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Single Umbilical Artery: Review

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Abstract

The umbilical cord is an important part of the fetoplacental unit. Single umbilical artery (SUA) is the most frequent anomaly, not only of the umbilical cord but probably among all birth defects. When there is only one umbilical artery in an umbilical cord it is called single umbilical artery. Many abnormalities of cord length and low placental weight have been associated with SUA. Fetal anomalies included are; diaphragmatic hernia, gastrointestinal, cardiac, musculoskeletal and renal anomalies. SUA is included in pathognomic feature of Sirenomelia. Fetal growth restriction is common in them due to anomalies, chromosomal abnormalities and impaired placental development and perfusion. Perinatal morbidity and mortality are increased mainly due to underlying structural and chromosomal anomalies. When SUA is diagnosed , prompt search should be made to detect other associated anomalies in the baby.

Keywords: Single Umbilical Artery; Congenital Anomalies; Ultrasonography.

Introduction

The umbilical cord is an important part of the fetoplacental unit. The presence of SUA is associated with poor perinatal outcome. The most common umbilical cord pathology in a human infant is SUA. The incidence of SUA ranges from 0.2% to 1.9%, with an associated anomaly rate of 67% and perinatal mortality of 20%. Even with the recent advances in ultrasonographic imaging, nearly two-thirds of all congenital malformations associated with SUA can be missed. The histopathological examination of the cord is considered to be the gold standard for diagnosis of SUA.[1]

Single umbilical artery (SUA) is the most frequent anomaly, not only of the umbilical cord but probably among all birth defects. Causes of Single umbilical artery:

1. Genetic
2. Environmental[2]

The umbilical cord forms between 13 and 38 days after conception and normally serves as the conduit for two umbilical arteries and one umbilical vein. The risk for congenital anomalies in infants with SUA also depends on the method of ascertainment, being highest at autopsy, but cases diagnosed at ultrasound or at term delivery still have an increased risk of anomalies compared to infants with two umbilical arteries. Observed anomalies most often involve the urogenital, gastrointestinal cardiovascular, respiratory, and central nervous systems, as well as the face.[3]

Counting cord vessel number is a standard component of anatomical evaluation during foetal sonographic examination and immediately after delivery. Embryos initially have 2 umbilical veins. In the first trimester the right vein typically atrophies to leave one large vein to accompany the two thick walled umbilical arteries. Four vessel cords are rare and often associated with congenital anomalies (Puvabanditsin 2011).

The most common aberration is that a Single Umbilical artery (SUA) with a cited incidence of 0.63% in live born neonates, 1.92% with perinatal deaths and 3% in twins. Foetuses with major malformation frequently have SUA. Thus it's identification often prompts consideration for targeted USG and Echocardiography. The frequent anomalies are cardiovascular and genitourinary (Hua 2010, Murphy-Kaulbeck, 2010). In an anomalous foetus a single artery greatly increases the aneuploidy risk, and amniocentesis is recommended (Dagklis, 2010; Lubusky, 2007). If target USG finds otherwise normal anatomy an isolated single artery in an otherwise low risk pregnancy does not increases the risk of aneuploidy. However as in isolated finding, it has been associated with foetal growth restriction and perinatal death in some but not all studies (Chetty–John 2010; Gutvirtz 2016; Hua 2010; Murphy Kaulbeck 2010; Voskamp 2013). Thus while clinical monitoring of growth is reasonable the value of sonographic surveillance is unclear.

A rare anomaly is that of a fused umbilical artery with a shared lumen. It arises from Failure of two arteries to split during embryological development. The common lumen may extend through entire cord, but if partial it may be found near placental insertion site (Yamada 2005). In one report these were associated with higher incidence of marginal or velamentous cord insertion, but not congenital foetal anomalies (Fujikura 2003).[4]

When there is only one umbilical artery in an unbilical cord it is called single umbilical artery. It is of 2 Types:

1. *Isolated SUA*: when SUA is isolated ultrasound finding without any other anomalies it is called Isolated SUA. However, there is some increase in incidence of some fetal growth restriction in some studies possibly due to suboptimal placental development and perfusion. Preterm delivery rate is slightly higher due to pregnancy complications associated with SUA. There is slight increase in perinatal mortality and morbidity in them due to associated pregnancy complications and reduction in Wharton's Jelly and abnormal cord twist. Long-term physical and neurological development of children with Isolated SUA is not affected.
2. *Nonisolated SUA:* It is associated with structural and/or Chromosomal abnormalities. karyotype abnormalities have been reported in 45% foetuses with nonisolated SUA with autosomal trisomy being the most common. Upto 20–30% SUA have major structural anomalies involving multiple organs with the most common being heart, gastrointestinal tract, CNS and renal system. Other less common anomalies include Diphragmatic hernia, Hydrops, Musculosceletal anomalies, cloacal exstrophy, Sirenomelia and VATER Syndrome.

Fetal growth restriction is common in them due to structural anomalies, chromosomal abnormalities and impaired placental development and perfusion. Perinatal morbidity and mortality are increased mainly due to underlying structural and chromosomal anomalies.[5]

Major Structural Anomalies

- Cardiovascular Anomalies
- Gastrointestinal Tract Anomalies
- Central Nervous System Anomalies
- Renal Anomalies

Other Anomalies

- Diphragmatic hernia
- Hydrops
- Musculosceletal anomalies
- Cloacal exstrophy
- Sirenomelia
- VATER Syndrome.

Significence of Single Umbilical Artery

May result from one of the following:

- Primary agenesis of one of the umbilical arteries
- Secondary atrophy or atresia of previously normal umbilical arteries
- Persistence of original single allantoic artery of body stalk

Associated with anomalies particularly:

- Genitourinary
- Cardiac
- Gastrointestinal

When not associated with anomalies, Good outcome

The outcome of SUA with associated anomalies or aneuploidy depends on the underlying chromosomal and structural abnormalities.

Foetal karyotype analysis should be offered when foetal anomalies are detected.

Diagnosis of SUA

Diagnosis of SUA is made or confirmed using Color Doppler ultrasonography at the level of the fetal abdominal cord insertion, by observing the absence of one of the two umbilical arteries (UAs), which normally encircle the fetal bladder.[7]

Management

When SUA is diagnozed, thorough prenatal ultrasound examination should be performed to all the anomalies but specially of heart and kidney including fetal echo cardiography. For isolated SUA, amniocentesis for karyotyping is not indicated. However, for non isolated SUA, amniocentesis for fetal karyotyping is indicated as aneuploidies are more common in them and may warrant fetal termination. Clinical examination and 4 weekly USG for fetal growth parameters should be performed. Prenatal fetal surveillance using weekly NST and Biophysical Scoring is indicated for non isolated SUA after 32 wks till delivery. Isolated SUA can have normal vaginal delivery. Timing and mode of delivery of non isolated SUA depends on specific diagnosis.[5]

Discussion

In the early periods of gestation, umbilical cord contains 4 vessels, 2 arteries and 2 veins. Under normal conditions the right umbilical vein regresses in the 2nd month of fetal life. The left umbilical vein and the 2 umbilical arteries become the vessels found in the normal umbilical cord. SUA cord is the result of agenesis, aplasia or atresia of one of the umbilical arteries.

Many abnormalities of cord length and low placental weight have been associated with SUA. Fetal anomalies included are diaphragmatic hernia, gastrointestinal, cardiac, musculoskeletal and renal anomalies. Cleft lip is observed in one of the baby. The rate of preterm delivery, IUGR, oligohydramnios and LBW were found to be statistically higher in cases with SUA, in a study conducted by Vinay Kumar.

During routine anomaly scan cross section of umbilical cord and after delivery cut section of cord should be examined for number of vessels. When there is a suspicion of SUA, it should be confirmed by histopathological examination. Also, the risk of preterm delivery, IUGR and LBW must be remembered in a case of SUA. When SUA is diagnozed, prompt search should be made to detect any other associated anomalies in the baby.[1]

Prevalence of SUA is 3 to 4 Times Higher in Fetal Conditions

- IUGR
- Prematurity
- Twinning
- Urinary track anomalies
- CVS anomalies
- CNS anomalies
- Perinatal death

SUA is Associated with Maternal Conditions

- Diabetes Mellitus
- Pregnancy Induced Hypertention
- Polyhydramnios
- Oligohydramnios

Advanced maternal age and associated chromosomal conditions are not a major contributor to the incidence of SUA and related anomalies among referral population in study conducted by Sandra Prucka, et al. Sandra Pruca, et al. explored 10% incidence of chromosome abnormalities in their SUA study group, with this finding they believe that amniocentesis should be offered to all patients with SUA on prenatal ultrasound.[3]

SUA is included in pathognomic features of Sirenomelia. Sirenomelia is synonymous with Mermaid, Monopodia, Symmelia, Sympus, Symposia, Uromelia.

Sirenomelia (Mermaid Syndrome) Features

- Renal Dysgenesis
- Absent external genitals
- 2 vessel umbilical cord (SUA)
- Imperforated Anus
- Malformations of lower limbs[8]

With the recent advent of color, power, and High Definition (HD) Doppler ultra-sound, the umbilical cord blood flow imaging makes the identification of which side of UA is missing much easier than ever before. Previous studies showed that left UA was absent more frequently than right UA.

Prenatal diagnosis of SUA is more common in second and third trimesters of pregnancy than in first trimester. Subsequent fetal echocardiography is indicated in cases of SUA. The increased incidence of SUA with chromosomal disorders has been also reported, especially in cases with additional malformation. Trisomy 18 is the most common

aneuploidy associated with SUA. The next most common types of aneuploidy associated with SUA are trisomy 13 and trisomy 21.[9]

Conclusion

Single Umbilical Artery is the most frequent anomaly, not only of the Umbilical Cord but probably amongst all birth defect. Genetic counseling should be advised to parents of SUA fetuses, especially those with additional congenital anomalies. When SUA is diagnozed, prompt search should me made to detect any other associated anomalies in the baby.

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Case Report

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Gravid Uterus in an Anterior Abdominal Wall Hernia and Successful Repair at the Time of Cesarean Section

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Abstract

This is a case report of a pregnant woman with previous 1 cesarean and 1 ectopic exploration whose uterus herniated in an incisional hernia of the anterior abdominal wall at 33 weeks + 6 days of pregnancy. Incarceration of the pregnant uterus in an incisional hernia is a rare but serious obstetric situation1. Treatment is conservative until term followed by delivery and repair of incisional hernia thus resulting in a successful outcome

Keywords: Incisional hernia; Cesarean section; Pregnancy complication.

Introduction

The herniation of a gravid uterus through an incisional hernia site is a rare occurrence.[2] Incisional hernia is a frequent complication of abdominal wall closure and the management of pregnancy with a large incisional hernia with a gravid uterus in its sac is challenging.

Following is a Case report of gravid uterus through an incisional hernia of a midline incision. 33 years old Gravida 3, Para 2, Live 1, Ectopic 1 with previous history 1 ectopic exploration and 1 cesarean section through midline incision was admitted due to lower abdominal pain on & off and ulceration of abdominal skin over incisional hernia at 33 weeks 6 days. She was booked case at government hospital, karaikal. Had her regular checkups until 2nd trimester there. At around 21 weeks she was diagnosed to have small anterior wall defect and referred. Came for 1st antenatal check up at 28 weeks to our hospital,was diagnosed as G3,P1,L1,E1 at 28, weeks with incisional hernia and was managed conservatively at subsequent antenatal visits. General surgeon opinion was obtained and planned for elective cesarean section after 37 completed weeks with incisional hernia repair. Her past obstetric history revealed that she had her ectopic exploration 4 years back and a cesarean section 3 years before because of post dated pregnancy. On both occasions she was operated on through infra umbilical midline vertical incision. There was no history of cesarean section wound infection during the postoperative period in the previous two pregnancies. On examination she was moderately built and nourished, there was mild pallor, her pulse rate was 80 beats per minute and her blood pressure was 120/80 mm Hg.

Abdominal examination revealed Distention of abdomen in central area. Uterus size could not be made out, uterus was felt just underneath with a complete lack of anterior abdominal wall. Fetal heart rate was 144 bpm, Huge defect on the anterior abdominal wall measuring 14*14 cm with the uterus herniating through the defect. Lie and presentation could not be made out. Superficial skin necrosis and Skin ulceration was present. Routine investigations