Prenatal diagnosis of fetal lower urinary tract obstruction presenting as an abdominal mass in a twin pregnancy using three-dimensional ultrasound with "Fly thru" technology: A case report*

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ABSTRACT

We report a case of a twin pregnancy, wherein one twin presented with an abdominal cyst since 12 weeks' gestational age. Upon referral at 21 weeks' gestational age, three-dimensional ultrasound with Fly thru technology was used to aid in the identification of the etiology and nature of the mass. Once megacystis was confirmed, serial vesicocentesis and urine biochemistries were used to direct the management. This shows the potential of Fly thru technology in aiding the clinician in studying fetal congenital anomalies. This can help guide the diagnosis and provide earlier and timely management of such cases.

Keywords: Fetal abdominal mass, fetal megacystis, Three-dimensional Ultrasound with Fly Thru technology, Case Report

INTRODUCTION

The presence of fetal abdominal masses, in general, is quite rare. Due to paucity of available data in literature, diagnosis and management is usually perplexing.[1]

In evaluating fetal abdominal masses, the suspected system from which the mass originates as well as the fetal gender should be identified in order to direct the management. The most commonly involved organ systems are the urogenital and gastrointestinal systems.[2]

Imaging modalities including ultrasound (two-, three- and four- dimensional sonography) and fetal magnetic resonance imaging (MRI) have traditionally been used in characterizing and diagnosing these lesions.

In cases wherein there is difficulty in diagnosis using the aforementioned modalities, 3D Fly Thru technology provides a novel way of imaging these lesions, allowing earlier diagnosis, directing management and improving the prognosis.

CASE

Patient is a 29-year-old woman, Gravida 3 Para 2, with two previous low transverse cesarean deliveries, who was diagnosed with a dichorionic-diamnionic twin pregnancy at 8 weeks and 6 days' gestational age (Figure 1). Her past medical and family histories were unremarkable. There was an episode of undocumented fever and generalized rashes during the early first trimester, which spontaneously resolved. The rest of her first trimester course was unremarkable.

At 12 weeks and 4 days' gestational age, there was an incidental finding of an intra-abdominal cystic mass in Twin A, measuring 5 centimeters in diameter. She was advised a repeat scan after a month. At 18 weeks and 1 day's gestational age, there was persistence of the intra-abdominal cyst (Figures 2 and 3), characterized as unilocular and sonolucent, measuring 5.57 x 4.36 x 4.69

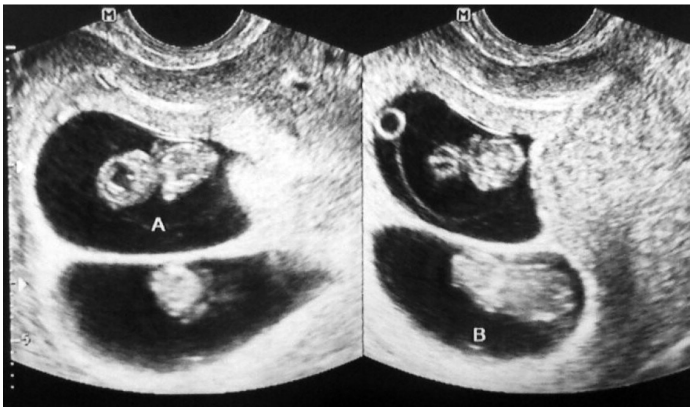

Figure 1. Transvaginal scan at 8 weeks and 6 days age of gestation showing two gestational sacs each with an embryo, with thick dividing membrane and lambda sign (signifying a dichorionic-diamnionic pregnancy). Embryos were assigned as Twin A and Twin B. There was no visualized intra-abdominal mass on either twin during this scan.

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cm (60 mL in volume). There was no color Doppler studies done on the mass. She was then referred to our service for further evaluation and management.

Patient was seen by our service at 21 weeks' gestational age. Transabdominal ultrasound revealed that Twin A had a cystic sonolucent mass occupying and distending the abdomen. It measured 14.3 x 12.7 x 14.1 cm (1.2 liters in volume), thick-walled, negative color flow, compressing the fetal lungs and heart (Figures 4 and 5). There was noted oligohydramnios (single vertical pocket of 0.8 cm) in the said twin. Due to the size of the abdominal mass and presence of oligohydramnios in Twin A, there was difficulty in evaluating the gender, extremities and other intra-abdominal structures. Twin B had no gross congenital anomaly and was noted to be male.

Given the shape, size and progressive enlargement of the fetal abdominal cyst starting at 12 weeks' gestation, the consideration at this time was a dilated bladder or megacystis versus an ovarian cyst. Three-dimensional volume data acquisition was done using Fly Thru navigation, utilizing the Aplio 500 system (Toshiba Medical Systems) equipped with a PVT-675MV 2.8-7.2 MHz probe.

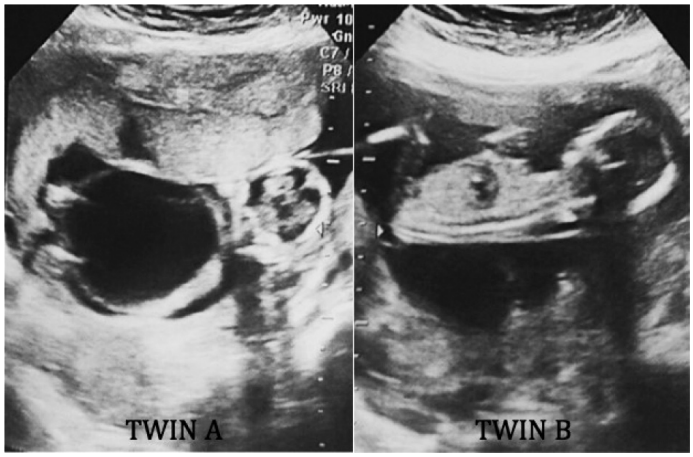

Figure 2. Transabdominal scan at 12 weeks and 4 days' gestational age showing Twin A with a cystic mass occupying the abdominal cavity and Twin B with no gross anomaly seen.

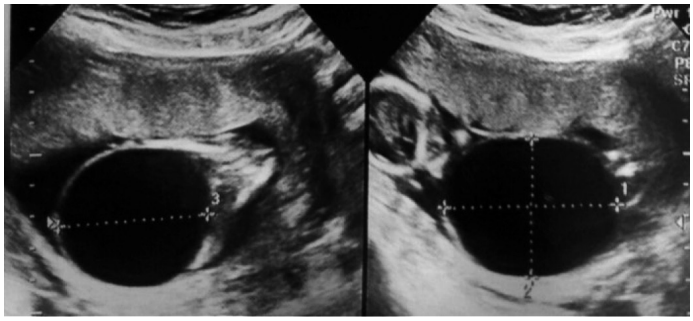

Figure 3. Transabdominal scan done at 18 weeks and 1 day's gestational age showing the measurement of the intra-abdominal mass in Twin A. The cyst was unilocular and sonolucent, measuring 5.57 x 4.36 x 4.69 cm (60 mL in volume). There were no Doppler studies done on the mass.

The arrow navigated in a cephalocaudad direction using the autopilot mode. The cystic mass had smooth inner walls. Upon approaching the most inferior portion of the mass, an opening was visualized. Upon navigation, this opening ended in a blind pouch. This was interpreted as a probable obstructed urethral os. (Figure 6) No other opening was visualized.

With the primary assessment of megacystis in Twin A, there was a consideration of lower urinary tract obstruction, the most common of which is a bladder outlet obstruction.[9] Primary considerations for the cause of the obstruction was posterior urethral valves or urethral atresia, given that megacystis presented in the early second trimester.[9] The gender of the affected fetus was still unknown at this time. The patient was advised vesicocentesis of Twin A to relieve compression of the fetal

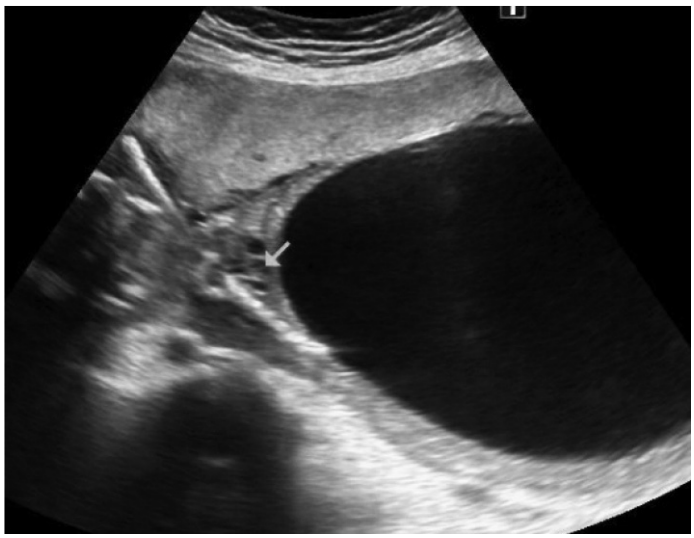

Figure 5. Transabdominal scan at 21 weeks' gestational age showing the cross section of the compressed fetal thorax of Twin A.

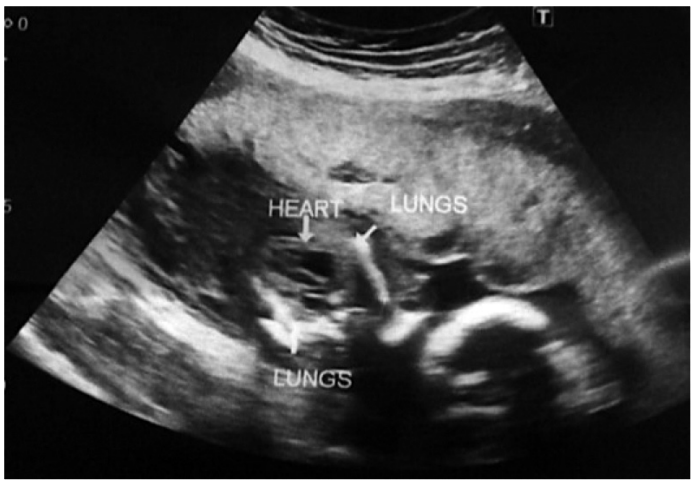

Figure 4. Transabdominal ultrasound at 21 weeks' gestational age, showing the sagittal view of the cystic sonolucent mass occupying and distending the fetal abdomen of Twin A. There was compression of the fetal thorax (small arrow pointing the fetal heart)

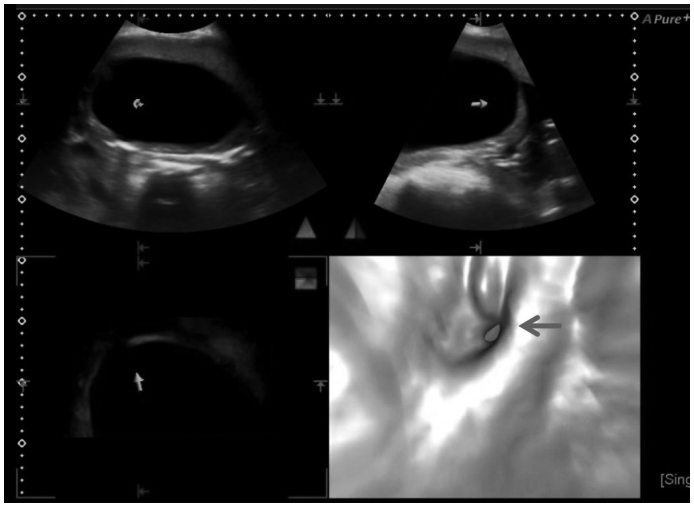

Figure 6. Three-dimensional volume data acquisition with Fly Thru navigation through fetal abdominal mass at 21 weeks' gestational age. The arrow navigated in a cephalocaudad direction using the autopilot mode. Upon approaching the most inferior portion of the mass, an opening was visualized (blue arrow). This was interpreted as the urethral os.

thorax and prevent pulmonary hypoplasia and to prevent mechanical injury to the fetal kidneys. Urine biochemistries would be used to evaluate the renal function, which would direct the management.

At 22 weeks' gestational age, vesicocentesis of Twin A was done. Pre-aspiration sonographic bladder volume of twin A (was 1.4 liters. Under aseptic technique, ultrasound-guided vesicocentesis was done using a gauge 21 spinal needle (Figures 7 and 8). The total fetal urine aspirated was 1.31 liters. It was clear light yellow to amber in color (Figure 9). Post-aspiration sonographic volume of the bladder was 75 mL.

The decompressed cystic structure, on color Doppler, revealed the presence of the umbilical arteries laterally, confirming that the pathologic structure was the fetal bladder (Figure 10). A congenital scan was done to investigate the other fetal structures. There was a questionable cardiac interventricular septal defect visualized in Twin A. The fetal spines were intact and the remaining intra-abdominal organs were grossly normal. The fetal thoracic circumference (measured at 13.12 cm) was at the 25th percentile for gestational age. There was still difficulty in identifying the fetal sex due to oligohydramnios.

Laboratory work-up of the fetal urine of Twin A were as follows: total protein 7.4 mg/dL, sodium 92 mmol/L, chloride 64.8 mmol/L, calcium 5.26 mg/dL, osmolality 187 mOsm/L and creatinine 0.44 mg/dL. These were consistent with good renal function. She was referred to a Pediatric urologist for insertion of a vesicoamniotic shunt. Inserting a shunt would decompress the fetal bladder, relieve compression of the fetal thorax and increase the

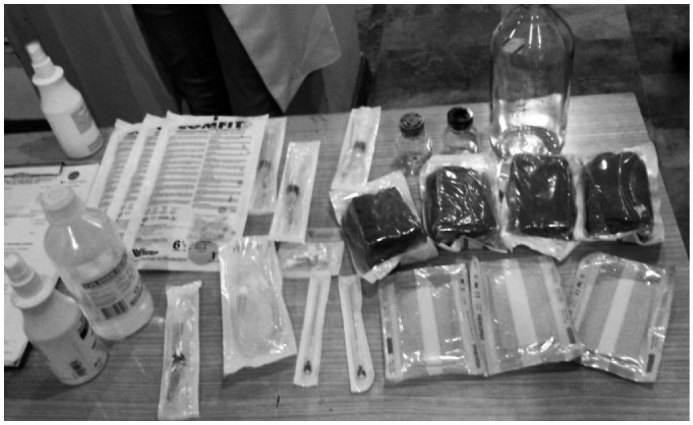

Figure 7. Materials used for the ultrasound-guided vesicocentesis.

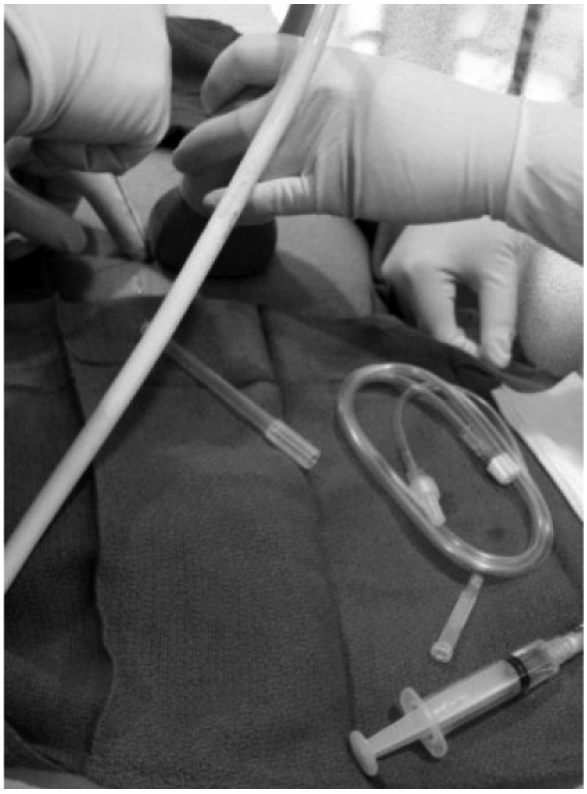

Figure 8. Ultrasound-guided vesicocentesis of Twin A under aseptic technique.

Figure 9. Fetal urine aspirated, showing clear light yellow to amber fluid.

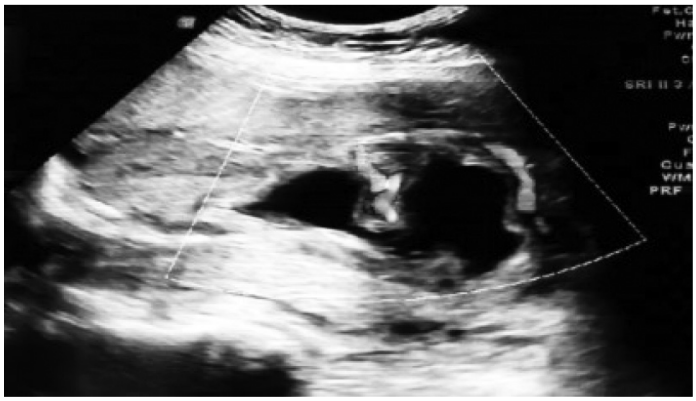

Figure 10. Ultrasound image of the fetal bladder of Twin A post-vesicocentesis. Post-aspiration picture demonstrates the two umbilical arteries on color Doppler, confirming that the structure aspirated was the fetal bladder.

amniotic fluid volume. At this age of gestation, the lung development was at the canalicular stage, thus, improving the amniotic fluid volume by inserting the shunt may help prevent pulmonary hypoplasia. Decreasing the distention of the bladder would also prevent mechanical injury to the fetal kidneys. These advantages of inserting a shunt would help improve the chances for fetal survival.

While awaiting the availability of the vesicoamniotic shunt, the patient was advised serial ultrasound to monitor the lower urinary tract obstruction, amnioinfusion to correct the oligohydramnios and possible serial vesicocentesis.

At 22 weeks and 3 days' gestational age, sonographic bladder volume of Twin A was 304 mL. There was also a note of a distended urethra, demonstrating the keyhole sign (Figures 11 and 12). It was also noted that right and left renal pelves were dilated, with the anteroposterior diameters measuring 9 mm and 8 mm, respectively. (Figure 13) A fetal 2D echocardiography was also done at this time to confirm the presence of a cardiac pathology in Twin A. It revealed a morphologically normal fetal heart with normal outflow tracts.

At 23 weeks' gestational age, bladder volume of Twin A was 599 mL. There was also visualization of the kidneys, which were both hyperechoic and multicystic. Twin A was also noted to have ascites and oligohydramnios (single vertical pocket 0.8 cm). There was no gross abnormality seen in Twin B.

On ultrasound-guided vesicocentesis, aspiration of 520 mL of clear yellow urine was done, followed by amnioinfusion of twin A. Due to difficulty in performing amnioinfusion, only 35 mL of warm saline was infused. Post-aspiration sonographic bladder volume was 74 mL. The single vertical pocket of Twin A post-amnioinfusion was measured at 1.1 cm.

Laboratory results of the fetal urine were as follows: creatinine 0.57 mg/dL, osmolality 195 mOsm/L sodium

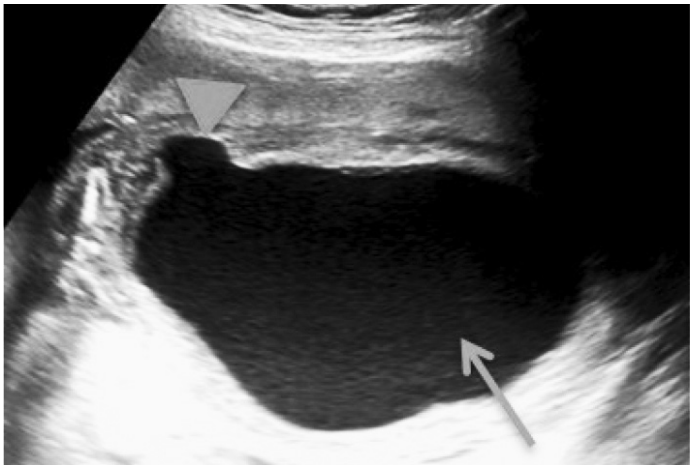

Figure 11. Transabdominal scan showing the sagittal view of Twin A at 22 weeks and 3 days' gestational age. The image shows the presence of megacystis (blue arrow) with what appears to be a local outpouching, the dilated urethra (red arrowhead), illustrating the so-called "keyhole sign".

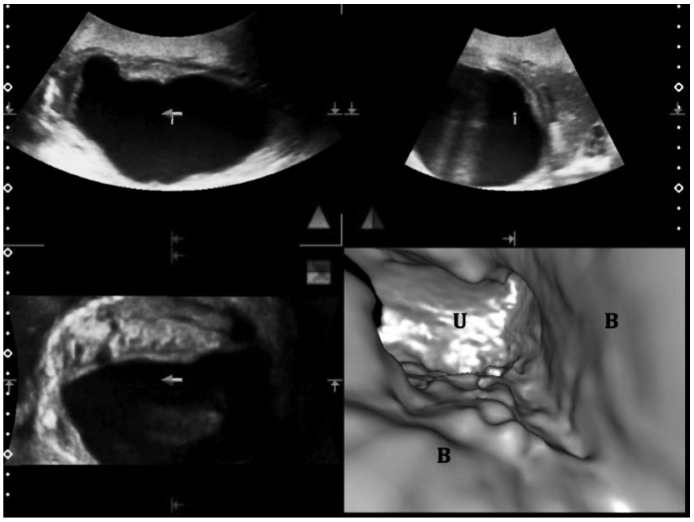

Figure 12. Three-dimensional scan with Fly thru at 22 weeks and 3 days' gestational age showing the bladder mucosa (B), with the navigation arrow pointing caudally, showing the 3D rendition of the distended urethra (U), illustrating the keyhole sign.

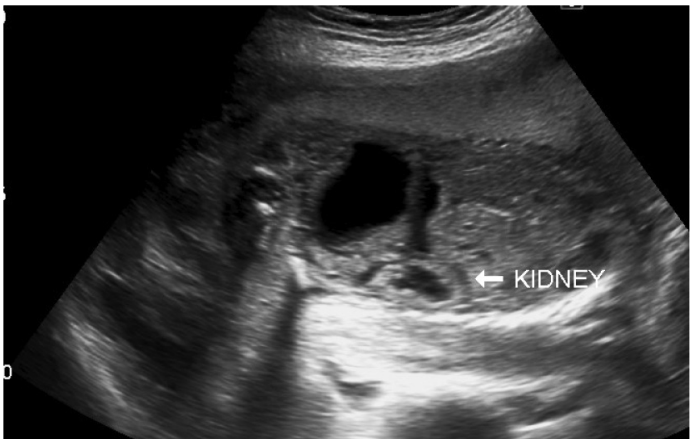

Figure 13. Sagittal view of Twin A at 22 weeks and 3 day's gestational age showing the dilated left renal pelvis. (white arrow pointing the left kidney

94 mmol/L, chloride 66 mmol/L, calcium 5.13 mg/dL, showing good renal function.

Six hours post-vesicocentesis, there was rupture of membranes, noted as presence of minimal watery discharge per vagina. On examination, fetal heart tones of both twins were normal, with absence of uterine contractions. Rupture of membranes was confirmed by a positive Ferning test. On internal examination, cervix was soft, long and closed. Assessment was preterm prelabor rupture of membranes. C-reactive protein (CRP) was initially elevated at 22 mg/L with normal white blood cell (WBC) count and differential count. She was started on intravenous Ampicillin and oral Erythromycin. Patient was given antenatal corticosteroids to accelerate fetal lung maturity. Serial WBC count and CRP showed decreasing trend, with normalization at the 5th hospital stay.

The plan at this time was for conservative management, continue oral antibiotics for 2 weeks and to prolong the pregnancy to increase the chances of survival for both fetuses. Serial antenatal ultrasound evaluation of the affected fetus was advised, with definitive treatment of the obstructive lesion to be done postnatally.

Interval history was unremarkable, with absence of signs and symptoms of maternal infection. Serial ultrasound showed progression of the degree of megacystis, bilateral hydronephrosis and cystic renal dysplasia in Twin A. Twin B showed normal interval growth, with no abnormalities noted. It was discussed with the patient that the prognosis for Twin A was not favorable and that the goal of prolonging the pregnancy would be more for improving the chances of survival for Twin B.

At 27 weeks' gestational age, patient noted presence of a cord-like structure per introitus. Cord prolapse was confirmed with an internal examination, revealing a cervical dilatation of 2 cm. Twin A was bradycardic at 77 bpm and Twin B had normal heart tones. She underwent emergency cesarean section. Upon delivery of Twin A, there was inadvertent rupture of the fetal abdomen, with egress of clear fetal urine. Twin A was delivered without cardiac activity, with a birthweight of 520 grams. Twin B was delivered alive, with birthweight of 860 grams, apgar score of 8,9, maturity of 26 to 27 weeks by Ballard scoring.

On gross examination of Twin A, the abdominal wall distended and thinned out. The bladder mucosa was distended, with smooth inner walls. The urethral os was visualized at the inferior end (Figure 17). Fetal fascies was flattened and fetal extremities were with contractures, both possibly secondary to the long-standing oligohydramnios. The fetus was noted to be male. No other gross congenital anomaly was noted.

The primary consideration for cause of lower urinary tract obstruction was urethral obstruction, secondary to either a urethral atresia or posterior urethral valves. There was no autopsy done to confirm the assessment.

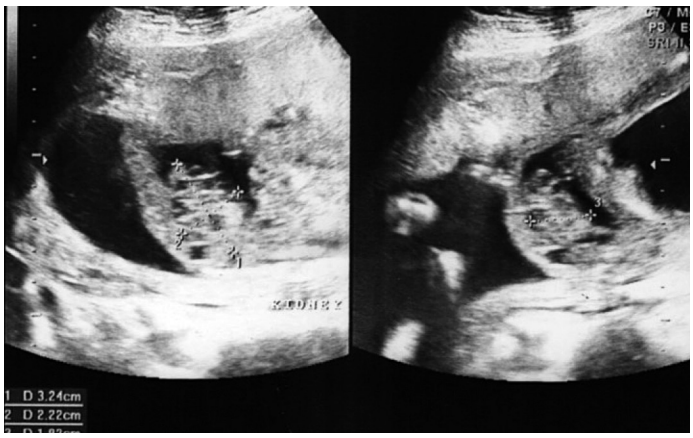

Figure 14. Transabdominal scan at 23 weeks (sagittal and transverse view) showing the right kidney of Twin A. Both kidneys were hyperechoic and multicystic.

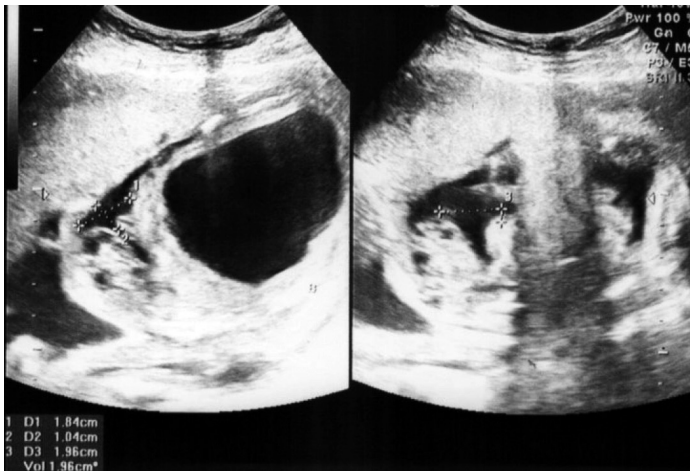

Figure 15. Transabdominal scan at 23 weeks (sagittal and transverse view) showing the presence of anechoic fluid in the abdominal cavity, signifying ascites, in Twin A.

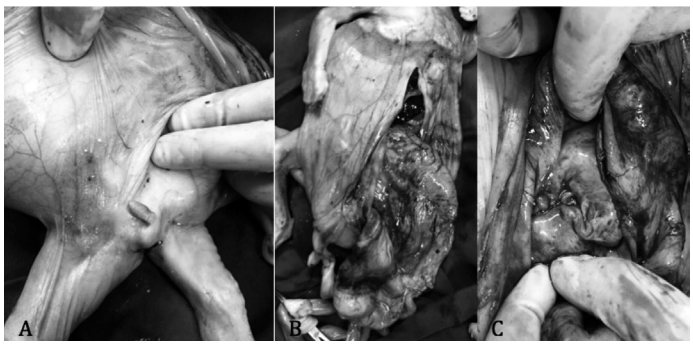

Figure 16. Gross picture of Twin A upon delivery: A showing the male genitalia, B showing the bladder that was stretched out and vascular and C showing the bladder mucosa and urethral os.

Twin B stayed at the neonatal intensive care unit, managed for sepsis, respiratory distress syndrome and bronchopulmonary dysplasia and was discharged stable at

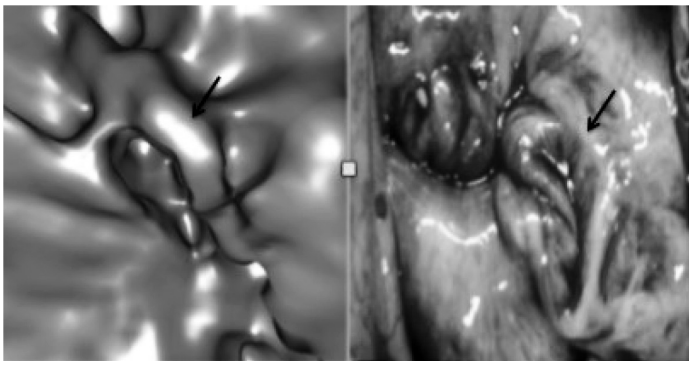

Figure 17. Comparison of the Fly Thru image (left) and the gross image (right) of the fetal bladder, from inside the bladder mucosa, showing the urethral os (black arrow).

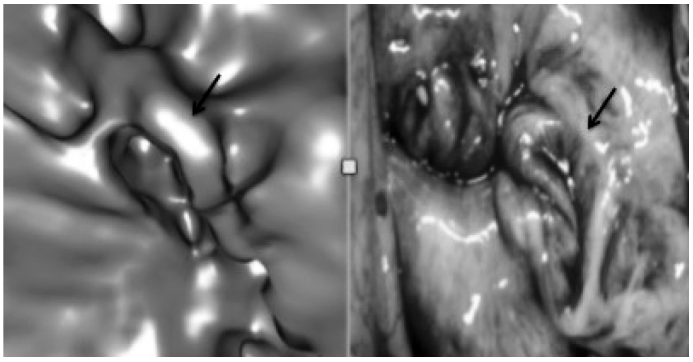

Figure 18. Comparison of Conventional 3D and Fly Thru perspective 3D imaging (From Toshiba Aplio 500 Brochure, Toshiba Medical Systems 2012)

the 10th week of life.

CASE DISCUSSION

In a fetus presenting with an abdominal mass, the key to differential diagnosis relies primarily on both the suspected system from which the mass originates and the fetal gender. In this case, both were not initially identifiable, making the diagnosis difficult.

Localizing the mass when it was still relatively small would have helped in the diagnosis, since the fetal bladder would initially be seen as early as the 9th to 10th week of gestation[9] in the lower abdomen, with the umbilical arteries found laterally on color Doppler. The relationship of the mass to the adjacent organs and normality of the other organs could also aid in the diagnosis. Upon reviewing the images of the previous scans done starting at the 12th week of gestation, the mass already occupied the whole abdominal cavity, obscuring the other abdominal organs, making it difficult to identify its etiology. There was no identification of the fetal sex during these scans.

At 21 weeks' gestational age, upon referral to our service, the morphologically normal twin was identified to be male, but the gender of the affected twin could not be determined due to presence of oligohydramnios. Taking into consideration the size, shape, and evolution of the abdominal cyst, the differential diagnoses at this time included a dilated fetal bladder or an ovarian cyst.

Antenatal ultrasound has high sensitivity in detection of intra-abdominal lesions, but has limited success in differential diagnosis of these masses.[3-6] Fetal magnetic resonance imaging (MRI) has superiority in elaborating equivocal sonographic findings and establish a differential diagnosis after the introduction of ultra-high-speed magnetic resonance imaging technique.[7]

Traditionally, if there is a dilemma in the diagnosis of a fetal abdominal mass on imaging studies, aspiration and chemistry of the contents of the mass may help in identifying the lesion. This holds possible complications, such as rupture in case the mass happens to be an ovarian cyst.

In the presented case, 3D ultrasound with Fly Thru technology (Toshiba Aplio 500, Toshiba Medical Systems) was able to strengthen the diagnosis of megacystis. The visualization of an opening at the caudal pole of the mass made ovarian cyst a less likely etiology of the mass. Once megacystis was diagnosed, the initial plan for the affected fetus was for insertion of a vesicoamniotic shunt if fetal urine biochemistries show good prognostic values, signifying good renal function.

Vesicoamniotic shunting which is performed under ultrasound guidance using a pigtail shunt, with one end of the shunt within the fetal bladder and the other in the amniotic space, would create a tract bypassing the lower urinary tract obstruction. This would relieve compression of the fetal thorax by decreasing the restrictive effect of the distended bladder and increase the amniotic fluid volume, preventing pulmonary hypoplasia, which largely contributes to the morbidity and mortality from fetal urinary tract obstruction.[8] Decreasing the distention of the bladder would also prevent mechanical injury to the fetal kidneys, preventing renal dysplasia and preserve renal function. These effects of vesicoamniotic shunt insertion would improve the chances for fetal survival and improve the prognosis.

While waiting for the availability of the shunt, serial vesicocentesis and amnioinfusion was planned. It was at the 23rd gestational weeks that there was premature prelabor rupture of membranes, precluding the insertion of the vesicoamniotic shunt. The plan at this time was serial antenatal ultrasound evaluation of the affected fetus, with definitive treatment of the obstructive lesion to be done postnatally.

Any dilatation of the urinary tract should be investigated to answer the following questions: the origin of the dilatation, presence of other congenital anomalies

and the prognosis of the malformation.[9] In cases wherein there is delay or failure to diagnose lower urinary tract obstruction, the bladder would be increasingly distended eventually filling the whole abdomen, resulting in retrograde pressure causing dilated ureters bilateral hydronephrosis and renal dysplasia.[10] Features signifying poor prognosis in urinary tract obstruction include early onset, persistently obstructed bladder, oligohydramnios and secondary lung hypoplasia.[9] These were noted in the presented case.

In a study done by Osborne et al., they retrospectively studied 20 cases of fetal megacystis diagnosed sonographically. They were able to differentiate posterior urethral valve, urethral atresia, prune belly syndrome and megacystis-microcolon-intestinal hypoperistalsis syndrome based on evidence of a keyhole sign, bladder thickness, amniotic fluid index, and fetal sex. They utilized 3D and 4D sonography, Doppler angiography, tomographic ultrasound imaging, virtual organ computer-aided analysis, and automatic volume calculation as part of the detailed fetal anatomic survey.[11]

Fly Thru is a new technology that adds cross-sectional ultrasound information to the plain surface data, making it a tool for exploring lesions and ingrowing masses. It can navigate manually or automatically through fluid-filled cavities, ducts and vessels. Compared to conventional 3D imaging, it utilizes perspective 3D imaging, producing greater depth perception which, together with the animated reconstruction, gives the appearance of virtual endoscopic imaging.[12] Its advantage includes the ability go around the area of interest. It was able to navigate inside the cavity, which in this case was the fetal bladder, making it a form of virtual fetal cystoscopy. It served as an adjunct in diagnosing the fetal megacystis, helping direct the management of the case.

On review of literature, since the advent of 3D Fly Thru technology, case reports and case series were done in the field of Obstetrics and Gynecology to study its use in investigating intrauterine masses[13,14], tube patency and the fluid-filled amnion with the embryo or fetus inside. Besides a case report studying the fetal trachea[11] using this technology, there is paucity of data in its use for studying congenital anomalies.

This form of technology may significantly improve prenatal diagnosis and subsequent management of fetal congenital anomalies. Early and timely proper diagnosis is important in managing many congenital anomalies, and in this case, Fly Thru technology was able to assist in directing the management.

SUMMARY AND CONCLUSION

In cases wherein there is difficulty in diagnosis of a fetal abdominal cyst, three-dimensional Fly Thru technology offers itself as an adjunct to the traditional methods of fetal imaging. It involves the creation and evaluation of fluid-filled structures using spatial information, producing a virtual view of the cavity (in this case, the fetal bladder) being explored. This new technology has the potential to provide information that may allow for earlier diagnosis and management of fetal congenital anomalies, and has the potential to significantly improve prognosis. ■

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