Umbilical Vein Varix

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Background
Fetal intra-abdominal umbilical vein (FIUV) varix is a focal dilatation of the umbilical vein of the fetus, just after entering the abdomen. Early case series have reported an association of FIUV varix with intrauterine death, and some investigators have suggested that it may represent a significant risk factor for intrauterine death. A correlation between FIUV varix with other anomalies, aneuploidies, fetal hydrops and other adverse pregnancy outcomes has also been suggested. Incidence is 1-2.8 per 1000.

Objective
• To guide the accurate diagnosis, investigation and management of women presenting with fetal umbilical vein varix.
• To provide a consistent approach to the care of women with fetal umbilical vein varix, which takes into consideration individual women’s views and wishes regarding care plan.

Definition
Dilated intra-abdominal, usually extrahepatic, portion of the umbilical vein, with diameter greater than 9 mm or enlargement of the varix to at least 50% more than the diameter of the intrahepatic umbilical vein.

Differential Diagnosis
Other sonolucent masses located in the same region of the fetal abdomen (distended gallbladder, choledochal cyst, urachal cyst, etc.).
**Important History**
Age, first trimester combined screening result.

**Ultrasound**
- FIUV varix is diagnosed on ultrasound examination by visualizing a dilated intra-abdominal, usually extrahepatic, portion of the umbilical vein. It can be differentiated from other sonolucent masses by colour-flow imaging. Displaying continuity with the umbilical vein aids in the diagnosis.
- The FIUV varix diameter is measured from one outer edge of the vein to the opposite inner edge with callipers, on axial images immediately cephalad to the insertion of the umbilical vein into the fetal abdomen. Diagnostic criteria include an umbilical vein diameter greater than 9 mm (measured without colour flow Doppler) or an enlargement of the varix to at least 50% more than the diameter of the intrahepatic umbilical vein.
- Careful examination for any other associated fetal structural abnormalities is essential.

**Investigation**
- Detailed anatomy scan indicated.
- Karyotyping to be considered, particularly if there are other markers of aneuploidy or other structural anomalies.
**Prognosis**

- Umbilical vein varix associated with structural anomalies (10-35%) and aneuploidy (6-12%, but this is lower if the finding is isolated).
- If no associated structural anomalies/ aneuploidy prognosis generally favourable
- However, FIUV varix is associated with an increased risk of fetal death before the onset of labour (4% in apparently isolated cases).
- There is a risk of thrombus formation in the varix, which has infrequently been associated with fetal demise or thrombosis of the portal system.
- With a protocol of intensified fetal monitoring fetal death occurs infrequently (about 20% of patients are induced for non-reassuring fetal condition).
- Mode of delivery is as per normal obstetric indications. Caesarean section is not indicated for reasons of FIUV varix.

**On-going Management**

- Advise once or twice weekly CTG and weekly scan of fetal wellbeing identifying amniotic fluid volume and fetal movements, with fortnightly growth scans in fetuses with an umbilical vein varix from gestational age of 32 weeks onwards, in Day Assessment Unit or in local hospital, under obstetrician led care (not MFM).
- Induction of labour at 39-40 weeks gestation.
References