

UNDERSTANDING VENTRICULOMEGALY

CARMEN AND JOHN THAIN CENTER FOR PRENATAL PEDIATRICS

What is ventriculomegaly?

Ventriculomegaly is the finding of abnormally dilated, or enlarged, ventricles in the fetal brain. Ventricles are normal structures in the brain that contain cerebral spinal fluid (CSF). There are two lateral ventricles (one on each side of the brain), a third ventricle, and a fourth ventricle.

In the second and third trimester of pregnancy (14 – 40 weeks), the lateral ventricles should measure less than 10 millimeters. Ventriculomegaly is diagnosed when these ventricles measure more than 10 millimeters in width. Ventriculomegaly can be unilateral (affecting only one side), or bilateral (affecting both sides). Ventriculomegaly may affect only the lateral ventricle(s), or the third and fourth ventricle as well.

What causes ventriculomegaly?

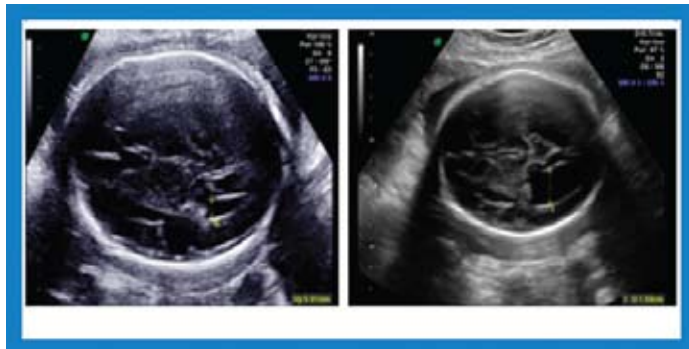
Ventriculomegaly has a number of different causes, but it can also be “idiopathic” with no identifiable cause. Some cases of ventriculomegaly are associated with chromosomal disorders or genetic syndromes, while others are associated with other structural malformations in the brain. Blockage of the connection between the third and fourth ventricles (also known as the Aqueduct of Sylvius) also leads to ventriculomegaly. This condition, known as “Aqueductal Stenosis”, results in a build-up of CSF in the lateral ventricles and in the third ventricle. Aqueductal stenosis can also be inherited (x-linked), but most cases are sporadic and result from an infection in the fetus (cytomegalovirus, rubella, or toxoplasmosis), from bleeding, or from a mass in the fetal brain.

How common is ventriculomegaly?

Ventriculomegaly is found in approximately < 2% of pregnancies. Aqueductal stenosis occurs in 1 in 2000 deliveries, and is more common in male fetuses than female.

How is ventriculomegaly detected during pregnancy?

The lateral ventricles can be easily seen and measured during obstetric ultrasound exams. Paying careful attention to the fetal head, physicians and sonographers can look for dilation of the ventricles and for



The images above demonstrate the measurement of the lateral ventricle. The left represents a normal lateral ventricle; the right represents an enlarged lateral ventricle.

evidence of other malformations that may explain the finding of ventriculomegaly. By performing repeated ultrasounds throughout the pregnancy, physicians can determine if the ventriculomegaly is stable or progressing, which may provide more information regarding the cause of the ventriculomegaly and the expected outcomes. Magnetic resonance imaging (MRI) can also be used in pregnancy to obtain more information about the structures of the fetal brain. MRI is safe for both mother and fetus, and this imaging can provide valuable information to aid in diagnosis and counseling.

How will my pregnancy be managed now that ventriculomegaly has been detected?

Our sonographers and physicians will perform another detailed ultrasound examination of the fetal anatomy to confirm the ventriculomegaly and to look for other anomalies in the fetal brain and elsewhere in the fetal body. We will also arrange for an MRI to further evaluate the anatomy of the fetal brain. A fetal echocardiogram (a detailed ultrasound of the fetal heart) will also be arranged to evaluate the cardiac anatomy and to exclude any associated major congenital heart disease. Because of the association of ventriculomegaly with infection and chromosomal disorders, we will recommend maternal blood tests and an amniocentesis to test for these conditions. You will also be referred to a Pediatric Neurologist and/or a Pediatric Neurosurgeon to discuss the management of this condition after birth and to discuss the outcomes associated with ventriculomegaly. Depending on the severity of the ventriculomegaly, we will arrange for repeated ultrasounds every 2-4 weeks to monitor for resolution or progression of the ventriculomegaly. (continued on next page)

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How will my pregnancy be managed? (continued)

Most patients with ventriculomegaly can be delivered at term. Cesarean delivery is not required except for standard obstetric complications and most patients can undergo a trial of labor. Sometimes, however, if the fetal head is enlarged or the fetus is in breech presentation, a cesarean section may be recommended by your physician.

What treatment will my baby have after birth?

Once you give birth, your baby will be carefully examined to look for causes of the ventriculomegaly and to look for other abnormalities. Infants diagnosed with ventriculomegaly will undergo further imaging (ultrasound, CT scan, MRI) after birth to confirm the diagnosis and to determine the severity of the ventriculomegaly. In cases of severe or progressive hydrocephalus, surgery is often required to decrease the pressure of the increased fluid and sometimes to remove the source of the obstruction. Pediatric neurologists and neurosurgeons will be involved in the care of your infant in the hospital, and follow-up with these doctors will be arranged for you for after your baby is discharged.

What is the long-term prognosis for babies with ventriculomegaly?

The long-term prognosis for infants with ventriculomegaly is determined by the cause of the ventriculomegaly and the presence/absence of other anomalies. Most infants with isolated mild ventriculomegaly have normal outcomes, although up to 20% have some degree of developmental delay. There is a lower rate of infant survival and higher rate of neurologic handicap in those infants with early onset or severe ventriculomegaly and in those with associated anomalies.

What are the chances I will have another baby with ventriculomegaly?

Most cases of isolated ventriculomegaly are sporadic, and have a low risk of recurrence. For example, isolated hydrocephalus is estimated to have recurrence risk of approximately 4%. Because of the possibility of an underlying genetic or inherited cause of the hydrocephalus, all patients with a history of an affected child should receive genetic counseling. In addition, patients with a previously affected pregnancy should have a comprehensive detailed ultrasound in all future pregnancies to evaluate for possible recurrence.

About the Carmen and John Thain Center for Prenatal Pediatrics

Complex pregnancies receive better care when specialists collaborate. The Carmen and John Thain Center for Prenatal Pediatrics is dedicated to helping pregnant women and their families when a birth defect or genetic syndrome is detected before the baby is born. The Center offers sensitive, complete, up-to-date information and testing, and an integrated approach to care that begins in the prenatal period and continues after birth with pediatric follow-up. A collaborative, coordinated program of care is created among specialists in perinatology, neonatology, genetics, pediatric cardiology, pediatric surgery and all pediatric subspecialties.