THE SURGICAL TREATMENT OF HYDROCEPHALUS WITH THE HOLTER VALVE

Herbert B. Eckstein, MA, MD, M. CHIR., FRCS.*

For many years surgeons have been attempting to treat hydrocephalus by operation. Essentially there are two ways of treating this condition, the first one being to reduce or eliminate the formation of cerebrospinal fluid, while the second one diverts the cerebrospinal fluid from the ventricular system. Destruction of the choroid plexus by diathermy or even excision is a major procedure which is technically difficult and often impossible to achieve. The results of this form of treatment have on the whole been highly unsatisfactory. Most surgical experiments have attempted to drain the ventricular system and in the past means were found to drain cerebrospinal fluid into the middle ear and then out through the eustachian tube, into the ureters and out from the bladder into the pleural and peritoneal cavities and even into the intestinal tract. Most of these procedures have had some success, but failures were more common and ascending infection and resulting meningitis or ventriculitis was a common sequel to most of these procedures.

The rational procedure is to divert the cerebrospinal fluid into the blood stream, but to achieve this a one way flow must be devised. Although a number of valves were produced previously, the ones by Holter and Pudenz were the first two successful valves to be used on any large scale.

I would like to discuss the management of hydrocephalus using the Holter valve and describe the various problems associated with this operation from my own experience.

As a pediatric surgeon the vast majority of my patients have had hydrocephalus associated with an Arnold Chiari malformation due to myelomeningocele. It is an important point to remember that I have treated relatively few patients with pure hydrocephalus.

* Consultant Surgeon, Queen Mary's Hospital for Children, Carshhaltan, Surrey.

First, the diagnostic measures to determine the cause of hydrocephalus become less important and second, many of my patients have had paraplegia and urinary incontinence, and one has perhaps subconsciously not tried quite as hard to keep these little patients alive, as one would with patients who had a totally correctible lesion. It is for this reason alone that I do not propose to give you statistics, as the treatment of hydrocephalus is only a small part in the treatment of the child with a myelomeningocele, and these children often die from a combination of different factors. My own experience with this operation is now well over 100 cases and I have had access to Mr. Macnab's patients at Great Ormond Street which total around 400. I have also carried out operations with a Pudenz valve on about 25 patients, but these have not been at all successful and have all failed ultimately for technical reasons associated with the tubing and the valve itself. It is only fair to point out, however, that surgeons in other centers are using the Pudenz type valve exclusively with apparently good results.

As far as the pre-operative investigations are concerned, we have reduced these to a reasonable minimum. It is important to know the approximate thickness of the cortex which can be determined by ventricular puncture, or even more accurately by doing a ventriculogram. We have done complete ventriculograms on many children using up to 100 cc of air, but an endotracheal anesthetic is almost essential for this procedure (Figure 1). At present we are doing a bubble ventriculogram using

Figure 1.

about 20 or 30 cc of air which does not require an anesthetic (Figure 2). Both types of ventriculogram will show the thickness of the cortex in different regions, but although it may show an aqueduct block, it is relatively useless to determine the exact etiology of the hydrocephalus. Cohen and

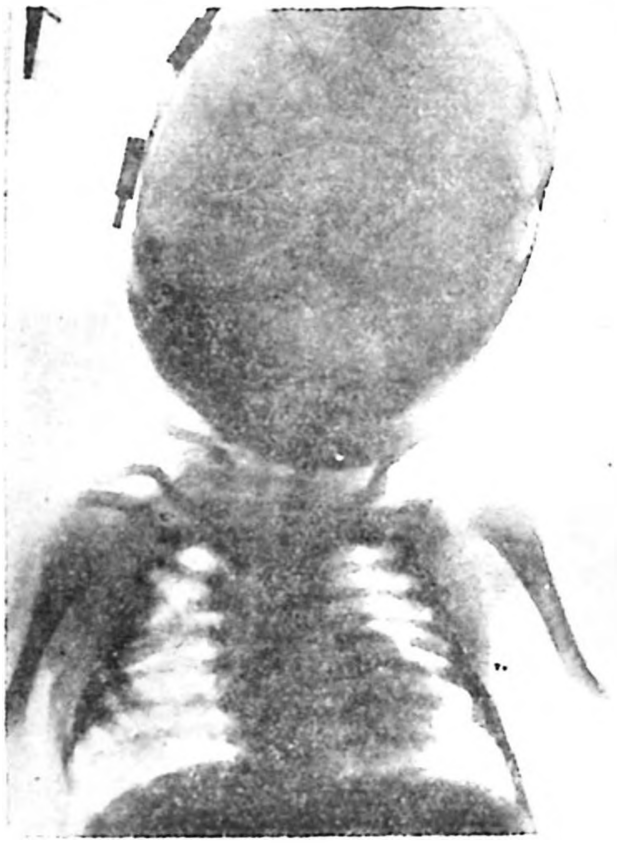

Figure 2.

Hoare, using a complete ventriculogram on 200 patients at Great Ormond Street, have shown that an aqueduct stenosis was present in about 25 per cent of their cases. However, such details are of little importance in the treatment, as the type of valve used is the same irrespective of the site of block. The cerebrospinal fluid must be examined microscopically for cells and also for its protein content. While in the past we been reluctant to insert a valve when the protein content was over 100 mg, I have now done operations on children whose cerebrospinal fluid had a protein content of 1000 mg and the valve appears to work quite satisfactorily. The highest protein content I know of in a successfully working valve is 4000 mg. The culture study pre-operatively of the cerebrospinal fluid is essential, as some children with a meningocele will develop meningitis, and obviously a valve cannot be inserted, until the cerebrospinal fluid has been sterilized. If the cerebrospinal fluid is infected, the child is treated by daily ventricular puncture with the installation of an appropriate antibiotic until the fluid becomes sterile. Continuous drainage of cerebrospinal fluid either by needle or tube is not recommended, as this tends to introduce infection more often than not.

I do not intend to discuss the diagnosis of hydrocephalus, but while the child with the grossly raised pressure and the enlarged head and down-turning eyes presents a classical picture, there is no doubt that considerable dilatation of the ventricular system can occur in children with myelomeningocele without an increase in the head circumference and without any of the classical signs of hydrocephalus. We are now tending to insert valves in these children before the head circumference reaches abnormal dimensions and are generally guided by the clinical appearance, the intraventricular pressure, and the bubble ventriculogram. A patent occipitomastoid suture is a most reliable sign of hydrocephalus even in children with normal-sized heads and normal-sized fontanelles.

The insertion of a Holter valve is a relatively simple operation. An endotracheal general anesthetic is essential, but I have discontinued routinely using intravenous cut down drips provided the child's hemoglobin is normal before the operation. I often use a small flap incision on the scalp and a burr hole and remove enough bone with a nibbler so that the upper end of the Holter valve lies in the groove of bone. The remainder of the valve should lie on top of the skull so that it can be felt easily after the operation. The distal catheter is threaded subcutaneously into the neck and pulled out through a short incision. Through this incision the internal jugular vein is exposed. Ideally the catheter is introduced through the common facial vein so that patency of the internal jugular vein is insured, but if this vein is small I do not hesitate to ligate the jugular vein itself, and to introduce the catheter directly into this. Having tried various ways of measuring the distance of the distal catheter I have come to the conclusion that just laying the catheter on the chest and cutting it off at the nipple will yield at least as good a result as any careful check of x-ray and measurement. It is important that the catheter should reach to the bottom end of the right atrium, or even into the upper part of the inferior vena cava so as to allow for the growth of the child (Figure 3). I have been using low pressure valves which were originally designed for subdural effusions almost exclusively in the past year with quite satisfactory results, and my impression is that these are more satisfactory than the standard medium pressure valves as the flow rate of cerebrospinal fluid is greater for a low pressure valve than for a medium pressure valve. Post operatively antibiotics are given routinely for one week and the children are then discharged home. A careful follow-up is essential so that any complications of the valve therapy can be detected early and remedied.

I have implied that the diagnosis and the actual treatment of hydrocephalus presents no great difficulty, and this indeed is so. The big problem

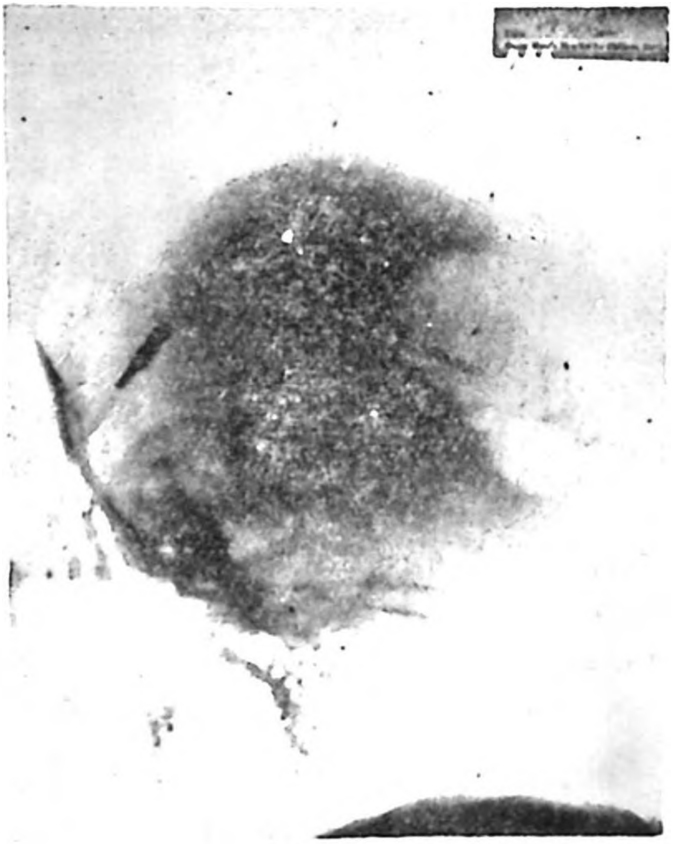

Figure 3.

in these children is the management of the various complications resulting after the insertion of a Holter valve, and I think that these deserve some discussion. Undoubtedly the commonest and most serious complication is a bacteremia usually caused by coagulase negative staphylocaci which has been shown by Stewart and others to be associated with a colonization of the valve itself.

I am sure that many normal people develop a temporary bacteremia which produces no symptoms at all, and the normal circulation can deal with this condition. However, if a foreign body is in the circulation, bacteria may become settled on this, and we have so far been unable to eradicate such a bacteremia with antibiotics alone. Even the installation of large quantities of antibiotics into the ventricular system with the hope that this antibiotic will reach the valve in a high concentration has been unsuccessful as a long term measure. While such a bacteremia does occasionally occur within a week or two of operation and may well be the result of contamination during operation, the majority of children develop this condition weeks or months later, and I am sure that this is not directly related to the operation. The only effective treatment for such a bacteremia is to dismantle the entire valve system, and my present policy is to exteriorize the caval cather for one week through a stab incision in the neck and allow the cerebrospinal fluid to drain into a closed bottle. The valve and all tubing is then removed and a further week is allowed to elapse with

the child on full appropriate antibiotic cover. Following this a second valve is inserted. Obviously it is essential to check the culture of the cerebrospinal fluid before the re-insertion of a valve, though it has been unusual to find infected cerebrospinal fluid, even in the presence of a bacteremia.

The second serious complication is a blocking of the distal catheter. This is in fact not due to a block of the catheter itself, but is due to the formation of a blood clot around the catheter sealing it off from outside. Such a block will not occur while the tip of the catheter is within the heart, but it will occur as the catheter rides out of the right atrium at the superior vena cava and is quite certain to occur when the tip of the catheter lies in the jugular vein itself (Figure 4). We now x-ray our patient at inter-

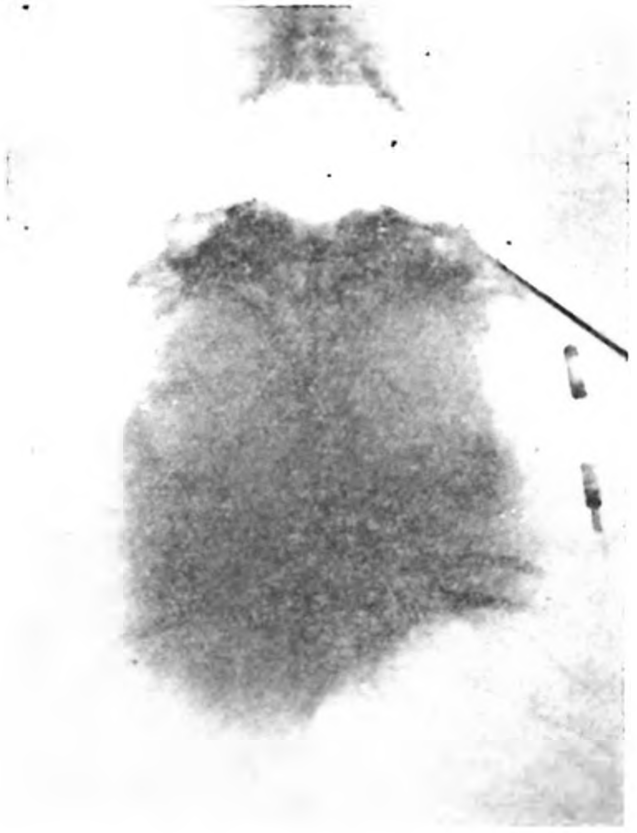

Figure 4.

vals of six months to determine the position of the distal catheter, and carry out prophylactic lengthening procedures if considered necessary. Once the catheter is blocked by a clot around it, it becomes much more difficult to re-insert a longer catheter. However, recently we have had more trouble from blockage of the ventricular catheter itself than from distal catheters, possibly due to the fact that we are inserting these valves with a much lesser degree of hydrocephalus. As the ventricles return to a nearly normal size following the insertion of a drainage system, the choroid plexus impinges upon the holes in the ventricular catheter and

Figure 5.

in fact is able to grow into these holes and to block the catheter completely. The differential diagnosis between a blocked upper and a blocked lower end is simple as attempted pumping of the valve will readily show which end is at fault. Replacement of an upper blocked catheter is a relatively minor and simple procedure. In a few children who have had meningitis we have found that the two ventricles no longer communicate with each other and that hydrocephalus may progress from the dilatation of one ventricle, while the other one is being drained satisfactorily. It is now possible to insert separate ventricular catheters into the two ventricles and to connect these through a T connection to one valve (Figure 6).

I have had experience with other less common complications. Craniostenosis, strange as it may sound, is becoming a more and more common problem, undoubtedly due to the fact that we are now inserting Holter valves at a much earlier age (my youngest patient was 4 days old). Generally speaking, in children with myelomeningoceles, the meningocele is treated as an emergency within hours of birth and a valve is inserted about two or three weeks later. Many of these children will have fused sutures by the age of six months, and so in fact finish with smaller heads than normal. Some of these children will undoubtedly require the formation of artificial sutures in due course, but I have as yet not had the occasion to do this. Pulmonary hypertension has occurred in a few of the older children

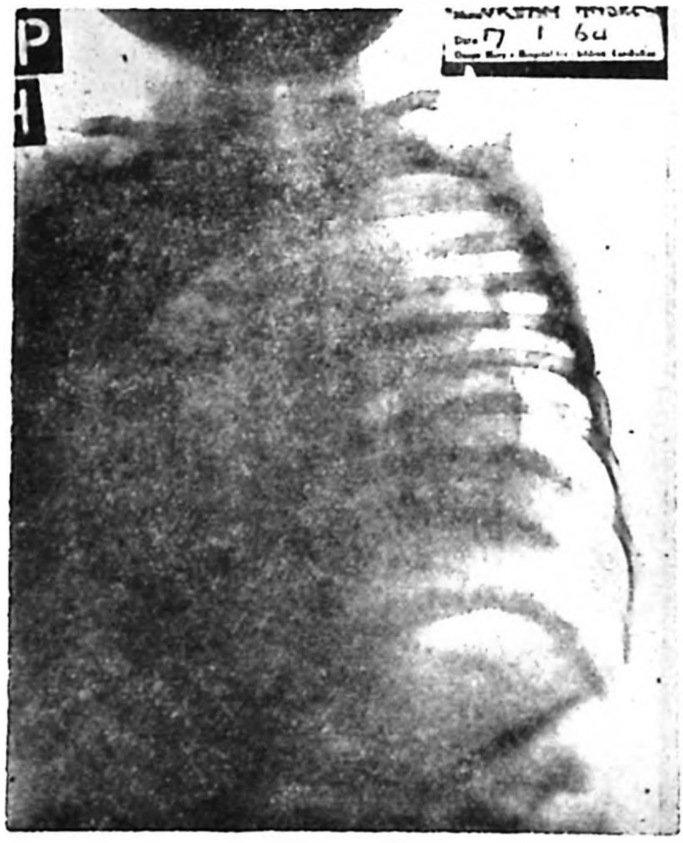

Figure 6.

who have had valves inserted as long as five years ago. This condition is undoubtedly due to the repeated falling off of small emboli which collect around the catheter in the right atrium, and there is at present no method of either preventing this, or of treating the condition. Another unusual complication on the same line is an embolic nephritis which is presumably due to the same cause. The last two complications would suggest that a valve should be removed as soon as spontaneous arrest has taken place. These complications are so rare that they have not tempted me to remove valves.

The most complicated problems associated with the Holter valve are those arising when the first valve has failed, either because of bacteremia or for technical reasons. As a second operation the valve can easily be inserted on the left side and the left jugular vein can be used, but the real problems from a technical point of view start after this. In the past I used to put the distal catheter either into the pleural cavity or into the peritoneal cavity as a last resort, but neither of these procedures have been successful in my hands. One child had a ventriculopleural shunt which worked quite satisfactorily for six months, after which the child was admitted as an emergency patient with severe respiratory distress, and a pleural cavity completely filled with crystal clear cerebrospinal fluid (Figure 7). Why such a shunt should work for a considerable time with satisfactory absorption of cerebrospinal fluid from the pleural cavity and should

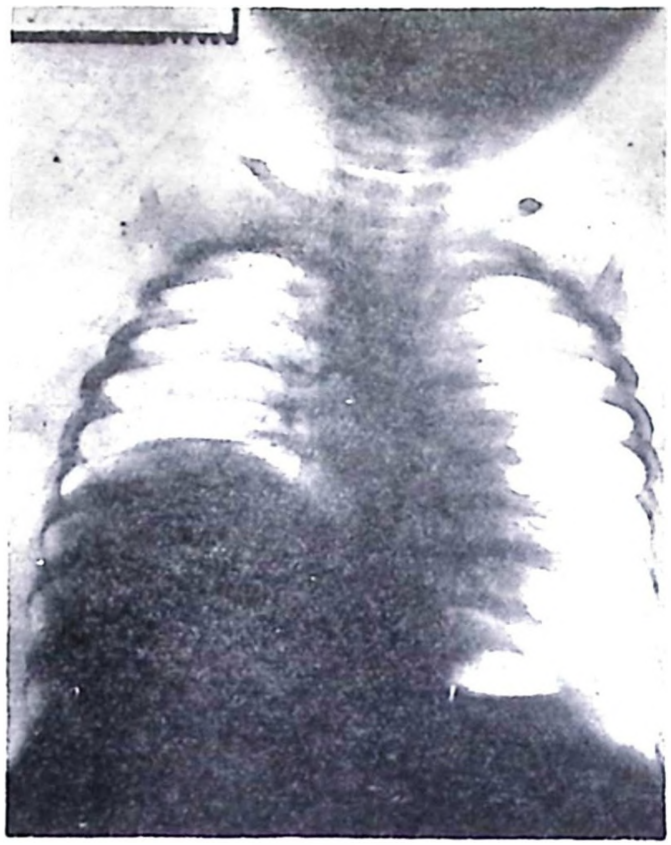

Figure 7.

then suddenly no longer function is not known. I am quite sure that it is essential to divert the cerebrospinal fluid into the blood stream itself and for this reason I now carry out a ventriculo trans–azygos shunt in those cases where both jugular veins have been used. The operation is not paticularly difficult, but it does involve a thoracotomy as well as the insertion of a new valve. The distal catheter can either be introduced into the azygos vein directly, or through the superior intercostal vein and can be guided by the finger into the heart itself.

Obviously on a relatively recent surgical invention like the Holter valve no long–term follow–up results can be quoted. The first valves were used in England in 1957, but it was not until 1960 that these were used on a really large scale. The recent suggestion by Sherard and Zachary that myelomeningoceles should be treated as surgical emergencies and closed within hours of birth has certainly shown that paralysis can be prevented in many of these children, but it has also led to a large increase in the number of cases surviving to develop hydrocephalus and requiring a shunt procedure. Laurence has shown that the cases of a large majority of children with with hydrocephalus will arrest spontaneously in time, but usually leaving a very large sized head. It is my impression that the condition in children treated with a valve will also arrest in time, but of course with a normal sized head. I think that there is little question of such a valve having to last a lifetime. I have seen a number of children of about five and six,

with valves that have obviously become blocked but these children are well and are not showing signs of progressive hydrocephalus. Our problem therefore is not such a big one. It is simply a question of controlling the hydrocephalus for a number of years. Unfortunately, although one can get statistics of the natural arrest of hydrocephalus in various age groups, one cannot predict the natural arrest in any particular child, and the case of each child must therefore be treated on its own merits. There is little doubt that the Holter valve has revolutionized the complete management of hydrocephalus in its various forms, and it is undoubtedly a more satisfactory form of treatment than any to which we have had access in the past.