HYDROCEPHALUS TREATED BY ARACHNOID-URETEROSTOMY

Report of 50 Cases

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It HAS been demonstrated in recent years that a variety of surgical procedures which shunt cerebrospinal fluid away from the subarachnoid spaces are technically feasible and may result in dramatic immediate arrest of progressive hydrocephalus and all of its distressing clinical complications. These surgical procedures include diversion of spinal fluid into the mastoid antrum, into the pleural and peritoneal spaces, into the bone marrow, and into the urinary tract.¹⁻⁴ It now seems evident that an objective analysis of longer term results of such operations with respect to growth and development of these children over a period of years is needed—not in any single isolated patient, but in a group of patients treated by a similar technic.

It is the purpose of this paper to report on experiences with 50 consecutive cases of communicating hydrocephalus in which one specific type of shunt, arachnoid-ureterostomy, or diversion of spinal fluid under pressure from the lumbar subarachnoid space to the urinary tract, has been carried out. No attempt is made in this report to compare the results of arachnoid-ureterostomy with those of other procedures but the material presented here becomes available for such comparisons with other experiences. At a later date the results from this clinic of such procedures as third ventriculostomy, ventriculocisternostomy, choroid plexectomy, ventriculo-peritoneal shunt, lumbar arachnoid-peritoneal shunt, and ventriculo-pleural shunt will be presented.

Operative Procedure

Certain principles previously tested in the laboratory in the treatment of experimental hydrocephalus⁵ were first applied to a patient with communicating hydrocephalus secondary to diffuse staphylococcus meningitis in 1948. In this patient, a seven year old girl, the left kidney was removed and a small calibre plastic (polyethylene) catheter was used to shunt fluid from the lumbar spinal fluid reservoir into the ureter.² The striking success of this procedure in an otherwise desperate situation prompted its early application to the discouraging problem of communicating hydrocephalus in early infancy.

Direct anastomosis of the ureter to the lumbar theca as introduced by Heile in 1925⁶ had been tried sporadically in this country and abroad with little success because of its technical difficulties and postoperative complications.⁹⁻¹¹ The operation used in the patients reported here was designed to obviate the complications of direct uretero-arachnoid anastomosis as far as possible. The operation is simple, well tolerated and easy to perform in one stage in patients of any age (three weeks to adult). The operative technic has not varied in this series of 50 children in any essential detail from that previously described.⁷

In all but 2 or 3 instances, the left kidney has been sacrificed simply because it is
slightly higher than the right and on the aortic side rather than that of the vena cava. The kidney is removed through a standard subcostal incision. Through a separate midline incision the spine and lamina of L₂ are removed exposing an area of dura about 1.5 cm. in length. A short incision is made in the dura in such a fashion as not to open the arachnoid. A pin-point hole only is then made in the protruding arachnoid and the polyethylene tube inserted through it as spinal fluid pours forth. This maneuver insures subarachnoid rather than subdural placement of the plastic tube, which is directed caudally for a distance of about 3 to 4 cm. so that it floats freely among the cords of the cauda equina. The tube is then tunnelled through the paraspinal muscles to the perinephric space and introduced into the ureter for 3 to 5 cm. Three or four silk sutures in the margin of its lumen are used to pull the ureter over the plastic tube and secure it firmly to the fascia of the paraspinal musculature. No ligatures are placed around the ureter at any time and an effort is made to leave its nerve and blood supply intact.

Material

The age distribution of the patients in this series is shown in chart 1. It will be noted that the majority were infants between 3 and 6 mo. of age. In a few instances, hydrocephalus was definitely known to be a sequel to generalized bacterial meningitis. In most of these patients, however, there were no clues in the history to suggest the etiology of the communicating hydrocephalus determined by procedure.
by the preoperative studies. A number of additional patients who have had arachnoid-ureterostomy performed as a palliative procedure to relieve pressure associated with inoperable tumors are not included in this report.

Preoperative studies include those necessary to establish the diagnosis of progressive communicating hydrocephalus as well as urine analyses, nonprotein nitrogen determination and intravenous urography to insure the presence of 2 normally functioning, uninfected kidneys. Communicating hydrocephalus is established by combined ventricular and lumbar puncture with hydrodynamic, tracer dye and air studies. The extent of the hydrocephalus is determined by ventricular air-bubble studies.

Arachnoid-ureterostomy has been performed only in children with a severe degree of hydrocephalus and only in children in whom it has been established that the hydrocephalus is progressive; that is, in patients in whom the head size is steadily increasing at an accelerated rate and in whom the rate of absorption and excretion of a tracer dye (phenolsulphonephthalein) placed in the subarachnoid space is markedly diminished. In this series of 50 patients, the tracer dye passed from the lateral ventricle to the subarachnoid space in 4 patients in less than 2 minutes, in 15 patients in between 2 and 4 minutes, in 19 patients in between 4 and 10 minutes, and in 7 patients in between 10 and 20 minutes; in 4 patients the test was not done or was unsatisfactory. The 12 hr. urinary excretion of the dye injected into the ventricle was less than 10% in 7 patients, from 10 to 20% in 29 patients and from 20 to 30% in 7 patients; normal excretion over a 12 hr. period with satisfactory urinary output should be 45 to 80%.

Many patients have been considered for surgery and regarded as unsuitable because ventricular air studies revealed the cortex compressed to a thickness of only a few millimeters, or because the baby appeared to be blind or otherwise irreversibly damaged. Among this series of 50 patients the cerebral cortex measured less than 1 cm. in thickness from the lateral ventricle to the surface in 12, between 1 and 2 cm. in 23, between 2 and 3 cm. in 14 and greater than 3 cm. in only 1 patient.

An effort has been made to evaluate each infant with hydrocephalus individually on the basis of all the information available and not on any single test; if there seemed to be reasonable possibility that with arrest of the hydrocephalus normal development might proceed, operation has been recommended. As will be pointed out subsequently, in only one of the surviving patients is it now felt that the baby was irreversibly damaged too severely prior to operation to permit a satisfactory result. It was, therefore, poor clinical judgment to operate on this patient.

**Postoperative Course**

The immediate postoperative course after arachnoid ureterostomy is usually smooth. Parenteral fluids are administered for 2 or 3 days routinely and subsequently only if the intake by mouth is inadequate. Ordinarily by the fourth or fifth postoperative day these infants exhibit a ravenous appetite and feedings must be repeatedly increased to keep them satisfied. At about this time also, it is noted that they become much more alert, smile and play, and lose their preoperative hyperirritability, disorganized motor activity and restlessness. There is transient rise in the blood nonprotein nitrogen level, usually to about 40 to 60 mg./100 cc. This returns to normal in 2 to 3 weeks and presumably indicates that the remaining kidney has satisfactorily met the additional burdens placed upon it.

After operation the anterior fontanelle remains depressed at all times (Fig. 1). If it does not do so, even when the infant is crying, it undoubtedly means that satisfactory drainage of spinal fluid is not taking place. The cranial bones overlap another to some degree so that ridges appear at the sites of the sutures. In infants, the head size usually diminishes in circumference by 1.0 cm. up to as much as 4.0 cm. depending upon the degree and rapidity of onset of the hydrocephalus prior to operation. From approximately two weeks after operation on, the head does not grow at all for a period usually of one to two years. At this time, or presumably whenever the infant’s body size has caught up to proper proportion to the head size, the circumference of the head again begins to increase, but now at a normal rate.

In those patients who have had lumbar or ventricular taps performed after surgery
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Fig. 1. Infant 3 1/2 mo. of age after arachnoid-ureterostomy showing persistent depression of anterior fontanelle and overlapping of the frontal and parietal bones at coronal suture line. Head became 2 cm. smaller in circumference after operation.

while the shunt was in satisfactory operation, the cerebrospinal fluid pressure has been from 50 to 90 mm. water. No sequelae of significance have been noted from this persistent intracranial hypotension. Three or four patients have had subsequent ventricular air studies for one reason or another and have shown a marked decrease in the size of the ventricular system. This has also been noted at postmortem examination in patients who have died a number of months after operation because of acute dehydration. It suggests, of course, that much of the thinning of the cerebral cortex is due to compression of interstitial fluid out of the brain by the expanding ventricles.

RESULTS

In this series there has been only one death during the postoperative period. This was a three months old infant in extremely poor general condition who developed postoperative aspiration pneumonitis and died suddenly from recurrent aspiration on the seventh day after operation. There have been no other deaths during the hospitalization in which the initial surgical treatment was carried out.

There have been no technical problems with establishment of a well functioning shunt at the initial operation with one exception. This was in a 2 1/2 week old, 3.0 kg. infant in whom the ureter was too small to receive the smallest size polyethylene catheter available. In two other infants, each 4 weeks of age and weighing about 3.2 kg., a small size catheter (internal diameter equal to a #19 intravenous needle) was satisfactory. In six patients obstruction developed in the shunt from 5 to 14 months following operation. These patients were all re-explored and in every case there was found to be a block in the tip of the tube within the subarachnoid space; this was apparently a plug of fibrin, arachnoid, or perhaps an adherent nerve root. In each of these six patients a new tube was inserted and has functioned satisfactorily for periods varying from 1 to 31 months to date. In no instance, either clinically or in patients examined at post mortem, has there been any demonstrable obstruction or reaction at the ureteral end of the plastic tube.

There has been no obstruction of the arachnoid-ureterostomy as a result of growth alone up to four years after operation, as far as is known. Since both the ureter and the

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structures within the spinal canal grow with the child and since there is 4 cm. of the plastic tube introduced into both the ureter and the spinal subarachnoid space, it is not anticipated that growth will ever in itself obstruct this type of shunt.

There has been no infection of either the nephrectomy or the laminectomy wounds in any of these patients. There has been early postoperative meningitis in two patients, in both due to B. coli. Both patients eventually succumbed after lingering illness of 4 and 6 months in spite of all antibiotic treatment and the persistent patency of the shunt. In only one of these was B. coli also cultured from the urine. Two other patients in this series died because of central nervous system infection. One was a child operated on ill-advisedly, early in the series, who had apparently an active ependymitis at the time and died two months after operation with an aqueduct block and extensive ventriculitis but never a positive spinal fluid culture. The other was a patient with salmonella meningitis, who was operated upon for hydrocephalus after the spinal fluid had been sterilized. The patient did well but remained a salmonella carrier in her intestinal tract and died of virulent recurrence of salmonella ventriculitis and meningitis eight months after arachnoid-ureterostomy. Five additional patients at various periods several months after operation have had fever, stiff neck, irritability and an increased number of white blood corpuscles in the spinal fluid. One of these only had a positive culture (B. proteus). These patients have all responded well to supportive and antibiotic therapy.

The most difficult problem postoperatively with these patients has been the control of their electrolyte and water balance during periods of intercurrent illness. As long as these children have been in good health, afebrile, maintaining a normal intake of food and fluids, and not vomiting or having diarrhea, there has been no problem in dehydration. However, eight infants, and perhaps one or two more, have died at periods from 3 weeks to 12 months after operation of acute overwhelming dehydration secondary to some variety of intercurrent illness, usually an upper respiratory or gastrointestinal infection.

The reasons for this bear some explanation. They are evident on the basis of metabolic balance studies done at the Children’s Medical Center under the direction of Dr. William Wallace. These studies were carried out in hydrocephalic infants before and after arachnoid-ureterostomy. The infants were placed on a virtually sodium-free diet but with a normal total fluid intake. On this diet, since the kidneys conserve sodium, a constant low level of sodium is reached in the urine within a few hours. Dehydration and weight loss do not occur. After arachnoid-ureterostomy, balance studies in the same patients were carried out on the same sodium-free diet and fluid intake. Although the remaining normal kidney continues to do its job of conserving sodium and fluid, there is now in the voided urine a quantity of spinal fluid from the shunt containing sodium and chloride at normal spinal fluid levels. If this loss of fluid and electrolytes is not replaced by adequate intake, the defence of normal plasma volume by transfer of interstitial fluid into the vascular system fails and may do so quite abruptly. The result may be profound peripheral vascular collapse and death in a clinical picture resembling that of an Addisonian or hypoadrenal crisis. A summary of one of these metabolic studies is charted in chart 2.

The clinical implications of these balance studies are obvious. In young infants, when the diet consists largely of cow’s milk, the sodium intake is low, in the neighborhood of only 1 to 2 gm. per day. The amount of sodium lost in the spinal fluid after arachnoid-ureterostomy in such an infant may represent, therefore, a significant fraction of the total
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EFFECT OF URETERO-ARACHNOID ANASTOMOSIS ON Na BALANCE

CHART 2. Metabolic balance chart on hydrocephalic infant placed on sodium-free diet before and after arachnoid-ureterostomy. On left (before operation) urinary excretion of Na drops to negligible amount, body weight does not change, and there is little change in serum Na concentration. On right (after operation) there is continued excretion of Na, body weight falls off sharply and serum concentration of Na diminishes; experiment was discontinued after 50 hr. because of patient's alarming clinical appearance.

intake. In other words, an infant on a milk diet is on the verge of negative sodium and fluid balance at all times if he is losing 150 to 250 cc. of spinal fluid daily. If then, such an infant develops an infection, vomits frequently, has diarrhea, or does not maintain a normal fluid and salt intake, acute dehydration accompanied by weight loss and profound shock may ensue. If treated soon enough, balance can be restored easily by intravenous infusion of fluids and salt.

In an attempt to obviate the danger of acute dehydration, all of these infants are placed on 2 gm. (1/2 teaspoon) of ordinary table salt per 24 hours after operation and this is continued indefinitely. This is given prophylactically even though it is not actually necessary as long as the child is in good health and eating and drinking well. In addition, all these infants should be offered extra fluids beyond that given with the regular formula. Much more important than the prophylactic use of salt and extra fluids, however, is the realization that critical dehydration may occur within a very few hours when one of these patients becomes ill and refuses his normal fluid intake, particularly if there is vomiting in addition. This problem is discussed with each patient's parents and with the pediatrician before the infant leaves the hospital. Parenteral fluids and salt should be
FIG. 2. Hydrocephalic infants followed from 2 to 4 yr. after arachnoid-ureterostomy. In following legends, (a) refers in each patient to preoperative photograph, (b) to preoperative ventricular air-bubble study and (c) to appearance of infant at period more than 2 yr. after operation.

1. (a) 5 mo. old male.
   (b) AP upright ventricular air-bubble study.
   (c) 3 6/12 yr. postoperative (now 4 3/12 yr. postoperative); normal intelligence, negative neurologic examination.

2. (a) 4 mo. old female.
   (b) AP brow-up ventricular air-bubble study.
   (c) 3 3/12 yr. postoperative (now 3 9/12 yr. postoperative); normal intelligence, negative neurologic examination.

3. (a) 4 mo. old male.
   (b) Lateral upside-down ventricular air-bubble study.
   (c) 3 yr. postoperative (now 3 5/12 yr. postoperative); normal intelligence, negative neurologic examination.

4. (a) 2 1/2 mo. old male.
   (b) AP upright ventricular air-bubble study.
   (c) 2 yr. postoperative (now 3 yr. postoperative); normal intelligence, negative neurologic examination.

5. (a) 5 mo. old female.
   (b) AP upright ventricular air-bubble study.
   (c) 2 6/12 yr. postoperative (now 3 yr. postoperative); probable slight mental retardation, mild left hemiplegia.

6. (a) 7 mo. old male.
   (b) AP upright ventricular air-bubble study.
   (c) 1 5/12 yr. postoperative (now 2 8/12 yr. postoperative); normal intelligence, negative neurologic examination.

7. (a) 1 mo. old male.
   (b) AP upright ventricular air-bubble study.
   (c) 2 2/12 yr. postoperative (now 2 7/12 yr. postoperative); probable normal intelligence; bilateral unsustained ankle clonus only positive neurologic finding.

8. (a) 2 1/2 mo. old female.
   (b) AP brow-up ventricular air-bubble study.
   (c) 2 6/12 yr. postoperative; normal intelligence, coordination poor but improving.
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given at once to any postoperative patient who vomits more than once or twice or fails to drink or appears at all dehydrated on examination of the skin and mucous membranes or estimation of the urine output.

As previously stated, at least 8 patients in this series, and perhaps 9 or 10, have died in this syndrome of acute dehydration secondary to infection. The first two died before the problem could be realized and the others in spite of efforts to impress on the family the seriousness of minor infections in these infants. In reviewing the deaths in those cases in which adequate information is available, it is felt that a fatality might have been avoided by earlier recognition and more vigorous treatment of the fluid balance problem in every instance. In an acute crisis, normal saline and one-half molar sodium lactate solution given intravenously rapidly is recommended. A vein should be exposed and cannulated if necessary since fluid and electrolytes cannot be replaced quickly enough by mouth or by subcutaneous administration in severe dehydration of rapid onset.

There will probably always be an irreducible minimum of critical situations where such a baby is in an isolated community or the family simply cannot be made to understand and cooperate; however, with intelligent, alert parents and astute pediatric care, it should be possible to avoid this complication in almost every instance.

To turn now to a pleasanter and more gratifying aspect of this report. Of the 50 children in this series, 33 are living. The encouraging feature to this author, however, is not that 33 out of 50 children with this severe degree of hydrocephalus are still alive, but that of these 33 only one is grossly retarded to the point of needing institutional care. Five other patients are definitely retarded behind a normal developmental schedule but are happy, asymptomatic children at home making progress. The most interesting feature of all is that 24 of the group are known to be in excellent health with normal or close to normal mental and motor development at periods ranging from a few months to over four years from operation. Three additional patients have not been followed long enough to be at all certain about. So that of the 33 living patients at the present time, 29, and perhaps as many as 31, can be classified as satisfactory to excellent results. On the basis of previous clinical experience, these are all children in whom the degree and rate of progression of hydrocephalus were such that, with the occasional unexplainable exception, they would have been dead or severely retarded with markedly enlarged heads if surgical treatment had not been carried out.

Thirty patients in this series have been followed longer than two years. Twenty of these are alive and as already implied, all but one doing well. A group of these is shown in figure 2, together with their preoperative photographs and ventricular air-bubble studies. This illustrates perhaps better than any verbal description that normal appearance and progressive development is possible with severe degrees of hydrocephalus in early infancy if definitive treatment is accomplished early enough.

SUMMARY

Parents of an infant with hydrocephalus are interested not in having their child kept alive but in giving it a chance for normal development. Therefore, once it is established that progressive hydrocephalus exists, temporizing measures and prolonged observation should be discouraged in favor of a definitive procedure which will immediately and continuously reduce spinal fluid pressure to within normal limits. This series of patients indicates that this objective can be accomplished by diversion of cerebrospinal fluid into the urinary tract. The complications of this procedure are three: (1) mechanical obstruction of the shunt, which has been seen in only a few cases and has always been remedi-
able; (2) meningitis, which has occurred in 8 out of 50 patients postoperatively and has been fatal in 3; and (3) acute dehydration secondary to intercurrent infection because of unreplaced loss of fluid and electrolytes through the shunt, which has been fatal in 8 to 10 of these patients.

Thirty-three out of 50 patients with severe communicating hydrocephalus treated by arachnoid-ureterostomy are living, and of these 31 are satisfactory to excellent results to date.* At least 24 of these children appear to be entirely asymptomatic with normal or close to normal mental and physical development at periods from a few months to over four years.

REFERENCES
5. Personal communication with numerous neurosurgeons about unpublished cases.

SPANISH ABSTRACT
Hidrocefalia Tratada por la Aracnoides-Ureterostomía

No es la vida del niño sino su desarrollo normal lo que interesa a los padres de un niño con hidrocefalia. Por lo tanto no debe realizarse una observación prolongada o practicarse métodos contemporizadores una vez que se ha establecido el diagnóstico de hidrocefalia progresiva, y sí decidirse por un procedimiento definido que reduzca inmediata y continuamente la presión cefalorraquídea a límites normales. Los pacientes de esta serie señalan que puede realizarse este objetivo al derivar el líquido cefalorraquídeo a las vías urinarias.

Las complicaciones de este procedimiento son tres:
1. Obstrucción mecánica de la derivación, observada en pocos casos y que siempre ha sido remediable;
2. Meningitis, que se ha presentado en 8 de los 50 casos en el período postoperatorio, habiendo sido fatal en 3;
3. Deshidratación aguda secundaria a infección intercurrente por no haberse restituido la pérdida de líquidos y electrolitos por la derivación, lo que ha sido fatal en 8 de 10 pacientes.

Actualmente viven 33 de los 50 pacientes y de ellos 31 tienen resultados desde satisfactorios hasta excelentes; los 50 casos presentaban hidrocefalia comunicante intensa y todos fueron tratados con la aracnoides-ureterostomía. 24 de los 31 niños con resultados satisfactorios se encuentran completamente asintomáticos, con desarrollo mental y físico normal o casi normal, en el período de observación postoperatorio de unos cuantos meses a más de cuatro años.

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* This series has been extended to 64 patients, of whom 45 are living and 42 are satisfactory results to date.
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