

## **A case of hydrometrocolpos in a neonate**

**Nimantha Dhananjaya Vithanage<sup>1</sup>, Kapilani Jayasekara<sup>1</sup>, \*Imalke Kankanarachchi<sup>2</sup>, Milinda Jayawardena<sup>1</sup>**

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### **Introduction**

Hydrometrocolpos is a rare congenital anomaly that is caused by an abnormally dilated vagina/uterus due to obstruction of the genital tract<sup>1</sup>. Dilatation is caused by fluid that is secreted by the cervical glands under the influence of maternal oestrogen<sup>1</sup>. This can be complicated with hydroureteronephrosis, bowel obstruction and lower limb oedema as a result of compression of the bladder, bowels and venous system. Here we report a case with hydrometrocolpos, ambiguous genitalia, and recto-vaginal fistula complicated with bilateral hydroureteronephrosis.

### **Case report**

A premature baby (34 weeks and 5 days) with ambiguous genitalia was born to a 31-year-old primigravida mother by an emergency lower segment caesarean section (LSCS) due to fetal distress. Baby was resuscitated at birth. Her antenatal period was complicated with polyhydramnios and fetal ultrasound scan showed a large abdominal cyst at the third trimester. Baby required five inflation breaths at birth and the 5 minute Apgar score was 6 and the 10 minute Apgar score was 10.

The baby was transferred to the neonatal intensive care unit (NICU), Teaching Hospital Karapitiya for further management of underlying problems. Her

<sup>1</sup>Teaching Hospital Karapitiya, Sri Lanka, <sup>2</sup>Faculty of Medicine, University of Ruhuna

\*Correspondence: [imalke462@gmail.com](mailto:imalke462@gmail.com)

 <https://orcid.org/0000-0002-9351-2966>

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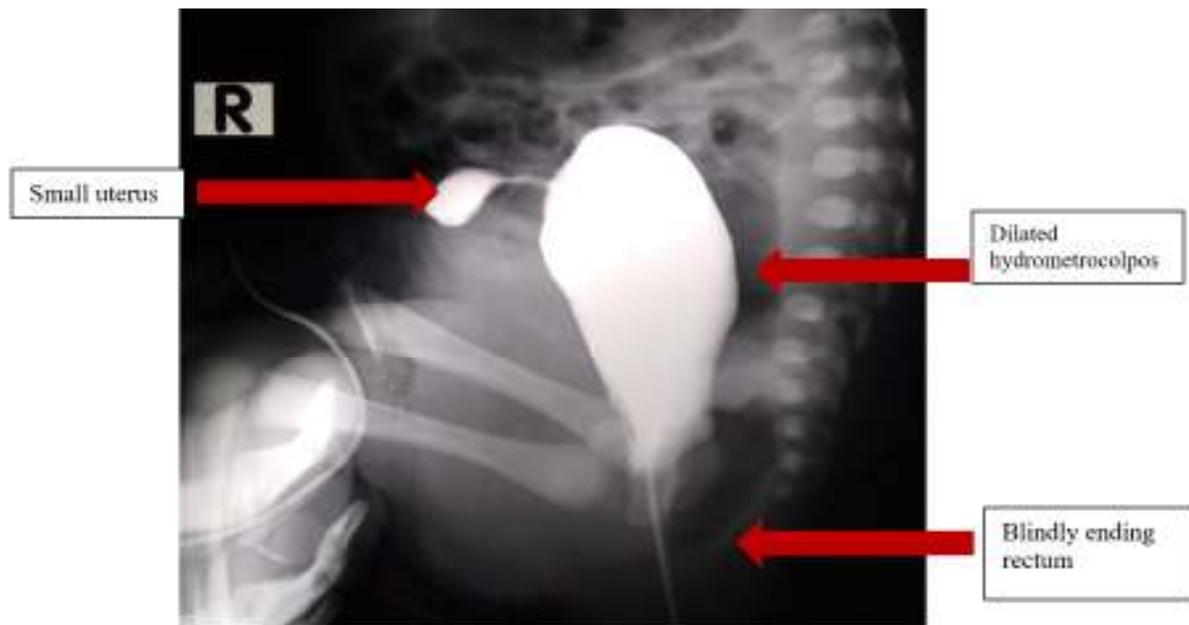
birth weight was 3.1kg and she did not have any facial dysmorphism. Baby had a 1cm sized phallus and the urethral meatus was situated in the base of the phallus. Scroto-vaginal folds were empty and bifid (Figure 1). No testicles were found even in the inguinal region. Perineal opening was situated more anteriorly. Straw coloured fluid was drained from both urethra and the perineal opening.



Figure 1: Baby with ambiguous genitalia

Post-natal ultrasound scan showed bilateral hydroureteronephrosis with a large pelvic mass. There was a difficulty in identifying the bladder, uterus and ovaries. Two 5 French gauge feeding tubes were inserted into urethral meatus and to the perineal opening and around 200ml of fluid was drained from the perineal opening. Subsequently, magnetic resonance imaging (MRI) of pelvis, micturating cystourethrogram (MCUG), intravenous urogram (IVU) and contrast study via perineal opening were done.

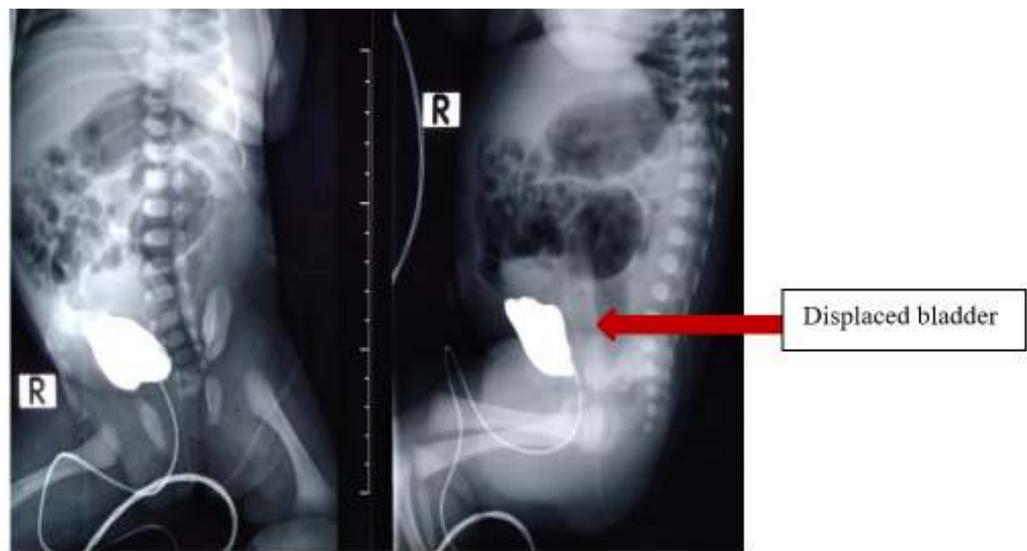
Contrast study through the perineal opening revealed a contrast filling large cystic lesion posterior to the bladder. However, contrast had not entered the rectum. (Figure 2)



**Figure 2:** Contrast study through perineal opening showing dilated hydrometrocolpos, small uterus and blindly ending rectum

Contrast study done through the urethral opening showed the anteriorly placed bladder. (Figure 3). Dilated vagina (hydrocolpos) was seen as the large cystic abdominal mass in the magnetic resonance imaging (MRI) of the pelvis and the uterus was seen as a small cystic structure superior to the vagina.

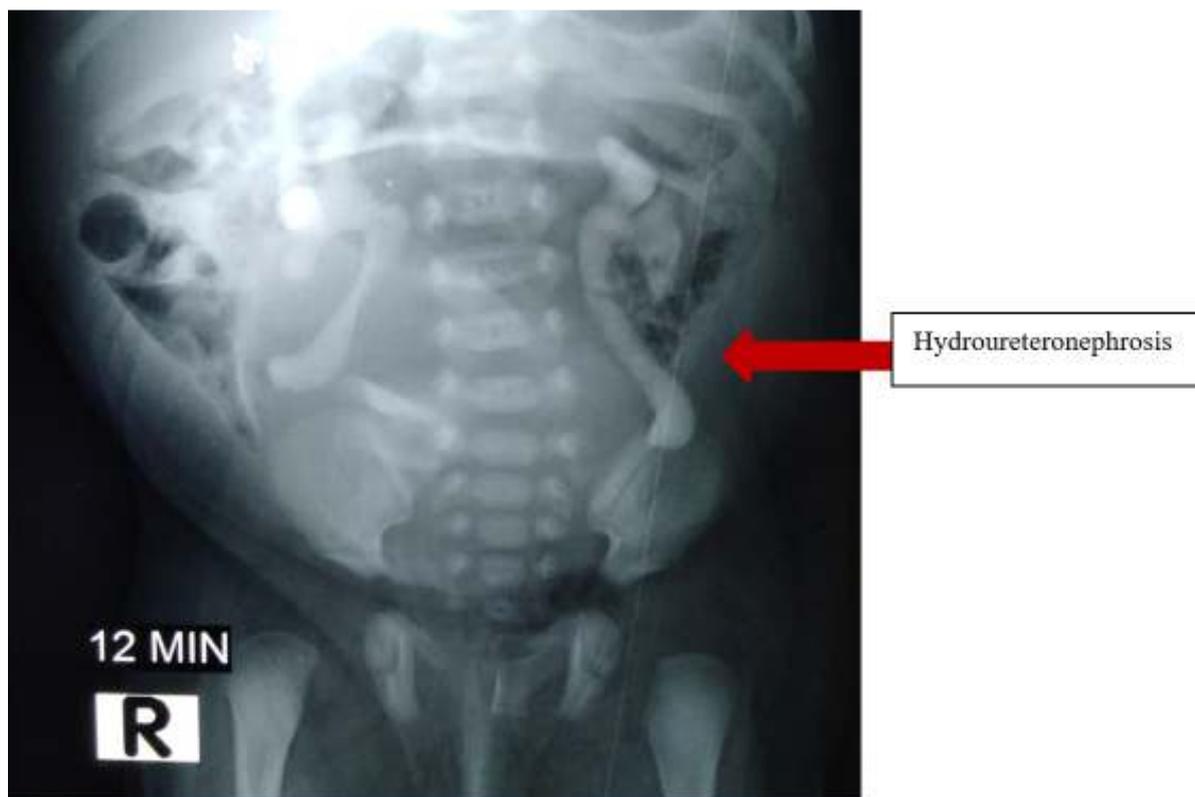
Bladder was anteriorly displaced and was compressed causing bilateral hydronephrosis. The rectum was not opening through an anal orifice and there was a recto-vaginal fistula into which it opened (Figure 4). IVU confirmed the presence of bilateral hydronephrosis (Figure 5).



**Figure 3:** Contrast inserted through urethral opening showing anteriorly displaced bladder



**Figure 4: Magnetic resonance imaging showing large hydrometrocolpos, small uterus and anteriorly displaced and compressed bladder**



**Figure 5: Intravenous urograph showing bilateral hydronephrosis**

Colostomy was performed at the age of 6 weeks and the baby is awaiting surgical reconstruction of the genital tract. Karyotyping revealed the genetic sex of the baby as XX. The cause for ambiguous genitalia was not identified and could be a structural malformation associated with hydrometrocolpos. We have excluded congenital adrenal hyperplasia

#### **Discussion**

Hydrometrocolpos is a rare disorder with genitourinary anomalies and a wide range of clinical presentations<sup>2</sup>. Most cases presented in the antenatal period with a pelvic mass in the ultrasound scan (USS). However, it is difficult to make a diagnosis only with an USS, a postnatal MRI being also needed<sup>1</sup>. Hydrometrocolpos is caused by the fluid

accumulation in vagina due to increased cervico-vaginal secretions under the influence of maternal oestrogen<sup>1</sup>. It is also associated with other malformations like imperforate hymen, imperforate anus, renal agenesis, cloacal anomalies, uterine anomalies and urogenital sinus<sup>3</sup>. Most common secondary complication is hydroureteronephrosis due to the obstruction of the bladder from the large mass<sup>2</sup>.

The definitive management requires early drainage of contents in the hydrometrocolpos in order to relieve the hydroureteronephrosis<sup>3</sup>. This could be done as an ultrasound guided procedure<sup>3</sup>. Stepwise surgical correction should be done later depending on the associated anomalies. The overall prognosis in hydrometrocolpos is good<sup>4</sup>. However, pyocolpos, sepsis, hydroureteronephrosis and bowel obstruction can cause increased mortality<sup>3</sup>. Early diagnosis and intervention is the key for better prognosis.

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