Intraventricular Hemorrhage and Periventricular Leukomalacia

Intraventricular Hemorrhage

Intraventricular hemorrhage (IVH) is bleeding inside the lateral ventricles. Bleeding frequently occurs in areas of high arterial and capillary blood flow, which most commonly occur in the subependymal germinal matrix of the brain in preterm infants. Bleeding occurs in the first 72 hours of life for about 90% of affected infants, with at least half of affected infants experiencing bleeding in the first 24 hours (Gardner, Carter, Enzman-Hines, & Hernandez, 2016). IVH is the most common type of intracranial hemorrhage present in infants.

Risk factors for IVH are prematurity and hypoxic events. Any event that results in hypoxia, alteration of cerebral blood flow, or intravascular pressure increases the risk of an infant developing IVH (Robinson, 2012). IVH also is associated with perinatal asphyxia, low Apgar scores, low birth weight, respiratory distress requiring mechanical ventilation, rapid volume expansion, and pneumothorax.

Depending on the degree of bleeding, infants with IVH may present with a range of symptoms. Some infants will not have a noticeable change in clinical condition; others will present with sudden deterioration or shock-like symptoms.

The extent of the bleed in the ventricles and brain will predict what future complications may occur. Bleeding may be confined to the germinal matrix or may enter the ventricular system. When blood enters the ventricular system, it can cause the ventricles to dilate due to increased pressure.

There are different grades assigned to IVH based on their severity. These include

- grade I (slight)—isolated germinal matrix hemorrhage
- grade II (small)—IVH with normal ventricular size
- grade III (moderate)—IVH with acute ventricular dilation
- grade IV (severe)—both intraventricular and brain parenchyma hemorrhage.

The diagnosis of IVH is determined via cranial ultrasound. For monitoring of an extensive bleed, serial ultrasounds may be used.

Periventricular Leukomalacia

Periventricular leukomalacia (PVL) refers to necrosis of white matter in the brain that occurs in a characteristic pattern. PVL is believed to be the long-term outcome of ischemia and injury to the fragile cerebral white matter in the premature infant. PVL can be caused by systemic hypotension, cerebral infarction and ischemia, and episodes of apnea and bradycardia (de Vries, 2015).

Additional complications that may arise from PVL depend on the size of the initial lesion and how much time has passed since the injury first occurred. Clinically, at about 6–10 weeks of age, an infant with PVL will present with irritability, hypertonicity, frequent tremors, and may have an abnormal Moro reflex. Diagnosis is made via cranial ultrasound, computed tomography scan, or magnetic resonance imaging. The long-term outcome of an infant with PVL may include spastic diplegia, motor deficits, intellectual deficits, visual impairments, upper arm involvement, and lower limb weakness.

Neonates who are born at younger than 30 weeks should be screened with cranial ultrasound at 7–14 days of age. Many units will rescreen again at 36–40 weeks of age to determine if PVL is present.

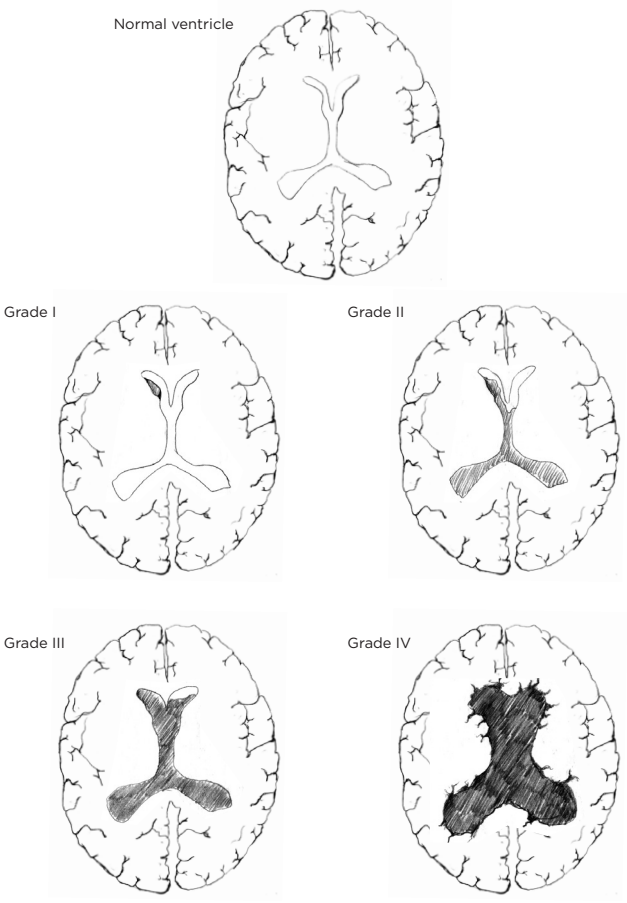
To help prevent IVH and PVL in the premature infant, care must be taken to avoid events that create swings in arterial and venous pressures. The immature neonatal brain does not have mature autoregulation of cerebral

circulation in place to compensate for changes in blood pressure (Ballabh, 2014). Prenatal interventions such as preventing preterm delivery, maternal transport to a regional neonatal center, and prenatal glucocorticoids may help prevent IVH.

Postnatal interventions include delayed cord clamping, minimized handling and suctioning, synchronized and gentle ventilation, prompt treatment of patent ductus arteriosus, maintaining normal O_2 and CO_2 levels, preventing apneic episodes and seizures, and correction of coagulopathies and bleeding disorders. The intervention that has shown the most benefit in preventing IVH is prenatal administration of glucocorticoids (Ballabh, 2014). Treatment for IVH and PVL is supportive in nature.

Hemorrhage alone will not account for all neurological deficits in the neonate with IVH. Ironically, half of premature infants with IVH will be free of neurologic symptoms. Outcome will depend on the severity of the hemorrhage. For a small hemorrhage, neurodevelopment disability is similar to that in premature infants without hemorrhage. For a moderate hemorrhage, major neurodevelopmental disability occurs in about 31% of infants; for severe hemorrhage, especially those diagnosed with post-hemorrhagic hydrocephalus and requiring shunt placement, major neurodevelopmental disability occurs in 80%–92% of infants (Ballabh, 2014). Other impairments include hearing and vision impairments. Hearing impairment, ranging from 2% to 6%, and visual impairment, ranging from 17% to 33%, may occur in infants with severe IVH (Patel, 2016).

Posthemorrhagic hydrocephalus (PHH) is a major complication of moderate to severe IVH and is defined as an abnormal accumulation of cerebrospinal fluid (CSF) in the brain that results in enlargement of cerebral ventricles. About 80% of CSF in the brain is produced by the choroid plexus within the four ventricles in the brain and the remaining 20% is produced by the brain parenchyma (Chen et al., 2017). CSF circulates to bathe the brain and spinal cord and is reabsorbed by the arachnoid granulations, or villi, within the subarachnoid space. In PHH, there are several mechanisms that result in PHH. Large

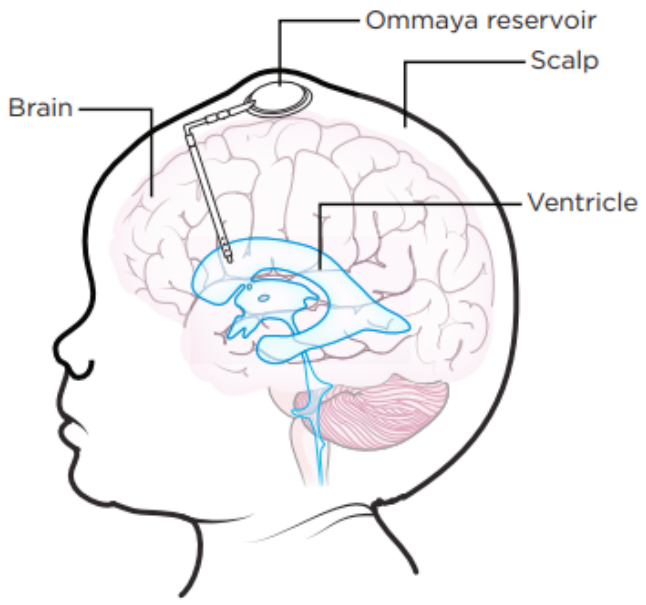


Cross-section view of brain: Grades of intraventricular hemorrhage.

blood clots can form and block the cerebral aqueducts and fourth ventricle outlet, resulting in ventricular dilatation. Small blood clots can enter the ventricular system and block the arachnoid villi, thereby reducing absorption of CSF and resulting in hydrocephalus (Chen et al., 2017). In the presence of a large bleed, lysis of the red blood cells causes the release of hemoglobin and iron. Both can lead to hydrocephalus (Chen et al., 2017). Currently, there is no effective means available to prevent PHH. Long-term outcomes for infants with PHH who require shunts may include neurocognitive impairment, motor dysfunction, and growth impairment.

Medical management to treat PHH has shown little success, and most infants will require surgical intervention. Serial lumbar punctures to remove CSF and prevent shunt placement is no longer recommended because it failed to reduce disability or the need for a shunt and

was associated with an increased risk of infection (Ellenbogan, Waqar, & Pettorini, 2016). In the past, diuretics such as acetazolamide and furosemide were used to treat PHH, but they did not prevent shunt placement and neurodevelopment outcomes were worse on the drugs, thus they should not be used (Ellenbogan, Waqar, & Pettorini, 2016). Streptokinase, a fibrinolytic, is ineffective and may increase risk of infection and secondary IVH, thus it should not be used to treat PHH (Ellenbogan, Waqar, & Pettorini, 2016).



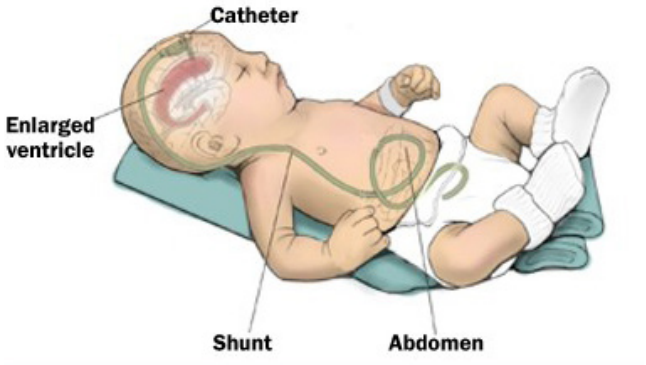
Placement of the Ommaya reservoir. ©Memorial Sloan Kettering Cancer Center. From www.mskcc.org/cancer-care/patient-education/about-your-ommaya-reservoir-placement-surgery. Reprinted with permission.

The most common surgical procedures to temporize PHH prior to insertion of a ventricular peritoneal (VP) shunt include ventricular access devices (VADs), external ventricular drainage (EVD), and ventriculosubgaleal shunts (VSGSs).

VADs are implanted with the tip of the catheter in the ventricle and a small bladder or reservoir directly under the skin. The reservoir can be tapped daily to remove 10–20 mL/kg of CSF. Complications of VADs include infection, skin breakdown, and CSF leaks (Ellenbogan, Waqar, & Pettorini, 2016). EVD has been used, but is not as common as VADs and VSGSs. EVD involves the placement of a catheter through the frontal horn of the lateral ventricle and connected to an external closed drainage system. Infection is the main complication. The benefit of EVD is questionable compared with VADs and VSGSs, therefore it may not be used often (Ellenbogan, Waqar, & Pettorini, 2016). VSGSs involve implanting a catheter from the ventricle leading into the subgaleal space. The VSGS provides consistent drainage to relieve hydrocephalus. Complications include infection and shunt failure (Ellenbogan, Waqar, & Pettorini, 2016). VADs and VSGSs are both very effective in reducing hydrocephalus. According to a recent study conducted by the Hydrocephalus Clinical Research Network, there was no difference in the proportion of infants who needed conversion to a VP shunt from a VAD or a VSGS (Wellons et al., 2017). Estimates are that 60%–80% of infants with PHH and a VSD or a VSGS will ultimately need a permanent VP shunt (Wellons et al., 2017).

VP shunts are the primary treatment to permanently relieve hydrocephalus resulting from moderate to severe bleeds in preterm newborns (Wellons et al., 2017). VP shunts involve surgically placing a catheter in the ventricle and connecting it to a tube that runs under the skin to the peritoneum. The shunt drains fluid from the distended ventricles to the peritoneum. Complications of a VP shunt include infection, obstruction, and primary shunt failure (Robinson, 2012). Studies indicate that for shunts placed before 1 year of age, up to 45% may require shunt revision within 9 months. For babies with poor peritoneal absorption, ventriculoatrial or ventriculopleural shunts may be used (Robinson, 2012).

Because of the high rate of shunt infection or failure, it is important to know the signs of shunt malfunction. Some of the most common signs of shunt malfunction or infection include fever of 38.3° C, distended veins on the head or scalp, irritability, redness or swelling at the incision site, vomiting, high-pitched cry, bulging anterior fontanel, and edema around the valve behind the ear and seizures. About 5% of premature infants with IVH will need a VP shunt (Rosenberg, 2016).



The long-term outcome of an infant with PVL may include spastic diplegia, motor deficits, intellectual deficits, visual impairments, upper arm involvement, and lower limb weakness.

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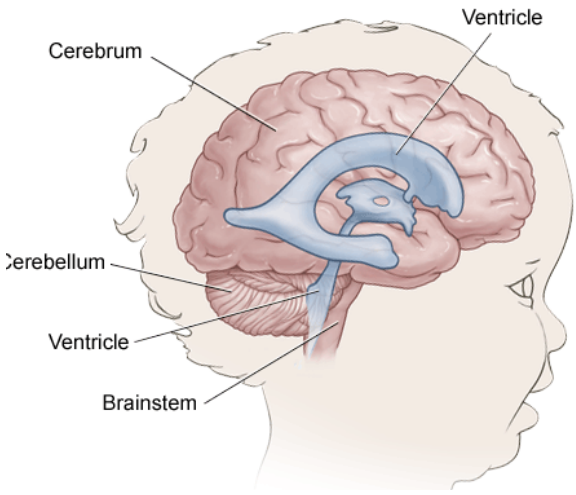
Intraventricular Hemorrhage and Periventricular Leukomalacia: Information for Parents

Your baby is very weak, including the way that his or her brain is forming. Right now, your baby's body is not able to control blood pressure changes in the same way that an older infant or adult body can. When the body can't control blood pressure changes, sometimes a baby's brain gets more blood than it needs, which can cause the vessels that carry the blood to rupture. When these vessels rupture, blood can build up inside your baby's brain and cause what is known as an intraventricular hemorrhage (IVH). The term *intraventricular* refers to the inside of the brain. The term *hemorrhage* refers to bleeding. Both terms together mean there is bleeding inside the brain.

There are different levels of bleeding in the brain with IVH. These levels also are called grades:

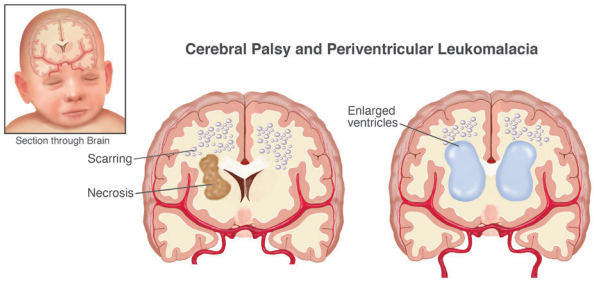
- Grade I means that there is a small bleed in the brain.
- Grade II means that there is a little more bleeding in the brain than Grade I, but it has not affected the inner part of the brain.
- Grade III means that there is bleeding that also has affected the inner parts of the brain. This includes the way that blood moves out of the brain.
- Grade IV means that there is more bleeding than in Grade III, and that the blood is pushing the brain against the bones of the head.

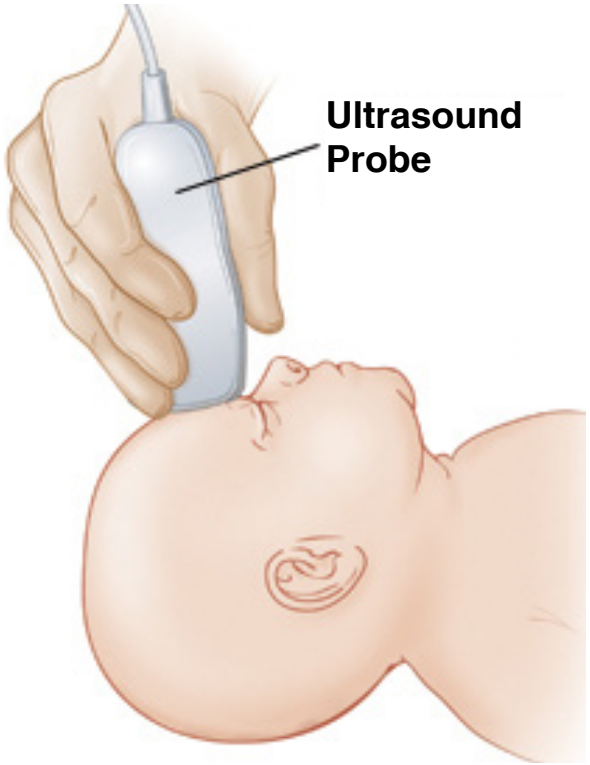
Periventricular leukomalacia (PVL) is different than IVH. *Periventricular* refers to the brain, whereas *leukomalacia* describes the way the baby's brain looks. PVL occurs when not enough blood gets to some parts of the baby's brain. These areas that have died leave little holes in the brain tissue (cysts). Babies with severe bleeding may develop a condition called posthemorrhagic hydrocephalus (PHH), which means the baby has a lot of fluid in the ventricles (where the bleeding was) or spaces in the brain. There may be so much fluid that it puts pressure on the brain tissue and may require a surgical procedure to drain the extra fluid.



Hemorrhage in the brain. © 2018 The StayWell Company, LLC. Reprinted with permission.

Your baby's provider will tell you if your baby has any bleeding in his or her brain (IVH) or if your baby has PVL. To test for IVH or PVL, your baby will need an ultrasound. An ultrasound is a painless test that uses a special wand with a jelly-like substance to take a video of your baby's brain. These videos can be broken down into pictures that a doctor will look at. Your baby's provider will then let you know the results of the test. Expect your baby to have an ultrasound after about a week of being in the hospital. Not all babies will have an ultrasound done. This is only done if your baby was born before 32–34 weeks. The ultrasound may be done again when your baby is close to 36 weeks gestation.





Ultrasound probe. © 2018 The StayWell Company, LLC. Reprinted with permission.

Your baby is at risk for bleeding in the brain because he or she was born early. Some other risks are if your baby had a low amount of oxygen during birth, had a low birth weight, or needs a machine to help with breathing.

If your baby is diagnosed with IVH or PVL, the outcome will depend on how much of your baby's brain is affected. Talk to your baby's provider to find out what the future needs will be for your baby. It is hard for your baby's providers to predict what will happen in the future for your baby, but it is important that you ask questions about any concerns that you have. Please ask your provider if you do not understand any part of the process of caring for your baby. It is OK to ask the same question more than once, so you get all the information you need to understand the diagnosis and treatment plan for your baby.