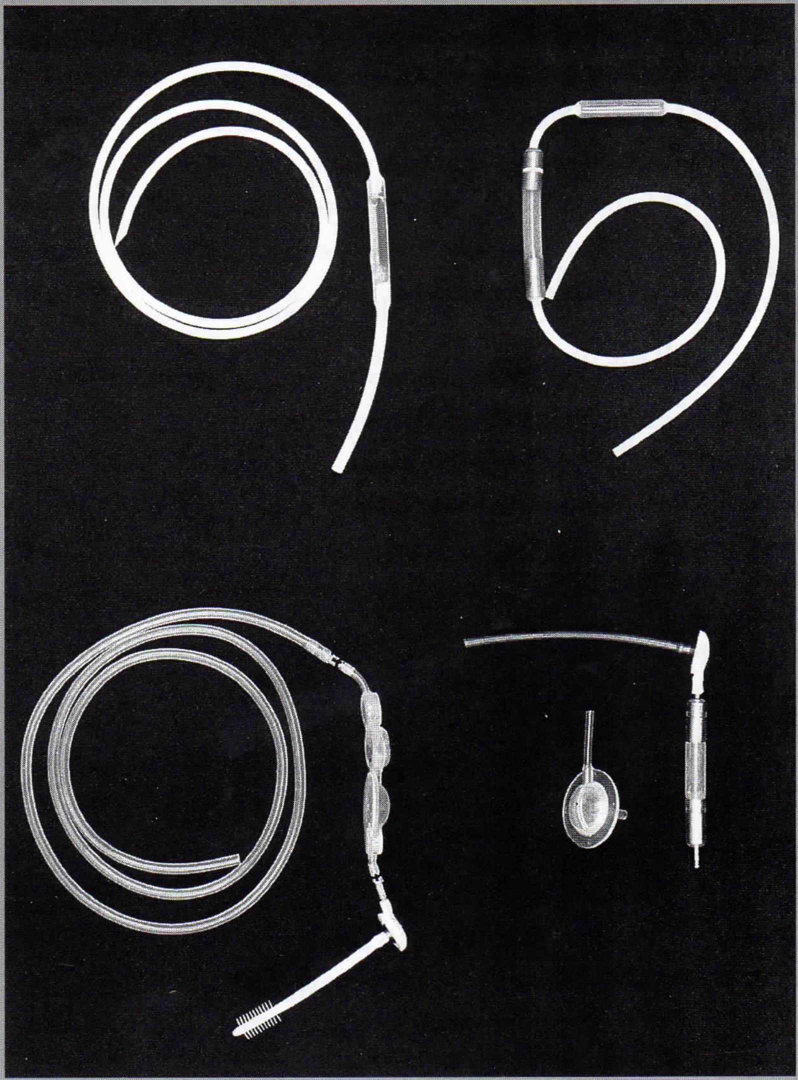

HYDROCEPHALUS

A series of three articles on hydrocephalus and its treatment which first appeared in *Hydrocephalus Network News* in 1995

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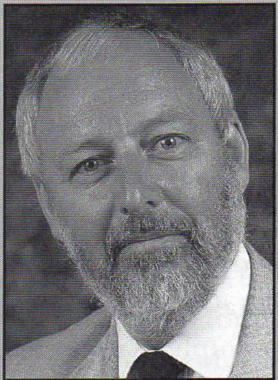


ASSOCIATION FOR SPINA BIFIDA AND HYDROCEPHALUS

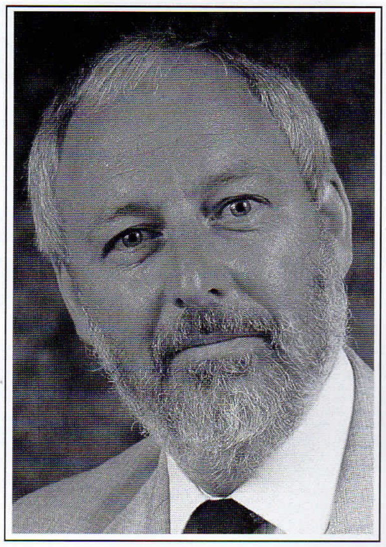
Main picture shows four types of 'shunts' used to treat hydrocephalus

PART I
Hydrocephalus: what it is and what causes it

THE TERM 'hydrocephalus' is from two Greek words meaning 'water in the head'. In fact the 'water' is cerebrospinal fluid (CSF), a clear liquid which is produced all the time in the cavities or ventricles inside the brain. It passes from one ventricle to the next (four in all) through narrow pathways, then circulates around the surface of the brain – a little also goes down the spinal cord – and is absorbed back into the bloodstream. The absorption takes place through specialised veins inside the skull which have a sieve-like surface. Though much slower than the circulation of the blood, the CSF is constantly being produced, circulated and reabsorbed.

Hydrocephalus can result when either too much CSF is produced (very rare), or when it is prevented from circulating or being reabsorbed. As in these circumstances CSF is constantly produced but cannot get out, it accumulates and causes raised pressure inside the brain. The ventricles swell and the brain tissue is stretched and squashed. The skull bones in babies and young children are not fixed together as they are in later life, and the pressure causes the head to increase in size. However it is important to realise that hydrocephalus can also arise in older children and in adults. A later article will deal with the effects of hydrocephalus on those who have it.

A number of conditions can give rise to hydrocephalus. Uncommonly it can be due to a genetic disorder, in which case other members of the family are usually affected. Non-genetic causes are more common. In babies who are born with hydrocephalus the condition is said to be congenital, and it must be realised that this means simply that it is present at birth, and not that it is hereditary. In congenital hydrocephalus the actual cause is usually impossible to determine but it is assumed to be due to events during the baby's development before birth, such as damage to local blood supply or infection. A known cause of congenital hydrocephalus, uncommon in this country, is toxoplasmosis. About 20 years ago the commonest form of hydrocephalus was that associated with spina bifida. At least 80% of people with spina bifida have some degree of hydrocephalus though it is not always in need of treatment. Here the interference with CSF flow is due to abnormalities of the brain structure at the back of the head which develop at an early stage of the pregnancy. This is often called the Arnold-Chiari malformation.



Dr Roger Bayston

In recent years far fewer babies have been born with spina bifida, and the largest number of cases of hydrocephalus in infancy occur in babies born prematurely, due sometimes to maternal ill-health or smoking. Even very small babies can now be made to survive, but their vital functions, normally taken care of during pregnancy by the mother, have to be controlled artificially. Unavoidable rapid changes in blood pressure can cause bleeding in the brain, and the blood from this haemorrhage blocks the sieve-like absorption system leading to post-haemorrhagic hydrocephalus. Brain haemorrhages in adults (stroke) is similar and can also lead to hydrocephalus in survivors. Head injury sometimes has the same effect.

Another way in which the CSF pathways can be blocked is by debris and pus in acute infections of the membranes surrounding the brain (meningitis). This can arise in previously healthy babies if they are unlucky enough to develop a serious bloodstream infection soon after birth, but it can happen to anyone in any age group. Fortunately, meningitis is not common and one major cause, haemophilus, has been greatly reduced by the introduction of the HIB vaccine. Other types of hydrocephalus can be due to obstruction by cysts in the ventricles. Dandy-Walker cysts, which block the fourth ventricle at the base of the brain are an example of these swellings. Brain tumours can obstruct the CSF flow, either by their bulk or by causing brain swelling. In adults, blockage of one of the connecting pathways between the ventricles (aqueduct stenosis) can cause the onset of hydrocephalus. Two other conditions, benign intracranial hypertension (not very benign for those who have it) in which the CSF pressure is raised, and normal pressure hydrocephalus in which, as the name suggests, the CSF pressure is not raised and which is probably not hydrocephalus at all, are also seen in adults.

Finally, hydrocephalus is often classified as either communicating or non-communicating. In the first type, the problem is usually failure to absorb the CSF at the end of the system, whereas in the second there is blockage of the CSF pathways within the ventricular system.

PART 2
THE EFFECTS OF HYDROCEPHALUS

IN THE first article in this series, I explained that hydrocephalus involves accumulation of cerebrospinal fluid (CSF) in the ventricles of the brain, with an increase in the pressure inside the head.

There are two sources of this pressure. One is that of the CSF itself, but a much higher pressure is produced by the heart in order to pump blood to the brain. If the CSF pressure rises, it eventually interferes with the blood supply to the brain, depriving it of oxygen and glucose which it needs in constant amounts to continue to function. Initially this causes tiredness, irritability and drowsiness, but if it progresses then loss of consciousness will result as the brain begins to shut down.

The immediate effects of this interference with the blood supply disappear if the CSF pressure is returned to normal, such as by ventricular tap or insertion of a shunt. However, in most cases the process has been continuing for some time before diagnosis of hydrocephalus is made. During this time the interference with the blood supply leads first to a 'dying back' of the very fine blood vessels in the brain. Even this process is largely reversible if prompt action is taken, but at this time there is often insufficient clinical evidence to suspect hydrocephalus. The next stages involve progressive damage to the actual nerve cells in the brain and to their eventual destruction, and this cannot be reversed.

Because of the areas of the brain most affected, functions associated with thought and learning as well as with co-ordinated skilled movement begin to deteriorate. The precise effects differ between individuals and are further complicated by other abnormalities as well as by the pre-existing degrees of ability and personality of each person affected. It is not surprising therefore that while, for instance, learning disorders are common amongst those with hydrocephalus, their exact effects vary considerably.

Much is said and written about intelligence, and particularly about IQ (intelligence quotient) in people with hydrocephalus. In fact this is far more complicated, and a good deal less informative, than many believe. The IQ is made up of several components which can be thought of as verbal and non-verbal, or performance-related tests. People with hydrocephalus generally score better on verbal IQ than on performance IQ and this is thought to reflect the distribution of nerve damage in the brain as described above. Certainly during periods of rising CSF pressure, such as in untreated cases or when a shunt is blocked, the effect on performance IQ is more marked. Generally speaking, people who have had hydrocephalus since birth or childhood have, as a group, a lower average IQ than a comparable group without hydrocephalus, but it is important to realise that there is a wide range in each group, and some people with hydrocephalus have very high scores.

The practical implications of these features of hydrocephalus are that there may be subtle problems of co-ordination of hand movements with what the person sees, as well as a degree of clumsiness, which make it difficult to perform certain tasks or do certain jobs. With regard to learning in the home or to education in school, there may be real problems with concentration and reasoning which require a sympathetic but skilled approach. For instance, it will often be necessary to teach simple every day tasks like getting out of bed, washing one's face, dressing and going downstairs as separate short items rather than all at once, and to keep them consistent and repetitive. This does not indicate 'stupidity' but is caused by damage to the nerves in the brain which normally allow us to learn very quickly how to do a complex series of things. Much can be done to help, and professional advice should be sought where needed.

Psychological development in children and adolescents with hydrocephalus may proceed normally, but sometimes the changes associated with puberty (breast development, body hair growth etc) appear much earlier than expected, and the intrusion of psychological aspects of sexual development into a mind which is emotionally still very immature can cause distressing problems. Again, specialist advice should be sought if necessary.

Other effects of hydrocephalus may also be seen, and some of these are difficult to explain. For instance, some people are very seriously distressed by every day noises such as vacuum cleaners or washing machines.

One effect of raised CSF pressure may be seen in the eyes, and this is why your doctor sometimes looks for 'papilloedema'. This is caused by pressure on the blood supply to the back of the eye. It is important to realise that it may not always be present, even when the pressure is high. If CSF pressure remains high for too long, damage to the optic nerves can become permanent resulting in blindness, though fortunately nowadays this is uncommon. Another appearance, particularly in babies, is the so-called 'sunset' eye sign, where the eyes are fixed in a downward position. This is due to CSF pressure affecting important nerves running from the brain which control eye movement.

If untreated the rise in CSF pressure can cause other serious problems in the brain, unrelated to blood supply. Many of our vital functions such as heart beat, breathing etc are controlled from the brain stem, a structure joining the spinal cord to the brain. Very high CSF pressure can compress this sufficiently to cause the heart and breathing to stop. Once again this is uncommon as signs of raised pressure are usually recognised before this. A similar problem might sometimes arise, particularly in those with spina bifida, due to compression of the cerebellum, a part of the brain lying at the back of the head. This can also give rise to breathing, speaking and swallowing difficulties.

Reading a catalogue of effects of hydrocephalus such as I have produced here can be very alarming. However it should be realised that some people with hydrocephalus may have very few of these problems, and hopefully none of the more serious ones. Also, many of those which I have described are found either in untreated hydrocephalus or when the treatment fails, and when successful treatment has been promptly introduced they often improve or sometimes disappear. On the other hand, the more subtle learning and reasoning problems are usually present in some degree and are very important where a child's development and education are concerned.

PART 3
THE TREATMENT OF HYDROCEPHALUS

IN THE first article of this series I explained that hydrocephalus, irrespective of its cause, was due to accumulation of cerebrospinal fluid (CSF) within the ventricles of the brain, resulting in raised pressure inside the head. In principle the solution is simple, that is to insert a tube into the swollen ventricles and drain off the excess fluid, thereby returning the pressure inside the head to normal again. Historically, many attempts to do this have been recorded but the results were usually disappointing, due to rejection of the tubing by the body's defences, to infection, or to blockage of the tubing. However, this began to change in the 1950's.

Charles 'Casey' Holter, the son of an American engineer, was found to have hydrocephalus. The surgeon explained that there was no satisfactory treatment. John Holter, the child's father, questioned the surgeon and gained an idea of the sort of treatment required, if only it existed. He experimented at home and soon developed such a device, made from silicone rubber which had never been implanted into people before. The surgeon decided to try this, with great success. The Holter valve soon became the treatment of choice for hydrocephalus, and it also revolutionised the treatment of spina bifida, removing the principal cause of death and encouraging orthopaedic and urological surgeons to develop procedures to improve mobility and continence. Other similar devices were developed soon afterwards, all made from the same material and having similar valve mechanisms.

In the 1960's investigations were proceeding into how CSF was produced, and drugs were tried experimentally to reduce the rate of CSF production. Some of these showed initially promising results, but they had unpleasant side effects and their use was abandoned. However, new investigations are now being undertaken on

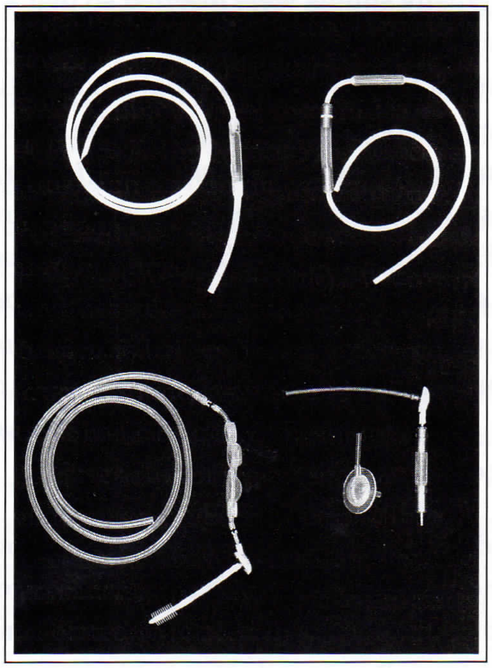


Figure 1: Different types of shunt in use. Due to revisions of part of the shunt, sometimes in several different hospitals, parts of different shunt systems can be found in the same patient. Included here are the Hakim, Denver, Holter, Accuflow and Multipurpose shunts, and Rickham reservoirs.

drugs which might reduce CSF production in the hope of avoiding a shunt, but even if this approach is successful it could be many years before it is used.

A relatively new approach, based on an old method, is to pass a tube (endoscope) into the ventricles and to use this to remove part of the CSF-producing mechanism (choroid plexus), or to make an opening in the wall of the ventricle to allow CSF to escape. Such techniques, if successful, avoid the use of a shunt but they do not always work, and some patients will have a type of hydrocephalus for which the treatment is not suitable.

Today there are numerous types of shunt (*Figure 1*) but while they all look different they work in a very similar way. None can be said to be significantly better or worse than another, and the shunt is usually chosen by the surgeon on the grounds of experience, cost and personal preference.

Originally shunts were inserted so that a tube drained CSF from the ventricles in the brain, through the valve and through another tube into a vein in the neck and then into the heart ***(ventriculo-atrial or VA shunt, Figure 2a)***. While these are still used, most nowadays drain the CSF into the abdomen ***(ventriculo-peritoneal or VP shunt, Figur e 2b)*** and the bottom tube can be felt over the ribs. Despite all these developments, shunting can have complications. These can be divided into under-drainage, over-drainage and infection.

Under-drainage, in which the fluid is not removed quickly enough and the symptoms of hydrocephalus return, is one of the commonest problems. It is usually due to blockage of the upper or lower tubes of the shunt tissue, though it can be due to the shunt breaking or its parts becoming disconnected from each other. It is rarely due to the valve itself, which usually continues to function in the same way for years. Pressure may sometimes build up rapidly, resulting in loss of consciousness, and treatment is required as an emergency. However, in most cases the onset is more gradual, and can follow a minor illness such as a cold. Headaches increase in frequency and severity, often worse on waking in the morning. Vomiting and dizziness also occur, and sometimes other symptoms which vary from patient to patient. In these cases the parents or carers will be able to recognise the symptoms from previous episodes. Specialist hospital staff are now fully aware of the various presentations of 'blocked shunt' but non-specialists and family doctors may not be.

Shunt blockage can also have much more subtle consequences and the headaches may be infrequent, the main problem being behavioural deterioration. In older children this might take the form of increased irritability, 'laziness', poor or disruptive school performance or even more antisocial activity. This may be very difficult to

Figure 2a: Route of a ventriculo-atrial (VA) shunt, from the ventricles in the brain to the atrium in the heart.

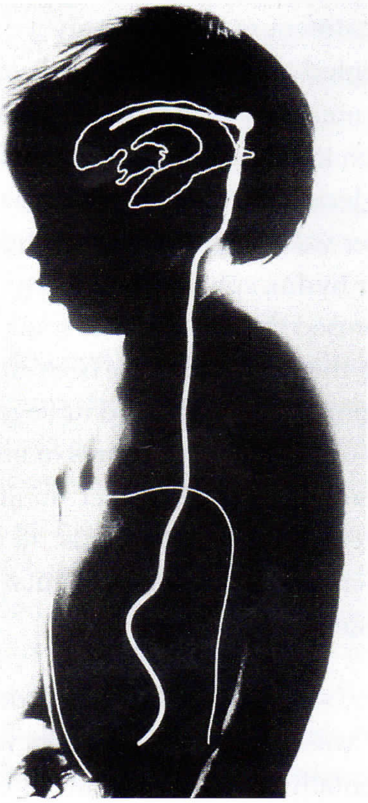


Figure 2b: Route of a ventriculo-peritoneal (VP) shunt, fr om the ventricles in the brain to the peritoneal cavity in the abdomen.

distinguish from the usual teenage angst but, if there is any reason to suspect that the deterioration in behaviour is not 'normal', assessment must be carried out by an experienced educational psychologist with a knowledge of hydrocephalus. The basis for the effects of high CSF pressure have been explained in the previous article. If the shunt is to blame, a dramatic improvement can result from appropriate treatment, though this form of shunt problem is particularly difficult to diagnose. It may be necessary to monitor CSF pressure, often over 24 hours. This can be done using a pressure monitor in the scalp connected to a recorder. In this way pressure can be recorded during sleep and changes in posture. Scans to show the size of the ventricles are particularly useful if they can be compared to previous scans, though in someone with clear symptoms of either high or low CSF pressure they may also serve to support the diagnosis.

In the case of over-drainage, the shunt allows CSF to drain from the ventricles more quickly than it is produced. If this happens suddenly, usually soon after the shunt is inserted, then the ventricles in the brain collapse, tearing delicate blood vessels on the outside of the brain and causing a haemorrhage ('subdural haematoma'). This can be trivial or it can cause symptoms similar to those of a stroke. The blood may have to be removed, and in some cases if this is not done it may be a cause of epilepsy later. If the overdrainage is more gradual, the ventricles collapse gradually to become slit-like ('slit ventricles'). This often interferes with shunt function causing the opposite problem, high CSF pressure, to reappear, but unfortunately the slit ventricles do not always increase in size again, producing the situation where there is very high CSF pressure with headache, vomiting etc but very small ventricles on scan.

The symptoms of over-drainage can be very similar to those of under-drainage though there are important differences. Headaches, dizziness and fainting occur and are often worse **after** getting up from lying down, whereas the headaches caused by high CSF pressure are often worse on waking, **before** rising in the morning. However the best way to diagnose the problem, having recognised that one exists, is to measure the CSF pressure over 24 hours. Underdrainage can be caused by blockage of the shunt tubing either at the top end, by tissue plugging the entry holes or by the position of the tube changing; or at the bottom end, by tissue in the abdomen sealing off the drainage tube. In VA shunts blockage can occur at the bottom end as the child grows, and a revision operation is sometimes necessary to lengthen the shunt at the age of 12-18 months. Over-drainage is a more difficult problem. There is no clear relationship between the type of valve (high or low pressure) or the brand, and over-drainage. A change of valve to a higher pressure cannot be relied upon to cure it, though it appears to do so in some cases. Studies have shown that the use of an 'antisyphon device', a small button inserted into the shunt tubing, will often solve the problem, but this does not always work. Some shunts have these built-in, but neurosurgical opinion varies as to whether they should be used. To change a valve pressure it is necessary to remove the valve and insert another. A relatively new shunt, the 'programmable' or adjustable shunt, is intended to allow adjustment of the working pressure of the valve without operation. The valve contains magnets which allow the setting to be changed by laying a second magnetic device on the scalp. This is undoubtedly useful where the need for a valve of a different pressure arises, but the adjustable valve is no less prone to over-drainage than any other and it cannot be used to treat this condition.

It has long been believed that a raised protein level in the CSF will block the shunt, and in babies with hydrocephalus shunting has been delayed until the protein level has fallen. Recent research has shown that a raised CSF protein level has no ill-effect on shunt function, nor does it increase the risk of infection, and there is now no reason to delay unless blood is also present.

The third complication of CSF shunting is infection. This is almost always due to bacteria from the skin getting into the CSF or shunt at operation, and is remarkably difficult to prevent. Antibiotics have not been shown to be of benefit for this purpose, and other measures often have only a temporary effect, though obviously the care and expertise of the surgical team is one of the most important factors in reducing the rate of infection to a minimum. However, even in the best of hands infection still occurs. One of our recent developments has been a process which makes shunts resistant to bacterial infection, and we hope that the current clinical trials will show that it is capable of reducing shunt infection by more than 80%.

In VP shunts infection will usually show itself within a few weeks or months of operation as a shunt blockage, though there may also be occasional fever and abdominal pain. Redness and swelling may be seen over the lower shunt tubing. It is important to distinguish between blockage of the VP shunt due to infection and that from other causes as the treatment is different. In VA shunts blockage due to infection is rare, and many months or years can go by before the infection becomes apparent. During this time there will be tiredness, irritability, poor appetite, various aches and pains, skin rashes and other signs but all of these can be due to common disorders. A blood test will usually reveal anaemia and this is an important though, on its own, not a specific indication of infection. Blood cultures and even CSF cultures can be negative. Later, blood may appear in the urine due to secondary kidney damage.

While shunt infections can sometimes be very easy to diagnose, they are often difficult and any delay increases the chance of further damage. Special blood tests have been developed which promise to allow a reliable diagnosis to be made early, but these are not in current use due to lack of funding.

VA and VP shunt infections are both treated in the same way. Until recently, this involved operations, long periods in hospital and disappointing relapses before a new 'clean' shunt could eventually be inserted. We have recently developed a new approach which shortens the time taken to treat most shunt infections to 7-10 days, with a very low relapse rate, and this is gradually being adopted by others. Unfortunately the infected shunt still has to be removed, though in future it may be possible to treat these infections successfully without taking out the shunt, and research into this aspect is continuing.

Though complications of shunting remain an important problem in the treatment of hydrocephalus, I hope I have managed to explain some of their mysteries, and to show that solutions are being found.

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