Hydrocephalus: old treatment, or new or none?

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HYDROCEPHALUS -- OLD TREATMENT, OR NEW, OR NONE?

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INTRODUCTION

The purpose of this paper is to discuss briefly the history and treatment of hydrocephalus, relating the methods used with the dynamics of the condition.

A series of untreated hydrocephalics who were studied exhaustively will be discussed for a comparison with the patients treated.

The varying means of relieving the condition will be discussed, and the outcomes of series of patients so treated will be reviewed.

The methods of treatment will be compared as to efficacy, risk, ultimate prognosis, and lack of complications.

The results will then be summarized.

A conclusion will then be drawn and an opinion rendered.
HISTORY

The discovery of the cerebrospinal fluid is generally ascribed to Cotugno, but the first clear description was provided some fifty years after Cotugno's report by Magendie (1825). One of the disorders of this fluid, hydrocephalus, has been known since Hippocrates, with Vesalius (1514-64) first describing internal hydrocephalus, including dilated ventricles and thinning of the corin substance over them [Russell (24)]. Very little was known of the physiology or pathology of hydrocephalus until the experiments of Dandy and Blackfan (5, 6). They defined two distinct types of hydrocephalus, obstructive or non-communicating, in which the fluid has no egress from the ventricular system because of blockage (mechanical), and non-obstructive or communicating, in which resorption of the fluid from the sub-arachnoid space is impaired. By obstructing the aqueduct of Sylvius, they produced internal hydrocephalus in experimental animals. Then Dandy (7) showed that occlusion of the foramen of Monro led to the distension of the corresponding ventricle. He then cast the first real light on the mechanism of the origin cerebrospinal fluid by demonstrating that if the choroid plexus in the ventricle were removed before occlusion of the foramen, no such distension occurred. The first real proof of the source of the fluid.

He also showed in subsequent experiments that the amount of fluid formed in the ventricles each 24 hours was approximately 800 to 1,000ML.; and that normal circulation time from ventricle to
subarachnoid space is about two to three minutes. He devised third ventriculostomy for the treatment of non-communicating hydrocephalus, thus creating an artificial passage out of the ventricular system to the subarachnoid space (8). He proposed for the treatment of non-obstructive hydrocephalus destruction of the choroid plexuses to reduce the formation of cerebrospinal fluid to an amount which could be handled by a compromised subarachnoid space. These two procedures were the first successful surgical treatments devised and form the framework for all procedures to follow, either directly or indirectly. They will be discussed in detail in the following pages.
NO TREATMENT

For the sake of comparison, the condition both treated and untreated should be evaluated. A complete study of untreated hydrocephalics should ostensibly show:

1. The incidence of spontaneous arrests;
2. The overall mortality associated with the condition; and cause of death;
3. Physical disability associated with the condition;
4. Intellectual ability in survivors;
5. Psychological status of survivors;
6. Any criteria by which good candidates for corrective procedures can be selected.

Such a study has been done.

Laurence (16) studied a series of 239 cases of hydrocephalus which were seen between January, 1938 and December, 1957. Of these cases, 57 patients came to surgery, leaving 182 patients for his study of untreated cases. His study showed:

Survival - At the end of 1958, 81 were alive and well with the hydrocephalus spontaneously arrested. Three were presumed alive, because their condition had arrested and they were doing well when they disappeared. In nine of the children, under five years of age, the condition was still progressive and the remaining 89 cases had died. This made a total of 93 cases living.
Laurence points out, however, that using actuarial methods, one finds that a hydrocephalic child who has survived to three months has a 26% chance of reaching adult life without surgery, while he has a 50% chance of doing so if he is between one and two years. In those who died, the causes were:

Table One [Laurence (16)]

<table>
<thead>
<tr>
<th>CAUSE OF DEATH</th>
<th>Numbers</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hydrocephalus</td>
<td>46</td>
</tr>
<tr>
<td>Acute hydrocephalus</td>
<td>7</td>
</tr>
<tr>
<td>Following investigation</td>
<td></td>
</tr>
<tr>
<td>Infection</td>
<td>16</td>
</tr>
<tr>
<td>Following investigation</td>
<td>4</td>
</tr>
<tr>
<td>Other related</td>
<td>22</td>
</tr>
<tr>
<td>Intraventricular</td>
<td>2</td>
</tr>
<tr>
<td>hemorrhage</td>
<td></td>
</tr>
<tr>
<td>Chest infection</td>
<td>16</td>
</tr>
<tr>
<td>Tumour extension</td>
<td>3</td>
</tr>
<tr>
<td>After operation for</td>
<td>1</td>
</tr>
<tr>
<td>spina bifida</td>
<td></td>
</tr>
<tr>
<td>Unrelated (gastroenteritis)</td>
<td>2</td>
</tr>
<tr>
<td>Uncertain (probably</td>
<td>3</td>
</tr>
<tr>
<td>hydrocephalus)</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>89</td>
</tr>
</tbody>
</table>

Natural Arrest - This was found to be a gradual process if it occurred. If head measurements remained the same for three months, coinciding with a noticeable improvement in the child's general condition, a reduction in fontanelle tension, a return of the orbital axes to normal, and if a commencement of rapid physical as well as mental development showed in the child, natural arrest had very likely occurred. In 47 cases, arrest took place between 5.
nine months and two years. In six cases, earlier arrest was noted, and in 27 cases, the condition smouldered on.

Physical disability of arrested survivors - Twenty-six were judged normal, 16 were slightly handicapped, 23 were severely handicapped, and 16 were incapacitated.

Intellectual Status - Those with normal IQ (85 and over) numbered 31 or 38%. Those of educable subnormality (50-85 IQ) included 28 or 35%. Twenty-two patients were judged uneducable (IQ less than 50), comprising 27%.

Psychological status: Thirty-five percent of the whole sample were found normal, 20% were disturbed, and 45% were unknown.

Criteria for good prognosis with correction: No correlation was found between intelligence, head size, cortical thickness, or duration of progressive disease, [Laurence (14)].

Laurence concluded that surgery should be done in rapidly progressive hydrocephalus, but that caution should be exercised in arrested or arresting cases.

In their study of 468 cases of mental retardation, McIntire and Adams (18) found hydrocephalus a primary diagnosis in 32 or 6.8% of the patients studied. Five clinical diagnoses, of which hydrocephalus ranked fourth, accounted for 45.9% of all cases studied, with 40 other conditions constituting the remaining 54.1%. From his study, Laurence noted that many of the children showed classic brain damage, which he attributed to the original brain insult rather than to cortical thinning.
THE OLD TREATMENT

Dandy's operation for obstructive hydrocephalus, third ventriculostomy, as he first performed it on six patients, required the deliberate section of a healthy optic nerve. He later modified the approach, eliminating this fault. The original six cases he reported in 1922, but it was not until 1945 (9) that he reported on 92 patients operated upon with the modified approach. In this series, operative mortality was 12% and there was arrest of the hydrocephalus in 50% of the cases for periods ranging from six months to 23 years. The average survival time was between seven and eight years. Re-operation was required in 7% of the 92 cases.

Stookey and Scarff (28) modified the Dandy ventriculostomy to a frontal approach and puncture of both the lamina terminalis and floor of the third ventricle. At that time they reported on six patients with one operative death, one failure to arrest, and four patients living and well at the time of report. Length of arrest varied from six months to three years. In 1951, Scarff (25) reported an additional 34 patients operated upon with an operative mortality of 12% and arrest of hydrocephalus in 54% of the patients, with length of arrest (all 54% initial arrest alive at report) varying from two months to fifteen years after operation.

Many more series of ventriculostomies and lamina punctures were reported, mostly showing good results. The results of these series will be shown in a following table.
Dandy's operation for non-obstructive or communicating hydrocephalus was basically sound in principle --- extirpation of the choroid plexuses, his technique was poor [Scarff (27)]. He emptied the third ventricles of fluid in order to expose the choroid tissue and the thinned cortex collapsed, which caused profound shock in his patients. Three of four patients so operated on at this time died, with the fourth living many years afterward.

Davidoff (10) reported a series of choroid plexectomies on 32 infants in which he had 43% operative mortality, 6% failure to arrest hydrocephalus, 50% successful arrest, and a follow-up of two months to four years. Sachs (as reported by Scarff (27) reported a series of 90 cases with 46% operative deaths and 54% operative recoveries.

The technique for plexectomy was improved by Putnam (22) and Scarff (26) who devised endoscopes through which the plexuses could be cauterized without collapse of the ventricular walls and cortex, which cut the operative mortality in their series far below their predecessors. Putnam reported 42 cases, with 25% hospital deaths, failures to arrest in 35%, and successful arrest of hydrocephalus in 40%.

Scarff reported two series of patients --- one in 1942, and one in 1952. In the first series (20 cases), he reported 15% hospital deaths, 35% failure to arrest hydrocephalus, and 50% success in arresting.
Scarff's second series (19 cases), resulted in only one hospital death (5%), failure to arrest in two (10%), and successful arrest of the hydrocephalus in sixteen patients (80%). The survival period in these series runs to 23 years.

Paine and McKissock (21) reported a series of 25 selected non neoplastic cases in which they had 16 successes and nine failures.

A table summary of the cases just discussed will be presented in following pages. The major attributes claimed for these two procedures by Scarff (27) are that they are physiologic in that they avoid the use of foreign substances in the body, and secondly, if arrest is achieved, need for subsequent revisions or secondary operations is minimal.
THE NEW TREATMENT

The "shunt procedures" are so called because they mechanically transport the cerebrospinal fluid from one place to another by means of a tube, usually. Most of the shunts to be discussed originate in the lateral ventricles of the brain. The intracranial spaces to which these shunts have transported fluid have been reported as subdural, transcallosal anterior, transcallosal posterior, chiasmatic cistern, mastoids, and aqueduct of Sylvius. The extracranial spaces to which fluid have been shunted include venous, cardiac, pleural, peritoneal, ureteral, cholecystostomy, salpingostomy, iliostomy, thoracic duct, Stenson's duct, and the epidural space. The tubes employed as shunts have been made from many materials, including venous tissue grafts, rubber, silver, plastics of all kinds, and possibly other materials.

The first of these procedures to be developed since World War II was Torkildsen's ventriculo-cisternostomy, in which a plastic tube is led from the lateral ventricle and skull, and then subcutaneously to the occipital bone and into the cisterna magna. This procedure, naturally, was devised for the relief of obstructive hydrocephalus, only.

Scarff (27) reports the results of the ventriculo-cisternostomy as performed by Torkildsen and other surgeons as 136 total cases, 30% operative mortality, initial arrest of hydrocephalus 58%, and follow-up periods of up to 22 Years.
All other intracranial shunts were reported by Scarff (27) as 118 total cases, 21% operative mortality, 65% success, and follow-ups of two to six years.

Ventriculo-pleural shunts yielded an initially high success rate, but many post-operative complications occurred. The results will be listed in a following table.

A shunt from ventricle to peritoneum was described in 1955 by Scott, Wycis, Murtagh, and Reyes [Scarff (27)] which resulted in 50% success in a 32-patient series. There were 10% hospital deaths. Later in 1955, Jackson and Snodgrass (13) reported 62 patients upon whom they performed 62 ventriculo-peritoneal and 50 lumbar-subarachnoid shunts. There was no operative mortality, but failure to arrest the hydrocephalus occurred in 44%, and 24 patients were alive one to four years after operation with arrest in 17 or 30% of the total. Other peritoneal series will be shown in a following table.

Shunts into epithelialized ducts did not show either enough cases or long enough follow-up times to be of any significance. These were the cases mentioned earlier which were shunts into the bile duct, salivary duct, fallopian tube, ileum, and thoracic duct.

Matson, in 1951, described a procedure in which a tube was led out of the ventricle, sub-cutaneously to the peri-renal area, through the abdominal wall, and into a ureter whose kidney had been sacrificed. This operation was designed for treatment of obstructive hydrocephalus. In seven cases treated this way, he reported an operative mortality of 11.
15% and apparent initial success of 85% (Scarff, 27). Later, he reported a similar but modified treatment for communicating hydrocephalus in which the proximal end of the shunt ran from the subarachnoid space to a ureter whose kidney had been sacrificed. His series of patients as reported in the New England Journal of Medicine, Nov., 1956, amounted to 108 patients. His operative mortality was only one percent with arrest of the hydrocephalus in 70 cases or 65% of the cases followed from three months to seven years after surgery. These procedures, however, carried two quite distinct and obvious disadvantages in them; first, a healthy kidney had to be sacrificed, and second, all the normal constituents of cerebrospinal, fluid such as electrolytes and proteins, were constantly draining to the outside of the body through the urinary tract and were lost.

Forrest, Laurence, and Macnab (11) tried shunting the fluid by ventriculo-subdural flow. They did the procedure on 70 cases in which 36 arrested, (52%), three progressed (4%), and 31 died (44%). These same investigators (12) devised a dye test for predicting successful outcome of their procedure. They injected a given amount of dye into the subdural space. If greater than 50% of the dye was excreted in six hours, it indicated a favorable prognosis.

A procedure by which fluid was shunted from the lateral ventricle to the right atrium of the heart via the superior vena cava was described in 1952 by Nulsen and Spitz - (Surg. Forum., 1951,2,399). This technique required a special valve which was designed by
Mr. John Holter. This valve is interposed in the course of the shunt tube, and is designed in such a way that fluid can pass from the ventricle of the brain into the atrium of the heart with no reflux from heart to ventricle. Nulsen operated on 70 patients, using the ventriculo-atrial shunt procedure and the Holter valve. The mortality rate was 0%. Approximately 70% were still alive in 1961, with survival times from six months to five years.

Dr. Spitz, in a communication with G. H. Macnab, stated that he had personally operated on 212 cases with 96% success. But he has not published any results since the initial case with Nulsen.

In 1960, Sayers (Scarff, 27) reported 156 cases in which he used the Holter valve and Spitz-Nulsen technique. His operative mortality was 18% and success rate 63% of the cases.

Carrington (4) reported 50 cases using the same technique. He achieved a success rate was 68% and mortality was 6%.

Another type of valve for use in ventriculo-atrial shunts was devised in 1957 by Border and Heyer. It was first used by Pudenz, Russell, Hurd, and Shelden on 15 patients in whom they achieved a success rate of 60% with no operative deaths.

Anderson (1) reported in 1959 that using the Heyer valve and Pudenz technique, he treated 48 cases of hydrocephalus. He showed an operative mortality rate of 6% and achieved arrest in 58% of his cases. His longest follow-up was two years.

Anderson, from his experiences, commented on ventriculo-atrial
shunts as follows:

1. The technique, while painstaking, is not difficult, and with experience, takes an hour with minimal blood loss.

2. Even in the young infant, the structures involved are large, making the procedure mechanically more easily done.

3. It is suitable for any type of hydrocephalus.

4. There is no loss of electrolytes, protein, or fluid from the body.

5. Reoperation, if necessary, is usually easy.

The complications specifically reported in all the cases will be shown in following tables, but two will be discussed later in more detail, because they are the most common and have been studied. These are septicemia and blockage of the atrial end of the shunt by clotting or thrombosis of the superior vena cava.
THE TWO MAJOR COMPLICATIONS AND THEIR POSSIBLE SOLUTIONS

Nulsen (20), in his series of 70 patients treated with Spitz-Holter ventriculo-atrial shunts, analyzed the causes both of revisions and failures.

Nineteen of the secondary operations in the 40 survivors were for venous recanalizations following jugular or caval occlusion. He notes that none were necessary if the catheter was properly placed in the right atrium, below the T-4 level. He has had to reoperate only very small infants whose growth pulled the catheter tip above this level.

Eighteen secondary operations were done because bacteremia developed. In the twelve patients in this group, it was noted in every case that the catheter tip was below the level of T-6 at the time endocarditis developed.

Nulsen concludes that proper placement of the cardiac end of the valve in the mid-atrium will obviate many of the possible complications. Radiographically, he considers this between T-4 and mid or high T-6. Other authors have agreed that the catheter ending at a point of most turbulent blood flow will result in less clot-formation and occlusion.

Dr. Spitz communicated with Macnab (17) in 1961 that he recently had tried to place the catheter in the inferior vena cava to lessen the possibility of clotting and blockage of the atrial end of the shunt as well as to allow for subsequent growth of the patient.
The means by which he placed the end of the catheter was the innovation. He ran a bent metal stylus the length of the caval catheter and used it as an EKG lead, noting that as the right atrium was entered, an enlarged P wave was recorded. As the inferior vena cava was entered, the P wave returned to normal.

Robertson, et al, (23) reported the same year that they had used a saline-filled catheter for the electrode and it had worked as an EKG lead.

Further work was done in 1962, when McLaurin et al (19) analyzed closely the specific EKG changes in 52 operations performed on 39 patients over an 18 month period. They checked the results with the radiographic findings in each case, with consistently good correlation in every case.

Their tracings, invariably the same in every case showed the following criteria by which the progress of the catheter can be traced electrocardiographically:

1. In the superior vena cava, inverted P waves show.

2. At the junction of superior vena cava and right atrium, P waves show the largest negative deflection.

3. Similar inverted and large P waves are also recorded in upper fourth of the right atrium.

4. At the desired point, the middle of the right atrium, an initial positive deflection appears so the P wave becomes biphasic.
### Table II -- Scarff (27)

**Comparison of Operative Results in the Treatment of Hydrocephalus**

<table>
<thead>
<tr>
<th>Type of Operation</th>
<th>No. of Cases</th>
<th>Initial Results</th>
<th>Total % Late Complications</th>
<th>Follow-Up Periods (Yr.)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Operative Mortality</td>
<td>Arrest of Hydrocephalus</td>
<td></td>
</tr>
<tr>
<td>OPERATIONS NOT REQUIRING MECHANICAL TUBES OR VALVES</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Third Ventriculostomy</td>
<td>529</td>
<td>15</td>
<td>70</td>
<td>2</td>
</tr>
<tr>
<td>Cauterization</td>
<td>91</td>
<td>15</td>
<td>60</td>
<td>3</td>
</tr>
<tr>
<td>Summary</td>
<td>620</td>
<td>15</td>
<td>65</td>
<td>2.5</td>
</tr>
<tr>
<td>OPERATIONS REQUIRING MECHANICAL TUBES AND VALVES (THE SHUNTS)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Intracranial Shunt (Torkildsen)</td>
<td>136</td>
<td>30</td>
<td>58</td>
<td>50</td>
</tr>
<tr>
<td>Intracranial Shunts (Other Authors')</td>
<td>118</td>
<td>21</td>
<td>60</td>
<td>--</td>
</tr>
<tr>
<td>Cardiac Shunts</td>
<td>345</td>
<td>6</td>
<td>62</td>
<td>46</td>
</tr>
<tr>
<td>Pleural Shunts</td>
<td>108</td>
<td>8</td>
<td>53</td>
<td>100</td>
</tr>
<tr>
<td>Peritoneal Shunts</td>
<td>230</td>
<td>13</td>
<td>55</td>
<td>58</td>
</tr>
<tr>
<td>Ureteral Shunts</td>
<td>108</td>
<td>1</td>
<td>65</td>
<td>44</td>
</tr>
<tr>
<td>Shunts into Epithelialized Ducts</td>
<td>29</td>
<td>6</td>
<td>50</td>
<td>.50</td>
</tr>
<tr>
<td>TYPE OF OPERATION</td>
<td>NO. OF CASES</td>
<td>INITIAL RESULTS</td>
<td>TOTAL % LATE COMPLICATIONS</td>
<td>FOLLOW-UP PERIODS (YR.)</td>
</tr>
<tr>
<td>-------------------</td>
<td>--------------</td>
<td>-----------------</td>
<td>---------------------------</td>
<td>-------------------------</td>
</tr>
<tr>
<td>OPERATIONS REQUIRING MECHANICAL TUBES AND VALVES (THE SHUNTS)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>SPINAL SUBARACHNOID TO SPINAL EPIDURAL SHUNTS</td>
<td>13</td>
<td>0</td>
<td>75</td>
<td>50</td>
</tr>
<tr>
<td>SUMMARY</td>
<td>1,087</td>
<td>10</td>
<td>60</td>
<td>57</td>
</tr>
</tbody>
</table>

1ESTIMATED.
5. As the junction of the inferior vena cava and lower right atrium are neared positive deflection is greater and the P wave is wholly upright in this position.

6. In the region of the tricuspid valve the P wave is upright, with about the same amplitude and contour of standard leads.

The immediate advantages of this means of placing the catheter are obvious. Accuracy of position are positively obtainable, time is saved from that consumed in the radiographic technique, some possibilities of infection through the use of the X-ray equipment are obviated, and the operator, patient, and operating personnel are not exposed to radiation. The long-range advantages might well be the lessening of number of revisions needed to secure a working shunt and the elimination of clotting, thrombus, and infection described earlier.
SUMMARY

In the untreated cases, only 93 of 182 cases were alive when the study was closed. Of these, only 38% were of normal intelligence.

In the series of patients in the Old Treatment section, a total of 527 third ventriculostomies were studied. The operative mortality was 15% and initial successes were 70%. Choroid plexectomy presented 91 cases with an operative mortality of 15% and successes claimed 65%. The New Treatment section presented 136 cases of ventriculocisternostomies with an operative mortality rate of 30% and an initial arrest of hydrocephalus in 58%. The 345 cases presented as ventriculo-atrial shunts showed operative mortality of 6%, with 66% arrest of hydrocephalus. Due to failure or complications, there were 698 operations in these 345 cases.

The major problems which caused a great many of these repeat operations are discussed in The Two Major Complications and Their Possible Solutions section.
CONCLUSION

As this paper took shape, the writer could not help but feel the force of dedication in each man whose procedure or series of patients were studied. Hydrocephalus, if one examines the ultimate survivor tables, or is faced with it clinically, is a saddening, disheartening experience.

The major work of Scarff asks very loudly the question, "Why isn't everyone doing it the old proven way?" And at present, this writer, going by the statistics, would have a difficult time explaining the inquiry away.

But taking into consideration the fact that even the great Dandy failed miserably at first in his own procedure, and the fact that the techniques, materials, and depth of experience in using the shunt are building and growing all the time, it is at least an even race between the old and new.

Much more work must be done along the lines of the magnificent efforts of Laurence -- a carefully worked out study of those children who have been treated by any procedure successfully, to see which offers the most to them -- the hydrocephalic children. They are the most important factor and every idea should grow out of a desire to do the best for them.

19.


12. _____, et al, (1957) Lancet 2, 827 Ventriculo Subdural Drainage in Infantile Hydrocephalus, Selection of Patients by Subdural Dye Test

20.


Treatment of Obstructive Hydrocephalus by Puncture of 
The Lamina Terminalis and Floor of The Third Ventricle

Hydrocephalus : Treatment by Endoscopic Cauterization 
of the Choroid Plexus, Long Term Results

27. _____ (1963) J. Neurol., Neurosurg., and Psychiat., 26,1 
Treatment of Hydrocephalus: An Historical and Critical 
Review of Methods and Results

28. Stookey, B., and Scarff, J.E.,(1936) Bull Neurol. Institute, 
N.Y., 5, 348 Occlusion of the aqueduct of Sylvius by 
Neoplastic and Non-Neoplastic processes With A Rational 
Surgical Treatment for Relief of The Resultant Obstruc-
tive Hydrocephalus