Fetal Intra-abdominal Umbilical Vein Varix (FIUVV)

Recommendations of Practice

Background:

- Fetal Intra-abdominal Umbilical Vein Varix (FIUVV) is a focal dilatation of the umbilical vein of the fetus, located just after entering the abdomen. Estimated incidence is 0.4-1.1 per 1000.
- Associations of FIUVV with intrauterine death, other anomalies, aneuploidies, fetal hydrops, venous thrombosis and other adverse pregnancy outcomes have also been reported.

Objective:

- To guide the accurate diagnosis, investigation and management of women presenting with FIUVV.
- To provide a consistent approach to the care of pregnancies with FIUVV, which takes into consideration the pregnant person and whānau wishes with respect to their pregnancy care.

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 non-dilated portion of the intrahepatic umbilical vein; or:
- Dilatation >2SD above the mean value for gestational age.

Differential diagnosis:

- Other sonolucent cystic masses located in the same region of the fetal abdomen e.g. dilated gallbladder, choledochal cyst, digestive duplication, mesenteric, hepatic or urachal cyst.

Important History:

- Age, combined first trimester screening or NIPT result.

Ultrasound:

- FIUVV 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 and pulsed Doppler to verify venous flow confirming the diagnosis.
- The FIUVV diameter is measured from one outer edge of the vein to the opposite inner edge with callipers, perpendicular to the lumen, on axial images immediately cephalad to the insertion of the umbilical vein into the fetal abdomen.

Diagnostic criteria:

- 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 non-dilated portion of the intrahepatic umbilical vein.
- Thorough examination for any other associated fetal structural abnormalities is essential.

Investigation:

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

Prognosis:

- FIUVV should be stratified into isolated and non-isolated.
- Isolated:
    - The prognosis is favourable with possibly an increased risk of aneuploidy or fetal death (0-4%).
- Non-isolated:
    - Associated structural anomalies (10-35%), and
    - Aneuploidy (6-12%).
- FIUVV is associated with an increased risk of fetal death before the onset of labour (0-4% in apparently isolated cases).
- There is a risk of thrombus formation in the varix, which has infrequently been associated with thrombosis of the portal system or fetal demise.
- With a protocol of intensified fetal monitoring (see below) fetal death occurs infrequently with approximately 20% of patients being delivered for non-reassuring fetal condition.
- Mode of delivery is as per usual obstetric indications. Caesarean section is not indicated for reasons of FIUVV.

Ongoing Management:

- For non-isolated FIUVV monitor from 32 weeks gestation onwards: Once or twice weekly CTG and weekly scan of fetal wellbeing identifying amniotic fluid volume and fetal movements. Fortnightly growth scans. This can be in the Day Assessment Unit or in local hospital, under obstetrician led care.
- For isolated FIUVV: Low risk of fetal demise (see above). Monitoring should be individualised. At a minimum this include growth scans with closer monitoring for vein patency from 36 weeks.
- Ultrasound may detect thrombus formation of the FIUVV. Suggested weekly from 36 weeks via colour flow assessment.
- Induction of labour by 40 weeks gestation.

This Recommendation of Practice was updated in March 2023 by Dr Jaynaya Marlow with input from members of Wāhi Rua NZMFM Network.

The most up to date version of this Recommendation of Practice can be found on Healthpoint Wāhi Rua: New Zealand Maternal Fetal Medicine Network (NZMFM) webpages: https://www.healthpoint.co.nz/public/wahi-rua-new-zealand-maternal-fetal-medicine/

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