Fetal Ventriculomegaly

New Zealand Maternal Fetal Medicine Network

This guideline was updated in May 2010 by Dr Chern Lo, with input from members of the New Zealand Maternal Fetal Medicine Network.

Background

Fetal ventriculomegaly is usually diagnosed at the routine anomaly scan, but can be identified later in pregnancy as an incidental finding.

There are a wide range of possible underlying conditions and the spectrum of outcome is also wide.

It is important that appropriate investigations are offered and that counselling is tailored to the likely diagnosis. If no underlying diagnosis is found then aspects of the ventriculomegaly can be used to offer some indication of prognosis.

Once the diagnosis is clarified (where possible) options can be discussed with the woman and her support. If the woman continues the pregnancy, continued care with fetal medicine input/ LMC is indicated.

Follow-up will depend on the situation.

Objective

To guide accurate diagnosis, investigation and management of women presenting with fetal ventriculomegaly.

To provide a consistent approach to the care of women with fetal ventriculomegaly taking into consideration the wide range of underlying causes and individual women's views.

Definition

- Lateral cerebral ventricular measurement increased above the normal cut-off
- Lateral ventricle is measured at the widest part of the posterior horn perpendicular to the long axis of the ventricle
- Callipers should be placed on the inner aspect of the wall of the ventricle or approximated to this position if the choroid plexus is filling the cavity

Normal Values

Normal	<10mm
Mild	10-11.9mm
Moderate	12-14.9mm
Severe	15mm+

Differential Diagnosis

- Aneuploidy
- Infection
- Syndromes
- Abnormal Neuroanatomy
- Cerebral haemorrhage

Important History

- Viral/bacterial illness
- Past obstetric history and any anomalies or neonatal thrombocytopenia in particular
- Any family history of note. In particular aneuploidy, syndromes
- Consanguinity

Ultrasound

1. Full anomaly screening: In particular spine, limb position and movement
2. Extended neurosonogram as described by ISUOG should be performed with the aid of 3D volumes if required
3. Lateral ventricular shape should be assessed
4. Colpocephaly (teardrop shaped): associated with agenesis of the corpus callosum.
5. Shape of the head and cerebellum: lemon head and banana cerebellum is nearly always associated with an open neural tube defect
6. Identification of the cavum septum pellucidum and corpus callosum
7. Posterior fossa views of the cerebellar hemispheres, vermis and cisterna magna should be sought
8. Fetal gender determination: as it can aid prognostication
9. Other markers:
    - Markers of infection (liver and brain calcification)
    - Markers of aneuploidy

Investigation

Maternal blood:

- CMV, Toxoplasmosis, Rubella, HSV and Parvovirus antibodies
- Anti-platelet antibodies

Amniocentesis:

- FISH full karyotype
- Alpha FetoProtein
- Store for PCR: CMV and Toxoplasmosis if maternal antibodies present

Prognosis

Dependant on the presence of an underlying aetiology and aspects of case.

Aneuploidy

- 5-17% will have aneuploidy. Prognosis as per the condition.

Infection

- CMV- very poor prognosis. Refer to CMV guideline.
- Toxoplasmosis- poor prognosis. Refer to Toxoplasmosis guideline.

Syndromes

- Prognosis as per the condition.

Abnormal Neuroanatomy

- Prognosis as per the condition. The neuronal migration disorders are not evident at
- 20 weeks. MRI or repeat USS at 28-30 weeks is recommended.

Cerebral haemorrhage

- If there is underlying NAIT, prognosis will depend on the size of haemorrhage and whether there is worsening of ventriculomegaly. Consideration may be given to maternal administration of IVIG/steroids and/or fetal blood sampling (FBS). The role of platelet transfusion is probably restricted to coverage of delivery and FBS. For future pregnancies refer to the NAIT guideline

Isolated Ventriculomegaly

Upon exclusion of other causes, the following figures can be offered to parents:

- Mild, unilateral, male + normal 30 week scan:
  - *Studies suggest normal outcome*
- Mild ventriculomegaly:
  - *5% require extra assistance at school*
- Moderate ventriculomegaly:
  - *14% some degree of handicap*
- Severe ventriculomegaly:
  - *48% handicapped*

A repeat USS at 30 weeks or an MRI may modify the prognosis.

On-going Management

Ongoing management depends on underlying aetiology. Clinicians from other disciplines may be involved antenatally e.g. geneticists, neonatologists, neurosurgeons, developmental paediatricians.

In isolated ventriculomegaly where the woman is continuing the pregnancy, further followup is recommended four weekly to identify a deteriorating situation.

References

Sonographic examination of the fetal central nervous system: guidelines for performing the 'basic examination' and the 'fetal neurosonogram'
(UOG Volume 29, Issue 1, Date: January 2007, Pages: 109-116