



A Guide to Hydrocephalus

Living Life to the Fullest

Published by the Spina Bifida and Hydrocephalus Association of Ontario





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2nd Edition

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A Guide to Hydrocephalus



This guide is intended to provide knowledge and understanding of hydrocephalus, its causes, diagnosis and treatments. It has been written for people with hydrocephalus, parents, caregivers and professionals, all of whom will find useful and relevant information in these pages.

The Spina Bifida & Hydrocephalus Association of Ontario (SB&H) is a not-for-profit organization committed to improving the quality of life of people living with hydrocephalus. A Guide to Hydrocephalus gives a general overview of the condition. More detailed information can be obtained by contacting SB&H.

This guide is intended to educate readers about hydrocephalus. The information should not be considered complete and should not be used in place of a call or visit to a healthcare or other professional, who should be consulted before you adopt any of the suggestions in this guide.





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Living with Hydrocephalus



Hydrocephalus is a complex condition affecting the brain that has a variety of causes. Because of this, those with hydrocephalus, or those assisting in their care, need to become familiar with the condition and its effects on the individual. It is extremely important that you understand the complications that can arise, and act on them immediately should any symptoms become apparent.

You will need to spend a lot of time to acquire the knowledge required for a full understanding of hydrocephalus. It is vital that you communicate with healthcare professionals every step of the way, and insist that all your questions are answered. You may also need to consult with education authorities, government departments and community agencies.

All this may be stressful work, and you will need to rely on the support of friends and family. The Spina Bifida and Hydrocephalus Association of Ontario also offers you support with extensive information, services and resources.

We hope this book will assist you on your journey as you understand and live with hydrocephalus.

Understanding Hydrocephalus



What is it?

Hydrocephalus comes from the Greek word “hydro,” meaning water, and “cephalus,” meaning head. It is a neurological condition that exists when excess cerebrospinal fluid (CSF) builds up in cavities, called ventricles, inside the brain.

Fluid accumulates in the ventricles when the body produces more CSF in a day than it can reabsorb. This accumulation causes enlargement of the ventricles, resulting in hydrocephalus.

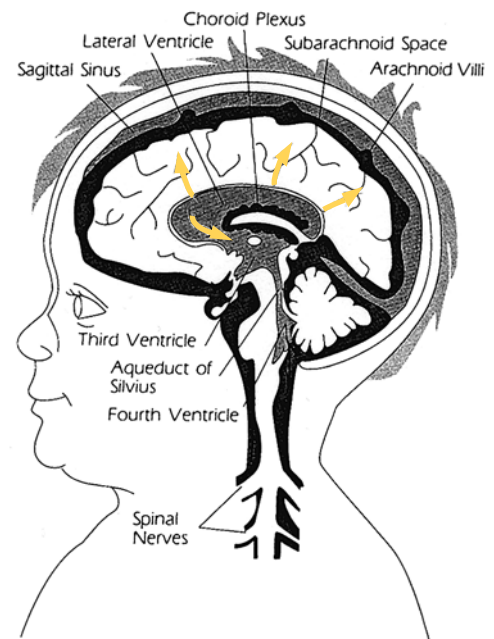
Hydrocephalus is usually treated by surgically implanting a *shunt* that takes excess CSF from the brain to another part of the body.

Who can have hydrocephalus?

Babies: Hydrocephalus may develop in the womb or after birth as a result of a congenital defect. This defect is not necessarily hereditary, but may result from something (including the condition known as spina bifida) that goes wrong during development of the fetus. Hydrocephalus may also result from complications associated with premature birth.

Children, young and middle-aged adults: Hydrocephalus may develop during these years as a result of intracranial bleeding, (stroke), brain injury, tumour growth, meningitis or other factors. Hydrocephalus that occurs after birth as a result of one of these factors is called “acquired hydrocephalus.”

Seniors: When it is diagnosed during these years, hydrocephalus is typically called “adult onset hydrocephalus.” It may take one of two forms: the common form of hydrocephalus which involves high intracranial pressure or normal pressure hydrocephalus (NPH).



Yellow arrows represent the normal direction of CSF flow.



What are the symptoms?

Infants:

- ❖ head enlargement
- ❖ fontanel (soft spot) is bulging when baby is upright and quiet
- ❖ prominent scalp veins appear unnaturally full
- ❖ fever (infection)
- ❖ vomiting (especially projectile)
- ❖ irritability
- ❖ sleepiness
- ❖ downward deviation of the eyes (sunset eyes)
- ❖ seizures

Toddlers:

- ❖ head enlargement caused by enlarged ventricles
- ❖ headache*
- ❖ vomiting (especially projectile)
- ❖ irritability and tiredness
- ❖ visual disturbances: blurred or double vision
- ❖ loss of previous cognitive or motor abilities, delayed development in walking and talking, or poor coordination or balance
- ❖ seizures
- ❖ lethargy/listlessness/sleepiness
- ❖ bowel and bladder incontinence
- ❖ change in personality, unable to concentrate

Older Children:

- ❖ headache*
- ❖ vomiting (especially projectile)
- ❖ visual disturbances: blurred or double vision
- ❖ personality change
- ❖ loss of coordination or balance
- ❖ seizures
- ❖ tiredness or difficulty staying awake
- ❖ difficulty waking up from sleep
- ❖ irritability
- ❖ incontinence
- ❖ impairment of mental or motor performance
- ❖ decline in academic or work performance

Young and Middle-aged Adults:

- ❖ chronic headaches; *
- ❖ incontinence;
- ❖ headache unrelieved by pain medication;
- ❖ visual disturbances, fainting;
- ❖ gait disturbances: clumsiness, difficulty walking on uneven surfaces and stairs;
- ❖ cognitive problems: dependent on lists, decline in academic or work performance.

* The headaches experienced by toddlers, children and adults are often at the front of the head on both sides. They are generally severe upon waking in the morning or following a nap, and may be relieved by sitting up.

If you are experiencing any symptoms related to hydrocephalus, seek immediate medical attention as the condition can become life-threatening.

Day-to-day management of hydrocephalus

Hydrocephalus is a complex condition that affects almost every aspect of life. To avoid feeling overwhelmed, target one problem at a time. Arm yourself with as many facts as you can, and talk to those assisting with your care in an open, friendly, informed manner. Communicate your expectation that change is possible. Your job will be much more difficult if you allow anger, frustration and a feeling of entitlement to enter the process.

Finding information may seem like a difficult task but SB&H can connect you with reliable sources to begin your search.

Tips for dealing with professionals

Decisions about your care are yours to make.

- ❖ Ask as many questions as you feel are necessary and write down the answers. (See *Appendix C and D*.)
- ❖ Take notes at all meetings. Include the date, those in attendance and their titles, any points made, any decisions made.

- ❖ Make sure someone is identified to perform each action agreed to at the meetings.
- ❖ Ask for copies of any reports, results or other paper-work related to you.
- ❖ Organize your information. Set up a filing system that will allow you to find relevant information easily.

Develop a support network

Dealing with a health problem can be frightening and stressful. Talking to someone with hydrocephalus, or their caregivers, is an easy way to share information and feelings. Contact SB&H to connect you with support groups and online communities.

Stay informed

Many people find that learning as much as possible about hydrocephalus, related complications and treatment options helps to relieve some of the feelings of helplessness that often accompany a diagnosis of hydrocephalus.



How is hydrocephalus diagnosed?

Whenever any, but not necessarily all, of the previously listed symptoms occur, hydrocephalus should be considered.

Before birth: Hydrocephalus can be detected as early as the latter part of the first trimester of pregnancy, and diagnosed by 13 weeks. It can be detected during routine ultrasounds that, by 20 to 24 weeks, can clearly show abnormal enlargement of the ventricles.

It is important for expectant parents to have access to specialized medical professionals, including a pediatric neurosurgeon, perinatologist, high-risk obstetrician-gynecologist, geneticist and other prenatal specialists. Counselling and support are also critical.

Premature infants: The doctor will:

- ❖ feel the fontanel, or soft spot, to see if it is fuller than normal;
- ❖ check the head circumference to see if it is within normal range;
- ❖ check muscle tone.

Full-term infants (birth through one year old): The doctor will check:

- ❖ the size of the head and fontanel for fullness;
- ❖ eyes, reflexes and “startle” response;
- ❖ developmental progress such as smiling, crawling, walking and rolling over.

Children: The doctor will check:

- ❖ that mental development milestones are being reached, such as verbal communication and memory;
- ❖ that physical development milestones are being reached, such as walking by age one, steady gait, balancing on one foot;
- ❖ tendon and muscle reflexes, visual impairments, motor skills.

In addition to a neurological examination, tests such as ultrasound, MRI and CT scans are commonly used to confirm a diagnosis of hydrocephalus.

Young and middle-aged adults:

Hydrocephalus may be diagnosed using a combination of brain scans (CT, MRI), monitoring of intracranial pressure, lumbar puncture, continuous lumbar CSF drainage and medical examination. A neuropsychological evaluation may be recommended. (See Neuropsychological evaluation in management of hydrocephalus, page 18).

Consequences of hydrocephalus

The cause, treatment and complications of hydrocephalus differ substantially from individual to individual. As a result, the consequences of the condition vary from individual to individual.

Physical consequences vary from weakness and spasticity to mild imbalance. Hydrocephalus can affect fine motor control and the movement of the hands. Other effects include headaches, nausea and tenderness around the shunt incision site.

Seizures may occur anywhere in the brain as a result of irritation caused by the shunt, shunt malfunction and compression of the brain due to abnormal tissue development. Seizures fall into two categories:

- ❖ *generalized*, which result from excessive electrical discharge throughout the whole brain;
- ❖ *partial*, which result from excessive electrical discharge in only one part of the brain.

Medication is the most common method used to treat seizures.

Sensitivity to external pressure (for example, weather changes) affects some individuals, resulting in headaches and varying degrees of disorientation.

Hearing sensitivity can cause some individuals to experience normal noises as irritating, uncomfortable or even painful.



How hydrocephalus affects others

Hydrocephalus affects siblings, other family members, friends, relationships and marriages.

Parent, or caregiver

Your time will be taken up by doctor visits and tests. There will be considerable stress and it is important to communicate with family and friends about your feelings. The more people understand about hydrocephalus, the more they will be able to help.

Siblings

During and after surgery, siblings may experience a range of emotions: worry, fear, resentment, jealousy, guilt. Talk to them. Explain what is happening to their brother or sister in easy-to-understand terms. Encourage them to ask questions.

It is important to let siblings know that they are loved and valued. They may feel isolated from the family because the individual with hydrocephalus gets so much attention, and you may observe emotional or behavioural problems. Include siblings in discussions about the individual with hydrocephalus. Involve them in family plans and make sure they have special quality time with family members.

Extended family

Others in your family may not know what to say or how to help. They will find it difficult to see you in distress. Talk to them and give them information. Suggest to grandparents ways in which they can help.

Visual problems can occur as a result of intracranial pressure damaging the optic nerves. A visual examination can sometimes determine the presence of intracranial pressure, as it causes noticeable swelling of the optic disc of the eye. If left untreated, this condition can result in impaired vision. Enlarged ventricles may cause compression or overstretching of the nerves that control eye movements.

Other visual problems include squinting; tunnel vision; perceptual problems; difficulty understanding spatial relationships and orientation of objects; difficulty estimating size, depth and distance; difficulty judging movement and speed; difficulty sorting out important information from the surrounding environment, such as seeing a word among others on a page (called figure-ground discrimination).

Constipation may be a problem and can be treated by a high-fibre diet. Consult a physician for other ways to manage this condition.

Intellectual consequences include confusion, forgetfulness, poor short-term memory.

People of all ages with hydrocephalus often experience additional consequences such as learning disabilities.



Treatment

Hydrocephalus is generally treated by implanting a *shunt*, a device that drains excess CSF from the brain to another part of the body.

Shunts

A shunt usually consists of three parts:

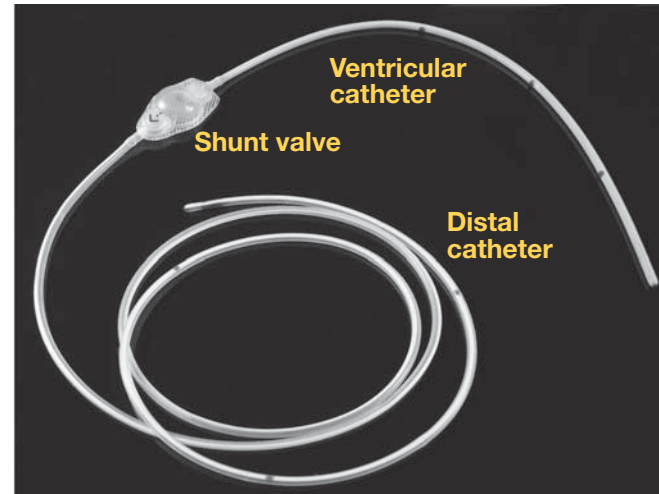
- ❖ **Ventricular catheter:** a small tube that leads from one of the ventricles, through the brain and into a valve. The catheter is passed through a region of the brain where it will least affect the individual's natural abilities.
- ❖ **Valve:** A device that controls how CSF flows through the shunt. It often includes a reservoir set against the skull and under the skin.
- ❖ **Distal catheter:** a long length of thin, flexible tube attached to the valve and placed under the skin.

Most common types of shunts:

- ❖ **Ventro-peritoneal (VP)** shunt diverts the CSF into the abdominal cavity, where it is reabsorbed by the membrane lining the stomach and intestines. This type of shunt is the most commonly used.
- ❖ **Ventriculo-atrial (VA)** shunt has the catheter in a vein leading to the right atrium of the heart. The CSF is carried from the ventricles directly into the bloodstream.
- ❖ **Lumbar-peritoneal (LP)** shunt drains from the bottom of the spine into the peritoneal cavity.

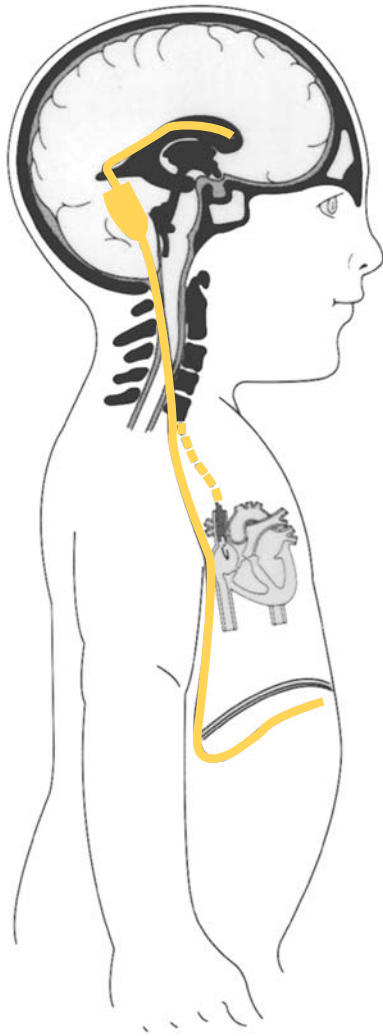
Most shunts have a reservoir that may be used to flush out the shunt; check if the shunt is working properly; and sample the CSF directly to test for infection.

Many different types and brands of shunt are available. Typically, each operates at low, medium or high valve pressure. The neurosurgeon will choose the most appropriate pressure for the circumstances.



Shunt valve and reservoir





An implanted shunt may lead to either the peritoneal cavity (abdomen or the atrium of the heart)

Advances have led to shunts with valves that can be programmed after shunt implantation surgery to adjust the opening pressure without the need to surgically replace the valve. They are called “programmable shunts” or “shunts with programmable valves.” Shunt technology continues to advance.

How do shunts work?

When CSF is produced in the ventricles, pressure builds up within the brain. Once this pressure exceeds a certain amount, the valve in the shunt opens and the excess CSF is drained to another part of the body. The individual generally does not notice the resulting minor fluctuations in pressure.

Since shunts rely on the difference in pressure between the brain and abdominal cavity, the position of the body affects the efficiency of the shunt.

If hydrocephalus has caused a substantial loss or compression of brain tissue, shunting can sometimes allow the brain to expand, decreasing the enlarged ventricular cavities.

Implanting the shunt

The surgical procedure to implant a shunt is relatively short.

- ❖ Some hair may need to be shaved to prevent infection.
- ❖ An incision is made on the scalp and a small hole is made in the skull. A tiny opening is made in the dura, a protective covering of the brain, to allow insertion of the ventricular catheter — the proximal end — into the lateral ventricle.
- ❖ Peritoneal or atrial catheters are tunnelled under the skin to an incision in the abdomen or neck.
- ❖ The end of the catheter is placed either in the peritoneal cavity or in a vein in the neck leading to the atrium of the heart.
- ❖ Once the shunt is in place, the one-way valve will automatically open to drain excess CSF.
- ❖ Following surgery, sterile bandages are applied to each incision.

Aftercare

Experiences vary, depending on the neurosurgeon, hospital and need for individualized care. In general, the following steps occur.

After surgery, the individual goes to the recovery room for observation. If no complications arise, he/she is transferred to a room. Length of stay in hospital varies.

If complications arise, the individual may be transferred to intensive care for closer monitoring.

After release from hospital, the individual will need to rest, although moderate activity is important to build up physical strength. The neurosurgeon or physician will provide advice about suitable levels of activity.

A post-operative appointment will be scheduled for several weeks after surgery. The individual should have questions or concerns ready. A CT or MRI scan may be required prior to the visit. The surgeon will check the wound and do a neurological exam to measure progress.

The next visit will be in another two or three months. Yearly visits are recommended initially but may be spaced out if all is going well.

The individual or caregiver must assume the responsibility for follow-up care and regular visits to help the neurosurgeon identify any subtle changes that may occur.



**Shunt failure or
“shunt attacks” are
medical emergencies!**

**If you are experiencing
any symptoms related to
hydrocephalus, seek
immediate medical
attention as the
condition can become
life-threatening.**

Problems with shunts

Shunts can stop working or malfunction. A shunt that stops working causes the same symptoms as untreated hydrocephalus. A shunt revision, similar to the implantation procedure, may be required to replace part or all of a broken or malfunctioning shunt. Some individuals may need few, if any, revisions. Others may have ongoing problems requiring many revisions. This cannot be predicted in advance.

The consequences of shunt malfunction vary greatly. In some cases, the shunt may totally cease to function without any adverse consequences. Or it may malfunction only occasionally, causing the individual few ill effects. However, quite often there are moderate to severe reactions, typical of excessive pressure in the brain. If shunt malfunction causes symptoms, then a revision will usually be done.

Individuals and families must be alert for signs and symptoms resulting from shunt complications. Early detection prevents emergency situations.

Blockage: Shunts may be blocked by tissue, blood cells or bacteria growths. Obstruction may occur in any of the shunt parts. The shunt may also be blocked if the parts become separated or if the position of the shunt changes.

Obstruction will produce signs and symptoms of increased pressure in the head. Partial obstruction may result in periodic headaches, nausea, vomiting, drowsiness, listlessness and decreased mental function.

Infection: Infection usually results from bacteria found normally on the skin, and occurs in 8 to 12% of all shunt surgeries. Infections may occur as long as six months after surgery.

Shunt infections can be life threatening and must be treated immediately to reduce the risk of brain damage or death.



Symptoms of a proximal catheter-end infection may include:

- ❖ headache
- ❖ fever
- ❖ swelling or redness along the shunt tract
- ❖ irritability
- ❖ meningitis (inflammation of the lining of the brain)
- ❖ stiff neck

Symptoms of a peritoneal catheter-end infection may include:

- ❖ mild to moderate abdominal pain
- ❖ fever
- ❖ change in bowel habits
- ❖ listlessness
- ❖ abdominal swelling

Shunt infections are generally treated in four steps:

- ❖ surgery to remove the infected shunt system;
- ❖ placement of a temporary external shunt system;
- ❖ antibiotics;
- ❖ surgical placement of a new shunt system.

Overdrainage: Shunts can sometimes drain too much CSF, causing the pressure in the ventricles to drop to below normal levels. This can cause the ventricles to shrink and become “slit-like.” In some situations the brain may pull away from the skull and cause bleeding that will require surgery. Individuals will generally experience a headache that is worse when standing and reduced by lying down.

Underdrainage: Shunts can sometimes drain too little CSF, usually because of blockage of the catheter. If pressure builds up rapidly, the individual may lose consciousness, in which case emergency treatment is required. However, in most cases, the consequences of underdrainage occur gradually. Symptoms may include headaches, vomiting and dizziness.





Other complications:

- ❖ Shunts are mechanical devices that can disconnect or break.
- ❖ Segments of tubing may separate at connection points. This tends to happen when scar tissue grows around the tubing, holding it stationary while the adjacent tubing moves. If this occurs, CSF may collect around the shunt and result in a soft, swollen area along the shunt tubing path.
- ❖ The shunt can become disconnected or fractured as a result of wear or a person's growth.
- ❖ The valve can fail due to mechanical malfunction.
- ❖ The peritoneal end of the shunt may be pulled out of the peritoneal cavity as a child grows taller. If this happens, the shunt may block.
- ❖ The shunt valve chosen by the surgeon may not have a pressure setting that successfully alleviates the symptoms.
- ❖ The type of shunt initially selected may not be working and may need to be replaced with a different type of shunt.
- ❖ The shunt may cause inguinal hernias in male infants. Portions of the intestines may protrude through a space in the muscle wall between the abdomen and scrota and down into the scrota.
- ❖ Shunting CSF into the abdominal cavity may cause the belly button to protrude. This is usually a temporary condition.

Precautions for those with shunts

Shunts are durable and can withstand the normal bumps and jars experienced by a child or adult. Once the scars have healed after surgery, a normal lifestyle and activities should be encouraged. Individuals wanting to play contact sports should first discuss this with their physician.

Individuals caring for a person with a shunt should be aware of its existence so that they may be alert to potential problems and take appropriate action.

Emergency procedures for those with shunts

Emergency room physicians are often general practitioners and may rarely see patients with hydrocephalus. They may be unfamiliar with specific procedures for checking for a malfunctioning shunt.

Because of this, you should always carry important information with you:

- ❖ a complete log of your prior medical history;
- ❖ the date of the shunt implantation, the type of shunt and the manufacturer;
- ❖ the number of revisions and dates;
- ❖ names of neurosurgeons and other specialists;
- ❖ a list of medications, dosages and allergies;
- ❖ a letter from your healthcare provider if there are procedures that should and should not be used during examinations.

It is also advisable to obtain and wear medical alert jewellery.



Dr. James Drake, Neurosurgeon-in-Chief,
Hospital for Sick Children, Toronto and Chair
of the SB&H Medical Advisory Board



Neuropsychological Evaluation in Management of Hydrocephalus

A neuropsychologist carries out tests and explains the interaction between the structure of the brain and the individual's abilities. The neuropsychologist describes how abilities, strengths and weaknesses show themselves in the short and long term; predicts the individual's progress; and helps the family and individual to identify appropriate resources.

Continuous testing may be needed as changes occur in the individual's condition, abilities, lifestyle and environment. A comparison of current and former evaluations will show whether the individual's shunt is working well.



Hydrocephalus in Children



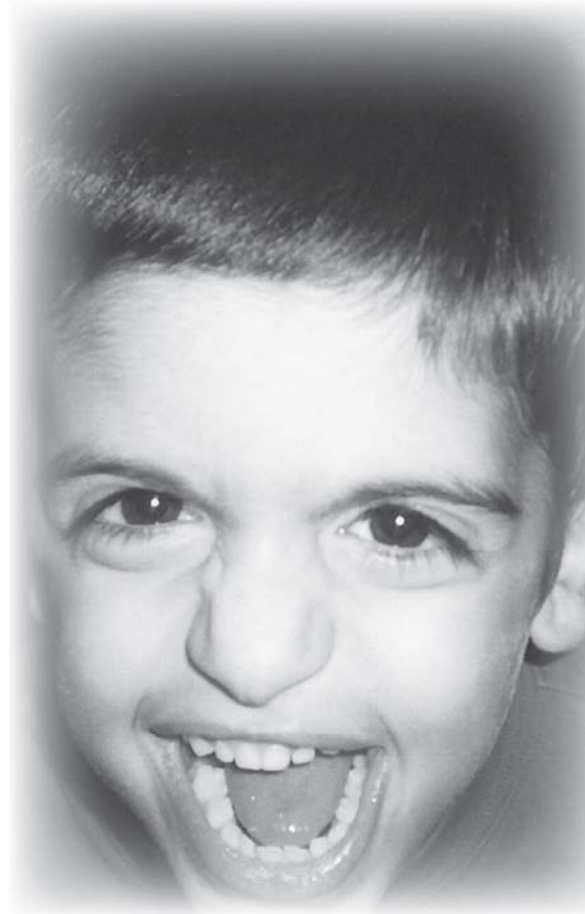
Parents should explain what hydrocephalus is to their child as soon as he/she is old enough to understand. This knowledge will give the child a healthier attitude toward the condition and allow him/her to cope better with any future complications or crises. Parents should also ask the child's neurosurgeon to talk to their child, explain the condition and allow the child to ask questions. This will help to dispel fears and misconceptions.

Parents should treat the child as they would any other. Shunts are durable and pose no special problems for childhood bumps and falls. Allow the child to live as normal a life as possible. Most children with hydrocephalus have normal intelligence, but face challenges with learning and development. Progress will depend on the nature of the problem causing the condition, and the degree of brain injury that occurred before treatment.

The child should be evaluated early on by a neuropsychologist, who can carry out neurological assessments to identify strengths and weaknesses in abilities. A child psychologist can help to maximize the child's physical, intellectual, emotional and social development. Regular evaluations are beneficial because it is believed that there are critical stages in development at which optimal learning takes place.

When travelling, parents should find out the names of medical resource personnel in the area they will be visiting, and take along copies of the most recent medical tests.

Development of independence and social skills should form part of the child's curriculum and home life. The child should be encouraged at an early age to form relationships outside the family. Independence should be emphasized. Giving the child tasks around the house will improve his/her potential to live independently in the future.



Issues such as employment and independent living will arise as the child grows into adolescence and then adulthood. Normally a difficult transition, it can be even more so for a child with hydrocephalus.

It is the parents' responsibility to pull together all the services the child needs and then begin to encourage the child to take more responsibility for his/her own life and decisions as appropriate. It is important to look to friends and family for emotional support during this time and to take time for oneself.



Congenital Hydrocephalus



Hydrocephalus may develop in the womb from a variety of factors including:

A birth defect or brain malformation

Aqueductal Stenosis: The most common form of congenital hydrocephalus, this involves an obstruction of the cerebral aqueduct — the long, narrow passageway between the third and fourth ventricle in the brain. Aqueductal blockage may result from narrowing of the aqueduct, infection, hemorrhage or tumour growth.

Arachnoid cysts: These abnormal, CSF-filled sacs are surrounded by the arachnoid membrane. Although they may occur anywhere in the brain, they are often found at the back, adjacent to the third ventricle, where they can obstruct the normal flow of CSF.

Dandy-Walker syndrome: This syndrome results from abnormal development of the brain. Typically, the fourth ventricle is enlarged as a result of a partial or full blockage of the opening through which CSF passes out of the fourth ventricle.

Porencephaly: Porencephaly is a rare condition in which cysts and cavities are present within the cerebrum. They may result from a destructive lesion caused by stroke, infection or abnormal development.

Spina bifida: Latin for “open spine,” this is a neural tube birth defect where the spine fails to develop properly, resulting in varying degrees of damage to the spinal cord and nervous system. Close to 90% of those born with spina bifida have some degree of hydrocephalus. This occurs as a result of Arnold-Chairi Malformation where the elongated cerebellum and brain stem protrude into the spinal column through the opening at the base of the skull.



X-linked hydrocephalus: This is a genetic disorder passed from the mother to the child on the X chromosome. The symptoms are usually exhibited in boys. There is also a small chance that first cousins of children with uncomplicated congenital hydrocephalus may inherit the disorder.

A variety of other disorders may also cause congenital hydrocephalus.



Acquired Hydrocephalus

Hydrocephalus can develop after birth for a number of reasons:

- ❖ **Intra-ventricular hemorrhage (bleeding into the ventricles):** can occur in premature babies when small blood vessels alongside the ventricles rupture. It may also result from a blow to the head, stroke or the malformation of blood vessels, or from a tumour. Development of hydrocephalus depends on the severity of the bleed and whether there is obstruction to the CSF pathways or reduction in the brain's ability to absorb CSF. Hydrocephalus from this cause is often mild and tends to stabilize.
- ❖ **Meningitis:** an infection, generally the result of a virus or bacteria, it causes inflammation of the membranes surrounding the brain and spinal cord and can scar the membranes lining the CSF pathway. If this scarring restricts or obstructs the CSF flow, hydrocephalus may develop.
- ❖ **Brain tumours:** may block the CSF flow, especially if they are adjacent to the ventricular system. Hydrocephalus may persist after removal of the tumour.
- ❖ **Severe head injury:** can cause hydrocephalus when brain tissue, nerves or blood vessels are damaged.



Additional Consequences of Hydrocephalus for Children



As well as the consequences listed on pages 9 and 10, children are vulnerable to other conditions or issues related to hydrocephalus.

Hormonal imbalance

Hormonal imbalance results from the production of abnormal levels of hormones. For example, it can shut down the growth plates of bones, causing short stature.

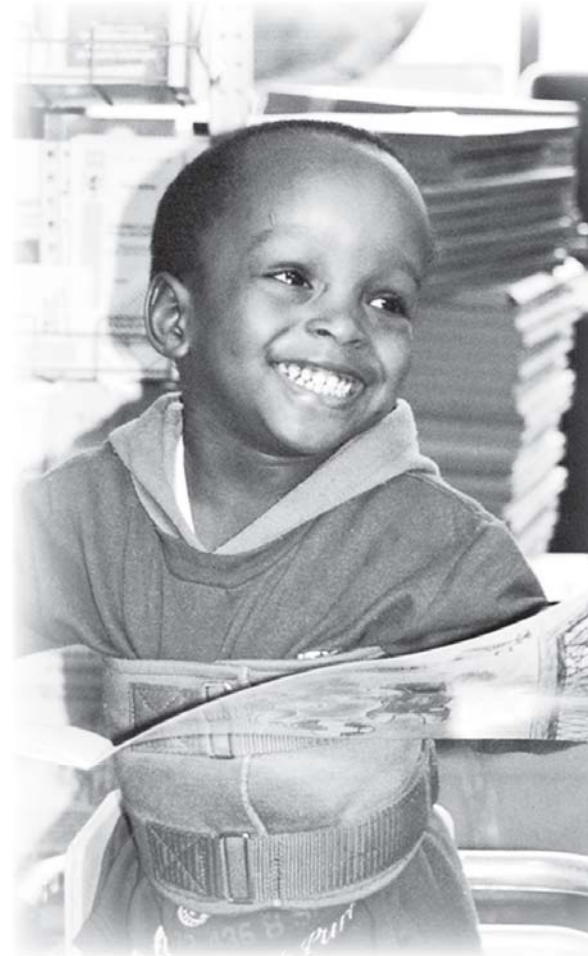
Increased intracranial pressure can damage the hypothalamus and pituitary glands, which control the release of hormones into the blood stream. Hormonal imbalance may also be linked to the number of shunt revisions prior to reaching the normal age of puberty.

The growth rates of children and adolescents should be tracked regularly. If a hormonal imbalance is suspected, the family doctor should be alerted. Medical tests may be conducted to determine if there is an imbalance — for example, an x-ray of the hands can determine if a child's bones are aging too quickly. Referral to an endocrinologist may also be needed.

If caught early enough, hormonal imbalance can be treated by hormone supplements.

The most common signs of hormonal imbalance in a child are:

- ❖ Noticeably shorter than classmates;
- ❖ Grows less than 5 cm (3 inches) a year between the ages of two and 14;
- ❖ Significant changes in the growth pattern;
- ❖ Failure to show signs of sexual development by age 13 for girls and 15 for boys;
- ❖ Entering puberty much earlier than usual (before age eight for girls, 10 for boys). This condition is common in children between the ages of five and 13 and while it affects both sexes, it is seen more often in girls.





- ❖ Diabetes insipidus, (not related to sugar diabetes) occurs when there is inadequate secretion of the hormone vasopressin, or when kidneys do not respond to stimulation by this hormone. Symptoms include constant thirst, frequent urination, abdominal bloating at night, dry hands, constipation.

Behavioural issues

Behavioural issues cover a range of abnormalities. Hyperactivity and inattentiveness may be associated with poor short-term memory. This may be improved with verbal or written reminders, memory games and short bursts of work.

Verbal aggression and swearing, bizarre behaviour that has no apparent trigger and obsession with objects, shapes, colour, or people can occur. The child may threaten suicide, engage in risky behaviour or have temper tantrums, particularly if routine is changed, if there is a perceived insult, or if there is a change in the weather. The child may exhibit inappropriate sexual behaviour.

The child's struggle with daily activities may lead to feelings of isolation and rejection at a time when physical activities are an important part of growing up and bonding with other children. The child may have no friends of his/her own age.

Once the behavioural issue is identified, look for reasons. Is the behaviour cyclical, that is, does it come and go regularly? Is it related to associated conditions, such as seizures, or to the starting or stopping of medications? Are herbal or recreational drugs being used? Is it related to short-term memory problems, or difficulties with spatial awareness or concentration? Is it stress-related? Are there new circumstances at home or school? Does it occur at the start or end of the day, indicating intracranial pressure changes? Is the shunt working properly?

The assistance of a specialized professional may be needed to deal with behavioral issues.

Learning Disabilities



The population of children with hydrocephalus includes individuals whose overall learning abilities range across the same broad span as that found in the general population of children — from those whose overall abilities are exceptionally strong to those who experience considerable limitations. Medical advances that enable earlier diagnosis, improved treatment and a reduction in the number of post-operative complications have improved the intellectual outcomes for children with hydrocephalus.

In the area of individual skills, children with hydrocephalus often show uneven developmental progress from one area to another. They may be able to gain a good level of skill development and understanding in one area with relative ease, but may have much more difficulty in another area. In this way, they are similar to other children who have specific learning disabilities. Taking note of each child's individual pattern of strengths and difficulties allows for the particular supports that they need to be put into place, to enable them to continue to grow and make progress in all areas.

Many children with hydrocephalus may have memorized important information and learned procedures for solving certain types of problems in the past. However, when faced with the need to solve a problem in a given moment, they are often slower in bringing to mind all of the relevant information and procedures that they have previously learned. It may not come naturally; they may really have to put effort into thinking about it. They often need more time to assemble and hold the information in their minds, evaluate it, decide what is most appropriate to the given situation and then act appropriately on it.

In addition to being slower in bringing information to mind, the number of things they can hold in their minds at one time may be constrained, necessitating their having to work harder at remembering. Because they



Aids to learning

In order for children with hydrocephalus to stay actively engaged in a learning process, the following features need to be embedded into all of their in-class work

- ❖ ample time to complete their thought processes;
- ❖ redirection to come back to the task when they have become distracted;
- ❖ reminders, either by being given the information in visual or written form or by verbal repetition, to help them keep in mind all the things they need to consider;
- ❖ strategies (such as pictorial or oral lists) to help them to remember;
- ❖ practice in implementing procedures so that they become more automatic;
- ❖ routines encouraging them to habitually check their own work;
- ❖ routines encouraging them to search for new strategies when indicated;
- ❖ strategies to deal with the stress and frustration that they may be feeling;
- ❖ reassurance and support.

may be slower at figuring out which stimuli are relevant to the task at hand and which are not, they may be more vulnerable to distraction. They may have difficulty in taking the initiative to solve problems, as they struggle to weigh the information at hand, generate ideas regarding possible courses of action and choose among these.

Having to put so much effort into solving problems that are more or less automatic for many other people can be very frustrating for these children. If they feel overwhelmed, they are very likely to respond with the first thing that comes to mind, or just give up and do something else. Often, they may be in a situation in which they are under pressure to respond before they have been able to complete all of this processing, and they may naturally respond by feeling more frustrated and overwhelmed. These emotions make it even more difficult to stay focused and concentrate.

Others may misinterpret the slowness of some children with hydrocephalus to respond. If one does not fully understand how difficult it is for such children to bring to mind and apply to a new situation what they have learned before, one may easily interpret their behaviour as resistant or lacking in motivation. Or, one may conclude that the children have not learned from past experience. Both of these interpretations could make it difficult to give these children the help they need to learn effectively.

Development of language is another area where children with hydrocephalus may differ from children in the general population. Often, verbal skills (such as talking, vocabulary, memorizing poems and songs) tend to develop better than “non-verbal” skills (such as drawing, colouring, cutting or putting puzzles together). This can be reflected in larger-than-normal differences between “verbal” and “non-verbal” IQ scores.

Once a child enters school, handwriting may stand out as a special difficulty. It is often slow. Letters may be large, uneven or poorly executed, and writing wanders from the horizontal. Use of large-barrelled pencils and lined paper help to minimize these problems. Children with these difficulties should be taught specific techniques for writing and given frequent, short practice periods. Their work should be marked for



content, not neatness, or else these two aspects of written work might be marked separately. For children with hydrocephalus, adaptive motor learning (for example, learning to trace an object) requires more time to complete a task and the child retains less information.

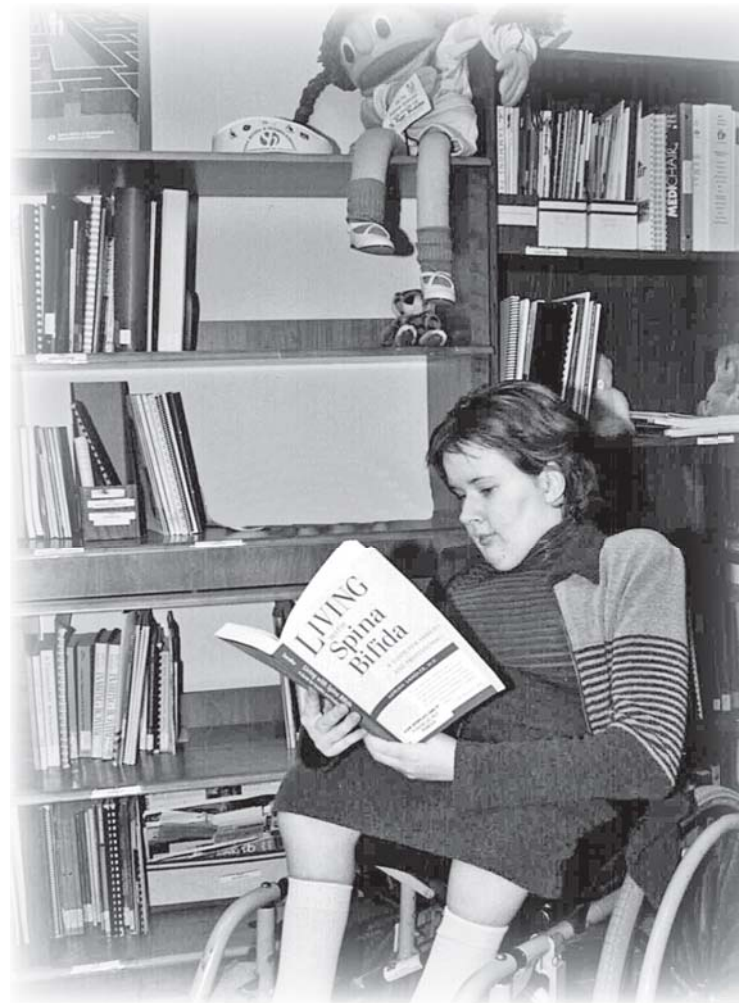
In later grades, reading comprehension is likely to lag behind word recognition. A child with hydrocephalus can build a strong vocabulary and produce grammatical speech. However, linking new ideas with learned knowledge can become difficult and drawing inferences and “reading between the lines” proves to be challenging.

Math may be another area of difficulty. It can be hard for young children with hydrocephalus to grasp concepts such as time, order and sequencing.

The reasoning required for understanding basic mathematics may be impaired in children who have not had the experience of manipulating small objects. Sometimes they have difficulty copying problems from the board to their paper because their visual-spatial processing is impaired. For the same reason, these children sometimes have difficulty keeping numbers in the proper columns or remembering where they were working on a page.

Frequent school absences, vision impairment and poor concentration and attention are other possible reasons for difficulty in mathematics. The more abstract and spatially loaded courses such as geometry and science may present special difficulties.

Organization may be a problem throughout school, but often becomes especially apparent in the later grades, when more independence and forward planning are required. In the junior high school years and beyond, study skills courses should be encouraged, as this will go far toward addressing the problems with organization. Only when the underlying cause is identified can reasonable remedial approaches be designed.





Development of social skills is another area where observation is required. Frequent absences from school sometimes make social contacts awkward and hard to maintain. In addition, some children with hydrocephalus may have difficulties “reading” subtle clues that rely on non-verbal interaction (for example, facial expression, body language, tone of voice) or aspects of communication such as irony and sarcasm. As a result, they may respond in ways that their peers find uncomfortable.

Conscious teaching of these types of interactions and skills is sometimes necessary. It is important to foster a child’s motivation to meet new people and develop new friendships. Children with hydrocephalus may have to face more teasing and misunderstanding. However, they should not be discouraged from forming close bonds with peers and expressing their feelings for others. There may be certain social difficulties to overcome relating to hydrocephalus, yet one must keep in mind that these children are still very verbose, opinionated and endearing, and have strong friendships to offer.

Each child with hydrocephalus is unique and obviously not every child will display all of these difficulties. In fact, some children may not display any form of learning problem at all. However, if parents are alert to the types of problems that may arise, they will be more sensitive to their presence. Should parents notice unevenness in their child’s development, or specific difficulties in any of these or other areas, it may be useful to obtain a psychological (neuropsychological or psycho-educational) assessment in order to document strengths and weaknesses and to suggest remedial approaches. (See Neuropsychological evaluation in management of hydrocephalus, page 18). This can be obtained privately or through the school system.

Hydrocephalus in Young and Middle-aged Adults



The causes of hydrocephalus in young and middle-aged adults may include:

- ❖ cysts or tumours
- ❖ meningitis
- ❖ encephalitis
- ❖ concussion or head injury
- ❖ certain strokes
- ❖ brain hemorrhages
- ❖ unknown causes (idiopathic)

Social and emotional issues

A diagnosis of hydrocephalus in the years from young adulthood to middle-age can present serious psychosocial and emotional problems. For parents, their parenting skills and confidence in these skills may be affected. There may be significant concerns about dying and leaving the child without financial security and support. The child, on the other hand, may worry about being a burden to the family, relying upon aging parents or busy siblings for long-term care and support.

There is a danger that hydrocephalus may go undiagnosed in this age group. Young and middle-aged adults are frequently told that their symptoms are “in their heads.” These symptoms are often dismissed as job dissatisfaction, midlife crisis, depression or other psychiatric problems. Those who have been diagnosed often feel great relief that their complaints are validated and that their symptoms are real.

A diagnosis of hydrocephalus in these years may lead to workplace concerns. There may be a decline in cognitive functioning that often manifests itself in decreased job performance. Forgetfulness makes it harder to do a job right, and is often perceived as mental vagueness.





Those with hydrocephalus may worry about job security and their future abilities — employers are not always familiar with hydrocephalus and may not readily believe that medical factors are behind the decrease in work performance.

Having a baby

Women who have hydrocephalus and become pregnant may experience complications related to hydrocephalus including: severe headaches; shunt blockage; bladder, kidney and other neurological problems. There is greater risk that women with hydrocephalus and other conditions may have a baby with a neural tube defect or hydrocephalus.

Important considerations for women

- ❖ Obtain preconception counseling.
- ❖ Have a preconception CT or MRI to give the doctor a scan for comparisons should a shunt malfunction occur during pregnancy.
- ❖ Take prenatal vitamins, containing folic acid, before and during the pregnancy.
- ❖ Consult a physician about taking higher doses of folic acid to minimize the risk of having a baby with a neural tube defect or hydrocephalus.
- ❖ Discuss current medications, including anticonvulsants, with a doctor. Pregnant women with shunts should not take some anticonvulsants.
- ❖ Consider having prenatal testing.
- ❖ Ensure that physical and neurological monitoring is conducted throughout the pregnancy.

Hydrocephalus in Older Adults



Adult onset hydrocephalus

When hydrocephalus is diagnosed in an adult, it is typically called adult onset hydrocephalus. It may be the common form of the condition, characterized by high intracranial pressure, or it may be Normal Pressure Hydrocephalus (NPH).

The causes of hydrocephalus in adults may be similar to the causes for all other ages. They include:

- ❖ cysts or tumours
- ❖ meningitis
- ❖ encephalitis
- ❖ concussion or head injury
- ❖ certain strokes
- ❖ brain hemorrhages
- ❖ unknown cause (idiopathic)

Hydrocephalus does not always appear immediately after these causes occur, and it may be years or decades before the symptoms become evident.





Normal Pressure Hydrocephalus



NPH is an accumulation of CSF that causes the ventricles in the brain to become enlarged, sometimes with little or no increase in intracranial pressure. It is most common among adults 55 years of age and older and, because of its symptoms, is often associated with the aging process. It is frequently misdiagnosed as dementia or Parkinson's disease (because of gait) or Alzheimer's disease (because of cognitive impairment).

What causes NPH?

The causes of most NPH cases are unknown. It may also develop as a result of the causes already listed.

What are the symptoms?

There are three classic symptoms:

Gait disturbances (difficulty walking) are often the most pronounced symptom and the first to appear. They range from mild imbalance to instability to inability to stand or walk. Gait is often wide-based, short-stepped, slow and shuffling. The individual has trouble picking up their feet and often trips over curbs and falls. It is often described as "feet stuck to the floor." Turning around is accomplished with many small steps.

Mild dementia is described as a loss of interest in daily activities, forgetfulness, difficulty performing routine tasks and short-term memory loss. Cognitive symptoms are often overlooked for years or accepted as a consequence of aging. Not all individuals have obvious cognitive impairment. They may retain conversational skills and their thinking abilities may be unchanged. Cognitive changes may be detectable only with formal neuropsychological testing.



Impaired bladder control can range from urinary frequency and urgency in mild cases to complete loss of bladder control in severe cases, when urinary urgency is strong and cannot be controlled. Bowel incontinence may occur. Not all individuals with NPH display signs of bladder problems.

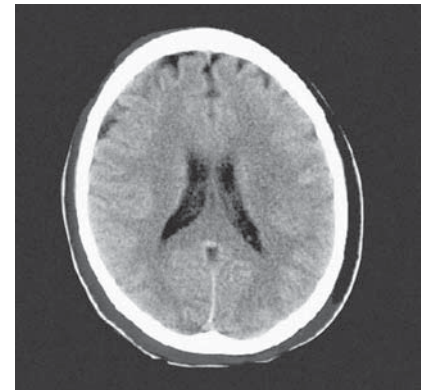
The symptoms of NPH seem to be progressive over time but the rate of progress varies. An individual often seeks evaluation and treatment only after a critical loss of function, or after a symptom becomes apparent. An early diagnosis of NPH seems to increase the chances of successful treatment.

How is NPH diagnosed?

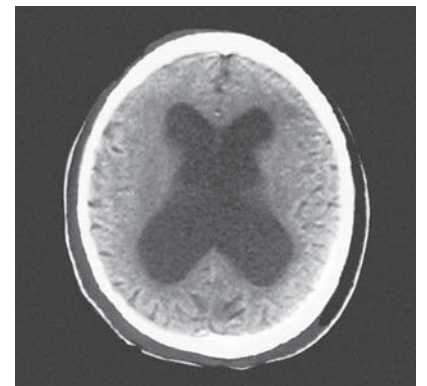
Once a physician suspects NPH, testing is usually carried out to confirm the diagnosis and assess the individual's suitability for shunting. It is important that a neurosurgeon or neurologist became part of the medical team at this point, to interpret test results and discuss surgery and its risks. The medical team will consider the pattern and severity of impairments, along with the results of other tests, in differentiating NPH from other conditions such as Alzheimer's or Parkinson's disease.

Physical and neurological examinations will be used to evaluate the symptoms. There will be discussion and observation of walking and turning to determine the extent and type of gait disturbance. Asking questions or administering neuropsychological evaluation will test cognition. Areas of examination include attention span, reaction time, memory, reasoning, language and emotional state. There will be verbal assessment of urinary urgency and frequency or incontinence.

An MRI or CT scan will examine the brain to detect enlarged ventricles. Other testing could include CSF tests such as lumbar puncture (spinal tap), external lumbar drainage, measurement of CSF outflow resistance and intracranial pressure.



Cross section of a normal brain



Cross section of a brain showing signs of NPH



Treatment

The seriousness of NPH symptoms may vary from day to day, but the progression is not likely to stop on its own. The most common, and usually the only available, treatment for NPH is surgical implantation of a shunt. (See Shunts, page 11.)

Appropriateness of shunt surgery

Many tests and evaluation criteria are considered but unfortunately, no single factor is reliable in predicting success from shunt implantation. However, the chance of a more complete recovery depends on the following factors:

- ❖ onset of gait disturbances is the first and most prominent symptom;
- ❖ monitoring of intracranial pressure or spinal fluid pressure shows an abnormal range or pattern of spinal fluid pressure or an elevated CSF outflow resistance;
- ❖ removal of spinal fluid gives dramatic temporary relief of symptoms;
- ❖ there is minimal evidence of a disease of the small blood vessels that nourish the brain.

Many people with NPH have additional medical or neurological problems. It is important to discuss with the medical team what outcomes can be expected in regaining motor skills or mental ability. It is also important to understand that shunting may improve all or none of the hydrocephalus symptoms. While everyone hopes for a full recovery, it is not often seen. Many individuals are satisfied with reduced levels of disability or dependence, or the prevention of further neurological deterioration.

Some individuals with enlarged ventricles show no symptoms and no neurological impairment, even when medically evaluated. This is often called compensated hydrocephalus. In such circumstances, there are no benefits that would offset the potential risks of treatment.

Success of shunt surgery

There is currently no way to predict how quickly or to what extent shunt implantation will relieve symptoms. Gait disturbance, mild dementia and bladder control problems may improve within days of surgery, or it may take months. There is also no way to predict how long these improvements may last. Some individuals reach a plateau, while others may improve for months and then decline again.

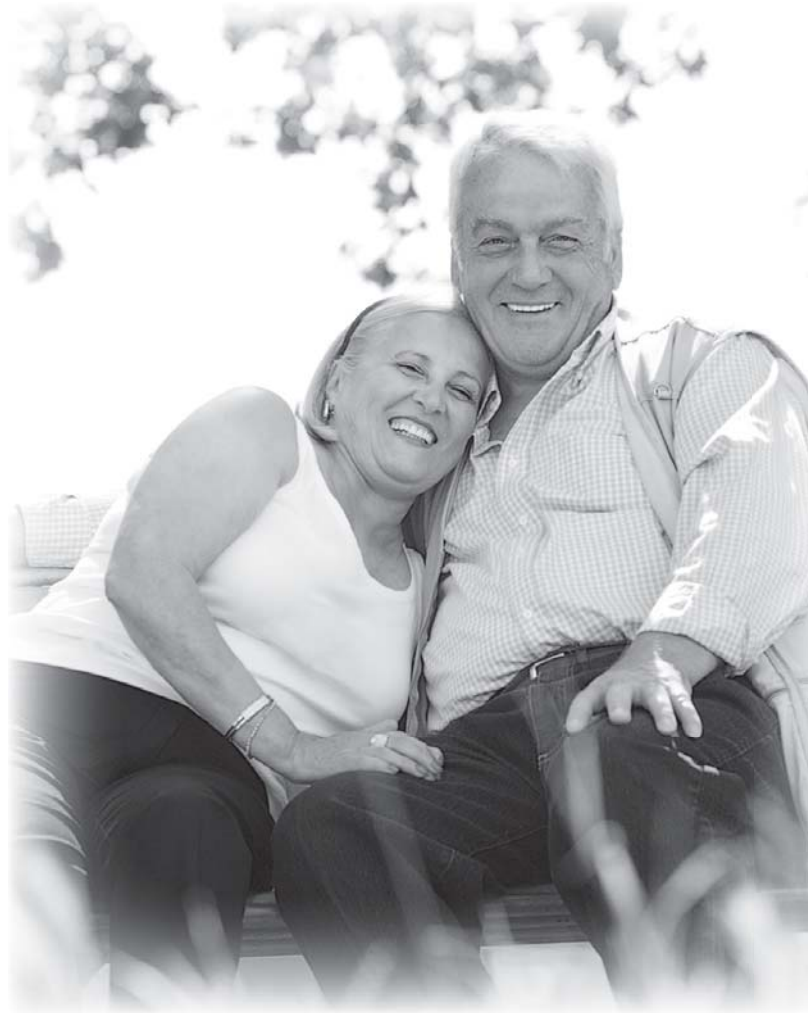
Shunt success may also be affected by the presence of other neurological or medical conditions. For example, high blood pressure on the brain can cause multiple tiny strokes in the same area of the brain as the hydrocephalus, causing virtually the same symptoms.

If there is a recurrence of symptoms, the individual should consult a doctor to ensure that there is no malfunction of the shunt, or other complications such as valve failure or the need for a higher or lower pressure valve.

Complications and risks of surgery

Complications of surgery include:

- ❖ obstruction and malfunction of the shunt;
- ❖ infection;
- ❖ a blood clot.





A Last Word

Hydrocephalus is a complex condition requiring care and vigilance. SB&H is there to help along the way.

We sincerely hope that you have found the material in the guide useful. We want to ensure that people affected by hydrocephalus have the support and information needed to make informed decisions for themselves or someone they love.



If you have questions about what you read in the guide, or anything related to hydrocephalus, contact the association. We will do our best to provide answers. You can also visit the SB&H website for additional information, useful links or to connect with others.

Clearly more progress needs to be made in the treatment of hydrocephalus. We are hopeful that medical technology and ongoing research will prove beneficial in the future.

Our goal is to improve the quality of life of all individuals with hydrocephalus.



Appendix A

Specialists who assist people with hydrocephalus



Notes

Treatment of hydrocephalus and the complications that may accompany it often requires the expertise of a number of professionals. Following is a brief list of those who provide this care.

- Anaesthesiologist:** a physician who is especially trained in administering anesthetics during a surgical procedure. The anaesthesiologist also monitors breathing and heart function while the individual is anaesthetized.
- Dietitian:** an individual trained in nutrition who can provide recommendations about diet that can enhance one's overall health, growth and development.
- Endocrinologist:** a physician who specializes in the diagnosis and treatment of disorders of the endocrine system (the system that produces the body's hormones).
- Gastroenterologist:** a medical specialist trained to diagnose and treat diseases and conditions pertaining to the digestive system.
- General Practitioner:** a physician usually responsible for the overall health care of an individual, including regular check-ups, vaccinations, and referrals to specialists.
- Gerontologist:** A specialist who works in the branch of science that deals with aging and the problems of aged persons.
- Geneticist:** a scientist who studies genetics, the science of heredity and variation of organisms.



Notes

Neurologist:	a physician who specializes in treating disorders of the nervous system (brain and spinal column). The neurologist may be involved in treating a seizure disorder and learning disabilities.
Neuropsychologist:	a doctor of psychology who is specially trained in treating disorders related to the brain and behaviour. The neuropsychologist can assess and provide remedial and academic recommendations for children and adults with a variety of disorders of the central nervous system. A neuropsychologist is not a medical doctor.
Neurosurgeon:	a physician specializing in the diagnosis and surgical treatment of central nervous system disorders such as hydrocephalus and spina bifida.
Nurse Practitioner:	a registered nurse with advanced training and often a master's degree. The NP is able to diagnose and treat certain illnesses, provide and order specific tests, and provide care and optimize an individual's overall health and well-being.
Occupational Therapist:	an individual trained to optimize the development and independence of persons with physical and mental limitations. Therapy for individuals with hydrocephalus may help to enhance fine and gross motor development, balance and hand-eye coordination.
Ophthalmologist:	a physician especially trained to diagnose and treat visual disorders and ocular diseases. The ophthalmologist prescribes glasses, treats eye muscle disorders and performs surgery.
Optometrist:	a doctor of optometry who specializes in the assessment of vision, checks general health of the eyes and prescribes glasses. An optometrist is not a medical doctor.

Orthopedic Surgeon:	a physician who specializes in the diagnosis and surgical treatment of disorders of the bones, muscles and joints.
Pediatrician:	a physician who specializes in the medical care of children. The pediatrician is usually responsible for the child's overall care, including regular check-ups, vaccinations and referrals to specialists.
Perinatologist:	An obstetrical sub-specialist who oversees the care of mother and fetus with higher-than-normal risks of complication.
Physiatrist:	a physician with special training in rehabilitation medicine. The physiatrist prescribes appropriate rehabilitation programs and works closely with occupational, physio- and speech therapists.
Physiotherapist:	an individual specially trained to evaluate and treat disorders of movement. The physiotherapist promotes mobility and works to maintain flexibility and a full range of motion in the joints and muscles.
Psychiatrist:	a physician who is able to diagnose and treat mental and emotional disorders. Psychiatrists are able to prescribe medications for treatments.
Psychologist:	a doctor of psychology who specializes in treating emotional, behavioural and mental development in children, adults and families. A psychologist may assess emotional and intellectual, as well as learning disabilities, and provide psychotherapy. Psychologists are not medical doctors.



Notes

Public Health Nurse: a registered nurse who provides care, information and support in the home environment. The public health nurse can help the patient and family make lifestyle adjustments to ease the transition between hospital and home. The public health nurse may also explain medical conditions and provide information about and referrals to relevant community services and programs.

Registered Nurse: an individual specially trained to provide care and to maintain, promote and restore health in the hospital, community, home and occupational setting.

Social Worker: an individual trained at the graduate level to provide assistance with personal, emotional and financial needs of the patient and family. The social worker can also provide appropriate community and agency referrals.

Speech Pathologist/Therapist: a professional trained to diagnose and treat abnormalities related to speech, communication, eating or swallowing. The speech pathologist/therapist may also provide speech therapy and may recommend specific teaching strategies that will assist your child's teacher and school.

Urologist: a physician who specializes in diagnosing and providing medical and surgical treatment of disorders of the kidney and bladder.

Appendix B

Commonly used medical terms



Notes

Angiogram:	a diagnostic test that provides an x-ray image of the blood vessels.
Aqueduct of Sylvius:	the narrow channel that connects the third and fourth ventricles.
Aqueductal Stenosis:	a condition wherein the aqueduct is narrowed, thereby blocking the normal flow.
Arachnoid mater:	the web-like tissue layer in the middle of the three meninges.
Arachnoid Cyst:	a sac filled with CSF.
Arachnoid Villi:	tiny, hair-like structures found within the arachnoid membrane that absorb CSF.
Arnold-Chiari Malformation:	a condition in which the poles of the cerebral hemispheres and the medulla protrude into the spinal column.
Brain Stem:	the lower part of the brain that connects to the cerebral hemispheres and extends to the spinal cord. The brain stem consists of three main sections: midbrain, pons, medulla oblongata.
Catheter:	flexible hollow tubing often made from silicon.
Cerebrospinal Fluid (CSF):	the fluid that is present within the ventricles, the subarachnoid space surrounding the brain and spinal column. One of its main functions is to act as a shock-absorber for the brain and spinal column.



Notes

Cerebrum/Cerebral Hemispheres:	the cerebrum is the name given for the main part of the brain occupying the upper part of the cranial cavity. The cerebrum consists of two cerebral hemispheres (halves).
Choroid Plexus:	tufts of tissue found in the ventricles that produce CSF.
Coma:	a state of unconsciousness wherein the patient cannot be aroused, even by stimulation.
Congenital:	a term used to denote a condition present at birth.
Cranium:	a part of the skull that encloses the brain. The cranium consists of the frontal, occipital, sphenoid, ethmoid, temporal and parietal bones.
Cranial Sutures:	the area where the bones in the skull fuse in early life to form a seam.
Dandy-Walker Syndrome:	a condition involving abnormal formation of the brain, which includes a full or partial blockage of the exits that carry away the CSF (called the foramina of Megendie and Luschka) and the enlargement of the fourth ventricles.
Distal:	any location situated away from the point of attachment, origin or centre (e.g. the hand is the distal part of the arm). In the case of a shunt, the distal end of the catheter is located in the abdomen or the aorta, depending on the type of shunt.
Dura Mater:	the tough, fibrous outer covering of the brain and spinal cord. It is external to the arachnoid and pia mater, and contains numerous blood vessels and venous sinuses.



Encephalitis:	the inflammation or infection of the brain. It may cause drowsiness, severe frontal headache, nausea, lethargy, seizures, fever, personality change and weakness. Encephalitis can lead to brain damage, coma and death.
Fit:	see seizure.
Fontanel:	two fibrous membranes or “soft spots” between an infant’s skull that have not yet come together or fused. These two soft spots are the anterior and posterior fontanel.
Foramen:	a hole or passage through a bone or other structure.
Foramen of Monro:	the passage and point of communication between the lateral ventricles and the third ventricle of the brain.
Hemorrhage:	bleeding.
Hemiparesis:	muscle weakness or partial paralysis on one side of the body.
Hemiplegia:	complete paralysis of one side of the body resulting from injury to the motor centres of the brain.
Hernia:	a protrusion of an organ from one compartment of the body to another.
Inguinal Hernia:	a protrusion of part of the intestine through the muscle wall in the groin area, often into the scrotum.
Intra-cerebral Haematoma:	an abnormal collection of blood in the brain as a result of a tear in a blood vessel. It may be caused by head injury, stroke or other mechanical injury.



Notes

Intracranial	within the cranium or skull.
Intra-ventricular Hemorrhage:	bleeding within a ventricle.
Ischemia:	a temporary condition in which an organ or tissue is deprived of blood supply.
Malfunction:	a failure to function in the normal manner.
Membrane:	a thin layer of tissue that covers an organ or structure.
Meningeal:	pertaining to the meninges.
Meninges:	a three-layered membrane covering the brain and spinal cord, consisting of, from the outside in, the dura mater, the arachnoid and the pia mater.
Meningitis:	an inflammation or infection of the meninges, particularly the pia mater and arachnoid. Meningitis is often caused by bacteria or virus, however, it can also be caused by other means.
Nervous system:	divided into the central nervous system (CNS), comprising the brain and spinal cord, and the peripheral nervous system (PNS), comprising the nerves connecting the spinal cord to the muscles.
Papilledema:	the swelling of the optic nerve at the point where it enters the eye (optic disc), caused by increased intracranial pressure. The presence of papilledema is used as a diagnostic sign.
Peritoneal Cavity:	a slit-like space containing fluid secreted by the serous membranes.

Pia Mater:	the innermost membrane on the brain and spinal cord. The pia mater is one of three meningeal membranes. The other two are the dura mater and the arachnoid.
Porencephalic Cyst:	abnormal CSF cavities in the brain. Some porencephalic cysts communicate with the ventricles.
Proximal:	situated near the point of attachment, origin or centre point.
Revision:	surgery to repair or replace a malfunctioning shunt.
Seizure:	one or a series of involuntary muscle contractions.
Shunt:	a device used to transport fluid from one area to another. In the case of hydrocephalus, it passes fluid from the ventricles to the abdominal cavity or heart.
Spina Bifida:	a congenital defect of the spinal cord.
Strabismus:	damage or weakness of the eye muscles. In individuals with hydrocephalus, this may be the result of damage to the optic nerve that controls the eye muscles.
Sunset Sign:	the term used to describe a state in which the eyes appear to be continually downcast and an abnormal amount of the white of the eye is visible above the iris.
Ventriculitis:	an infection of the ventricular system.





Appendix C

Questions to ask before surgery

Notes

These are questions you should have answered after speaking with your health care team. You may want to write any additional ones down, record the answers in writing or on tape, or bring a friend or relative with you in case you forget to ask something:

- ❖ Why do I have hydrocephalus, and what are the various treatment options available?
- ❖ What are the risks and benefits of these treatment options, and why is one particular one being recommended?
- ❖ What are the common complications of this particular procedure?
- ❖ What are the more rare and serious complications?
- ❖ What steps are being taken to reduce the complications as low as possible?
- ❖ How long is the operation, and how long will I be in hospital?
- ❖ How long will I need to recover at home, before I go back to work or school?
- ❖ What will the follow up process be?
- ❖ Who can I contact if there are any problems or in an emergency once I leave the hospital?



Notes

- ❖ How much experience do you have with a case like mine, and do you think there is anyone else I should see?
- ❖ Is this operation absolutely necessary?
- ❖ What type of anaesthetic will be used?
- ❖ Can anaesthetic cause problems with other drugs?
- ❖ Is the surgical staff aware of latex allergy and how to deal with it?
- ❖ I'm afraid of more surgery. Is there anything you can tell me about your personal experience with this procedure that will make me less scared?
- ❖ If I require surgery, will you be the surgeon. If not, can I meet with the other surgeon?
- ❖ Will you be using sutures, staples or stitches?
- ❖ How much hair will be shaved or clipped?
- ❖ Should it be cut prior to hospitalization?
- ❖ How soon can I have visitors?
- ❖ Will I need physiotherapy?
- ❖ When can I resume normal activities?
- ❖ Whom can I call if I have questions or suspect a problem?



Notes

Appendix D

General questions to ask healthcare providers

- ❖ Do you see many patients with hydrocephalus in your practice?
- ❖ Could I connect with them?
- ❖ Can you provide me with the name of a support organization?
- ❖ I have this type of shunt (VA, VP, programmable valve). Are you familiar with it?
- ❖ In case of emergency who do I contact? You, your nurse?
- ❖ If I have to go to a hospital emergency room, which one should I go to?
- ❖ Should I take anything with me, like previous scans?
- ❖ I've had complications in the past. How would you handle these situations (such as infection, shunt failure)?
- ❖ Thank you for taking the time to answers these questions. If I think of more, can I contact you or should I talk to someone else in your practice?



Notes





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