

Strategy for the treatment of Hydrocephalus

Adaptation for Developing Countries - (Dr. BC Warf)

1. What is hydrocephalus?

In the center of the brain are fluid-filled spaces called ventricles. The fluid (cerebrospinal fluid) is continuously produced at the rate of about 20cc per hour, and under normal circumstances exits the ventricles and is absorbed outside the brain at the same rate. Hydrocephalus results when the normal exit or absorption of this fluid is This leads to progressive accumulation of cerebrospinal fluid within the impaired. ventricles (the normal spaces inside) of the brain, resulting in progressive damage to the developing brain with associated mental retardation and visual impairment. In the infant it also leads to progressive abnormal enlargement of the head (macrocephaly). Onset in the older child leads to progressive pressure within the head, which can end in coma and death. It may exist as an isolated congenital condition or it may be a common accompaniment to other congenital anomalies such as spina bifida and encephalocele. In the case of spina bifida aperta (myelomeningocele), hydrocephalus is present in 85-90% of the cases. Hydrocephalus may also arise secondary to intraventricular hemorrhage in the premature neonate, infection (e.g. bacterial meningitis), head trauma, or a brain tumor. With appropriate and timely treatment, hydrocephalus is completely relieved, and children in which hydrocephalus is an isolated problem can be normal. When left untreated, the results are devastating.

2. Ventriculoperitoneal Shunting

Currently, ventriculoperitoneal shunting is the most common way of treating hydrocephalus.

Newer methods (i.e. third ventriculostomy) could help some patients, but skills and technology are not yet widespread, this method is most appropriate only for certain types of hydrocephalus (noncommunicating hydrocephalus), and the technique can be technically more difficult in myelomeningocele patients. VP shunting will doubtless be the mainstay of treatment for some time.

In ventriculoperitoneal (VP) shunting, a tube is inserted through a small hole in the skull into the spaces (ventricles) of the brain which contain the cerebrospinal fluid (CSF). This tube is connected to another tube which runs underneath the skin down to the



abdomen, where it enters the abdominal cavity (the peritoneal cavity). The shunt allows CSF to drain out of the ventricles and into the abdominal cavity where it is absorbed. Typically, a valve in the system helps regulate the flow of fluid.

3. What is necessary for successful shunting?

- Motivated parents
- A curve and tape measure
- Basic laboratory facilities
- A surgeon properly trained in shunt placement and management of shunt problems and complications
- Pre- and postoperative care
- Anaesthesia
- An operating theatre
- A shunt (can be provided by if)
- Antibiotics

4. How is hydrocephalus diagnosed?

In Developing Countries (in other countries too, of course) we should avoid unnecessary investigations. This increases the cost dramatically and also the pressure on the family and the child. Very expensive investigations like CT and MRI scans, although ideal, are not essential to the diagnosis and management of hydrocephalus in infants, in whom the clinical signs are usually immediately apparent. (Imaging studies are, however, essential to the management of hydrocephalus in older children and adults).

The key clinical sign of hydrocephalus in infants is accelerated head growth. This is determined by serial measurements of the infants head which are recorded on a growth curve. Accelerated head growth is demonstrated when the head circumference measurements are not following one of the normal growth curves. It is important to note, however, that accelerated head growth is not always caused by hydrocephalus. "Benign macrocephaly" is a common condition in which the baby's head is large and grows at an accelerated rate in the first year or so of life. This sometimes runs in families (mother or father were told they had a big head when they were children). Therefore, it is very important that, in the absence of an imaging study, the diagnosis of



hydrocephulas is made in combination with other clinical signs as well. The baby's soft spot (anterior fontanelle) is usually full or bulging, or even tense, due to the increased pressure inside the head. The spaces between the bones of the baby's skull (which don't normally fuse together until later) may be widened because the bones are being pushed apart. Sometimes (but certainly not always) the baby's eyes will appear to be looking downward all the time, or may look crossed.

As long as the soft spot (fontanelle) is open, an ultrasound study of the brain can be performed. This is an excellent way to image the spaces (ventricles) of the brain, and is very helpful in confirming the diagnosis of hydrocephalus. Ultrasound is less expensive and more readily available than CT or MRI.

5. Treatment

The shunts used most commonly in North America and Europe are very expensive. However, studies demonstrating a significant advantage in these shunt systems over simpler, less expensive shunt systems have not been performed. A large experience suggests less expensive shunt systems are adequate. Factors other than the type of shunt used are probably much more important to the treatment of hydrocephalus. These include early diagnosis and treatment; the experience, skill, and judgement of the surgeon; the environment for surgery and postoperative care; skilled clinical follow up of the patient; and, observant, informed, and concerned parents.

IF can provide good quality shunts if acceptable standards of services are available. In several developing countries, surgeons have developed a 'home-made shunt' from feeding tubes with rather good results.

6. Selection for shunting

It is important to select the children with hydrocephalus who can benefit from shunting. **A shunt gives children a solution for their problem, but it is not a total cure**. It helps and it avoids additional handicaps, but it can cause complications. When the selection or the shunting is not done in a proper way, it can cause more complications than it prevents. So it is important to make the right selection and to make sure that the conditions are of acceptable standards before investing in shunting.



7. When is shunting NOT wanted?

(This does not mean that the child and the family do not need help)

- In children with **very big heads**, there is an increased risk that shunting can cause intra-cranial bleeding. Also, the end result of shunting (in regard to brain development) is less hopeful.
- Children with **stabilised (arrested) hydrocephalus** (no signs of high pressure and the head is not enlarging) do not require treatment.
- Children with **infections**, especially infection involving the CSF, should be treated before shunting.
- **Distance**: Children who live too far from surgical facilities. Shunts can block and then the service should be reachable in at least 24-48 hours. Otherwise a good, operated child could die when shunt complications occur. Therefore, reasonable access to care should be a consideration. A shunted child requires maintenance.
- When the **general condition** (malnutrition, anaemia, malaria, fever,...)of the child is not good enough for surgery, preoperative treatment is necessary.

8. Pre and Post Operative Care

- Listen to the story of the mother and take note of her information. She knows her child and will often see changes in the child even before objective symptoms occur.
- -She has the information about the past and about the actual situation. When did she see the problem for the first time? How do her relatives react? What is her financial situation? Where does she live? ...etc
- Record the child's head-size on the attached curve. The curve is a way of measuring the evolution of the head-circumference. In small children (till 20 months) the head will enlarge when they develop Hydrocephalus. Only when the circumference of the head is increasing, shunting is required.
- Preoperative tapping of the water in the brain might be necessary to exclude infection.
- This should only be done by a skilled person.
- If water is not tapped before the operation the surgeon should always take some water during the operation. This will be a help if infections appear after surgery.
- As in each surgical intervention the general condition of the child has to be evaluated. If shunting needs to be postponed ventricular tapping (or a spinal tap,



when communicating hydrocephalus is present) can relieve high pressure for a few days. (Repeated ventricular taps can be harmful.)

- Pre- and postoperative antibiotics can be useful and necessary depending on the conditions.
- Make sure that the surgeon is reachable the first days after the operation.
- Teach the mother how to handle the child and what to do with shunt complications

9. Counselling parents

- Teach the parents to keep records of the size of the head of their child.
- Inform them about possible complications that can appear
- Tell them what to do when high pressure occurs again
- Give them the IF information kit (tape measure, curve, and leaflet)
- They should be informed but not scared. It is better not to dramatise the operation. It is a simple and easy operation. It is important that parents understand how the shunts work. It is helpful to show them a shunt.

10. What to do when shunting is not wanted

When hydrocephalus develops or is already developed and shunting is not wanted, you know that the child and the family have to face a longstanding difficult situation. Help will be needed in assisting the relatives (mother..) and the person with hydrocephalus for comfort and quality of life.

What can be done?

- assisting the mother
- helping with aids (sitting and walking aids)
- supporting the child in its daily activities (occupational therapy)
- arrange schooling or labour
- physiotherapy
- etc

11. Chhabra shunt



In most countries or projects, availability of shunts is a first problem to come across.

IF has 10 years of experience in using the Indian manufactured Chhabra shunt and promotes the use of this type of shunts. A recent scientific study of Dr B. Warf, an IF medical advisor, showed that the Chhabra shunts has the same quality outcome as the more expensive American Codman shunt.

IF can help in providing the Chhabra shunt at a rate of 45 € per shunt (price Codman shunt 650 \$)