The arrest of treated hydrocephalus in children

A radionuclide study


T. Y. Nelson Department of Neurology and Neurosurgery, Department of Nuclear Medicine, and Childrens Medical Research Foundation, Royal Alexandra Hospital for Children, Camperdown, New South Wales, Australia

A prospective study was made of the incidence of arrest of treated non-neoplastic hydrocephalus in 30 neonates and infants over a 5-year period. Radionuclide assessment of shunt function and cerebrospinal fluid (CSF) dynamics was carried out at intervals over this period, using a method that allowed injection of the radionuclide into the ventricular system independent of the shunt apparatus. The radionuclide scanning results were correlated with computerized tomography and clinical findings. Of 24 patients still available for analysis at the end of 5 years, four patients showed restoration of CSF circulation independent of the shunt apparatus and, in three of these, the shunt has either been clipped or clipped and removed without any ill effects. Clinical details of these three patients are provided. Two initially had communicating hydrocephalus, and one had probable aqueduct stenosis.

KEY WORDS • arrested hydrocephalus • shunt function • radionuclide scanning • cerebrospinal fluid

SHUNTING is now well established as the appropriate treatment for neonatal and childhood hydrocephalus. Despite the morbidity and mortality rates associated with such a procedure, the beneficial effects are well documented.2,3 A major problem persists, however, in identifying those patients who have developed spontaneous arrest of hydrocephalus and therefore no longer require a shunt. Despite the maxim "once a shunt, always a shunt," it has been suggested that between 9% and 20% of hydrocephalic children will, in fact, become independent of their shunts.6,7 Close monitoring of these patients is required to determine whether true shunt independence has occurred, if the deleterious effects of untreated hydrocephalus are to be avoided.3,10

This paper presents the initial results of a continuing prospective study designed to assess shunt function in previously treated neonatal and childhood hydrocephalic patients. A new technique has been employed utilizing an indwelling catheter, separate from the shunt apparatus, which allows computerized analysis of ventricular radionuclide clearance.9 Computerized tomography (CT) scanning, clinical assessment, and psychometric studies have been performed to relate clinical and developmental progress to the investigative findings. The initial results provide data on the incidence of shunt independence in the first 5 years of life in previously shunt-dependent hydrocephalic children.

Clinical Material and Methods

A consecutive series of 30 neonates and infants, first treated for progressive non-neoplastic hydrocephalus in 1977 to 1978, forms the basis of the study. There were 14 males and 16 females and all were under 1 year of age at the time of initial shunt insertion (Fig. 1). All patients were investigated in the conventional manner to diagnose hydrocephalus and its severity, site of cerebrospinal fluid (CSF) obstruction, and etiology where possible (Table 1). A number of children had other extraneural developmental abnormalities.

Following the diagnosis of progressive hydrocephalus, a ventriculoperitoneal (VP) shunt (with a Hakim pediatric medium-pressure apparatus) was inserted on the right side. At the same procedure, a right frontal burr-hole was made, and a right frontal ventricular
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Fig. 1. Age ranges of the 30 patients in the study at the time of insertion of the ventriculoperitoneal shunt and Rickham system.

Table 1

<table>
<thead>
<tr>
<th>Etiology</th>
<th>No. of Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>aqueduct stenosis</td>
<td>7</td>
</tr>
<tr>
<td>idiopathic</td>
<td>7</td>
</tr>
<tr>
<td>myelomeningocele</td>
<td>1</td>
</tr>
<tr>
<td>acquired</td>
<td>4</td>
</tr>
<tr>
<td>communicating hydrocephalus</td>
<td>2</td>
</tr>
<tr>
<td>idiopathic</td>
<td>2</td>
</tr>
<tr>
<td>myelomeningocele</td>
<td>2</td>
</tr>
<tr>
<td>acquired</td>
<td>2</td>
</tr>
<tr>
<td>Dandy-Walker syndrome</td>
<td>3</td>
</tr>
<tr>
<td>4th ventricular outflow obstruction</td>
<td>2</td>
</tr>
<tr>
<td>posterior fossa arachnoid cyst</td>
<td>2</td>
</tr>
<tr>
<td>total cases</td>
<td>30</td>
</tr>
</tbody>
</table>

Radionuclide Studies

Serial CRSS's of ventricular clearance (or shunt function studies in those patients of whom the Rickham apparatus had been removed) were performed in 24 (80%) of the 30 patients. An average of 3.8 isotope studies per patient has been performed over the 5-year period. In 20 (83%) of the patients, intraventricular radionuclide rapidly cleared down the shunt systems, and these patients remained either totally or partially shunt-dependent. Four (17%) of the patients, of whom three had communicating hydrocephalus and one had aqueduct stenosis, demonstrated progressively less radionuclide clearance down the shunt system with circulation via the basal cisterns. Three of these eventually showed no clearance of radionuclide down the shunt system. Of these, none had clinical stigmata of intracranial hypertension or progressive developmental delay; all three had their shunts clipped and, in two cases, subsequently removed. The fourth patient remains under observation.

During the period of the study, CRSS's were also used to assess shunt patency in 10 children who presented with a clinical syndrome suggestive of shunt malfunction. Shunt obstruction was found in four of these, and in each case the obstruction was confirmed at operative revision. The remaining six patients, all with normal CRSS's, recovered uneventfully.

Neuroradiological Studies

Serial CT scans were performed in 21 of the 24 patients who underwent sequential CRSS's. In 17 of these children, ventricular size was reported as normal or small with a radionuclide-determined patent shunt system. Four patients initially had persistence of ventricular dilatation after shunt insertion despite good radionuclide clearance. In all four, ventricular size decreased within 12 months without any shunt revision. Two patients had normal ventricular size and one had dilatation of the frontal horns, despite radionuclide studies showing unequivocal shunt blockage. In all of these children, ventricular size remained unchanged following clipping or removal of the shunt system. One
I. H. Johnston, R. Howman-Giles and I. R. Whittle

**Case Reports**

**Case 1**

This 10-month-old baby girl had a 6-week history of rapid increase in head circumference following initially normal development. A CT scan showed severe symmetrical hydrocephalus associated with a posterior fossa cyst (Fig. 2a). Air ventriculography revealed an anterior cortical thickness of approximately 1.5 cm and also showed that what appeared to be a posterior fossa cyst was probably a dilated fourth ventricle. This latter finding and communication of the cyst with the spinal subarachnoid space was confirmed by pneumoencephalography. A VP shunt and Rickham reservoir were inserted, and a subsequent CRSS revealed rapid clearance of the isotope down the shunt system.

One year later the patient had normal neurological development and absence of any clinical signs of intracranial hypertension; however, a CT scan showed persistent ventricular dilatation (Fig. 2b). On CRSS there was passage of radionuclide into the basal cisterns, posterior fossa cyst, and spinal subarachnoid space, with good clearance at 24 hours but no evidence of flow along the shunt system. On the basis of these studies the shunt was revised. The opening pressure of the peritoneal catheter was found to be 18 cm H$_2$O, and this catheter was replaced. Eighteen months later, there had been continuing normal development, no change in head circumference, and no evidence of intracranial hypertension. A CT scan still showed quite marked dilatation of the frontal horns (Fig. 2c), and the CRSS again demonstrated slow clearance of the isotope without flow along the shunt. Psychometric assessment in summary revealed a mental age of 4 years 11 months (at a chronological age of 4 years 1 month), and an estimated intelligence quotient (IQ) of 110. There were, however, some difficulties in comprehension of language and drawing construction. No further action was taken at this stage.

The patient was readmitted 1 year later when a CT scan again showed quite marked symmetrical dilatation of the frontal horns (Fig. 2d) despite good placement of the shunt apparatus. There had, however, been a small decrease in ventricular size since the previous study. A CRSS again demonstrated egress of fluid into the basal cisterns and clearance over the hemispheres with no flow along the shunt. The shunt was then clipped and the child has remained well over a 6-month follow-up period.

**Case 2**

This 10-week-old baby boy presented with a history of irritability and increasing head circumference. Investigations showed symmetrical hydrocephalus (communicating) with an anterior cortical thickness of 2 cm. A VP shunt and frontal catheter with Rickham reservoir were inserted. A subsequent CRSS showed rapid clearance of the radionuclide along the shunt apparatus with no flow along normal pathways. Studies were repeated in 1980 and 1981 with essentially similar findings. Throughout this period developmental progress was normal. At 4 years of age, a CRSS showed normal ventricular size but no flow of isotope along the shunt apparatus. The isotope cleared via the basal and spinal subarachnoid spaces and over the cerebral hemispheres. The child was clinically well with normal developmental progress and low average intelligence on Wechsler Preschool and Primary Scale Index (WPPSI). The Peabody Picture Test showed a mental age of 3
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FIG. 3. Computerized tomography scans in Case 2. Left: Scan at age 4\nyears 8 months, prior to removal of the ventriculoperitoneal shunt. The position of the two ventricular catheters is well seen. Right: At age 5 years, 5 months after shunt removal, there has been no change in ventricular size. The ventricular catheter attached to the Rickham reservoir is still in place.

years 9 months at a chronological age of 4 years 4 months.

In view of the above findings, the shunt apparatus was removed. Follow-up CT scan, 5 months after the shunt removal, showed no change in ventricular size (Fig. 3) and the child has remained clinically well.

Case 3

This 3-week-old baby girl had a history of rapidly increasing head circumference. Initial studies showed mild symmetrical hydrocephalus. No treatment was initiated. Further studies after several weeks showed quite marked progression of the hydrocephalus with evidence of aqueduct stenosis (Fig. 4 left). A right VP shunt and frontal catheter were inserted in April, 1978. Postoperative CRSS showed good clearance along the shunt.

The child was readmitted 8 months later for further assessment. Developmental progress had been normal, and there was rapid clearance of radionuclide along the shunt system on CRSS but there was also some activity in the basal cisterns and passage of radionuclide over the convexity of cerebral hemispheres after 24 hours. A CT scan showed a small ventricular system. Similar findings were obtained on repeat studies in 1979 and 1980, at which time development remained normal.

In November, 1980, an elective CRSS showed no evidence of flow along the shunt while the ventricular size on CT scanning remained small (Fig. 4 center). Following this the peritoneal catheter was clipped and a CT scan 2 months later showed no change in ventricular size. This remained the case over a further 12 months, during which period development continued to be normal (Fig. 4 right). The shunt system was then removed. The child has remained clinically well and is performing with her peer group in primary school.

Discussion

Arrested hydrocephalus is a subject of some complexity, with problems relating to incidence, recognition, and mechanisms. The situation is relatively clear with regard to incidence in untreated patients, \(^5\) but with the recognition of the deleterious effects of untreated hydrocephalus and the ready availability of shunts, most patients with any evidence of progression are now treated aggressively. The figures with regard to arrest in treated patients are unsatisfactory. The studies are retrospective and inadequately controlled.

Utilization of techniques involving injection of radionuclide into the shunt apparatus is open to objection

FIG. 4. Computerized tomography scans in Case 3. Left: At age 3 months and prior to shunting there is moderate, asymmetrical hydrocephalus. Center: At age 3 years 9 months, there has been resolution of the hydrocephalus despite radionuclide evidence of shunt nonfunction. The position of the ventricular catheters is well seen. Right: At age 4 years 9 months, there has been no change in ventricular size 1 year following removal of the ventriculoperitoneal shunt.
since these methods may not give a satisfactory measurement of CSF pressure, nor may they give a rate of clearance of CSF from the ventricular system. Clinical experience has repeatedly shown that shunt obstruction and intracranial hypertension can exist at the time of an apparently satisfactory radionuclide study. In addition, the absence of the usual clinical signs of intracranial hypertension cannot be relied upon as a basis for the diagnosis of arrested hydrocephalus. Up to 83% of such patients may still have significant intracranial hypertension.1

Difficulties in determining the incidence and in recognizing the existence of arrested hydrocephalus are reflected in the figures and statements available in the literature. Thus, two retrospective studies using radionuclide clearance methods have shown an incidence of arrested hydrocephalus in 9% and 20% of patients, respectively.6,7 On the other hand, the maxim "once a shunt, always a shunt" is still regarded as sound, and Milhorat12 has recently remarked that "spontaneous arrested hydrocephalus is rarely an optimum physiological state if the ventricles are enlarged and there is no evidence of a primary atrophic process."

The present study is an attempt to address the problems of recognition of arrested hydrocephalus prospectively in shunted patients. The technique described in detail elsewhere9 allows a quantitative evaluation of CSF clearance from the ventricular system as well as a spot measurement of CSF pressure. Combined with serial CT scanning and detailed clinical and psychological follow-up monitoring, it provides a comprehensive evaluation of shunt function over an extended period. The results indicate that hydrocephalus is arrested, with restoration of normal patterns of CSF circulation, in up to 17% of patients.

Little can be said with regard to the mechanism of arrested hydrocephalus on the basis of the present study. In general terms, it may be due to the reestablishment of normal pathways, utilization of alternative pathways, or changes in CSF production. The evidence from this study seems to favor restoration of normal pathways as the mechanism for arrest, and this may be more likely to occur in patients with communicating hydrocephalus. It is known from experimental and other studies that alternative pathways of CSF circulation do exist.5,13,14 It may be possible with refinement of radionuclide techniques to examine this question more fully. Nothing can be learned of CSF production from the present results, although experimental work suggests changes in patterns of production related to changes in CSF pressure.11 Whether there could be a long-term resetting of CSF production levels following a period of shunt obstruction and intracranial hypertension is not known.

It does seem from the present study that there is a significant incidence of arrest of hydrocephalus in previously shunted patients. The method described provides the best available technique for following progress and identifying arrest when it does occur. In addition, by leaving the Rickham reservoir and frontal ventricular catheter in place, it is possible to follow these patients after shunt removal. This is particularly pertinent in view of the possibility of reversal of arrest reported elsewhere.7 Results of this initial study have encouraged us to continue and indeed to expand the study to include a greater number of patients and also to prolong the follow-up period of those patients who have had their shunts removed on the basis of serial investigations.

References

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Address reprint requests to: Ian H. Johnston, F.R.C.S., Department of Neurology and Neurosurgery, Royal Alexandra Hospital for Children, P.O. Box 34, Camperdown, New South Wales 2050, Australia.