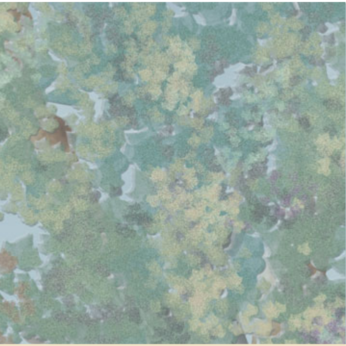
Medtronic

HYDROCEPHALUS THERAPY

Living with Hydrocephalus



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The purpose of this booklet is to help patients and their families gain an understanding of hydrocephalus and its treatment. Although an ideal treatment is not yet available, numerous advances have contributed to more effective treatment within the past few decades.

Contents

What Is Hydrocephalus?

The term hydrocephalus is derived from two Greek words: *hydro* for water and *kephale* for head. Hydrocephalus implies an excessive amount of cerebrospinal fluid (CSF), within the cavities of the brain known as ventricles. This excessive amount of CSF can result from a blockage in the brain's ventricular system which prevents the normal flow of the CSF, or as the result of a problem with CSF absorption.

In most instances, hydrocephalus is a lifelong condition since the patient is treated rather than "cured." Treatment, by shunting (sending) the CSF to another area of the body through a small tube, generally allows people with hydrocephalus to lead full and active lives.

People with hydrocephalus and their families, however, should be aware of the potential complications and their signs and symptoms so that medical care can be sought in a timely manner when necessary. This booklet is intended to help provide that information.

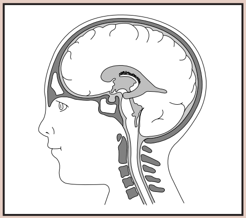


Fig 1. Normal Ventricles

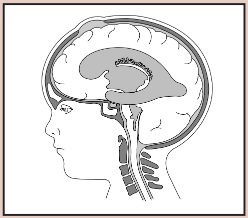


Fig 2. Hydrocephalic Ventricles

Types of Hydrocephalus

Obstructive or **non-communicating** hydrocephalus occurs when CSF flow is blocked within the ventricular system.

Non-obstructive, or **communicating**, hydrocephalus occurs when the CSF leaving the fourth ventricle is restricted in its flow over the surface of the brain, or if the sites of absorption are not functioning adequately. Hydrocephalus may also be due to overproduction of CSF by a rare tumor known as a choroid plexus papilloma.

Obstructive or non-obstructive hydrocephalus can be a **congenital** or an **acquired** condition. **Congenital** hydrocephalus simply means that the condition existed before birth, such as aqueductal stenosis or spina bifida. **Acquired** hydrocephalus develops after birth, for a variety of reasons, such as trauma, a brain tumor, scar tissue formation, or meningitis.

Normal Pressure Hydrocephalus (NPH) is an increase in the amount of CSF in the ventricles with little or no increase in the pressure inside the head. It is most often seen in adults over 60 years of age. There are three classic symptoms associated with NPH: difficulty walking, mild dementia, and impaired bladder control.

Hydrocephalus can be treated, allowing people to lead full and productive lives.

Anatomy and Physiology

The illustration below shows a view of the center of the brain with the ventricles and surrounding structures. The solid arrows show the major pathway of CSF flow. The broken arrows show additional pathways.To better understand hydrocephalus, a basic knowledge of the anatomy of the skull and brain, and the formation and absorption of CSF is helpful.

The brain occupies most of the space inside the skull and is surrounded by a cushioning layer of CSF. This fluid is primarily produced and circulated within the four interconnecting ventricles of the brain.

The ventricles contain delicate tufted structures known as choroid plexus. These structures produce most of the CSF, approximately 500 ml (about one pint) per day. The fluid is in continuous circulation and contains many substances essential for nourishment and normal function of the nervous system. CSF also provides a protective cushion within and around the brain.

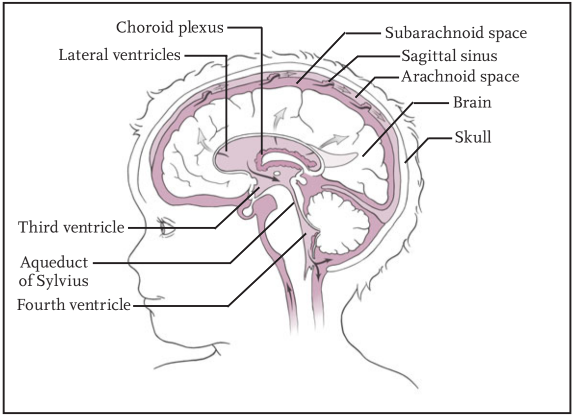
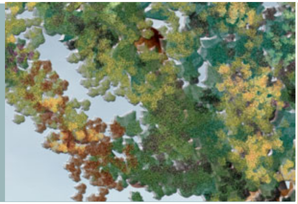


Fig 3. Cerebrospinal Fluid (CSF) Circulatory Pathway

CSF flows through the ventricular system, and out three small openings in the fourth ventricle before entering the subarachnoid space surrounding the brain and spinal cord. The fluid then flows over the surfaces of the brain and spinal cord, and is eventually absorbed into the blood stream through valve-like structures called pacchionian granulations. Thus, CSF is in a continuous process of production, circulation, and absorption. Under normal conditions, there is a delicate balance associated with this process in order to keep the amount of CSF at a constant level.

Hydrocephalus develops when CSF cannot flow through the ventricular system, or when absorption into the blood stream is not the same as the amount of CSF produced.

Diagnosis



In infants and toddlers, the bones of the skull are not yet closed and hydrocephalus may be obvious. The child's head will enlarge, and the fontanel (soft spot) may be tense and/or bulging. The skin may appear thin and shiny, and the veins of the scalp may appear full or engorged. Symptoms may include vomiting, poor feeding, listlessness, irritability, constant downward gaze of the eyes, and at times, seizures.

In older children and adults, the bones of the skull have closed. These patients have symptoms of increased intracranial pressure due to ventricular enlargement (from the extra CSF) which causes compression of the brain tissue. Symptoms may include, but are not limited to, headache, nausea, vomiting, visual disturbances, poor coordination, personality changes, lack of concentration, and lethargy.

The signs and symptoms of increasing intracranial pressure are likely to change over time, as the cranial sutures (the joints between the bones of the skull) begin to close in the infant and toddler and are fully closed in the school age child.

Signs and symptoms of increased intracranial pressure are useful in the initial diagnosis of hydrocephalus and also when there is a shunt malfunction or infection as will be discussed later.

In adults with normal pressure hydrocephalus, the symptoms are usually difficulty in walking, mild dementia, and urinary incontinence.

Diagnostic Studies

There are several diagnostic tests that can help in diagnosing hydrocephalus. These same studies can also help evaluate the shunt system in case of malfunction or infection.

Ultrasound is a sophisticated method of outlining structures within the head using high frequency sound waves. This procedure can be used only in infants whose fontanels are open, since the skull otherwise blocks sound waves.

Computerized Tomography (CT Scan) is a technique in which tiny beams of x-ray outline the skull, brain, ventricles, and subarachnoid space. In addition to visualizing the size and shape of the ventricles, abnormalities such as tumors, cysts, and other pathology can also be seen.

Magnetic Resonance Imaging (MRI) is a non-invasive diagnostic tool that uses radio signals and a magnet to form computer images of the brain, its ventricular system and coverings, and pathological lesions.

Cisternography is a test requiring injection of a small amount of radioactive material into the CSF. This test differentiates communicating from obstructive hydrocephalus, and determines CSF flow.

Air Studies are done much less frequently today than in the past. Injection of air into the ventricles, either by direct puncture or through a spinal needle, may be necessary in certain instances.

Angiography is a specialized technique in which contrast material is injected into the arteries supplying the brain. Abnormal blood vessel problems or pathological lesions can be detected with this technique.

Neuropsychological Tests are a series of questions and answers used to determine if there is decrease in brain functioning due to hydrocephalus.

Lumbar Infusion Test is a specialized technique in which fluid is injected into the lumbar subarachnoid space. This procedure is used in NPH patients to determine their CSF absorptive capacity.

Controlled Lumbar Drainage is a technique used to externally drain CSF. The test is used to determine if a patient with NPH will improve with shunt placement

Treatment

Regular visits to your neurosurgeon will be necessary.

At this time, the standard treatment for hydrocephalus is surgery. There is no long term medical treatment. The surgical procedure usually involves diverting (sending) CSF to either the abdominal cavity (a ventriculo-peritoneal or VP shunt), or to a chamber of the heart called the right atrium (a ventriculo-atrial or VA shunt). Occasionally, the CSF is shunted (sent) into the pleural cavity (ventriculo-pleural shunt) or from the lumbar spine (lower back) to the abdominal cavity (lumbo-peritoneal or LP shunt).

In order to divert the CSF, the surgeon will insert a shunt system made from silicone and polypropylene plastic. All components of the system are placed under the skin. There are no parts on the outside of the body.

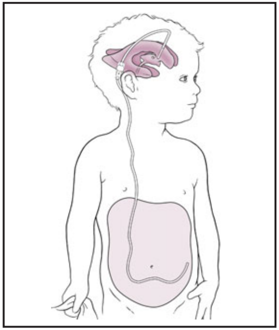


Fig 4. Ventriculo-peritoneal (VP) Shunt

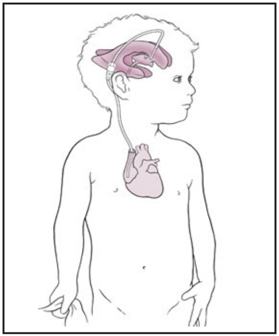


Fig 5. Ventriculo-atrial (VA) Shunt

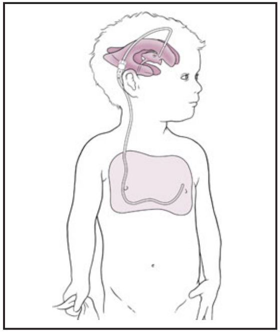


Fig 6. Ventriculo-pleural Shunt

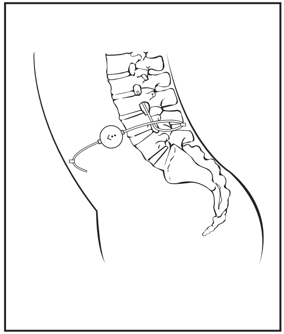


Fig 7. Lumbo-peritoneal (LP) Shunt

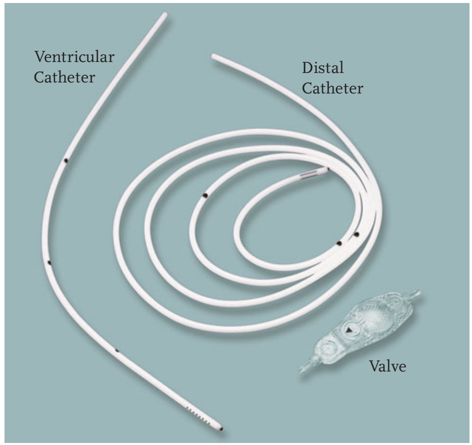


Fig 8. Shunt Components

Shunt Components

The components of a shunt system usually include two catheters and a one-way valve. The catheter placed in the ventricle of the brain is called the proximal catheter because it is closest to the ventricles. The catheter placed in either the peritoneal cavity (abdomen) or, occasionally, in the right atrium of the heart, is called the distal catheter because it is the catheter farthest way from the ventricles. Both catheters are attached to a one-way valve used to regulate the amount, direction, and pressure of CSF flow out of the ventricles. There are several different kinds CSF valves. Each valve is designed to operate at a different pressure/flow range or performance level. The surgeon's choice of valve is based on an evaluation of the type of hydrocephalus and the individual needs of the patient.

In some shunt systems a reservoir is included in the design. A reservoir can be used for a variety of reasons. By flushing the reservoir, shunt function can be tested. Also, samples of CSF for lab studies can be obtained through a reservoir. Patients and their families are discouraged from pressing the reservoir in an attempt to "test" the shunt. This maneuver can be dangerous unless done under explicit instructions from a physician.

Patients with obstructive hydrocephalus must have one catheter inserted into the ventricle. Patients with communicating hydrocephalus, however, may have the CSF drained from the subarachnoid space of the lumbar spine to another cavity of the body, usually the peritoneal cavity. This is known as a lumbo-peritoneal shunt (LP shunt)

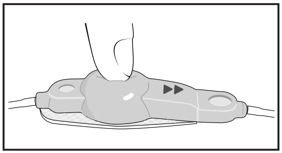


Fig 9. Valve Pumping

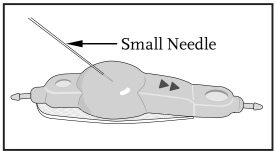


Fig 10. CSF Sampling

and is usually reserved for, but not entirely limited to, the adult population.

The type of shunt and the placement of the shunt is based on what the neurosurgeon determines is best for the patient based on the type of hydrocephalus and any other medical conditions the patient may have.

Valve Types

Some of the shunt types available include fixed pressure valves, valves with overdrainage protection (Delta® valves), or valves that can be adjusted to different pressure settings after surgery (Strata® valves).

The fixed pressure valves include a single valve mechanism that regulates the shunt flow rate. The valves are typically available in three pressure ranges: low, medium or high.

The Delta valves include an overdrainage device at the valve outlet. The purpose of the overdrainage device is to minimize excessive drainage due to gravity causing more fluid to drain when the patient is in the upright position.

The adjustable Strata valve includes a mechanism that can be non-invasively adjusted magnetically. This gives the doctor the ability to change the valve pressure setting in his office without using a surgical procedure. Since the valve includes a magnet, special precautions must be observed when being around strong magnetic sources such as retail security scanners, metal detectors, and some audio headphones. Common everyday household equipment such as microwave ovens, telephones, and computers are not strong enough to affect the valve. For patients that are undergoing an MRI procedure, it will be necessary for the doctor to check the valve pressure setting afterwards and readjust it if necessary.

Fig 12. Delta® Valves with Siphon Control

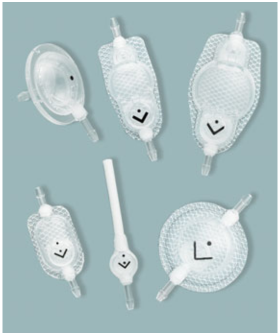


Fig 11. Fixed Pressure Valves

Fig 13. Adjustable Strata® Valves

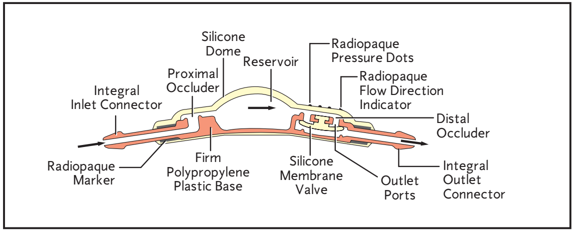


Fig 14. Cutaway of a Fixed Pressure Valve

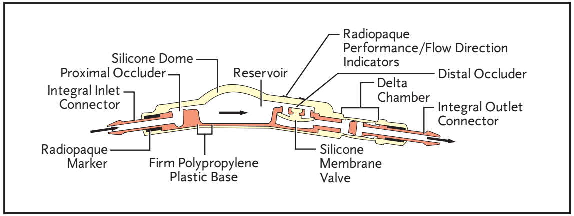


Fig 15. Cutaway of a Delta Valve

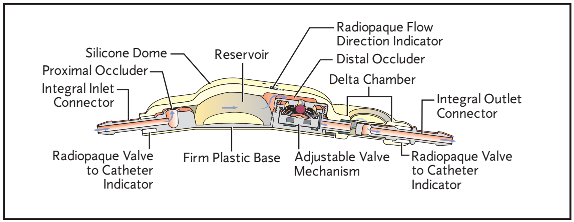


Fig 16. Cutaway of a Strata Valve

Surgery and Hospitalization

The surgical procedure is carried out under sterile conditions in the operating room. Although the operation is relatively short, careful preparation for the surgery adds extra time. In order to help prevent infection, some of the hair on the head may need to be shaved. The head and body are washed with special soap. Sterile linen is used to cover the patient and to maintain the sterile environment throughout the surgery.

A small incision (cut) is made in the scalp (the skin covering the head). A small hole is then made in the skull. A tiny opening is made in the dura, a protective covering of the brain. These openings are made to accommodate the ventricular catheter (proximal catheter) being placed into the lateral ventricle. The neurosurgeon then makes two or three small incisions in order to place the shunt valve (usually above or behind the ear). The peritoneal or atrial catheter (distal catheter) is tunneled under the skin to the abdominal or neck incision. Finally, the end of the catheter is carefully placed either in the peritoneal cavity or in a vein of the neck leading to the atrium of the heart. Following the operation, small sterile bandages are applied to each incision.

Immediately after surgery, the patient will go to the post-anesthesia care unit. The patient will remain there for close observation for about an hour and then go to his/her room. The length of hospitalization varies from patient to patient. Most patients leave the hospital within two to seven days, depending on their clinical progress.

Although this is the usual procedure when a shunt is placed, each patient may have a slightly different experience based upon their neurosurgeon, hospital, and the need to individualize the care for the patient.

Participating in Follow-Up Care

Most patients with hydrocephalus have every right to look forward to a normal future. However, because this condition is "on-going," patients do require long-term, follow-up care by a neurosurgeon. Having medical check-ups at intervals recommended by the neurosurgeon is sensible. The patient, or his/her family, must assume a share of the responsibility for follow-up care. The neurosurgeon will also keep a watchful eye on the patient and pick up subtle changes that may indicate a shunt malfunction.

Complications

Children and adults who have shunts in place will normally require surgical shunt revisions to replace a part of the shunt that is no longer working. Children can physically outgrow a shunt; any patient may eventually need a different pressure valve. CT or MRI scanning provides the physician with the tools necessary to make an early and noninvasive evaluation of the shunt's performance. In addition to regular follow-up visits to the neurosurgeon, family members should watch for symptoms of shunt complications. Immediate and accurate reporting of health related problems is very important. Flu symptoms, for example, mimic those of shunt obstruction. The early detection of complications allows revisions to be scheduled, preventing emergency situations.

Patients and their families must be alert for the signs and symptoms resulting from shunt complications. The major complications of shunting are obstruction, infection and overdrainage.

Obstruction

The most common complication of shunting is obstruction of the system. Obstruction may occur at any point along the shunt system. The openings of the ventricular catheter may become plugged with brain or choroid plexus tissue. They may become plugged due to an excessive reduction in size of the ventricular cavity (slit ventricle) due to overdrainage of CSF. The peritoneal end may become surrounded by loops of bowel, other structures, or by scar tissue. Shunts in the right atrium of the heart may be obstructed by the clotting of blood around the end of the tube. Shunt tubing may become plugged with blood elements, brain fragments, or tumor cells. The shunt may also become obstructed if the components become separated, or if the position of the shunt changes with the growth of the infant or child, either in the proximal or the distal end.

Obstruction of the shunt will produce signs and symptoms of increased pressure in the head. These will vary depending upon the degree of obstruction and age of the patient. Partial, or intermittent, obstruction may result in periodic headache, nausea and vomiting, along with drowsiness, listlessness, and decreased mental function. Poor performance in school or at work is common under these conditions.

If complete obstruction occurs, there will be a rapid development of the signs and symptoms — headache, nausea, vomiting, blurring of vision, loss of coordination, and deterioration of consciousness. The patient may become stuporous or comatose. Should this occur, emergency hospitalization for observation and treatment is required. The neurosurgeon will run tests to determine the location and degree of shunt obstruction. Removal and replacement of the obstructed part of the shunt system may be necessary.

Infection

The second most common complication of CSF shunting is infection. This hazard is present in all surgical operations — particularly when a foreign body such as a shunt system is implanted. Infection should be suspected if there is unusual redness or swelling of the wounds or along the length of the shunt system (the shunt track). These changes should be called to the attention of the neurosurgeon. If not treated, infection can lead to the wound

opening up or, more seriously, to systemic infection with chills and high fever. Infection usually requires removal of the shunt system. In some cases, the infection can be controlled with intensive antibiotic therapy, without removing the shunt.

Since the shunt system is a "foreign body," a patient may develop an allergic or inflammatory reaction to it at any time. Inflammation or open sores over any part of the implanted system should be brought to the attention of the neurosurgeon immediately.

Overdrainage

Overdrainage is generally caused when gravity drains too much fluid while the patient is upright. Overdrainage of CSF may produce a variety of signs and symptoms. Patients generally experience a headache that is worse when standing and reduced by lying down. Additional symptoms are nausea, vomiting, drowsiness and changes in vision, particularly double vision. Overdrainage should be suspected in school age children if their performance in school is declining.

Precautions

In addition to recognizing the signs and symptoms of shunting complications, common sense precautions should always be taken. Since a shunt system is implanted in the attempt to regulate CSF flow and intracranial pressure, participation in activities that might upset the delicate balance should be limited. In particular, rough contact sports and diving should be discussed in advance with the neurosurgeon.

Check with your health care team about specific physical activities you would like to do after receiving your shunt implant.

The following table summarizes many of the signs and symptoms of a shunt malfunction based on the patient's age. Each patient is an individual. Therefore, not everyone will have all of these symptoms, and some patients may have other symptoms not listed here.

It is important to remember that many other common illnesses may have these same symptoms. But since these symptoms could also be a shunt malfunction, it is very important that the patient be evaluated immediately by a neurosurgeon.

Symptoms of Shunt Malfunction or Infection

Infants	Toddlers	Children and Adults
Enlargement of baby's head	Head enlargement	Vomiting
Fontanel full and tense when infant is upright and quiet	Vomiting	Headache
Prominent scalp veins	Headache	Vision problems
Swelling along shunt tract	Irritability and/or sleepiness	Irritability and/or tiredness
Vomiting	Swelling along shunt tract	Personality change
Irritability	Loss of previous abilities (sensory or motor function)	Loss of coordination or balance
Sleepiness	Fever*	Swelling along shunt tract
Downward deviation of eyes	Redness along shunt tract*	Difficulty in waking up or staying awake
Poor feeding		Decline in academic performance
Fever*		Fever*
Redness along shunt tract*		Redness along shunt tract*

*Fever and redness along the shunt tract both indicate infection.

The information in this table is courtesy of the Hydrocephalus Association, www. hydroassoc.org. This list of symptoms is for your reference only and is not a diagnostic aid. If you are in doubt about your child's medical condition, consult your neurosurgeon immediately.

Emotional Support

The physical aspect of hydrocephalus is only one part of dealing with this condition. Emotional factors must also be considered, for the patient as well as the family.

Although the surgical procedure is likely to control the hydrocephalus, those involved may be upset, fearful, depressed, angry or frustrated. If the patient is a child, keep in mind that children have feelings similar to adults, and may be suspicious that there is a serious problem.

Since the child may not feel well, may have had some unusual tests, and is visiting the doctor more than usual, this suspicion is understandable. Rather than allowing the child's fright to escalate, and the imagination to create unrealistic, unnecessary fears, the child's anxiety may be relieved with an explanation. Knowing what to expect increases the cooperation of the child. Children, like adults, generally don't like surprises.

Explain hydrocephalus to the child in terms the child can understand. A quiet, calm atmosphere with a loving, supportive family, and as few distractions as possible, is the best environment for such a discussion.

Young children, up to about 3 years of age, are unlikely to understand. They will mostly be concerned with the here and now, especially separation from parents. In this age group, crying is quite typical as a means of attempting to gain control.

Older children, up to about 10 years of age, are usually satisfied with simple explanations and honesty. Acknowledge the child's feelings and allow the child to express them. Reassure the child that when the needles hurt, it's okay to cry and be comforted. Although going to the hospital may not be an experience a child looks forward to, telling the child the truth will help to establish and maintain trust.

Children over the age of 10 are usually able to understand more complex concepts. They can associate a variety of signs and symptoms with their condition, and can better accept limitations placed upon them.

Children of all ages are curious, and eventually ask questions. Since parents know their children and have their trust, those questions will probably be directed at Mom and Dad. Honesty is the best policy, in order to maintain the established trust. Most children's hospitals have a child life specialist on staff that can help explain about hydrocephalus and the surgery on the child's developmental level. Also, there are numerous children's books available to help children better understand their visits to the hospital.

Expressing feelings to the doctor will help him/her with professional guidance. Many people can handle their emotions with the help of relatives and friends, but for others, professional help is necessary. Health professionals caring for the patient are interested in a total well-being. Their goal is to do what is best for the patient and his/her family.

As a patient, or as the parent of a patient, take control over the dialogue with the doctor. Be an active participant in the communication process, so that the caregivers understand what kind of help is needed.

Glossary

Abdominal Cavity: The area of the body between the chest and pelvis containing the liver, intestines, kidneys and other organs

Angiography: X-ray examination of blood vessels using a contrast dye material (giving the vessels visibility)

Antibiotic: Any substance (such as Penicillin) which destroys or inhibits the growth of bacteria

Atrium: One of the two upper chambers of the heart

CSF: The cerebrospinal fluid

Cerebrospinal Fluid: The fluid filling the ventricles of the brain and surrounding the brain and spinal cord

CAT or CT Scan: Abbreviation for computerized axial tomography, a special x-ray technique which outlines the ventricles and other structures of the brain in cross section

Choroid Plexus: Delicate structures in the ventricles of the brain that produce CSF

Cisternography: Special x-ray technique using a small amount of radioactive material for seeing the ventricles and CSF filled spaces at the base of the brain

Comatose: A state in which the patient does not respond to stimulation

Congenital: A condition present since birth

Diagnosis: Determination of a patient's problem

Distal Catheter: Shunt catheter that is farthest from the ventricles, usually in the peritoneum or atrium

Dura: The fibrous membrane that surrounds the brain and spinal cord (also dura mater)

Fistula: An abnormal passage between two structures or organs

Fontanels: The spaces between the growing bones of the skull of the infant, commonly called the soft spot

Foreign Body: An object, such as an implant, introduced into a living body from the outside

Hydrocephalus: Excessive build-up of CSF in the ventricles of the brain, causing head enlargement and brain compression

Hydrocephalus, Acquired: Hydrocephalus developed after birth

Hydrocephalus, Communicating: Hydrocephalus in which there is no obstruction between ventricles and subarachnoid space

Hydrocephalus, Congenital: Hydrocephalus existing before or at birth

Hydrocephalus, Non-Communicating or Obstructive: Hydrocephalus in which there is obstruction of CSF flow between ventricles

Isotope: A radioactive material used for determining spinal fluid flow and shunt function

Lumbar Spine: The area of the spine in the small of the back

Meninges: The coverings of the brain and spinal cord

Meningitis: Inflammation or infection of the meninges

MRI: Abbreviation for Magnetic Resonance Imaging. By means of magnetic energy, images are taken, showing the ventricles and other structures within the brain

Peritoneum: Lining of the abdominal cavity

Pleura: Lining covering the lungs in the chest cavity

Pleural Space: The space between the pleura and the chest wall which contains a small amount of fluid

Pneumoencephalogram: X-ray done after filling the ventricles or subarachnoid space with air, or another gas, injected through a spinal needle

Polypropylene: Plastic used in the manufacture of shunt systems

Proximal Catheter: Shunt catheter that is in the ventricle

Shunt (noun): A system of tubing used to drain CSF from the ventricles or subarachnoid space into another area of the body

Shunt (verb): Surgical procedure during which a shunt system is implanted

Silicone: A polymer characterized by inertness in the body tissues and used in the manufacture of shunt systems and other medical devices

Slit Ventricle: Excessive narrowing of the lateral ventricle due to overdrainage of CSF

Spinal Cord: The elongated structure of the nervous system lying within the spine

Stuporous: A semi-conscious condition, in which the patient is very sleepy

Subdural Hematoma: A collection of blood between the skull and the brain

Ultrasound: Sound waves of high frequency used to outline structures within the brain

Ventricles: The four cavities (two lateral, one third, and one fourth) lying within the brain

Ventriculogram: An x-ray technique to visualize the ventricles by filling them with air or another gas

Resources

Hydrocephalus Association
1-888-598-3789
www.hydroassoc.org

Medtronic
www.medtronic.com

Spina Bifida Association of America
1-800-621-3141
www.sbaa.org

Hydrocephalus: A Guide for Patients, Families & Friends
Chuck Toporek and Kelli Robinson

Medtronic Neurosurgery
125 Cremona Drive • Goleta, California 93117-5500 USA
(800) 468-9710 USA/Canada
(901) 344-0645 International
(800) 468-9713 FAX
(901) 396-2698 FAX International

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Refer to product package insert for instructions, warnings, precautions and complications.