Ventriculomegaly is defined as dilation of the fetal cerebral ventricles and is a relatively common finding on prenatal ultrasound. Prenatally detected fetal ventriculomegaly. When enlargement of the lateral ventricles (>10 mm) is identified, a thorough evaluation should be performed, including detailed sonographic evaluation of fetal anatomy, amniocentesis for karyotype and chromosomal microarray analysis, and a workup for fetal infection. In some cases, fetal magnetic resonance imaging may identify other central nervous system abnormalities and should be considered when this technology as well as expert interpretation is available. Follow-up ultrasound examination should be performed to assess for progression of the ventricular dilation. In the setting of isolated ventriculomegaly of 10–12 mm, the likelihood of survival with normal neurodevelopment is >90%. With moderate ventriculomegaly (13–15 mm), the likelihood of normal neurodevelopment is 75–93%. The following are Society for Maternal-Fetal Medicine recommendations: We suggest that ventriculomegaly be characterized as mild (10–12 mm), moderate (13–15 mm), or severe (>15 mm) for the purposes of patient counseling, given that the chance of an adverse outcome and potential for other abnormalities are higher when the ventricles measure 13–15 mm vs 10–12 mm (GRADE 2B); we recommend that diagnostic testing (amniocentesis) with chromosomal microarray analysis should be offered when ventriculomegaly is detected (GRADE 1B); we recommend testing for cytomegalovirus and toxoplasmosis when ventriculomegaly is detected, regardless of known exposure or symptoms (GRADE 1B); we suggest that magnetic resonance imaging be considered in cases of mild or moderate fetal ventriculomegaly when this modality and expert radiologic interpretation are available; magnetic resonance imaging is likely to be of less value if the patient has had a detailed ultrasound performed by an individual with specific experience and expertise in sonographic imaging of the fetal brain (GRADE 2B); we recommend that timing and mode of delivery be based on standard obstetric indications (GRADE 1C); we recommend that with isolated mild ventriculomegaly of 10–12 mm, after a complete evaluation, women be counseled that the outcome is favorable, and the infant is likely to be normal (GRADE 1B); we recommend that with isolated moderate ventriculomegaly of 13–15 mm, after a complete evaluation, women be counseled that the outcome is likely to be favorable but that there is an increased risk of neurodevelopmental disabilities (GRADE 1B).

Key words: dilated cerebral ventricles, fetal brain, fetal magnetic resonance imaging, hydrocephalus, ventriculomegaly

Introduction

Ventriculomegaly is characterized by dilation of the fetal cerebral ventricles and is a relatively common finding on prenatal ultrasound. Prenatally detected fetal
ventriculomegaly is typically categorized in 1 of 2 ways: mild (10–15 mm) or severe (>15 mm); or as mild (10–12 mm), moderate (13–15 mm), or severe (>15 mm).\textsuperscript{1,2} Although mild fetal ventriculomegaly is often incidental and benign, it also can be associated with genetic, structural, and neurocognitive disorders, and outcomes range from normal to severe impairment. Hydrocephalus is one cause of ventriculomegaly and is defined as pathologic dilation of the cerebral ventricular system due to increased pressure, usually caused by obstruction. In general, severe ventriculomegaly is more likely to be associated with obstruction and to represent hydrocephalus than mild ventriculomegaly, which rarely represents obstruction. This consult reviews the diagnosis, evaluation, and management of mild to moderate fetal ventriculomegaly.

How is ventriculomegaly defined?

Fetal cerebral ventriculomegaly is defined as an atrial diameter of ≥10 mm on prenatal ultrasound.\textsuperscript{3–5} The atrium of the lateral ventricle is the part at which the body, posterior horn, and temporal horn converge (Figure); the atrial diameter remains stable between 15–40 weeks of gestation. The mean diameter of the lateral ventricle has been reported to range from 5.4–7.6 mm, and a measurement of 10 mm is 2.5–4 SD above the mean (3–6). An appropriately obtained sonographic measurement of <10 mm should be considered normal.\textsuperscript{6} We suggest that ventriculomegaly be characterized as mild (10–12 mm), moderate (13–15 mm), or severe (>15 mm) for the purposes of patient counseling, given that the chance of an adverse outcome and potential for other abnormalities are higher when the ventricles measure 13–15 mm vs 10–12 mm (GRADE 2B).

It is important that the lateral ventricle be measured correctly, as small differences in technique can result in false-positive or false-negative results. Substantial interobserver variability in interpretation can occur, particularly at borderline ventricular diameters (ie, about 10 mm).\textsuperscript{7} The atrium of the lateral ventricle should be measured in the transventricular (axial) plane at the level demonstrating the frontal horns and cavum septi pellucidi, in which the cerebral hemispheres are symmetric in appearance. The calipers should be positioned on the internal margin of the medial and lateral walls of the atria, at the level of the parietooccipital groove and glomus of the choroid plexus, on an axis perpendicular to the long axis of the lateral ventricle (Figure and Box).\textsuperscript{6}

The incidence of mild to moderate fetal ventriculomegaly is approximately 1%.\textsuperscript{3,5,6} Asymmetry of the lateral ventricles is common, and ventriculomegaly can be unilateral or bilateral. Unilateral ventriculomegaly is present in approximately 50–60% of cases, and bilateral ventriculomegaly occurs in approximately 40–50%.\textsuperscript{8,9} Although mild ventriculomegaly is more common in male fetuses, accounting for approximately 65–75% of cases, there are no data indicating that the prognosis for this finding differs by fetal sex.\textsuperscript{10,11}

What are the causes of ventriculomegaly?

The differential diagnosis of ventriculomegaly is extensive and includes a normal variant as well as disorders associated with severe impairment. A thorough evaluation is critical to make the correct diagnosis and to provide an accurate prognosis.

Normal variation

Measurements that are closer to 10 mm are more likely to represent a normal variant, particularly when isolated, and fetuses with a ventricular atrial diameter of 10–12 mm are found to have a normal postnatal evaluation in >90% of cases.\textsuperscript{2} Mild ventriculomegaly is likely to represent a normal variant if no other structural abnormalities are noted and if aneuploidy screening or diagnostic genetic testing results are normal. The chance that mild ventriculomegaly represents a normal variant decreases with increasing degrees

BOX

Criteria for appropriate measurement of lateral cerebral ventricle

1. Head is in axial plane
2. Image is magnified appropriately, so that fetal head fills majority of image
3. Focal zone is at appropriate level
4. Cerebral ventricles are symmetric in appearance
5. Midline falx is imaged
6. Atrium and occipital horn of lateral ventricle are clearly imaged
7. Atrium of lateral ventricle is measured at level of parietooccipital groove
8. Calipers are placed on medial and lateral walls of atrium perpendicular to long axis of ventricle
of dilation, and 75–93% of fetuses with moderate ventriculomegaly (lateral ventricles measuring 13–15 mm) are found to be normal after birth.1,10,12–14

Approximately 7–10% of fetuses with apparently isolated mild ventriculomegaly are found to have other structural abnormalities on examination after birth.1,14,15 Because it is not possible to determine with certainty that mild ventriculomegaly is truly isolated during pregnancy, normal variation is a diagnosis of exclusion that cannot be made with certainty until after birth.

**Structural abnormalities**

Ventriculomegaly can be associated with a number of underlying central nervous system (CNS) abnormalities. Some structural CNS anomalies, such as holoprosencephaly, hydranencephaly, porencephaly, or schizencephaly, and cystic lesions, such as arachnoid cysts, result in abnormal fluid collections in the fetal brain that may be misdiagnosed as ventriculomegaly, although these anomalies do not truly represent dilation of the ventricular system.

Structural abnormalities that can lead to dilation or enlargement of the lateral ventricles include agenesis of the corpus callosum, Dandy-Walker malformation, neural tube defects, cortical defects, and migrational abnormalities or heterotopia. The most common cause of severe ventriculomegaly is aqueductal stenosis, which results from narrowing of the cerebral aqueduct of Sylvius located between the third and fourth ventricle leading to progressive dilation of the lateral and third ventricles.16 Aqueductal stenosis can be genetic (see below) or can result from fibrosis secondary to fetal infection (eg, cytomegalovirus [CMV], toxoplasmosis, or Zika virus) or bleeding (eg, intraventricular hemorrhage). In many cases, the cause of aqueductal stenosis is unknown.

A mass or congenital tumor can also lead to compression of the aqueduct with resultant ventriculomegaly. In rare cases, a tumor or choroid plexus papilloma may result in overproduction of cerebrospinal fluid with resultant ventriculomegaly.17 Large isolated choroid plexus cysts may transiently dilate the fetal cerebral ventricles. Although limited data are available on outcomes of such cases, choroid plexus cysts are typically benign, and the associated mild ventriculomegaly is unlikely to be clinically significant.18

**Infection**

Approximately 5% of cases of mild to moderate ventriculomegaly are reported to result from congenital fetal infections, including CMV, toxoplasmosis, and Zika virus.14,18 Sporadic cases of ventriculomegaly associated with other viruses have also been reported (mumps enterovirus 71, parainfluenza virus type 3, parovirus B19, and lymphocytic choriomeningitis virus).20,21 Congenital infection may cause isolated ventriculomegaly due to cerebral atrophy, aqueductal stenosis due to ependymal fibrosis, or communicating hydrocephalus due to inflammation of arachnoid granulations and excess production of cerebrospinal fluid.

Many cases of ventriculomegaly associated with congenital infection demonstrate other sonographic features, including fetal growth restriction; periventricular, hepatic, and other intraabdominal calcifications; echogenic fetal bowel; hepatosplenomegaly; ascites; meconium peritonitis; polyhydramnios, and microcephaly. However, these features may not be evident until later in gestation, and not all infected fetuses will have other sonographic signs.

**Genetic disorders**

Approximately 5% of fetuses with apparently isolated mild to moderate ventriculomegaly have an abnormal karyotype,22 most commonly trisomy 21. Another 10–15% have abnormal findings on chromosomal microarray.7,9,22,23

Although hydrocephalus is a component of several congenital syndromes, there are relatively few genetic causes of isolated ventriculomegaly or hydrocephalus.24 In male fetuses, the most common inherited form of hydrocephalus is caused by a variant in the L1CAM gene, which accounts for up to 30% of males with X-linked idiopathic hydrocephalus.25 A number of other syndromes have been associated with hydrocephalus, including Walker-Warburg, Bardet-Biedl, Meckel, Joubert, and hydrocerebellar lethal syndromes.24 These conditions are typically associated with more severe ventriculomegaly as well as additional abnormalities that may be identified sonographically or by fetal magnetic resonance imaging (MRI) (see below).

**How should a fetus with mild or moderate ventriculomegaly be evaluated?**

When mild or moderate ventriculomegaly is detected (ie, when the lateral ventricle[s] measure 10–15 mm), further evaluation is indicated. Such evaluation is focused on determining whether additional structural (CNS and non-CNS) anomalies, genetic abnormalities, or congenital infection, are present.

**Ultrasonography**

The incidence of additional CNS and non-CNS sonographic abnormalities identified in fetuses with mild or moderate ventriculomegaly ranges from 10–76%, but appears to be <50% in most studies.1,2,11,26 When ventriculomegaly is identified, a detailed ultrasound should be performed by a practitioner experienced in the diagnosis of fetal anomalies. Careful attention should be given to intracranial anatomy including the lateral, third, and fourth ventricles; corpus callosum; thalami; germinal matrix region; cerebellum; and the cerebellar vermis. Ventriculomegaly is a nonspecific finding and careful attention to all fetal anatomic structures, both CNS and non-CNS, is important. The fetal heart should be carefully examined, and fetal biometry should be assessed for evidence of growth restriction. Finally, a thorough inspection should be performed for signs of fetal
infection, including intracranial or extracranial calcifications, hepatosplenomegaly, ascites, and fetal growth restriction.

**Testing for genetic disorders**

Fetal aneuploidy and copy number variants are both associated with mild ventriculomegaly. **We recommend that diagnostic testing (amniocentesis) with chromosomal microarray should be offered when ventriculomegaly is detected (GRADE 1B).** Aneuploidy screening, including cell-free DNA testing, screens for only a limited number of the most common fetal aneuploidies. Such screening assesses the risk for trisomy 21, 18, and 13 but not for other potentially important chromosomal abnormalities or other genomic variants. Cell-free DNA screening can be considered for women who decline diagnostic testing after counseling about the limitations of this approach. In women with prior normal screening test results, including cell-free DNA, diagnostic testing should still be offered due to the higher diagnostic yield.

**Testing for fetal infectious etiologies**

Congenital fetal infections, including most commonly CMV, toxoplasmosis, and Zika virus, have been associated with mild ventriculomegaly, and a history of potential exposures and symptoms of maternal infection should be elicited. The woman’s history should be reviewed for symptoms suggestive of CMV infection, and exposure to potential sources of toxoplasmosis (eg, outdoor cats, gardening, consumption of undercooked meat) and Zika virus should be assessed. **We recommend testing for CMV and toxoplasmosis when ventriculomegaly is detected, regardless of known exposure or symptoms (GRADE 1B).** Testing can include maternal serology or polymerase chain reaction (PCR) on amniotic fluid. Because the latter is more accurate, we recommend that PCR for CMV and toxoplasmosis be included when amniocentesis is performed and be offered to women during counseling regarding the benefits of diagnostic testing. For women with risk factors for Zika virus, testing is recommended per current guidelines, which are rapidly evolving.

For women who decline amniocentesis, serum testing for CMV includes IgG and IgM, as does screening for toxoplasmosis. Negative IgG and IgM results for CMV and toxoplasmosis suggest no prior exposure, which excludes these infections as the cause of ventriculomegaly; a positive IgG and negative IgM results suggest prior infection and immunity, making congenital infection unlikely as the cause of ventriculomegaly. In women with a positive CMV IgM result, IgG avidity testing is recommended; a low avidity IgG and positive IgM indicates infection within the previous 3 months. A positive toxoplasmosis IgG and IgM result may indicate a recent infection or a false-positive result. A positive IgM toxoplasmosis antibody result should be followed by IgG avidity testing and repeat IgM testing in a reference laboratory. As with CMV, high avidity IgG suggests that infection predates the pregnancy. In contrast, low avidity toxoplasmosis IgG is more difficult to interpret, because some individuals have persistent low IgG avidity for many months after infection.

For women who undergo amniocentesis, the amniotic fluid should be tested by PCR for CMV and toxoplasmosis. Amniocentesis with PCR performed <21 weeks of gestation has a 45–80% sensitivity for CMV; therefore, a negative result does not exclude CMV infection. PCR performed on amniotic fluid >21 weeks of gestation or >6–7 weeks from maternal primary infection has a higher sensitivity and a specificity between 97–100%. The positive predictive value of the test approaches 100%, although false-positive CMV by PCR results have been reported. PCR for toxoplasmosis performed on amniotic fluid has a sensitivity of 64%, negative predictive value of 87%, and positive predictive value of nearly 100%.

A detailed travel history should be included in the evaluation of ventriculomegaly. For women with a history of travel to any Zika-endemic area, testing according to the Centers for Disease Control and Prevention guidelines is recommended. The prevalence of Zika infection in women with mild ventriculomegaly but no known exposure to Zika virus is unknown, but it is likely that Zika is very rare in such cases, and therefore testing is not routinely recommended. The diagnostic accuracy of serum or amniotic fluid PCR for Zika virus infection is also unknown at this time.

**What is the role of fetal MRI?**

Fetal MRI can be useful in the evaluation of ventriculomegaly because this modality can identify significant abnormalities not easily detected by ultrasound. Diagnoses such as cortical malformations and migrational abnormalities are rarely detected by ultrasound but can be associated with mild or moderate ventriculomegaly and identified by MRI. In the setting of ventriculomegaly, the chance that MRI will identify additional abnormalities varies widely and ranges from 5–50% in reported series. The added value of MRI depends in part on the degree of ventricular dilation, as well as on the quality of the original ultrasound and whether a detailed neurosonography examination was performed by a provider with specific expertise. Not all additional findings are clinically significant or change the counseling regarding prognosis; the incidence of important additional findings detected by MRI in fetuses with mild or moderate ventriculomegaly has been reported to range from 1–14%. The most common abnormality detected on MRI but missed on fetal ultrasound is agenesis of the corpus callosum; this diagnosis is associated with a variable prognosis from subtle to severe; this depends in part on other associated brain abnormalities.

MRI is most useful at >22–24 weeks of gestation, as milestones of CNS development become more evident.
with advancing gestation. MRI is generally not useful in cases of fetal aneuploidy, as the neurologic outcome is almost certainly abnormal regardless of the results of the imaging test. However, MRI may be of benefit in assessing the extent of destructive injury in fetuses with known infection, hemorrhage, or ischemia, and when other sonographically evident CNS malformations, such as agenesis of the corpus callosum or Dandy-Walker malformation, are present.

Confirmation that mild ventriculomegaly is isolated increases the likelihood that long-term neurodevelopment will be normal, and identification of other CNS malformations makes it more likely that the fetus will have neurologic abnormalities, including developmental delay. However, there is no consensus regarding the clinical utility of MRI in this setting, which also depends on the expertise of the examining sonologist. In addition, the availability of fetal MRI varies geographically and is often institutionally dependent. Nevertheless, given the potential for detection of clinically important fetal CNS abnormalities, we suggest that MRI be considered in cases of mild or moderate ventriculomegaly when this modality and expert radiologic interpretation are available; MRI is likely to be of less value if the patient has had a detailed ultrasound performed by an individual with specific experience and expertise in sonographic imaging of the fetal brain (GRADE 2B). It is important to note that the width of the lateral ventricle is often slightly larger when measured by MRI, and the ultrasound measurement should be used for prognosis and counseling.

**What is the appropriate antenatal management of a pregnancy after the detection of mild to moderate ventriculomegaly?**

Follow-up ultrasound after initial detection of fetal ventriculomegaly is helpful to assess progression, stability, or resolution. Ventricular dilation is progressive in approximately 16% of cases; evidence of progression can change both the diagnosis and prognosis. Conversely, if the ventriculomegaly remains stable or resolves, the prognosis generally improves. The optimal timing and frequency of follow-up ultrasound examinations in the setting of mild to moderate ventriculomegaly is dependent on the initial gestational age at diagnosis as well as other clinical factors. Multiple serial exams are unlikely to be helpful if an initial follow-up ultrasound demonstrates stable findings, while a follow-up ultrasound in the third trimester to assess head circumference and rule out significant progression is reasonable.

Women should receive counseling from a health care provider, such as an obstetrician, radiologist, maternal-fetal medicine specialist, genetic counselor, or a pediatric neurologist or neurosurgeon with specific expertise in the prenatal diagnosis and prognosis of fetal ventriculomegaly. Women should be informed that the prognosis varies widely based on the exact findings of the complete prenatal and postnatal evaluation. If ventriculomegaly is progressive, consultation with a pediatric neurosurgeon may be useful, as some neonates may require postnatal surgical intervention, such as ventriculoperitoneal shunting. Overall, the likelihood of mild to moderate ventriculomegaly requiring surgical intervention after birth is low.

Antepartum fetal testing is not likely to be beneficial in the setting of mild to moderate ventriculomegaly, as this abnormality is not typically associated with placental insufficiency, unless other abnormalities such as fetal growth restriction or amniotic fluid abnormalities are present.

**What is the optimal timing and mode of delivery for fetuses with ventriculomegaly?**

There is no evidence that preterm or cesarean delivery improves maternal or neonatal outcomes in the setting of mild to moderate ventriculomegaly. Macrocephaly is rare, and we recommend that timing and mode of delivery be based on standard obstetric indications (GRADE 1C). Given the potential for mild to moderate ventriculomegaly to be associated with long-term adverse neurodevelopmental outcomes, the primary pediatrician should be made aware of this prenatal finding.

**What is the prognosis for infants with mild ventriculomegaly?**

The prognosis for infants with mild to moderate ventriculomegaly is widely variable and depends on the presence or absence of structural or genetic abnormalities, fetal infection, and the severity of ventricular dilation. If the ventriculomegaly is mild and isolated, the outcome is most commonly normal. In a recent meta-analysis, the rate of neurodevelopmental delay in truly isolated mild ventriculomegaly was 7.9%, which is similar to the background rate. Importantly, however, postnatal imaging revealed previously undiagnosed findings, some of which would impact prognosis, in 7.4% of patients.

Outcome data, particularly long-term neurocognitive outcomes, are limited by the heterogeneous nature of the studies, differences in prenatal and postnatal evaluation, inclusion or exclusion of children with other abnormalities, and the duration of pediatric follow-up. With these limitations in mind, current evidence suggests the following regarding prognosis.

**Isolated mild ventriculomegaly (10–12 mm)**

Survival for newborns with isolated mild ventriculomegaly is high, with reported rates of approximately 93–98%. The likelihood of normal neurodevelopmental outcomes is >90% and may not be different from general population rates. We recommend that with isolated mild ventriculomegaly of 10–12 mm, after a complete evaluation, women be counseled that the outcome is favorable, and the infant is likely to be normal (GRADE 1B).
Isolated moderate ventriculomegaly (13–15 mm)

Newborns with prenatal detection of isolated moderate ventriculomegaly are somewhat more likely to have adverse outcomes than those with mild ventriculomegaly. Survival for newborns with isolated moderate ventriculomegaly is reported to range from 80–97%, 1,60 and the likelihood of normal neurodevelopmental outcomes is reported to range from 75–93%. 4,11,50 We recommend that with isolated moderate ventriculomegaly of 13–15 mm, after a complete evaluation, women be counseled that the outcome is likely to be favorable but that there is an increased risk of neurodevelopmental disabilities (GRADE 1B).

In the setting of mild to moderate ventriculomegaly with associated abnormalities, the prognosis primarily depends on the specific abnormality rather than the degree of ventricular dilation.50 Outcomes are also associated with progression, and in cases in which ventriculomegaly progresses, the rate of adverse outcomes is reported to be as high as 44%, while outcomes are normal in >90% of cases in which ventriculomegaly improves.10 Recurrence risk of isolated ventriculomegaly in future pregnancies in most cases is low. In cases with an underlying cause, such as a chromosomal or genetic condition, the recurrence risk will depend on the specific diagnosis.

### Summary

When ventriculomegaly is identified, a thorough evaluation should be performed including detailed sonographic evaluation of fetal anatomy, amniocentesis for assessment of chromosomal abnormalities, and a workup for fetal infection. Fetal MRI may identify other abnormalities and can be considered when such imaging and expert interpretation are available, although MRI is not likely to add useful diagnostic information beyond that obtained with detailed neurosonography by a provider with specific experience and expertise. Follow-up ultrasound examination should be performed to assess for progression of the ventricular dilation. In the setting of isolated mild ventriculomegaly (10–12 mm), the likelihood of survival with normal neurodevelopment is >90%.

### REFERENCES


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