

Mild fetal ventriculomegaly: diagnosis, evaluation, and management

Check for updates

Society for Maternal-Fetal Medicine (SMFM); Nathan S. Fox, MD; Ana Monteagudo, MD; Jeffrey A. Kuller, MD; Sabrina Craigo, MD; and Mary E. Norton, MD

The practice of medicine continues to evolve and individual circumstances will vary. This opinion reflects information available at the time of acceptance for publication and is neither designed nor intended to establish an exclusive standard of medical practice. This publication is not expected to reflect the opinions of all members of the Society for Maternal-Fetal Medicine.

Ventriculomegaly is defined as dilation of the fetal cerebral ventricles and is a relatively common finding on prenatal ultrasound. The purpose of this document is to review the diagnosis, evaluation, and management of mild fetal ventriculomegaly. When enlargement of the lateral ventricles (\geq 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

Corresponding author: Society for Maternal-Fetal Medicine: Publications Committee. pubs@smfm.org

Received April 19, 2018; accepted April 19, 2018.

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).^{1,2} Although mild fetal ventriculomegaly is often incidental and benign, it also can be associated with genetic, structural, and neuro-cognitive 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 \geq 10 mm on prenatal ultrasound.^{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.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).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 parietal-occipital groove and glomus of the choroid plexus, on an axis perpendicular to the long axis of the lateral ventricle (Figure and Box).⁶

The incidence of mild to moderate fetal ventriculomegaly is approximately 1%.^{4,6} 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%.^{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.^{10,11}

FIGURE

Correct technique for measurement of lateral ventricle



Normal fetal brain in transventricular (axial) plane. Photograph courtesy of Alfred Abuhamad, MD.

CSP, cavum septi pellucidi.

Society for Maternal-Fetal Medicine. Mild fetal ventriculomegaly. Am J Obstet Gynecol 2018.

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.³ 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 dilatation 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.^{14,19} Sporadic cases of ventriculomegaly associated with other viruses have also been reported (mumps enterovirus 71, parainfluenza virus type 3, parvovirus 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 karyo-type,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 hydro-lethalus syndromes.²⁴ These conditions are typically associated with more severe ventriculomegaly as well as additional abnormalities that may be identified sono-graphically 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 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.^{27,28} 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.^{29,30} A positive toxoplasmosis IgG and IgM result may indicate a recent infection or a falsepositive 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.³¹

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%, $^{32-34}$ although false-positive CMV by PCR results have been reported. 32 PCR for toxoplasmosis performed on amniotic fluid has a sensitivity of 64%, negative predictive value of 87%, and positive predictive value of nearly 100%. 35

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.³⁶ The prevalence of Zika infection in women with mild fetal 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.³⁷

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.^{38–40} 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.⁴¹

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.^{2,38-48} 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%.45,46 The most common abnormality detected on MRI but missed on fetal ultrasound is agenesis of the corpus collosum²; 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.⁴⁵ 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 fetal 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.49

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.¹⁰ Conversely, if the ventriculomegaly remains stable or resolves, the prognosis generally improves.^{1,11} 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, maternalfetal 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.¹⁵ 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%.^{1,11,50} The likelihood of normal neurodevelopmental outcomes is >90%^{1,10,15} 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).

Number	Recommendations	GRADE
1	We suggest that ventriculomegaly be characterized as mild $(10-12 \text{ mm})$, moderate $(13-15 \text{ mm})$, 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 $10-12 mm$.	2B Weak recommendation, moderate-quality evidence
2	We recommend that diagnostic testing (amniocentesis) with chromosomal microarray should be offered when mild ventriculomegaly is detected.	1B Strong recommendation, moderate-quality evidence
3	We recommend testing for CMV and toxoplasmosis when ventriculomegaly is detected, regardless of known exposure or symptoms.	1B Strong recommendation, moderate-quality evidence
4	We suggest that MRI be considered in cases of mild or moderate fetal 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	2B Weak recommendation, moderate-quality evidence
5	We recommend that timing and mode of delivery be based on standard obstetric indications.	1C Strong recommendation, low-quality evidence
6	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.	1B Strong recommendation, moderate-quality evidence
7	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.	1B Strong recommendation, moderate-quality evidence

SMFM Consult Series

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

1. Gaglioti P, Oberto M, Todros T. The significance of fetal ventriculomegaly: etiology, short- and long-term outcomes. Prenat Diagn 2009;29:381-8.

2. Griffiths PD, Reeves MJ, Morris JE, et al. A prospective study of fetuses with isolated ventriculomegaly investigated by antenatal sonography and in utero MR imaging. AJNR Am J Neuroradiol 2010;31:106-11.

3. Farrell TA, Hertzberg BS, Kliewer MA, Harris L, Paine SS. Fetal lateral ventricles: reassessment of normal values for atrial diameter at US. Radiology 1994;193:409-11.

4. Alagappan R, Browning PD, Laorr A, McGahan JP. Distal lateral ventricular atrium: reevaluation of normal range. Radiology 1994;193: 405-8.

5. Salomon LJ, Bernard JP, Ville Y. Reference ranges for fetal ventricular width: a non-normal approach. Ultrasound Obstet Gynecol 2007;30: 61-6.

6 International Society of Ultrasound in Obstetrics and Gynecology Education Committee. Sonographic examination of the fetal central nervous system: guidelines for performing the 'basic examination' and the 'fetal neurosonogram.' Ultrasound Obstet Gynecol 2007;29: 109-16.

7. Faas BH, Feenstra I, Eggink AJ, et al. Non-targeted whole genome 250K SNP array analysis as replacement for karyotyping in fetuses with structural ultrasound anomalies: evaluation of a one-year experience. Prenat Diagn 2012;32:362-70.

8. Falip C, Blanc N, Maes E, et al. Postnatal clinical and imaging follow-up of infants with prenatal isolated mild ventriculomegaly: a series of 101 cases. Pediatr Radiol 2007;37:981-9.

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%,^{1,50} and the likelihood of normal neurodevelopmental outcomes is reported to range from 75–93%.^{1,11,50} We recommend that with isolated moderate ventriculomegaly of 13–15 mm, after a complete evaluation,

9. Griffiths PD, Brackley K, Bradburn M, et al. Anatomical subgroup analysis of the MERIDIAN cohort: ventriculomegaly. Ultrasound Obstet Gynecol 2017;50:736-44.

10. Melchiorre K, Bhide A, Gika AD, Pilu G, Papageorghiou AT. Counseling in isolated mild fetal ventriculomegaly. Ultrasound Obstet Gynecol 2009;34:212-24.

11. Sethna F, Tennant PW, Rankin J, C Robson S. Prevalence, natural history, and clinical outcome of mild to moderate ventriculomegaly. Obstet Gynecol 2011;117:867-76.

12. Vergani P, Locatelli A, Strobelt N, et al. Clinical outcome of mild fetal ventriculomegaly. Am J Obstet Gynecol 1998;178:218-22.

13. Weichert J, Hartge D, Krapp M, Germer U, Gembruch U, Axt-Fliedner R. Prevalence, characteristics and perinatal outcome of fetal ventriculomegaly in 29,000 pregnancies followed at a single institution. Fetal Diagn Ther 2010;27:142-8.

14. Pasquini L, Masini G, Gaini C, et al. The utility of infection screening in isolated mild ventriculomegaly: an observational retrospective study on 141 fetuses. Prenat Diagn 2014;34:1295-300.

15. Pagani G, Thilaganathan B, Prefumo F. Neurodevelopmental outcome in isolated mild fetal ventriculomegaly: systematic review and metaanalysis. Ultrasound Obstet Gynecol 2014;44:254-60.

16. Shaheen R, Sebai MA, Patel N, et al. The genetic landscape of familial congenital hydrocephalus. Ann Neurol 2017;81:890-7.

17. Anselem O, Mezzetta L, Grangé G, Zerah M, et al. Fetal tumors of the choroid plexus: is differential diagnosis between papilloma and carcinoma possible? Ultrasound Obstet Gynecol 2011;38:229-32.

18. Fong K, Chong K, Toi A, et al. Fetal ventriculomegaly secondary to isolated large choroid plexus cysts: prenatal findings and postnatal outcome. Prenat Diagn 2011;31:395-400.

19. Devaseelan P, Cardwell C, Bell B, Ong S. Prognosis of isolated mild to moderate fetal cerebral ventriculomegaly: a systematic review. J Perinat Med 2010;38:401-9.

20. Jamieson DJ, Kourtis AP, Bell M, Rasmussen SA. Lymphocytic choriomeningitis virus: an emerging obstetric pathogen? Am J Obstet Gynecol 2006;194:1532-6.

21. Bonthius DJ, Wright R, Tseng B, et al. Congenital lymphocytic choriomeningitis virus infection: spectrum of disease. Ann Neurol 2007;62: 347-55.

22. Donnelly JC, Platt LD, Rebarber A, Zachary J, Grobman WA, Wapner RJ. Association of copy number variants with specific ultrasonographically detected fetal anomalies. Obstet Gynecol 2014;124: 83-90.

23. Shaffer LG, Rosenfeld JA, Dabell MP, et al. Detection rates of clinically significant genomic alterations by microarray analysis for specific anomalies detected by ultrasound. Prenat Diagn 2012;32:986-95.

24. Kousi M, Katsanis N. The genetic basis of hydrocephalus. Annu Rev Neurosci 2016;39:409-35.

25. Finckh U, Schröder J, Ressler B, et al. Spectrum and detection rate of L1CAM mutations in isolated and familial cases with clinically suspected L1-disease. Am J Med Genet 2000;92:40-6.

26. Gaglioti P, Danelon D, Bontempo S, Mombrò M, Cardaropoli S, Todros T. Fetal cerebral ventriculomegaly: outcome in 176 cases. Ultrasound Obstet Gynecol 2005;25:372-7.

27. Brasil P, Pereira JP, Gabaglia CR, et al. Zika virus infection in pregnant women in Rio de Janeiro—preliminary report. New Engl J Med 2016;375: 2321-34.

28. Papageorghiou AT, Thilaganathan B, Bilardo CM, et al. ISUOG interim guidance on ultrasound for Zika virus infection in pregnancy: information for healthcare professionals. Ultrasound Obstet Gynecol 2016;47:530-2.

29. Society for Maternal-Fetal Medicine (SMFM), Hughes BL, Gyamfi-Bannerman C. Diagnosis and antenatal management of congenital cytomegalovirus infection. Am J Obstet Gynecol 2016;214:B5-11.

30. American College of Obstetricians and Gynecologists. Cytomegalovirus, parvovirus B19, varicella zoster, and toxoplasmosis in pregnancy. Practice bulletin no. 151. Obstet Gynecol 2015;125:1510-25.

31. Centers for Disease Control and Prevention Division of Parasitic Diseases and Malaria. DPDx laboratory identification of parasitic diseases of public concern, 2015. Available at: https://www.cdc.gov/dpdx/. Accessed 5/11/18.

32. Enders G, Bäder U, Lindemann L, Schalasta G, Daiminger A. Prenatal diagnosis of congenital cytomegalovirus infection in 189 pregnancies with known outcome. Prenat Diagn 2001;21:362-77.

33. Liesnard C, Donner C, Brancart F, Gosselin F, Delforge ML, Rodesch F. Prenatal diagnosis of congenital cytomegalovirus infection: prospective study of 237 pregnancies at risk. Obstet Gynecol 2000;95: 881-8.

34. Donner C, Liesnard C, Brancart F, Rodesch F. Accuracy of amniotic fluid testing before 21 weeks' gestation in prenatal diagnosis of congenital cytomegalovirus infection. Prenat Diagn 1994;14:1055-9.

35. Romand S, Wallon M, Franck J, Thulliez P, Peyron F, Dumon H. Prenatal diagnosis using polymerase chain reaction on amniotic fluid for congenital toxoplasmosis. Obstet Gynecol 2001;97:296-300.

36. American College of Obstetricians and Gynecologists and Society for Maternal-Fetal Medicine. Practice advisory: interim guidance for care of obstetric patients during a Zika virus outbreak. September 2017. Available at: https://www.acog.org/About-ACOG/News-Room/Practice-Advisories/Practice-Advisory-Interim-Guidance-for-Care-of-Obstetric-Patients-During-a-Zika-Virus-Outbreak. Accessed 5/11/18.

37. Eppes C, Rac M, Dunn J, et al. Testing for Zika virus infection in pregnancy: key concepts to deal with an emerging epidemic. Am J Obstet Gynecol 2017;216:209-25.

38. Griffiths PD, Bradburn M, Campbell MJ, et al. MERIDIAN Collaborative Group. Use of MRI in the diagnosis of fetal brain abnormalities in utero (MERIDIAN): a multicenter, prospective cohort study. Lancet 2017;389: 538-46.

39. Whitby EH, Paley MN, Sprigg A, et al. Comparison of ultrasound and magnetic resonance imaging in 100 singleton pregnancies with suspected brain abnormalities. BJOG 2004;111:784-92.

40. Paladini D, Quarantelli M, Sglavo G, et al. Accuracy of neurosonography and MRI in clinical management of fetuses referred with central nervous system abnormalities. Ultrasound Obstet Gynecol 2014;44:188-96.

41. Rossi AC, Prefumo F. Additional value of fetal magnetic resonance imaging in the prenatal diagnosis of central nervous system anomalies: a systematic review of the literature. Ultrasound Obstet Gynecol 2014;44: 388-93.

42. Benacerraf BR, Shipp TD, Bromley B, Levine D. What does magnetic resonance imaging add to the prenatal sonographic diagnosis of ventriculomegaly? J Ultrasound Med 2007;26:1513-22.

43. Morris JE, Rickard S, Paley MN, Griffiths PD, Rigby A, Whitby EH. The value of in-utero magnetic resonance imaging in ultrasound diagnosed fetal isolated cerebral ventriculomegaly. Clin Radiol 2007;62:140-4.

44. Valsky DV, Ben-Sira L, Porat S, et al. The role of magnetic resonance imaging in the evaluation of isolated mild ventriculomegaly. J Ultrasound Med 2004;23:519-23.

45. Parazzini C, Righini A, Doneda C, et al. Is fetal magnetic resonance imaging indicated when ultrasound isolated mild ventriculomegaly is present in pregnancies with no risk factors? Prenat Diagn 2012;32: 752-7.

46. Salomon LJ, Ouahba J, Delezoide AL, et al. Third-trimester fetal MRI in isolated 10- to 12-mm ventriculomegaly: is it worth it? BJOG 2006;113: 942-7.

47. Tercanli S, Prüfer F. Fetal neurosonography: ultrasound and magnetic resonance imaging in competition. Ultraschall Med 2016;37:555-7.

48. Malinger G, Ben-Sira L, Lev D, Ben-Aroya Z, Kidron D, Lerman-Sagie T. Fetal brain imaging: a comparison between magnetic resonance imaging and dedicated neurosonography. Ultrasound Obstet Gynecol 2004;23:333-40.

49. Behrendt N, Zaretsky MV, West NA, Galan HL, Crombleholme TM, Meyers ML. Ultrasound versus MRI: is there a difference in measurements of the fetal lateral ventricles? J Matern Fetal Neonatal Med 2017;30:298-301.

50. Beeghly M, Ware J, Soul J, et al. Neurodevelopmental outcome of fetuses referred for ventriculomegaly. Ultrasound Obstet Gynecol 2010;35:405-16.

All authors and committee members have filed a conflict of interest disclosure delineating personal, professional, and/or business interests that might be perceived as a real or potential conflict of interest in relation to this publication. Any conflicts have been resolved through a process approved by the Executive Board. The Society for Maternal-Fetal Medicine has neither solicited nor accepted any commercial involvement in the development of the content of this publication.

This document has undergone an internal peer review through a multilevel committee process within the Society for Maternal-Fetal Medicine (SMFM). This review involves critique and feedback from the SMFM Publications and Document Review Committees and final approval by the SMFM Executive Committee. SMFM accepts sole responsibility for document content. SMFM publications do not undergo editorial and peer review by the American Journal of Obstetrics & Gynecology. The SMFM Publications Committee reviews publications every 18-24 months and issues updates as needed. Further details regarding SMFM Publications can be found at www.smfm.org/ publications.

All questions or comments regarding the document should be referred to the SMFM Publications Committee at pubs@smfm.org.