pyelectasis (Figure 2).

Due to the findings described above, it was decided to perform surgery to solve the tumor problem and renal decompression by means of a laparoscopic approach. The finding was a very distended uterus without the presence of the referred cyst (Figure 3), so the intervention was converted to a mini-laparotomy, which found the hydrometrocolpos. The uterus was punctured and mucoid material was obtained, which allowed its decompression (figure 4).

FIGURE 1. NEONATAL ULTRASOUND WITH DIAGNOSIS OF OVARIAN CYST.



FIGURE 2. NEONATAL ULTRASOUND SHOWING BILATERAL PYELECTASIS.



FIGURE 3. LAPAROSCOPIC FINDING OF HYDROMETROCOLPOS.



At the end of the operation, the vaginal study was completed, with the finding of imperforate hymen. The evolution was clinically satisfactory, with remission of pyelectasia in the ultrasound control at 5 postoperative days.

## DISCUSSION

The first case of hydrometrocolpos was published in 1856, by Godefroy, who described it associated with urinary and intestinal obstruction<sup>(3)</sup>.



FIGURE 4. DRAINAGE OF THE HYDROMETROCOLPOS.



Imperforate hymen corresponds to 0.1% of girls born at term<sup>(4)</sup>, but it is rarely associated with hydrocolpos or hydrometrocolpos, closely related to estrogen levels, which usually remain low in the perinatal period and infancy, responsible for the production of mucus from the cervical glands; this only increases in adolescence, when it is diagnosed<sup>(2,4-7)</sup>. Additionally, hydrometrocolpos has been described due to urine accumulation secondary to vesico-vaginal fistulas<sup>(4)</sup>. Many times, the finding is associated with fluid protrusion through the obstructed vaginal introitus.

The most common neonatal diagnosis is congenital ovarian cyst<sup>(3,8)</sup> and, although ultrasound is the diagnostic tool of choice, images can be inconclusive, sometimes requiring the request of an MRI. Pelvic cystic images may correspond to gynecological (hydrometrocolpos, hydrocolpos, ovarian cyst, ovarian torsion), urinary (megacystitis) or intestinal (imperforate anus, gastrointestinal duplication) malformations<sup>(2,3,5)</sup>.

Imperforate hymen causes 50% of vaginal obstructions<sup>(4)</sup> and is associated with duplication of the ureter, polydactyly, imperforate anus, renal dysplasia, ectopic ureter and other malformations<sup>(7)</sup>.

The compressive effect is often associated with hydronephrosis and rectal obstruction for defecation, and even compression of the inferior vena cava. Any pelvic tumor in a newborn should lead to a detailed genital inspection<sup>(6,8)</sup>. An attempt has been made to look for a suspicious sign of hydrometrocolpos, "The Fjord sign" in Norwegian, which is described as a narrow

channel separating the dilated vagina from the uterus<sup>(5)</sup> and which would be differential diagnosis of ovarian cysts.

Hydrometrocolpos is sometimes associated with an autosomal recessive disorder, called Mckusick-Kaufman syndrome<sup>(3)</sup>.

Because of its low incidence, it is difficult to find worldwide series. Okoro reported 7 cases in Nigeria, from 7 to 27 days of life, associated with other malformations. Some patients underwent laparotomy and some underwent hymenotomy. Thirty percent of the cases were diagnosed intraoperatively, due to the low index of suspicion<sup>(7)</sup>.

As a final suggestion, every newborn should have a thorough vulvovaginal evaluation, especially if a congenital malformation of the genitourinary system is found.

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