Looking through the keyhole at Fetal Megacystis

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INTRODUCTION

Megacystis is a sonographic feature of an abnormally large fetal bladder for gestational age. The incidence is estimated to be 0.38% globally with predominantly male fetuses affected. Even with treatment options and early diagnosis this condition has a relatively poor prognosis.

The fetal kidneys produce urine from around 10 weeks. During the first trimester from 10-14 weeks the normal sagittal diameter of the bladder is <7mm. A measurement above this is classified as megacystis. The diagnosis is made in the second and third trimester when there is an enlarged fetal bladder that fails to empty over a 45-minute period or a fetal bladder that measures >10% of the craniocaudal length of the fetus. The prognosis and outcomes of megacystis are varied because of the wide spectrum of aetiologies involving the kidneys and other structures of the urinary system.

This following is a case of megacystis caused by lower urinary tract obstruction (LUTO).

PATIENT BACKGROUND

37-year-old Para 0+0 presented with vaginal bleeding in early pregnancy at 10 weeks. She had a history of asthma and a BMI of 37. There was no other relevant medical, surgical or gynaecological history. Trans Vaginal ultrasound (TVS) was the modality of ultrasound in view of early gestation and BMI.

CAUSES OF MEGACYSTIS

Posterior urethral valves **(PUV's),** a congenital obstruction of the posterior urethra (also referred to as lower urinary tract obstruction or **LUTO**), account for **47%** of overall cases of megacystis**.**

Other causes are chromosomal (Trisomy's 18 and 21) /genetic and other associated conditions (**33%),** Vesico urethral reflux (**VUR**), congenital malformation of the vesico urethral junction (**19%)** and **1%** of cases occur with an unknown cause and these can spontaneously regress.

***Figure 1*. The Fetal bladder is prominent at 10 weeks gestation in the presence of a cord cyst, a follow up scan was arranged for 12 weeks.**

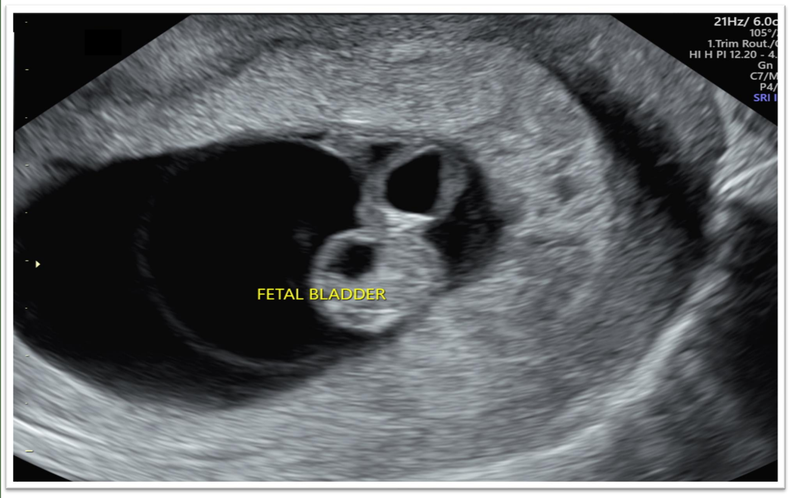

***Figure 2*. Transverse view of cord insertion site. Cord cyst and fetal bladder, note echogenic area in the base of the cord insertion site.**

ULTRASOUND EXAMINATION

TVS was performed at 10 weeks for bleeding in early pregnancy and showed a viable intrauterine pregnancy, equal to dates. Fetal bladder was visible, with a cord cyst present with echogenic area below it suggestive of bowel herniation. Nuchal translucency measured 1.7mms (normal). Figures 1, 2 and 3.

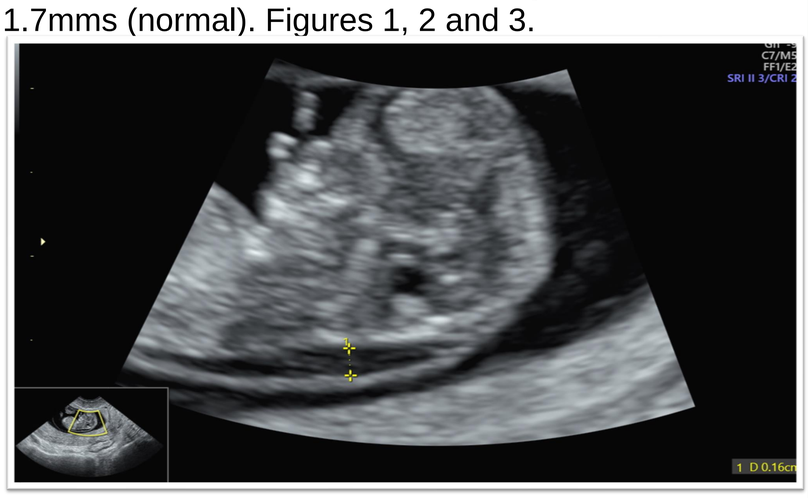

***Figure 3*. Nuchal translucency measurement of 1.7mm which is normal.**

A further scan at 12 weeks showed a large fetal bladder measuring 22mm x 16mm x 15mm, with the cord cyst larger than previous scan. Fetal stomach and kidneys were visualised. Bowel appeared to be echogenic. No evidence of bladder exstrophy noted at cord insertion site. Figures 5,6 & 7.

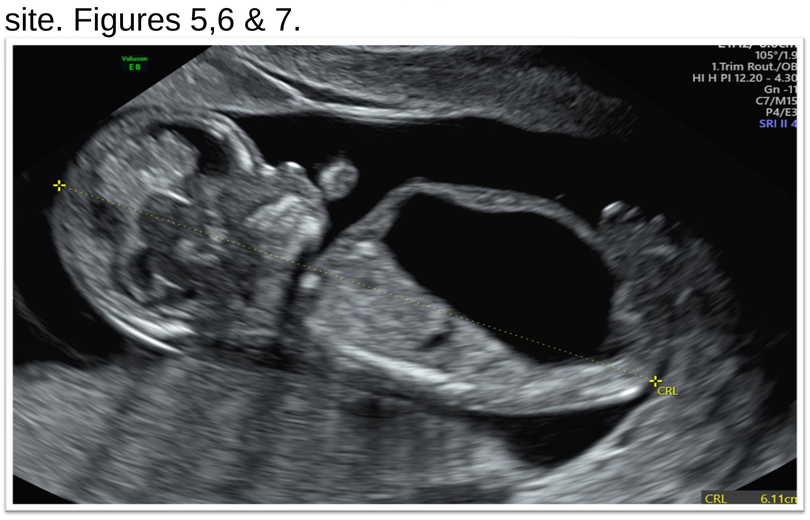

***Figure 4.* sagittal view of fetus at 12 weeks showing bladder encroaching into thoracic cavity.**

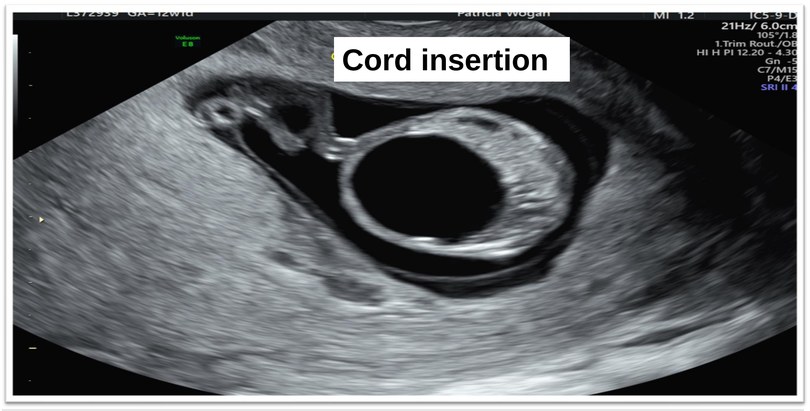

***Figure 5*. Cord insertion at 12 weeks fetal kidneys visible and note surrounding echogenic bowel.**

DIFFERENTIAL DIAGNOSIS & PROGNOSIS

Cloacal malformation during early development with other associated abnormalities of the urinary and gastrointestinal tract can also be a cause. There are other rare conditions where megacystis can be present like Prune belly syndrome and Megacystis microcolon hypoperistalsis syndrome.

The prognosis is determined by the gestational age at discovery, the cause and other associated conditions. As gestation advances there is continued unrelieved obstruction of the urinary tract as the fetal bladder gets bigger. The renal architecture can be irreversibly damaged and the longer this is happening the worse the outcome for the fetus. A combination of renal damage, oligohydramnios and underdevelopment of the lungs leading to severe lung hypoplasia can mean a very poor prognosis. Amniotic fluid volume is the most valuable predictor of renal function and fetal survival. Sonographic features along with karyotyping can determine diagnosis and prognosis.

ULTRASOUND EXAMINATION

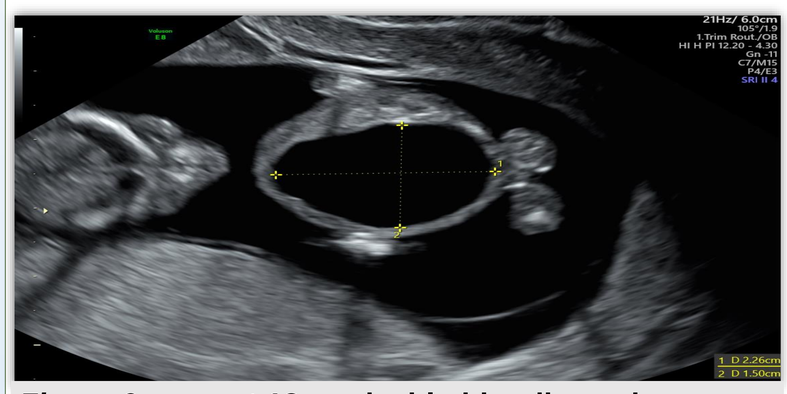

***Figure 6.* scan at 12 weeks bladder dimensions 22mm x 15mm x 16mm.**

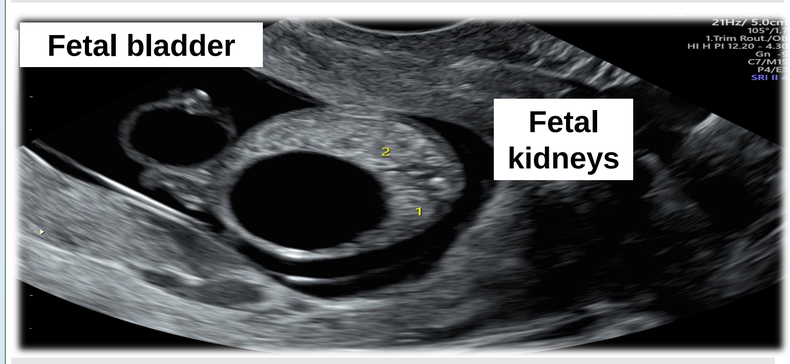

***Figure 7*. Transverse view of fetal abdomen at 12 weeks with bladder, bowel and echogenic kidneys in view**

The patient was referred to a fetal medicine specialist at 15 weeks. Bladder noted to be very large with a keyhole appearance typical of LUTO seen in figure 8 below.

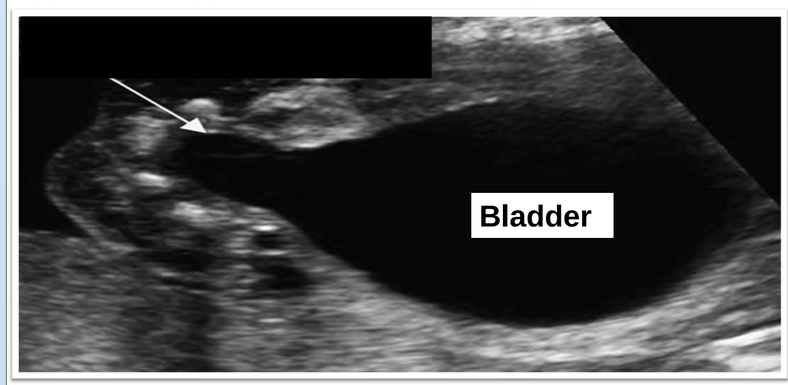

***Figure 8.* Keyhole shape at uteric junction**

Following tests no chromosomal abnormalities were detected. A scan at 16 weeks with the FMS showed **severe oligohydramnios**. Bladder noted to be very large at this scan >40mm with a keyhole appearance. The appearance of the fetal bladder is suggestive of LUTO and a poor prognosis expected, and options discussed with parents. This was a difficult scan due to oligohydramnios and the maternal BMI. The poor quality of the images in figure9 and 10 are reflective of this.

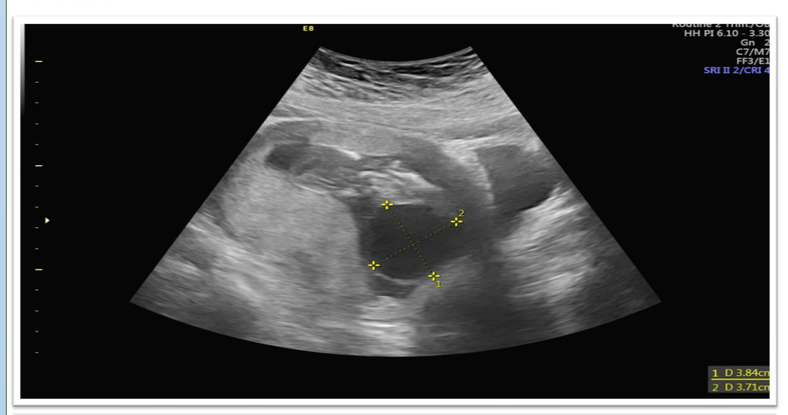

***Figure 9*. Large fetal bladder & oligohydramnios**

***Figure 10*. Difficult to assess biometry**

Following counselling the patient opted for a termination of pregnancy and she travelled to the UK at 17 weeks for the procedure.

References

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