DR. HATCHER: But the whole point of our discussion is that the best for an individual baby and his family may be to let him die within a month.

DR. SLATER: This seems so clear that I'm delighted to hear it said outright.

DR. HATCHER: Many of the problems we have raised would be solved by the early involvement of a good and compassionate paediatrician.

References

1 Slater, E., *British Medical Journal*, 1971, 4, 734.
2 Eckstein, H. B., London Medical Group Symposium on Perinatal Paediatrics in Relation to Ethics and Moral Problems, February, 1973.
3 Lightowler, C. D. R., *British Medical Journal*, 1971, 2, 385.
4 McLachlan, G., (Ed.), *Patient, Doctor, and Society*. London, Oxford University Press, 1972.
5 Department of Health and Social Security, *Care of the Child with Spina Bifida*. London, H.M.S.O., 1973.
6 Lorber, J., *Developmental Medicine and Child Neurology*, 1971, 13, 279.

Hospital Topics

Ventriculoperitoneal Shunting for Hydrocephalus

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British Medical Journal, 1973, 2, 289–292

Summary

Revision rate and complications were reviewed in 297 patients of all ages subjected to ventriculoperitoneal shunting and adequately followed. Both operative mortality and deaths due to complications of this type of shunt were much lower than after ventriculocaval shunts. Although 44% required revision a third of the cases had a period of three years without revision; two-thirds of these went for three years after the initial operation without need for operation. This shunt should be considered both in children and in adults because it is no more likely to block than ventriculoatrial, is easier to revise, and the other complications are fewer and much less serious.

Introduction

The introduction of various shunting and bypassing operations has transformed the management of hydrocephalus. As the variety of sites for drainage and the number of devices for implantation have increased there have inevitably been competing claims for various techniques. Hydrocephalus, even in fancy, is of heterogeneous origin and the morbidity and mortality may depend more on the cause of the hydrocephalus than on the particular technique employed. Another difficulty in comparing different procedures, particularly in respect of revision rate and complications, is the failure of many authors to quote the duration of follow-up in meaningful terms; a range of follow-up duration is of little use, and an *average* follow-up is almost valueless. Moreover, the population at risk is seldom known because no adjustment is made for early deaths.

Ventriculoatrial shunting was introduced in 1952 and has been widely adopted as the standard treatment for hydrocephalus. All reported series, however, show a high postoperative morbidity and it is necessary to set aside a large number of paediatric surgical beds to deal with complications of this operation. The ventriculoperitoneal shunt has been used intermittently since it was first introduced in 1905,[1] but this is commonly dismissed because of its supposed high failure rate from blockage.[2] Nevertheless, this shunt does not produce the serious and often fatal cardiopulmonary complications and septicaemia which occur in every sizeable series of ventriculoatrial operations. There has therefore been a revival of interest in the ventriculoperitoneal method,[3 4] and we decided to look critically at the efficacy of this procedure in this Institute in order to discover whether its bad reputation was deserved.

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Methods

Operative Technique.—A burr hole is made 2 cm above and 1 cm behind the tip of the right pinna; this allows the tube to lie along the length of the ventricle for some distance. The lower margin of the burr hole is nibbled away and the dura is opened by a single vertical slit. Small-bore silicone tubing with one or two side-holes is introduced well into the lateral ventricle and secured by a single flange to the dura. The tubing is led subcutaneously to the abdominal wall towards the right iliac fossa; from there it is taken subcutaneously in a wide curve to a right upper paramedian muscle slide incision whose upper end curves into the xiphoid notch. The tubing is then taken through a stab in the rectus muscle and a small peritoneal incision to the right side of the falciform ligament so as to lie over the upper surface of the liver below the diaphragm. A flange may be used to secure the tubing to the peritoneum. No valves or connecting junctions are used.

Follow-up Procedure.—This report is concerned with the frequency with which revision was required and with other complications. It was not our practice to undertake elective revision but to revise the shunt only if symptoms suggestive of blockage developed. Only patients known to have survived for definite periods were analysed for this purpose, but in them it was decided to consider periods free of complications as indicative of a successful shunt. It is, of course, possible that in some of these cases hydrocephalus became spontaneously arrested. An attempt was made to review all 404 patients subjected to ventriculoperitoneal drainage between 1955 and 1968. However, no postoperative information could

be obtained on 107 patients, and these were therefore excluded. This report thus deals with the 297 patients on whom adequate information was available.

Results

Causes of Hydrocephalus.—The majority of cases in most published series are infants with spina bifida or other malformations. In this series 131 (44·1%) of the patients were aged under 1 year, 77 (25·9%) were aged 1 to 15 years, and 89 (30·0%) were aged over 15 years, and meningitis and tumour were each as common a cause as myelomeningocele (table I). Total mortality was about 50% of those traced, and this was similar for all age groups. It was highest in those with tumours, and it is clearly important to take account of the diagnosis when considering the overall mortality, which is more likely to reflect the nature of the primary disorder than the method of treatment.

TABLE I—*Cause of Hydrocephalus*

	Proportion of Patients	
	No.	%
Postmeningitic	64	21·5
Tumours	62	20·9
Myelomeningocele/Arnold-Chiari	55	18·5
Aqueduct stenosis	27	9·0
Various	89	30·0
Total	297	100·0

Operative Mortality.—Operation accounted for only 14 (9·9%) of the 142 deaths. Of these 14 patients 8 died of unrelieved (or recurrent) raised intracranial pressure, 5 died of postoperative infection, and the remaining patient collapsed and died during the course of a revision operation. This gives a case mortality of 4·7%, but when the original and revision operations are considered together it is 2·1% of 674 operations on the 297 patients (table II).

TABLE II—*Operative Mortality*

Cause of Death	No. of Cases	As Percentage of:		
		297 Patients	674 Operations	142 Deaths
Postoperative infection	5	1·7	0·7	3·5
Blocked shunt: acute pressure	8	2·7	1·2	5·6
Death on table	1	0·3	0·1	0·1
Total	14	4·7	2·1	9·9

Operative Morbidity.—The overall infection rate was 14·1% (table III); it was twice as great (19·7%) in cases that ultimately proved fatal as in the survivors (9·0%). Septicaemia was never encountered, and many of the infections were confined to the subcutaneous track. Fatal postoperative infection occurred in only 0·7% of operations and 1·7% of patients. No case of peritonitis resulted from an infected shunt, and one child who died after three days of peritonitis as a consequence of a ruptured appendix had no sign at necropsy of intracranial infection although the shunt was patent. Bowel perforation was not seen but abdominal pseudocyst was encountered once in this series. One subdural haematoma and three subdural hygromas were observed, the latter probably due to cerebrospinal fluid running alongside a blocked ventricular tube into the subdural space.

TABLE III—*Postoperative Infection Rates*

	Survivors (n = 155)	Fatal Cases (n = 142)	Total (n = 297)
No. (%) infected	14 (9·0)	28 (19·7)	42 (14·1)

Revision.—Of the 297 patients 132 (44·4%) required revision, but if those lost to follow-up within a month of operation are excluded the rate goes up to 50·8% (Table IV). The overall revision rate for fatal cases was lower, but this was because of the high mortality in the first month, itself the result of rapidly progressive conditions. In these circumstances even if blockage is suspected it may not be considered worth while to reoperate. In those who survived more than a month but eventually died the revision rate was appreciably higher than in long-term survivors. This shows the importance of basing the revision date only on patients with adequate survival and follow-up. This is more dramatically shown for the series as a whole when the revision rate is calculated on the assumption that the 107 patients about whom there was no postoperative information neither died nor required revision; instead of 50·8% the revision rate becomes 32·7%. In a third of the patients requiring revision more than two operations after the original one were needed.

TABLE IV—*Revision Rate*

Duration of Follow-up	No. of Cases	Proportion Revised	
		No.	%
>1 Month	250	127	50·8
>3 Months	208	109	52·4
>1 Year	162	84	51·9
>3 Years	116	57	49·1
All cases	297	132	44·4

This multiple revision rate remained similar in various follow-up groups (table V). More of the fatal cases had required revision, and in them revision was more often required repeatedly than in the non-fatal cases with revisions.

TABLE V—*Rate for More than Two Revisions at Different Follow-up Periods*

Duration of Follow-up	Proportion of Whole Series Followed-up		Revised Cases	
	Total No. of Cases	No. (%) with >2 Revisions	Total No. of Cases	No. (%) with >2 Revisions
>1 Month	250	43 (17·2)	127	43 (33·9)
>3 Months	208	37 (17·8)	109	37 (33·9)
>1 Year	162	29 (17·9)	84	29 (35·4)
>3 Years	116	17 (14·7)	57	17 (29·8)
All cases	297	43 (14·5)	132	43 (32·6)

Time of Revision.—More than 80% of the patients who required revision had their first revision within the first year, and 40·9% within a month of the original operation. Out of 162 patients followed for more than a year 99 (61·1%) had not had a revision within the first year; 21·2% of these 99 subsequently needed revision. Out of 116 followed for more than three years 66 (56·9% had gone three years from the original operation without revision; 7 (10·6%) eventually required revision.

Revision-free Periods.—A third of the whole series had a three-year period without revision, and two-thirds of these went for three years from their original operation without complications If those who died or were lost to follow-up in the first three months are excluded almost half of the patients had a three-year remission (table VI). Infants more often needed revision within the first month, but thereafter age had no influence on long remission, suggesting that body growth is not a crucial factor in determining the need for revision when this type of shunt is used (table VII).

Site of Blockage.—Half of the revision operations were for blockage at the abdominal end of the shunt only; 13% were

TABLE VI—*Revision-free Periods at Follow-ups of More than Three Months, More than One Year, and More than Three Years*

	No. of Cases	Total (% of 297)	>3 Months (% of 208)	>1 Year (% of 162)	>3 Years (% of 116)
No revision in first year ..	99	33·3	47·0	61·1	85·3
No revision in first 3 years	66	22·2	31·7	40·7	56·9
At least one 3-year period without revision	97	32·7	46·6	59·9	83·6

TABLE VII—*Effect of Age on Revision Rate*

	Age at Operation				P
	<1 Year (n=131)		>1 Year (n=166)		
	No.	%	No.	%	
Revision within one month	37	28·2	17	10·2	<0·001
One revision-free period of 3 years	45	34·4	52	31·3	Not significant

for ventricular replacements, and in 37% the whole drain was revised. Abdominal revisions were more often repeated than ventricular.

Discussion

This report is concerned with the narrow issue of the safety and efficacy of ventriculoperitoneal drainage as a technical procedure. In a heterogeneous group of patients, in respect of age and primary disease, it is not appropriate to look to the quality of survival or even its duration as an indication of the effectiveness of the drainage procedure. Indeed it is doubtful how appropriate such measures are, even with infantile hydrocephalus, because survival seems more likely to reflect the total management (including the timing of surgery or elective revision), and the drainage procedure itself is only one component of this. Comparison is therefore confined to revision rates and complications (table VIII).

Scarff[2] undertook an exhaustive review of the treatment of hydrocephalus up to 1963. In aggregating large numbers of reported cases from different sources he was inevitably reduced to quoting maximum and estimated average follow-up periods, and his figures must be interpreted with this limitation in mind. Comparison with ventriculoatrial (ventriculojugular) shunts only is attempted, because the wide acceptance of this technique in the past 15 years or so makes it the standard against which other procedure will be judged in most centres.

An overall complication rate for ventriculoatrial shunts is almost impossible to derive owing to the variety of ways in which reports are written. In two large series, however, *valve complications* were the cause of 56% and 69% of deaths respectively.[5,6] *Infection rates* mostly lie between 10 and 20%; in Forrest and Cooper's[5] large series 11% of the patients died of septicaemia, and in the smaller one of Merrill *et al.*[7] 31% died of infection within one year. In Scarff's aggregate series septicaemia or meningitis occurred in 12%, but in some reports infection occurred in over 20%. The duration of follow-up is closely associated with the infection rate and with the passage of time any population of shunt-dependent patients will have an increasing case infection rate.[8] *Cardiopulmonary* complications occurred in every series of ventriculoatrial shunts; Scarff's overall figure in 345 collected cases was 6%. A study of 65 cases at necropsy showed 57% with some thromboembolic complication, but relatively few showed evidence of pulmonary hypertension.[9] The *revision rate* quoted by Scarff for his combined series was 28%, with an average follow-up of one and a half years; most reports, however, gave higher rates (42% in 9 months, 61% in 1 year, or 77% in 1-6 years). It is not clear whether these were revisions after block only or whether they included planned prophylactic revision. Because of the difficulty of revising venous shunts once thrombosis has occurred some reports have recommended frequent planned revisions, even monthly or six monthly at first in infantile cases.[10]

In our hands the ventriculoperitoneal technique has a very much lower mortality rate attributable to the shunt itself. The overall infection rate is comparable with the best ventriculoatrial series, but there are no instances of septicaemia. Cardiopulmonary complications do not occur whatever the underlying disease. The question, then, is whether the revision rate is unacceptably high, so that ventriculoatrial shunt might still be justifiable in spite of its attendant mortality and morbidity. Because our revision rates refer only to patients who survived and were traced the figures are biased to the high side; it is likely that in many cases no news is good news, but we have not made this assumption. That more than 20% had no revision in the first three years and more than 30% had at least one three-year period without revision is encouraging, although comparable figures cannot be derived from published reports on ventriculoatrial shunting. What is certain is that revision of a ventriculoperitoneal shunt is very much easier to do; indeed it is the difficulty, sometimes the impossibility, of re-establishing a blocked lower end of a ventriculoatrial shunt which makes some clinicians advocate planned revision, which inevitably increases the rate.

One significant advantage of ventriculoperitoneal drainage is that a valve is not required. The valve itself is commonly the site of blockage, and septicaemia can result from colonization of the valve, which must then be removed; a valve also entails junctions, with the risk of disconnexion. In spite of a valve most series of ventriculoatrial shunts include a number of subdural haematomas. Intraperitoneal pressure is above atmospheric and is therefore higher than intrathoracic; even in the erect posture there is likely to be much less negative pressure draining the ventricle than after ventriculoatrial shunting. This may account for the low incidence of subdural haematoma even though no valve is used.

References

1 Kausch, W., *Archiv für klinische Chirurgie*, 1908, **87**, 709.
2 Scarff, J. E., *Journal of Neurology, Neurosurgery and Psychiatry*, 1963, **26**, 1.

TABLE VIII—*Rates of Infection and Revision in Some Reported Series of Ventriculoaterial shunts*

Authors and Year	No. of Cases	Infection Rate	Revision Rate	Duration of Follow-up
Jackson and Snodgrass (1955)[11]	62		61%	1 Year
Scarff (1963)[8]	345	12%	28%	1½ Years (average)
Foltz and Shurtleff (1963)[12]	65	22%	42%	9 Months
Yashon and Sugar (1964)[13]	31		21% (26 Valve removals)	6-24 Months
Friedman *et al.* (1964)[9]	65		45%	
Merrill *et al.* (1965)[7]	42	31%	88%	1 Year
Overton and Snodgrass (1965)[14]	48	15·6%	40%	>10 Months
Forrest and Cooper (1968)[5]	455	20% (11% septicaemia)	77%	1-6 Years
Guthkelch (1967)[6]	166		27%	
Becker and Nulsen (1968)[10]	64	7%	207 ops. on 64 patients	
Shurtleff *et al.* (1971)[8]	102	14%	61% (299 ops. on 102 patients)	12 Years
Illingworth *et al.* (1971)[15]	99	9%	46% (Mean)	

[3] Ames, R. H., *Journal of Neurosurgery*, 1967, **27**, 525.
[4] Weiss, S. R., and Raskind, R., *International Surgery*, 1969, **51**, 13.
[5] Forrest, D. M., and Cooper, D. G. W., *Journal of Neurosurgery*, 1968, **29**, 506.
[6] Guthkelch, A. N., *British Journal of Surgery*, 1967, **54**, 665.
[7] Merrill, R. E., McCutchen, T., Meacham, W.'F., and Carter, T., *Journal of the American Medical Association*, 1965, **191**, 21.
[8] Shurtleff, D. B., Christie, D., and Foltz, E. L., *Journal of Neurosurgery*, 1971, **35**, 686.
[9] Friedman, S., Zita-Gozum, C., and Chatten, J., *Journal of Pediatrics*, 1964, **64**, 305.
[10] Becker, D. P., and Nulsen, F. E., *Journal of Neurosurgery*, 1968, **28**, 215.
[11] Jackson, I. J., and Snodgrass, S. R., *Journal of Neurosurgery*, 1955, **12**, 216.
[12] Foltz, E. L., and Shurtleff, D. B., *Journal of Neurosurgery*, 1963, **20**, 1064.
[13] Yashon, D., and Sugar, O., *Archives of Disease in Childhood*, 1964, **39**, 58.
[14] Overton, M. C., and Snodgrass, S. R., *Journal of Neurosurgery*, 1965, **23**, 517.
[15] Illingworth, R. D., Logue, V., Symon, L., and Uemura, K., *Journal of Neurosurgery*, 1971, **35**, 681.

Occasional Survey

Multiple Sclerosis: A Review

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British Medical Journal, 1973, 2, 292–295

While basic research on multiple sclerosis has underlined the complex nature of its immunological background the clinician, and the neurologist in particular, has become aware of a lack of unanimity on the subject of diagnostic criteria and of its effect on the validity of recent studies on the course of this ubiquitous disease. With the general practitioner especially in mind an attempt is made here to summarise certain aspects of a disease whose origins and behaviour continue to defy clinician, epidemiologist, and laboratory worker alike.

The relative role of heredity and environment is a bone of contention in a number of diseases whose aetiology is obscure, none more so than multiple sclerosis. Certain genetic data are beyond dispute—the risk to a first-degree relative is many times greater than to the general population; the closer the relationship the higher the risk; high-risk families exist.[1] As yet no definite genetic pattern has been proved.

Turning to the role of environment, attention is at present focused on the possibility that the primary cause is to be sought in an infection occurring in childhood followed by a latent interval of years.[2-5] From this premise arise two questions: firstly, what may be the frequency of this specific neural sensitization in a population exposed to the appropriate type of infection? secondly, what could be the incidence of multiple sclerosis among sensitized individuals? Since the antigen or antigens responsible for the immunological changes in the central nervous system have not yet been identified[6] both suppositions are at present of academic interest only. The second query does, however, raise two further propositions, the first in part hypothetical and the second factual—that in some subjects the disease may remain latent or "silent" during life[7-8] or until one or more environmental factors "trigger off" the initial symptoms; and the fact that a benign form of the disease does exist, proved by clinical observation and supported by Bornstein's concept of "reversibility" of the early lesion based on his study of demyelination in tissue culture models.[9]

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Precipitating and Aggravating Factors

The following briefly summarizes present knowledge.[10] (1) Infection is an unusual precursor to the onset, except in children, but it is not uncommonly related to relapse or progression. (2) Trauma is now recognized as an occasional precipitant, the strongest evidence coming from its localizing effect on the initial symptoms—for example, injury to a hand causing sensory disturbance at the same site a few hours or days later. (3) Pregnancy, the subject of special study, with results suggesting that the risk is maximal during the puerperium. Measures to lessen fatigue and emotional stress during this three-month period are thus clearly indicated. (4) Abnormal reaction to foreign substances covers a wide field, including certain foods, drugs, vaccination, and inoculation, particularly in subjects with an allergic history. (5) Emotional stress, difficult of statistical proof but whose relation to onset symptoms in some patients is clear, while being generally recognized as a common cause of temporary exacerbation of existing symptoms. (6) Exertion and fatigue may bring to light the first hint of the disease—for example, blurred vision after exercise, indicative of early optic neuritis. (7) Changes in temperature are of theoretical as well as of practical interest. Symptoms and signs—for example, blurring of vision, paraesthesiae, and extensor plantar response—may appear for the first time or be aggravated by a hot bath (see below).

Clinical Picture

Certain pathological features have a bearing on symptomatology: (1) a predilection for certain sites—for example, periventricular region, optic pathways, brain stem, and cervical portion of the spinal cord; (2) a tendency towards symmetry of plaques; (3) diffusion or spread of the early plaque, described by Lumsden[6] as "a radial spread outwards from venules" and "by coalescence of contiguous sleeves."

"Symptoms and signs"; "Signs and Symptoms." The order in which these nouns are placed reflects a distinct and important difference in the approach to diagnosis. For too long dogmatic insistence on the presence of certain signs has hampered recognition of the disease in its early stage. Furthermore, this attitude of mind, while tending to exclude cases running a benign course, has also cast doubt on the validity of many studies on the age of onset, character of presenting symptoms, and course of the disease.[10]