Objectives

1. Discuss the incidence and definition of hydrocephalus and ventriculomegaly, and how cerebral spinal fluid is formed, its flow, and its reabsorption.
2. Describe how the ventricles can grow to an abnormal size and the three major types of hydrocephalus.
3. Discuss the ultrasound findings and potential methods for making a diagnosis of hydrocephalus / ventriculomegaly.
4. Discuss the prognosis and some of the management options that may be utilized once the diagnosis is made.

Introduction

Using epidemiologic surveys, the incidence of congenital hydrocephalus ranges from 0.3 to 1.5 per 1000 births in the United States. Hydrocephalus refers to an increased intracranial content of cerebral spinal fluid (CSF). The term is generally used to refer to a situation in which an abnormal accumulation of cerebral spinal fluid results in an enlargement of the ventricular system.

The term "ventriculomegaly" is often used synonymously with hydrocephalus. However, ventriculomegaly means enlargement of the ventricles, which may be caused by increased intraventricular pressure secondary to obstruction of cerebral spinal fluid flow (for which "hydrocephalus" is the proper term), or it may be the result of passive enlargement caused by atrophy of the brain parenchyma. With atrophy of brain parenchyma, the ventricles basically enlarge to fill the voided space. Over time, it has become increasingly clear that a variable degree of enlargement of the ventricles is shared by a wide variety of anomalies different from obstructive hydrocephalus and that ventriculomegaly can be regarded as a potential marker of abnormal brain development.

Hydrocephalus almost always is due to an obstruction of cerebral spinal fluid flow, resulting in an increase in intracranial pressure. In rare instances, it may be the result of an increase in cerebral spinal fluid production or a relative decrease in the amount of CSF resorption. Hydrocephalus is said to be "isolated" when the fetus is free from anomalies that are not the direct result of the ventricular enlargement and intracranial pressure.

Etiology

Cerebral spinal fluid is mainly formed by the choroid plexus, which are infoldings of blood vessels covered by a thin
tissue called pia and are located in the four ventricles of the brain – the largest of which are found in the lateral ventricles. This CSF flows or communicates between the lateral ventricles and the third ventricle by way of the interventricular foramen (also called the foramen of Monro). The cerebral spinal fluid then slowly flows from the third ventricle to the fourth ventricle through the aqueduct of Sylvius. From the fourth ventricle, the CSF then passes through the foramen of Luschka and the foramen of Magendie to enter the subarachnoid space that externally bathes the cerebral structures of the brain and spinal cord. Flowing along the subarachnoid cisterns, the CSF is then reabsorbed by the arachnoid granulations (also called the granulations of Pacchioni) that are mainly distributed along the superior sagittal sinus.

Basically, there are four ways that the ventricles can grow to an abnormal size:

- Obstruction of CSF flow through the interventricular system and/or outflow obstruction into the subarachnoid space
- Impaired resorption of cerebral spinal fluid by the arachnoid granulations
- Overproduction of cerebral spinal fluid
- Underdevelopment or destruction of cortical tissue with a relative increase in the size of the ventricles

Hydrocephalus can be categorized into three major types:

1. Aqueductal stenosis, which accounts for about 45% of cases.
2. Communicating Hydrocephalus, which accounts for about 40% of cases.
3. Dandy-Walker Malformation, which accounts for the remaining 15% of cases.

Aqueductal stenosis is an obstruction of the flow of CSF at the level of the aqueduct of Sylvius (between the third and fourth ventricles in the brain) (figure 1). It has numerous causes including genetic (especially the X-linked recessive form), infectious (toxoplasmosis, syphilis, cytomegalovirus) (figure 2), teratogenic (radiation), and neoplastic (tumors). Communicating hydrocephalus is caused by an obstruction of cerebral spinal fluid flow that is outside of the ventricular system. The most common cause for this form of hydrocephalus is spinal defects, such as meningomyeloceles (figure 3). A meningocele can result in an Arnold-Chiari malformation (where the cerebellar vermis and fourth ventricle protrude down into the spinal canal through the foramen magnum) that obstructs the normal flow of CSF around the brain and spinal cord. Other causes for communicating hydrocephalus include obliteration of the superior sagittal sinus (where most of the arachnoid granules are located), absence of the subarachnoid granules, subarachnoid hemorrhage (clotted blood may block the subarachnoid granules), and choroid plexus papilloma. Hydrocephalus due to a Dandy-Walker Malformation is caused by a blockage of CSF flow from the fourth ventricle out through the foramen of Luschka and the foramen of Magendie. In this disorder, the fourth ventricle connects to a posterior cyst that protrudes through the cerebellar vermis (resulting in hypoplasia of the cerebellar vermis). The exact etiology is unknown; however, it is associated with several genetic disorders (autosomal recessive, autosomal dominant, X-linked, and multifactorial), some chromosomal anomalies, alcohol abuse, and possibly some viral infections.

Ultrasound Findings

The optimal approach to the antenatal diagnosis of hydrocephalus with sonography is still an unresolved issue. The first attempts to identify hydrocephalus in utero were made by visualizing a gross enlargement of the fetal head (or macrocrania). However, hydrocephalic fetuses usually do not develop macrocrania until late in gestation. Therefore, head measurements are unreliable for an early diagnosis. Presently, the identification of fetal hydrocephalus depends on the direct demonstration of an enlargement of the ventricular system with ultrasound. There have been qualitative and quantitative criteria developed to suggest the recognition of ventriculomegaly.

Regarding the qualitative criteria, this evaluation depends on the observation that in a normal fetus, the large choroid plexus almost entirely fills the lumen of the lateral ventricle at the level of the atrium and is closely apposed to both the medial and lateral walls of the ventricle. In early hydrocephalus, the choroid plexus is shrunken and anteriorly displaced, thus being clearly detached from the medial wall of the ventricle (figure 4).

For the quantitative criteria, there are normograms available for the normal size of the frontal horns, bodies, temporal horns, and atria of the lateral ventricles. A commonly used measurement is the ratio of the lateral ventricular width to that of the hemispheric width at a level just superior to or cephalic to the level of the biparietal diameter (BPD); however, the value of this determination has recently been questioned. Many diagnosticians favor the measurement of
the atria of the lateral ventricles. The internal diameter of the atrium does not vary in the second half of gestation. Thus, from 16 weeks to term, a measurement of 1 cm or less is indicative of normal brain development.

The following methods have been used to potentially make a diagnosis of and follow the progression of hydrocephalus:

- **Width of the antrum of the lateral ventricle** – a measurement of greater than 1 cm is considered abnormal. The antrum is the area where the ventricular lateral body, temporal horn, and occipital (posterior) horns are joined. The best image to measure the antrum is slightly cephalic to the plane used to measure the BPD. The ventricle on the side away from the transducer is usually easiest to see. The near field ventricle can be difficult to visualize or measure depending on the transducer, the woman's body habitus, the contrast resolution of the equipment, and/or the skill of the sonographer (figures 5 and 6).
- **Lateral ventricular width/hemispheric width ratio** – a measurement is taken of the transverse view of the head just above the plane used for the BPD in which two parallel lines can be seen. The lines are thought to be white matter tracts and not the walls of the lateral ventricles; however, this method is not considered to be very accurate.
- **Coronal view** – evaluation of the fetal head showing the antrum of the lateral ventricles and the choroid plexus. This view can be difficult to obtain due to fetal position. It may be more helpful to view the ventricular bodies and temporal horns of the lateral ventricles in the coronal view.
- **Cavum Septum Pellucidum and Cavum Verga** – the cavum septum pellucidum is a fluid-filled cavity that lies in the midline immediately anterior and superior to the thalamus. It has been called the fifth ventricle and in some cases may be enlarged; however, it normally does not exist after birth. Likewise, the cavum Verga (sometimes called the sixth ventricle, located between the corpus callosum and the fornix) may also be prominent.
- **Dangling choroid plexus** – with an enlarged ventricular space, the choroid plexus can drop to the posterior or dependent aspect of the ventricle. The result of this movement is that the anterior attachment of the plexus remains in place near the midline while the posterior portion is free to respond to gravity. Therefore, the dependent choroid plexus dangles from the attachment as a result of gravity.

**Associated Anomalies**

Hydrocephalus can sometimes be an “isolated” ultrasound finding, but is often seen with other anomalies. If hydrocephalus is detected, it is crucial that a complete detailed anatomical survey be performed of the fetus because a variety of other organs and systems may be involved.

The most common concurrent anomaly is spina bifida, probably due to its frequent association with the Arnold-Chiari malformation. Approximately one-third of all cases of hydrocephalus are associated with myelomeningocele or encephalocele. The hydrocephalus is again the result of the Arnold-Chiari malformation.

Up to 40% of hydrocephalic cases have associated anomalies of the cranial and intracranial anatomy. These may include hypoplasia of the corpus callosum, cephalocele, holoprosencephaly, hydranencephaly, cloverleaf skull, arteriovenous malformation, and arachnoid cyst. Extra-cranial anomalies can be found in up to 60% of hydrocephalic cases. They may include cardiac anomalies, gastrointestinal anomalies, cleft lip and/or palate, Meckel-Gruber Syndrome (posterior meningoencephalocele, polycystic kidneys, and polydactyly), gonadal dysgenesis, sirenomelia (mermaid syndrome), arthrogryposis, and dysplastic phalanges. Chromosomal anomalies are present in approximately 10% of cases, primarily including trisomy 13, 18, 21, translocations, and mosaicism.

**Prognosis**

For many cases, the prognosis remains uncertain. Ultrasound findings are often very limited and brain function and mental intelligence cannot be accurately determined prenatally. Unfortunately, this results in complex and difficult dilemmas for the parents.

Approximately half of the severe cases result in a stillbirth delivery or a neonate that dies shortly after delivery. One-fourth of live born hydrocephalic babies survive infancy untreated, though many are severely mentally retarded. Of those who undergo ventricular drainage procedures, about 50% survive and one-third of these have normal intelligence.

The prenatal ultrasound evaluation of hydrocephalus / ventriculomegaly, which can affect the management, prognosis, and fetal outcome, most often presents in three ways. These are:
Ventriculomegaly associated with other congenital anomalies that would be fatal (such as Meckel-Gruber Syndrome, renal agenesis, etc.). Delivery is often chosen regardless of gestational age.

Marked ventriculomegaly detected late in gestation that is associated with other abnormalities that are severe but not invariably fatal. Vaginal delivery usually occurs, though neonatal death is common.

Isolated ventriculomegaly in cases where no other anomalies are detected sonographically, or if detected, are minor. This category is the most difficult to manage and further investigation is needed. The outcome is variable and is usually uncertain.

Obstetrical Management and Care

When the diagnosis of hydrocephalus is made prior to viability, many parents choose to terminate the pregnancy. If the diagnosis is made after viability or the couple chooses to continue with the pregnancy, the following procedures can be performed (depending on the circumstances) in conjunction with patient counseling:

- Fetal echocardiogram to check for cardiac anomalies
- Amniocentesis to analyze the fetal karyotype
- Maternal testing to check for recent or current infections
- Serial ultrasound scans to follow the progression and/or regression of the hydrocephalus
- Genetic counseling: X-linked recessive aqueductal stenosis carries a 1 in 4 risk of recurrence for future pregnancies and a 1 in 2 risk for male fetuses. Cerebellar agenesis with hydrocephalus is rare but may also be sex-linked and thus have a similar recurrence risk.
- Attempt to carry the pregnancy until fetal lung maturity is achieved and then proceed with delivery followed by prompt neurologic treatment. This will potentially maximize the chances of survival and (hopefully) normal development.
- If the hydrocephalus is rapidly progressing and delivery is necessary prior to lung maturity, administer maternal corticosteroids to potentially decrease the severity of respiratory distress syndrome.
- Elective cesarean section may be indicated if macrocrania is present
- Ventriculo-amniotic shunt (the placement of a tube between the fetal ventricular system and the amniotic cavity to potentially reduce pressure) – preliminary experiments on human fetuses are not encouraging.
- Cephalocentesis prior to delivery – done to reduce the cranial size and potentially allow for vaginal delivery. This is associated with significant fetal/neonatal morbidity and is indicated only in cases where the prognosis is thought to be extremely poor.
- Shunting after birth – the prognosis is usually improved when this occurs. The outcome is better if performed before 6 months of age. If operative treatment is not delayed, most cases of hydrocephalus are compatible with normal physical development and normal head size.

The in utero treatment of ventriculomegaly can be very challenging due to many different factors. The fetus with isolated, progressive ventriculomegaly, who is most likely to benefit from in-utero shunting, is rarely identified. In addition, even experienced diagnosticians may not identify some of the associated systemic and/or central nervous system anomalies. When associated anomalies are present, the prognosis is not as favorable, which makes the decision for possible in utero treatment more difficult. Finally, current research has not identified the group of fetuses for whom in utero shunting would clearly be beneficial.

Summary

The ultrasound finding of hydrocephalus can be very devastating to parents and their families. Since the degree of ventriculomegaly is not uniformly predictive of fetal outcome and since the cause is often unknown, the decision to continue or terminate the pregnancy can be very difficult. The involved healthcare provider needs to understand these issues and be supportive of whatever decision is made by the parents.

Figures:

1 A case of hydrocephalus in a male fetus caused by aqueductal stenosis. Note the dilation of the third ventricle along with the lateral ventricles. After genetic counseling, it was determined to be the X-linked recessive disorder.

2 A case of aqueductal stenosis hydrocephalus caused by a severe in utero infection with cytomegalovirus.

3 A fetus that was diagnosed with a sacral neural tube defect that resulted in communicating hydrocephalus due to an
Arnold-Chiari Malformation. Note the "lemon sign" of the fetal cranium and the dependent choroid plexus.

4 A case of hydrocephalus that demonstrates the gravity dependent choroid plexus with cerebral spinal fluid located medially.

5 A case of hydrocephalus that demonstrates the difficulty in visualizing the near field ventricle.

6 A case of hydrocephalus that demonstrates the difficulty in visualizing the near field ventricle.
References or Suggested Reading:


About the Author(s)

Susan Riordan received her Bachelors degree in nursing in 1979 and has been a full time nurse for 21 years. She has worked in labor and delivery, postpartum, and the newborn nursery. She has been a high-risk obstetrical sonographer for the past 10 years and received her RDMS in Obstetrics and Gynecology in 1997. She currently works at Norton Suburban Hospital in Maternal-Fetal Medicine under the direction of Dr. Jonathan Weeks.