Fetal ventriculomegaly

Etiology

This occurs in around 1% of pregnancies.

Fetal warfarin syndrome: including scoliosis, brachydactyly, vertebral column calcifications, ventriculomegaly, agenesis of the corpus callosum) and spasticity/seizures and eye defects after the 1st trimester.

Arachnoid cysts are often associated with ventriculomegaly (in 64% of supratentorial and 80% of infratentorial cysts), in such cases probably the best overall treatment. For shunting into the peritoneum, use a low-pressure valve. If there is concurrent ventriculomegaly, one may simultaneously place a ventricular shunt (e.g. through a “Y” connector).

Classification

When this measurement is between 10 and 15 mm, the ventriculomegaly may be described as mild to moderate.

When the measurement is greater than 15 mm, the ventriculomegaly may be classified as more severe.

Enlargement of the ventricles may occur for a number of reasons, such as loss of brain volume (perhaps due to infection or infarction), or impaired outflow or absorption of cerebrospinal fluid from the ventricles. Often, however, there is no identifiable cause.

Isolated mild fetal ventriculomegaly

Diagnosis

Fetal ventriculomegaly (VM) refers to the enlargement of the cerebral ventricles in utero. It is associated with the postnatal diagnosis of hydrocephalus. VM is clinically diagnosed on ultrasound and is defined as an atrial diameter greater than 10 mm. Because of the anatomic detailed seen with advanced imaging, VM is often further characterized by fetal magnetic resonance imaging (MRI). Fetal VM is a heterogeneous condition with various etiologies and a wide range of neurodevelopmental outcomes. These outcomes are heavily dependent on the presence or absence of associated anomalies and the direct cause of the ventriculomegaly rather than on the absolute degree of VM. In this review article, we discuss diagnosis, work-up, counseling, and management strategies as they relate to fetal VM. We then describe imaging-based research efforts aimed at using prenatal data to predict postnatal outcome. Finally, we review the early experience with fetal therapy such as in utero shunting, as well as the advances in prenatal diagnosis and fetal surgery that may begin to address the limitations of previous therapeutic efforts [1)].

Accurate measurements are paramount in the diagnosis and subsequent counseling in fetal ventriculomegaly. The standardized method for ultrasound measurements based on five anatomic or technical requirements put forth by Guibaud et al. is meant to reduce the variation in cerebral ventricular measurements between centers and to decrease the number of false-positive results. We add that fetal magnetic resonance imaging (MRI) provides improved clarity of ventricle borders and the axial plane, which are sources of potential error in ultrasound-based measurements. Also, several anatomic landmarks that do not change position with the degree of ventricular enlargements, such as the parieto-occipital sulcus, are readily identified on MRI. In addition, segmentation techniques applied to fetal MRI yield volume measurements of all cerebrospinal fluid spaces, which may provide useful prognostic information when combined with currently used linear measurements.

As stated by Drs. Guibaud and Lacalm, the diagnostic algorithm they put forth to determine the etiology of ventriculomegaly based on ultrasound imaging patterns may be similarly applied to MRI. We agree that gathering information related to the underlying pathology of ventriculomegaly is useful for counseling. Furthermore, we concur that the term “severe,” in regard to ventricular dilation, is a historical term used for classification and should not be used for prognostic purposes, especially when counseling families. We add that the outcome depends both on the severity of ventriculomegaly as well as associated anomalies.

The commentary and additional references included by Drs. Guibaud, Lacalm, and Rault highlight the complexity and nuances involved in the prenatal evaluation of fetal ventriculomegaly, an understanding of which will enable practitioners to provide the highest level of counseling and care to patients [2)].

Treatment

Fetal ventriculomegaly treatment.

Outcome

In many cases of mild ventriculomegaly, however, there is a resolution of ventriculomegaly during the pregnancy.

To determine whether ventriculomegaly is associated with ongoing intracranial hypertension, physicians often rely on corroborative imaging features such as altered periependymal signal, distortion of ventricular shape, subarachnoid space flattening, and an increase in ventricular size over time.

A 2nd-trimester maternal serum alpha-fetoprotein (MSAFP) level >3.8MoM in a fetus with an open neural tube defect (ONTD) is associated with mid-gestation ventriculomegaly [3)].

Prospective observational study

A prospective observational study of patients referred with fetal ventriculomegaly from January 2011 to July 2020. Data were obtained from the hospital medical database and analyzed to determine the rate of isolated ventriculomegaly, associated structural abnormalities, chromosomal/genetic abnormalities, and survival rates. Data were stratified into three groups; mild(Vp=10-12mm), moderate(Vp=13-15 mm) and severe(Vp >15mm) ventriculomegaly.

There were 213 fetuses included for analysis. Of these 42.7% had mild ventriculomegaly, 44.6% severe and 12.7% had moderate ventriculomegaly. Initial ultrasound assessment reported isolated ventriculomegaly in 45.5% of fetuses, with additional structural abnormalities in 54.5%. The rate of chromosomal/genetic abnormalities was high,16.4%. After all investigations, the true rate of isolated VM was 36.1%. The overall survival was 85.6%. Survival was higher for those with isolated VM across all groups(P<0.05).

Ventriculomegaly is a complex condition and patients should undergo genetic counseling that even with apparently isolated VM, there remains the possibility of additional genetic and/or structural problems being diagnosed in up to 10% of fetuses [4].

1)
Pisapia JM, Sinha S, Zarnow DM, Johnson MP, Heuer GG. Fetal ventriculomegaly: Diagnosis, treatment, and future directions. Childs Nerv Syst. 2017 Jul;33(7):1113-1123. doi: 10.1007/s00381-017-3441-y. Epub 2017 May 16. PMID: 28510072.
2)
Pisapia JM, Heuer GG. In reply to “Letter to the Editor: Fetal ventriculomegaly: Diagnosis, treatment, and future directions.” Childs Nerv Syst. 2017 Aug 8. doi: 10.1007/s00381-017-3563-2. [Epub ahead of print] PubMed PMID: 28791427.
3)
Corroenne R, Zhu K, Orman G, Huisman TAGM, Mehollin-Ray AR, Johnson E, Johnson RM, Andrucioli A, Espinoza J, Nassr AA, Belfort M, Donepudi R, Shamshirsaz AA, Aagaard K, Whitehead WE, Sanz Cortes M. Maternal serum alpha-fetoprotein level and the relationship to ventriculomegaly in fetal neural tube defect: A retrospective cohort study. Eur J Obstet Gynecol Reprod Biol. 2021 Apr;259:185-190. doi: 10.1016/j.ejogrb.2021.02.010. Epub 2021 Feb 16. PMID: 33684673.
4)
Ryan GA, Start AO, Cathcart B, Hughes H, Denona B, Higgins S, Corcoran S, Walsh J, Carroll S, Mahony R, Crimmins D, Caird J, Robinson I, Colleran G, McParland P, McAuliffe FM. Prenatal Findings and Associated Survival Rates in Fetal Ventriculomegaly: A Prospective Observational Study. Int J Gynaecol Obstet. 2022 Apr 3. doi: 10.1002/ijgo.14206. Epub ahead of print. PMID: 35373343.