THE TREATMENT OF CONGENITAL HYDROCEPHALUS

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This paper deals with our experience in the treatment of congenital hydrocephalus by means of the ventriculo-auricular-shunt operation, which was introduced into this country about 3 or 4 years ago.

Hitherto, the treatment of congenital hydrocephalus has given uniformly and universally poor results. No fewer than 30 different methods of surgical approach to the problem are known and none enjoyed popularity for any length of time. In fact it has taunted the medical profession since the days of Hippocrates. The multiplicity of methods of approach to this problem is an index of how difficult this neurosurgical problem actually is.

ANATOMICAL AND PHYSIOLOGICAL CONSIDERATIONS

Experimentally, it has been shown that the cerebrospinal fluid (CSF) is not a simple filtrate, but is actively secreted by the choroid plexus. The absorption of the CSF, on the other hand, is a combination of filtration and osmosis. CSF is continually secreted by the choroid plexus in the ventricles and circulates from the lateral ventricles through the foramen of Monro into the third ventricle. From there it passes through the aqueduct of Sylvius into the fourth ventricle, from where it escapes through the medially placed foramen of Magendie and the laterally placed foramina of Luschka into the subarachnoid space. About one-fifth of the total quantity

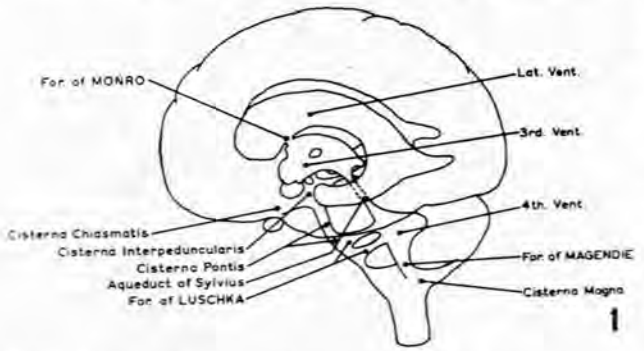


Fig. 1. Diagram of the ventricular system of the brain.

circulates over the spinal cord and is absorbed by the perineural lymphatics. The major portion finds its way into the basal cisterns, from where it is 'pumped' over the surface of the brain and absorbed by the arachnoid villi. Fig. 1 is a diagrammatic representation of the ventricular system in the brain.

PATHOLOGY

Pathologically, we recognize 2 types of hydrocephalus:

1. *Compensatory Type*

In this type there is usually no enlargement of the head and no increase in the CSF pressure. In the newborn it is caused by agenesis of the brain, and its counterpart is found in the adult as cerebral atrophy. There is passive distention of the ventricular system and laking in the subarachnoid space. It is of very little importance clinically to us and is not amenable to treatment.

2. *Hypertensive Hydrocephalus*

This is caused by: (*a*) excessive formation of CSF, (*b*) obstruction to its circulation, or (*c*) defective absorption.

According to Russell, the majority of cases in this group are due to some form of obstruction. Here, there are 2 sub-groups:

A. *The communicating type of hydrocephalus.* This is readily recognized clinically by the amount of CSF obtained by lumbar puncture. If more than 3 - 4 ml. are obtainable, then this is a communicating type of hydrocephalus. When neutral phenolsulphophthalein is injected into the ventricles, it will appear in the lumbar CSF in 3 - 5 minutes. The pneumo-encephalogram in this type of case shows that the ventricular system fills readily, and if the subarachnoid space is filled, the level of the obstruction can sometimes be easily demonstrated. The obstruction is usually at the incisura tentorii, the interpeduncular or chiasmatic cisterns.

B. *The non-communicating type of hydrocephalus.* This is practically always caused by some obstruction of the foramen of Magendie or stenosis of the aqueduct of Sylvius. Usually less than 3 - 4 ml. of CSF is obtained at lumbar puncture and neutral phenolsulphophthalein injected into the ventricles will not readily appear in the lumbar CSF. A lumbar pneumo-encephalogram will either fill the fourth ventricle only, in the case of aqueduct stenosis, or will not fill the ventricular system at all, in the case of obstruction at the foramen of Magendie.

CLINICAL PICTURE

The diagnosis of hydrocephalus is made without difficulty because of the obvious enlargement of the head and bulging of the anterior fontanelle. The enlargement is confirmed by measuring the occipito-bregmatic circumference, which will be above the normal average. As the hydrocephalus becomes more marked, the disproportion between the normal face and large cranium becomes more striking. The fontanelle and

sutures widen, the scalp veins become distended, the eyes are displaced downwards from pressure on the orbital plate, and the sclera becomes visible above the iris. The patient becomes irritable and has a high-pitched cry. Feeding becomes a problem, and there is relative weight loss of the body as the head grows in size, even up to a point where there is frank emaciation and dehydration.

Optic atrophy and mental retardation are in inverse proportion to the rapidity of the production of the hydrocephalus, but we are all familiar with the mild hydrocephalic with an I.Q. above the average.

Some hydrocephalic enlargements become spontaneously arrested weeks or months after onset. Every case should be assessed on its own merits to decide when to operate. It is our practice to observe the baby for at least 2 weeks, when circumstances are favourable, and if there is a steady and regular increase in the size of the head it is unlikely that spontaneous arrest will ensue.

All the patients must be subjected to investigations, e.g. X-rays of the skull, subdural taps, and pneumo-encephalography.

SURGICAL APPROACH

Fifty years ago, Payer attempted a ventricular-venous anastomosis. All 3 of his patients died within 4 months.

Six years ago, Pudenz *et al.* developed a ventricular-venous shunt by introducing one end of a catheter into the lateral ventricle and the other end into the right auricle of the heart (Fig. 2). The distal tip, which is lodged in the auricle (Fig. 3), has a slipcore-valve mechanism which will allow unidirectional flow of CSF. Since this method is an imitation of nature to shunt the CSF into the venous system, it is more likely to meet with success than other methods whereby the CSF is shunted into one or other body cavity, e.g. Eustachian tube, extradural space, pleural cavity, peritoneal cavity, lesser sac, ureter, duodenum or colon.

Fig. 2. Diagram showing the ventricular-venous shunt *in situ*.

The method described by Pudenz involves joining the appropriate length of the proximal and distal ends of the catheter by means of a metal connection in the neck.

Causes of Failure

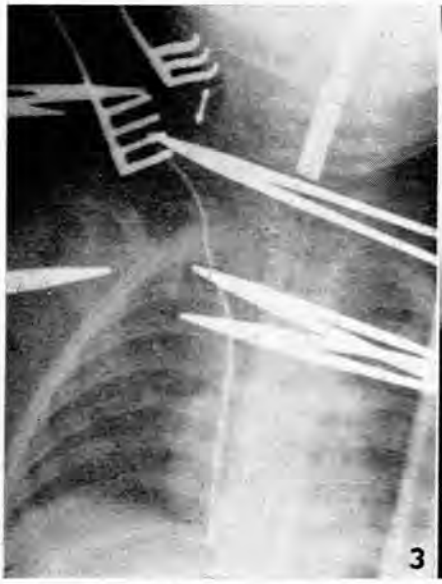
Four of our first 8 patients had to be re-explored at various times, but since we introduced a modification of the Pudenz method, whereby the junction of the proximal and distal ends of the shunt is made immediately below the burr hole or eliminated altogether, the recurrence rate has dropped to less than 25% in 20 successive cases. In 4 of the patients needing re-exploration we found:

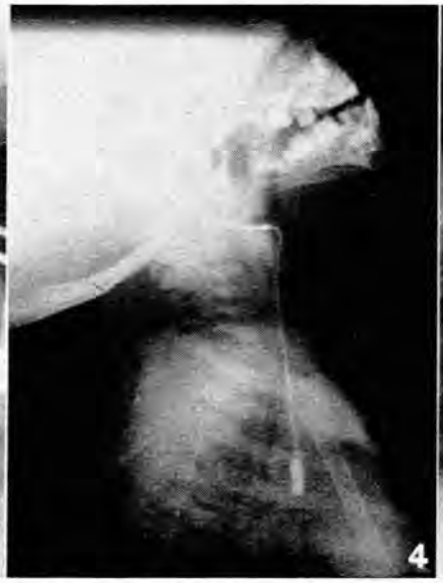
(*a*) a kink in the tubing at the metal junction (associated in 2 cases with a crack in the tube at the bend so produced);

(*b*) blocking of the lumen of the tubing by brain tissue;

(*c*) blocking or sealing off of the valve by fibrous tissue from the wall of the superior vena cava caused by faulty introduction—the tip of the shunt was not introduced into the right auricle, but was still in the superior vena cava (Fig. 4). The tube can also migrate upwards towards the head from the auricle (Fig. 5).

In a few cases the shunt becomes blocked by particulate matter. A flushing device has recently been introduced which already shows marked improvement in the ultimate results. It consists of a flanged silicone capsule and a diaphragm valve, which is introduced into the shunt where it comes through the burr hole immediately under the skin.





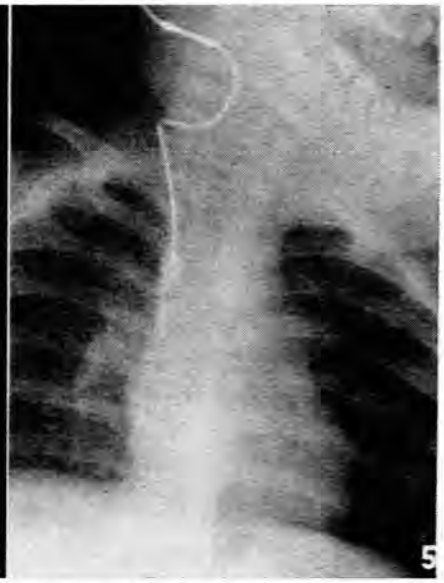


Fig. 3. X-ray showing the distal end of the tube in the right auricle (note the metal connecting-piece in the neck).

Fig. 4. X-ray showing an obstruction distal to the tip of the catheter at the level of the superior vena cava.

Fig. 5. X-ray showing cephalic migration of the catheter (the connecting-piece is again well shown).

When the capsule is compressed by pressure exerted on the overlying skin, the diaphragm closes the ventricular inlet and the CSF in the capsule is forced through the cardiac tube.

The operation, as such, is technically easy to perform and hardly disturbs the baby. No baby requires transfusion, and it should be avoided for fear of over auto-transfusion from a large reservoir in the head.

Results

We had 1 postoperative death in 30 cases and 36 operations. At postmortem examination, a massive intraventricular haematoma was found, associated with anaemia and collapse of the cerebrum. The greatest danger is intercurrent infection and septicaemia.

SUMMARY

The horrifying condition of congenital hydrocephalus can now be successfully treated in many cases, and the foetus with the large head need not be destroyed by the obstetrician, nor need the hydrocephalic patient, discovered after birth, be left to die a slow death.

Our experience in 30 cases over a period of 3 years is most encouraging, and should the shunt need to be replaced or adjusted as the child grows, this should offer no difficulty to either surgeon or patient.