

POSTER - USG (USP)

A Rare Case of Meconium Peritonitis Due to Ileal Atresia with Volvulus with Intrauterine Perforation with Hemoperitoneum

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Case: Mrs. J.M 22 years admitted with complaints of 37 weeks of amenorrhoea and decreased fetal movements. She was booked antenatally and her midtrimester ultrasonography at 20 weeks was normal. Her antenatal period was uneventful. At 37 weeks she c/o decreased fetal movements.

USG(20/12/2004)- single live fetus 38 weeks, 3.3 kg, and oligohydramnios with AVI of 7, slightly dilated bowel loops, Cyst is seen in abdomen (?meconium cyst is to be considered).

In view of decreased fetal movements and persistent non reactive NST, she was taken up for LSCS. Alive Female baby, weight 2635 grams was born. Baby was well at birth, no active resuscitation was required. Abdomen was distended, 20 ml bilious fluid aspirated through nasogastric tube.

X-Ray Abdomen showed air fluid levels with meconium peritonitis. The baby was immediately taken up for surgery. Laparotomy followed by resection and anastomosis was done.

Post operative period was uneventful. The gastric aspirate decreased gradually, stool passed after 8 days, feeds tolerated well and was discharged in stable condition. Baby is fine and is now eleven months old.

Incidence ranges from 1/1500 to 1/2000, If, without prenatal diagnoses and planned post natal treatment, the mortality is as high as 62%. But with improved antenatal diagnosis, survival rate has improved to 90% with operative mortality < 1%. The literature is reviewed.

We were happy that with good antenatal diagnosis and planned postnatal management we could give a healthy baby.

Prenatal Diagnosis of Harlequin Fetus by Three Dimentional Ultrasonography – A Rare Case Affecting Three Consecutive Pregnancies

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Introduction: Herlequin ichthyosis is a congenital keratinizing disorder which is lethal in almost all cases. Three dimentional (3-D) ultrasonography is non invasive upcoming modality to diagnosis this devastating condition in utero.

Case Report: A 24 year old, 3rd gravida presented at 16 weeks gestation with history of previous two female babies with herlequin ichthyosis. Both babies died after 12 hours. In the present pregnancy, level II ultrasound at 17 and 22 weeks did not reveal any malformation. Repeat 2-dimentional ultrasound at 26 weeks showed oedematous eyes and persistent open mouth. 3-D ultrasonography revealed multiple dysmorphic features as ectropion – cutaneous thickening of eyelids, eclabium – thickened & fixed lips, short phalanges with persistent flexion of fingers and toes, short foot length and polyhydramnios. Woman had preterm labour at 34 weeks and delivered a male baby weighing 1.7 kg with features of Herlequin ichthyosis. Baby received isotretinoin 2mg/day; however he died on day 9.

Discussion: Herlequin ichthyosis is an autosomal recessive condition with 25% risk of transmission; although in our case all three consecutive babies were affected which is very rare. Ultrasound features appears as late as 26 weeks. Early diagnosis by skin biopsy is possible at 20 weeks, but it is invasive and has other limitations. By 3-D ultrasound, early definitive diagnosis is possible so that termination becomes possible.

Conclusion: Early screening by three dimentional ultrasound should be done in cases with history of affected babies.

A Newborn with Prenatal Diagnosis of Cardiac Rhabdomyoma in Mother with Tuberous Sclerosis

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Introduction: Fetal rhabdomyoma is not a common cardiac tumor. Association of this tumor with Tuberous Sclerosis is reported up to 81% of the cases. Autosomal dominant pattern of tuberous sclerosis put the inherited risk of 50% however; isolated case reports or small series are reported suggesting mother having tuberous sclerosis.

Case-Report: We report on a newborn with antenatal diagnosis of multiple rhabdomyoma by fetal echocardiography arising from either side of intraventricular septum. Echocardiography of newborn confirmed the diagnosis of multiple rhabdomyomas. This male baby also had cutaneous lesions suggestive of tuberous sclerosis like ash leaf macule on left scrotum and hypopigmented macule on lower back but there was no neurological deficit. There was history of tuberous sclerosis (with epilepsy, mental retardation, bipolar affective disorder and adenoma sebaceum) in mother. Since birth the infant's intracardiac tumor has been regressed and the baby has shown good growth and development without surgery in one-year follow-up.

Discussion: Association of Rhabdomyoma with tuberous sclerosis is common but it is difficult to diagnose antenally. Cardiac tumor is the only sign, which may be seen on careful evaluation on ultrasound. Fetal echocardiography in case with family history of tuberous sclerosis is warranted to detect the lesion early so that mothers can be referred for delivery in the centre with facilities of qualified professionals and suitable equipments. Even though cardiac rhabdomyomas occasionally induce poor outcome they may regress in early infancy, remain unchanged during childhood, and then again regress in adolescence. Thus surgery is needed only on the patient's clinical presentation.

Conclusion: Offering fetal echocardiography in pregnant women with tuberous sclerosis may diagnose the affected baby in utero.

Antenatal Diagnosis and Management of Ebstein's Anomaly- A Case Report

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Objective: To present a case report of antenatally diagnosed Ebstein's anomaly.

Introduction: Ebstein's anomaly (EA) is a rare but fascinating congenital heart disorder which occurs in about 1–5 per 200,000 live births, accounting for <1% of all congenital heart disease. This anomaly is currently most easily diagnosed by echocardiography.

Case report: A 25 year old, X, primigravida presented to us at 32 weeks POG in antenatal clinic with an fetal echocardiographic diagnosis of fetus having Ebstein's anomaly. USG was showing IUGR. Parents were counseled about risk and prognosis of this anomaly. They wanted to continue the pregnancy. Patient was kept on regular antenatal surveillance. Pregnancy continued uneventfully till term. She was induced with cerviprime at 39wks with consent for caesarian section if necessary for maternal-fetal indication. She delivered vaginally a female baby (wt 2.06 kg) on 11/01/06. After delivery, baby had cyanosis but no features of congestive cardiac failure. Baby was shifted to ICU. Echocardiography was done. It was showing grossly displaced tricuspid valve with thickened tricuspid leaflets. It was also showing markedly dilated right atrium with small functional right ventricle. Surgery was advised for the baby with explaining prognosis to the parents.

Discussion: EA occurs in about 2.5% of autopsy series in patients with congenital heart disease, males and females being affected equally. EA is a malformation of the tricuspid valve and right

ventricle marked by a spectrum of several features which include: 1) adherence of the tricuspid valve leaflets to the underlying myocardium (failure of delamination); 2) downward (apical) displacement of the functional annulus (septal>posterior>anterior); 3) dilation of the “atrialised” portion of the right ventricle with variable degrees of hypertrophy and thinning of the wall; 4) redundancy, fenestrations, and tethering of the anterior leaflet; and 5) dilation of the right atrioventricular junction (true tricuspid annulus).

Celermajer et al. described an echocardiographic grading score for neonates with EA, GOSE (= Glasgow Outcome Score Extended) Score Grade 1–4. For this score the ratio of the combined area of the right atrium and atrialised right ventricle to that of the functional right ventricle and left heart in a four-chamber view at end-diastole was calculated (ratio <0.5, grade 1; ratio 0.5–0.99, grade 2; ratio 1.0–1.49, grade 3; and if ratio $\geq$ 1.5, grade 4).

The cardinal symptoms in EA are cyanosis, right-sided heart failure and arrhythmias. The clinical presentation is dependent on age at presentation and the degree of haemodynamic disturbance, which in turn is dependent on the extent of displacement of the tricuspid valve leaflets, the size and function of the right ventricle, right atrial pressure and degree of right-to-left interatrial shunting.

In utero, severe EA may lead to cardiomegaly, hydrops and tachyarrhythmias. EA is a common lesion referred for foetal echocardiography by the obstetrician. The intrauterine mortality rate is high in the severe forms of EA. Neonates with EA may present with congestive heart failure due to tricuspid valve regurgitation, cyanosis, and marked cardiomegaly caused by right heart dilation. 20–40% of all neonates diagnosed with EA will not survive 1 month, and fewer than 50% will survive to 5 years of age. If neonates are symptomatic their prognosis is almost always very poor. In subjects <2 years old at presentation, a haemodynamic problem is more common than in older patients (72% versus 29%, $p < 0.01$).

Celermajer et al. reviewed 220 cases with 1–34 years’ follow-up. Actuarial survival for all live-born patients was 67% at 1 year and 59% at 10 years. Predictors of death were echocardiographic grade of severity at presentation (relative risk increased by 2.7 for each increase in grade, CI 1.6–4.6), foetal presentation (6.9, CI 1.6–16.5), and right ventricular outflow tract obstruction (2.1, CI 1.1–4.4). J Am Coll Cardiol 1994;23:170–6.

Observation alone is advised for asymptomatic patients and symptomatic patients with no right to-left shunting and only mild cardiomegaly. Surgery should be considered if there is objective evidence of deterioration such as progressive increase in heart size on chest radiography, progressive right ventricular dilatation or reduction of systolic function in echocardiography, or appearance of premature ventricular contractions or atrial tachyarrhythmias. Once symptoms progress to NYHA class III or IV, medical management has little to offer, surgical risks increase sharply and surgery is clearly indicated. Biventricular reconstruction is usually possible, but if advanced cardiomyopathic changes have occurred, especially involving the left ventricle, cardiac transplantation is the only option.

Conclusion: A case of Fetal EA with planned delivery is reported. EA is a complex form of congenital heart disease. No two hearts with EA are the same. Precise knowledge of the different anatomical and haemodynamic variables, associated malformations and management options, is essential.

Nuchal Translucency: Frequency Distribution and Factors Affecting Its Measurement

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Aim: To determine the influence of period of gestation and other conditions like position of fetal neck on nuchal translucency measurement.

Method: A cross-sectional study. In nine hundred & sixty-five women between 10-13⁺⁶ weeks, Nuchal translucency was measured to calculate frequency distribution and mean NT at each week. 100 pregnant women between 11-13⁺⁶ weeks, nuchal translucency was measured in the mid-sagittal plane, with the fetal neck in the flexed, neutral and extended positions. Mean nuchal translucency measurement at different periods of gestation was compared. Differences between the extended and

neutral positions (D extended nuchal translucency) and the flexed and neutral positions (D flexed nuchal translucency) were calculated. The repeatability coefficient for the measurements in all the three positions was computed. Statistical analysis was done.

Results: Nuchal translucency increased with gestation. The mean D flexed value was 0.233 mm + 0.133 lesser than the neutral value [$p < 0.0001$]. The mean D extended NT was 0.305 mm + 0.155 greater than the neutral value [$p < 0.0001$].

The repeatability coefficient was lowest in the neutral position (0.17 mm in the neutral position, 0.28 in the flexed position and 0.41 mm in the extended position).

Conclusion: Period of gestation and fetal neck position can make a significant difference to nuchal translucency measurement. Repeatability of measurement is more accurate with the fetal neck in the neutral position.

Keywords: Nuchal, translucency, gestation, neck position, and repeatability.

Congenital Chylous Ascites: A Case Report

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Congenital chylous ascites is a rare but vexing clinical problem condition. The most common etiology in the fetus and newborn being congenital malformation of the lymphatic system. For such situation the term “leaky lymphatic” has been used. We had a patient who presented to us at 22 weeks of gestation with a large fetal abdominal cyst. Multiple paracentesis were done under ultrasound guidance initially to establish a diagnosis and prevent pulmonary hypoplasia, and later to avoid dystocia and obstructed labour. However, a prenatal diagnosis could not be established. The patient delivered at 38 weeks. The newborn underwent tapping of fluid, still no diagnosis was made. But on 5th day of life when a paracentesis was done – a diagnosis of congenital chylous ascites was established. Treatment modalities may be either medical or surgical. We present a case of congenital chylous ascites with its successful antenatal, intrapartum and postnatal management.

Perinatal Outcome in Growth Restricted Fetuses with Absent/ Reversed End Diastolic Flow in Umbilical Artery and Those with Normal Flow

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Objective: A prospective study was conducted to compare perinatal outcome in growth restricted fetuses retaining normal umbilical artery Doppler flow and those with absent/reversed end diastolic flow.

Methods: Fifty pregnant women with growth restricted fetuses were followed with anthropometry and serial umbilical artery Doppler flow measurements as per protocol. Outcome measures included diagnosis to delivery interval, perinatal morbidity and mortality and NICU stay. Outcomes were compared among small for date fetuses with normal umbilical artery Doppler (25) and those with absent/reversed end diastolic flow (25)

Results: Mean interval between the diagnosis to delivery was measured and was 1.6 ± 0.7 days for fetuses with reversed/absent end diastolic flow versus 17 ± 7.8 days for fetuses with normal Doppler flow. Also the duration of ICU stay was significantly higher in the reversed/absent flow group (54.6 ± 19.5 days) as compared to the group with normal flow (21.4 ± 8.7 days). Perinatal morbidity in terms of growth restriction, oligohydramnios, and prematurity was more in fetuses with compromised flow.

Conclusions: Growth restricted fetuses with reversed/absent end diastolic flow in umbilical artery are at great risk of complications. Early delivery of such fetuses advocated to prevent intrauterine deaths.

Growth restricted fetuses with normal doppler wave forms growth are at significantly less risk than their counterparts with abnormal umbilical artery Doppler.

Prenatal Detection of Fetal Congenital Malformations: Dilemma to patient and Physician

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Introduction: Ultrasound is an excellent modality to detect malformations in fetus, but to counsel for invasive testing and take right decision for continuation vs. termination is a dilemma for the patient and physician.

Materials and Method: Time of detection by ultrasound and type of malformations was noted for two years from January 2004. Patients were counseled and advised invasive testing if required. Patients' decision and fetal outcome were assessed.

Results: Total 56 cases with congenital malformations were assessed. Four cases had multiple defects. Defects found were musculoskeletal in 10(17.8%) patients, genitourinary in 10(17.8%), Cardiovascular in 9(16.1%), intestinal in 8(14.3%), neural tube defects in 7(12.5%) facial defects in 6(10.7%), diaphragmatic hernia in 3(5.3%), pleural collection, abdominal wall defects and ambiguous genitalia in 2(3.5%) patients each, skin disorder in 1(1.7%). Diagnosis was made by ultrasound before and upto 20 weeks in 21(37.5%) patients, >20-28 weeks in 20(35.7%) and > 28 weeks in 12(21.4%) patients. Diagnosis was missed in 3(5.3%) whereas in 4(7.1%) no abnormality was found after delivery. Total 28 cases were offered invasive testing. Nine had testing with all normal report, except one. Out of 56 cases, 6 were lost to follow up. Ten (20%) opted for abortion, 2(4%) had missed abortion, 4(8%) had intrauterine death, 11(22%) had neonatal death, 6(12%) were operated, 13(26%) were abnormal and 4(8%) were normal at delivery. Eventually perinatal loss occurred in 17 out of 40(45%).

Conclusion: After diagnosis of congenital malformations, it is difficult to convince patient for invasive testing. In view of high perinatal loss, recommendation of termination after detection of malformations should be revisited so the woman can be saved from trauma to carry abnormal pregnancy till term.

Prenatal Detection of Short Limbs: Counseling for the Present and Future Pregnancies

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Lethal skeletal dysplasias form a genetically and phenotypically heterogeneous group of disorders. Many of them have associated abnormalities of other systems but definite diagnosis of the type of skeletal dysplasia depends on postnatal radiograph. We have compared prenatal ultrasonographic (USG) findings and differential diagnosis based on them, with postnatal findings and final diagnosis. Out of 21 cases referred for short femur or long bones on prenatal USG, 18 had severe limb shortening while in 3 cases the length of long bones was around 5th centile. Two of the 3 cases with borderline shortening of limbs had associated findings giving diagnostic clues. Third case with isolated short femora was detected to have Trisomy 21 in amniotic fluid analysis.

Of all 21, narrow thorax was present in 17 cases. Associated USG findings other than limb shortening and narrow thorax were prenatally detected in 11. These were polydactyly, predominant mesomelic shortening, multiple fractures, ventriculomegaly and hydrops fetalis. Based on prenatal USG findings it was possible to limit the differential diagnosis to one or few types in 8 cases. These diagnoses matched with the final diagnosis based on post termination radiograph.

Severe shortening of long bones confirms the diagnosis of skeletal dysplasia with universal lethality and this information is sufficient to counsel for planning the current pregnancy. Prenatal USG also can provide clues to the specific type of skeletal dysplasia in some cases but confirmation of specific type of skeletal dysplasia need to be done by post termination/ post natal radiograph. This is essential to provide accurate risk of recurrence for next pregnancy. In cases with borderline short limbs it is essential to look for associated malformation, narrow thorax, and chromosomal anomalies to differentiate from isolated IUGR.

Which Syndrome is this?

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Mrs. N.N a 29 years Muslim female with history of consanguinous marriage, primigravida at 24 weeks +4 days presented to our OPD in July 2004 with complaints of inability to feel fetal movement and with USG report suggestive of fetal hydrops. She was investigated thoroughly including Level II USG and fetal echo. Level II USG was done by two consultants suggestive of Pena Shokeir syndrome with non-immune fetal hydrops. However she went into preterm labour and delivered a preterm still borne malformed male baby weighing 500gms. Baby was sent for autopsy and fetal blood for karyotyping was done. Autopsy was suggestive of Pena shokeir syndrome and karyotyping revealed Trisomy-20. Parvo IgG was positive and karyotyping of both parents was normal. The details about these syndromes will be discussed with special reference to preconception Counselling for future pregnancy

Fetal Hydrometrocolpos: Perinatal Diagnosis and Treatment

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Introduction: Fetal hydrometrocolpus with urogenital sinus and congenital vesicovaginal communication is a rare occurrence. Diagnosis is difficult on antenatal ultrasound especially in late gestation. Neonatal hydrometrocolpos is a surgical emergency and immediate drainage is required.

Case Report: We report two cases of female newborns with urogenital sinus (with no ambiguous genitalia) and hydrometrocolpos in whom prenatal diagnosis was made on ultrasound in the late third trimester as cystic dilatation in the female pelvis. There was no associated anomaly except fetal hydronephrosis in first fetus. After birth, newborns presented with abdominal distension. There were female genitalia and a single opening between the labia. Ultrasonography was inconclusive to clinch the definitive diagnosis. Karyotypes in both cases were normal. On operation, there was urogenital sinus. There was a communication between the vagina and bladder through a small orifice resulting in filling of vagina and uterus with urine. Both babies required primary drainage of hydrometrocolpos and vaginostomy through abdominal route. The neonates were discharged in good condition.

Discussion: Hydrometrocolpos is an uncommon fetal genitourinary anomaly occurring 1 in 30,000-60,000 birth. It is characterized by cystic dilatation of uterus and vagina mainly due to vaginal obstruction and excessive accumulation of cervical and endometrial mucus. However, accumulation of urine in the genital tract is very rare. Careful examination of fetus and newborn is needed as this anomaly may be associated with other malformations such as postaxial polydactyly, anal atresia, esophageal atresia, renal agenesis, genital abnormalities, cardiopathy or autosomal recessive disorders. Abdominoperineal vaginal pull through operation is recommended as definitive treatment but accumulated fluid to be drained in neonates to avoid respiratory distress and to reduce back pressure on urinary system and on inferior vena cava.

Conclusion: Prenatal counseling, targeted ultrasound and institutional delivery to be offered in fetus showing pelvic cyst.