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Primary ventriculo-peritoneal shunts in treatment of hydrocephalus associated with myelomeningocele

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Stark, G. D., Drummond, M. B., Poneprasert, S., and Robarts, F. H. (1974). *Archives of Disease in Childhood*, **49**, 112. **Primary ventriculo-peritoneal shunts in treatment of hydrocephalus associated with myelomeningocele.** In 50 children suffering from open myelomeningocele, a ventriculo-peritoneal shunt was employed as the primary procedure for control of hydrocephalus. In 11 cases the system was ultimately replaced by a ventriculo-atrial shunt, though this may not always have been necessary. In the remaining 39 patients satisfactory control of hydrocephalus was achieved. Obstruction, particularly of the peritoneal catheter, occurred frequently in the first 6 months but no revisions were required after 2 years. It is concluded that while ventriculo-peritoneal and ventriculo-atrial shunts achieve comparable success in controlling hydrocephalus, the former, due to the relatively innocuous complications, offers significant long-term advantages.

In most centres the standard procedure for the control of hydrocephalus associated with myelomeningocele is the insertion of a ventriculo-atrial shunt. Other procedures tend to be used only when the ventriculo-atrial shunt has failed. There are few reports of the ventriculo-peritoneal shunt being used as a primary procedure, and those dealt with hydrocephalus of mixed age and aetiology (Villani, Paoletti, and Gaini, 1971; Weiss and Raskind, 1969; Murtagh and Lehman, 1967; Ames, 1967). This report documents the results of the ventriculo-peritoneal shunt as a primary procedure in children suffering from hydrocephalus associated with myelomeningocele.

Patients and methods

The series comprises 50 patients (22 male, 28 female) suffering from hydrocephalus associated with open myelomeningocele and includes all such children treated in the Spina Bifida Unit of the Royal Hospital for Sick Children, Edinburgh, by the insertion of a primary ventriculo-peritoneal shunt between January 1965 and December 1969, irrespective of the age at insertion. With the exception of one child who required insertion of his first shunt at age 9 years, several weeks after a late repair of his back lesion, all shunts were inserted within the first year of life, the majority (82%) within the first 3 months.

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Since the unit opened in 1965, a policy of selection of infants for early back closure has been adopted, depending on the size and site of the lesion, the severity of lower limb paralysis and hydrocephalus, and the presence of other congenital abnormalities or severe birth injury (Stark, 1971, Stark and Drummond, 1973). In the exceptional case of infants who survive despite nonclosure of the back lesion, hydrocephalus is treated if necessary by insertion of a shunt. Late repair of the back lesion may subsequently be carried out for cosmetic reasons. The majority of infants with unrepaired lesions do not, however, survive, the one-year survival for operated and unoperated cases being 77 and 12%, respectively.

Of the patients in the present series, 39 had early operative closure of the back lesion, all within 58 hours of birth, the majority (90%) within 24 hours. In 5 others the lesion was repaired at ages ranging from 8 weeks to 9 years. In 6 patients no repair was carried out.

Air ventriculography was carried out in 44 patients before insertion of the shunt. In these cases the mean thickness of cerebral mantle measured from the roof of the lateral ventricle to the skull vertex was 21·4 mm ± SD 4·2 mm.

In 22 patients the primary CSF shunt was preceded by insertion of a ventriculostomy reservoir (Rickham, 1964), usually on account of ventriculitis, an unhealed back wound, or the presence of blood in the CSF. In 7 of these patients the primary shunt was connected to the reservoir which, in the remainder, was removed. The technique of the ventriculo-peritoneal operation was based on that of Murtagh and Lehman (1967). In 41 children a Pudenz flushing device was attached to the ventricular catheter: in 9, including all of those in whom the reservoir was retained, a Spitz-Holter valve was used. In all cases the silastic distal catheter terminated in a Pudenz-Heyer slit valve and radio-opaque tip that was placed either over the bare area of the liver or deep in the pelvis.

All patients were followed up regularly at a spina bifida clinic where attention was paid to occipitofrontal circumference, shunt function, and clinical signs of increased intracranial pressure. Follow-up ranged from a minimum of 6 months after insertion of the primary shunt to a maximum of 5½ years. Revisions were carried out as indicated by signs of acute or chronic increase in intracranial pressure. At revisions carried out for distal block, the lower catheter was usually replaced in the peritoneal cavity. In 11 cases (22%), however, usually after repeated lower catheter obstruction, conversion to ventriculo-atrial shunt was carried out at some time within the follow-up period.

No attempt was made to assess developmental progress in relation to control of hydrocephalus as it was felt that too many other factors such as birth injury, ventriculitis, and cerebral palsy could influence the final intellectual attainment.

Results

Mortality. 11 of the 50 patients (22%) died within the follow-up period. This is, however, a crude way of expressing mortality, as the duration of follow-up varied from 6 months to 5½ years. This problem can be circumvented by examining the mortality in consecutive follow-up periods, expressed as a monthly mortality rate. It can be seen that the highest mortality rate (Table I) occurred in the first 3 months after insertion of the primary shunt. Thereafter, there was a substantial drop in mortality. The results expressed in this manner become less meaningful as the number of children followed up diminishes, e.g. the increase in monthly mortality rate to 1·6% in the 3- to 4-year follow-up period refers to the death of 1 child. The cause of death in each of the 11 children is shown in Table II.

TABLE I

Mortality and monthly mortality rate in consecutive follow-up periods after insertion of primary ventriculo-peritoneal shunt

Period of follow-up from insertion of primary shunt (yr)	Total possible survivors	Deaths in follow-up period		Mortality rate per month (%)
		No.	%	
0–3/12	50	6	12	4
3/12–6/12	44	1	2·3	0·77
6/12–1	38	2	5·3	0·88
1–2	29	1	3·5	0·29
2–3	15	0	0	0
3–4	5	1	20	1·6
4–5	2	0	0	0

TABLE II

Cause of death in 11 fatal cases

	Case no.	Age at death (mth)	Shunt *in situ*	Cause of death
Selected for early operation	1	10	V-P	Cyclical neutropenia; pancreatic hypoplasia
	2	3	V-A	Endocarditis on tricuspid valve, multiple pulmonary infarcts, bronchopneumonia
	3	4	V-A	Spinal meningitis
	4	3¾ yr	V-A	RV failure, pulmonary hypertension, multiple pulmonary infarcts, *Staphylococcus albus* septicaemia
	5	2	—	Brain stem compression; shunt removed during treatment of ventriculitis
	6	10	V-P	Respiratory infection; child resident in mental deficiency hospital
No early operation	7	3	V-P	Ventriculitis, no antibiotic therapy
	8	2	V-P	Respiratory arrest during late repair of back lesion
	9	1½ yr	V-A	*Staph. pyogenes* septicaemia, bacterial endocarditis, ventriculitis—no antibiotic therapy
	10	2	V-P	Ventriculitis, peritonitis—no antibiotic therapy
	11	9	V-P	Respiratory infection; no antibiotic therapy

V-P, ventriculo-peritoneal shunt; V-A ventriculo-atrial shunt.

Effectiveness. The effectiveness of a shunting system must be assessed both by the necessity for *further surgical intervention* and by the *degree of control* achieved, as shown, for example, by serial measurements of ventricular size, ventricular pressure, or occipitofrontal circumference.

Necessity for further surgical intervention.

(a) Alive and unrevised primary shunt (no further surgical intervention). Because of the variation in duration of follow-up, consecutive follow-up periods are again used. Fig. 1 shows, for each follow-up period, the proportion of survivors in whom the primary shunt continued to function. Nearly two-thirds of the patients had already required revision of some part of their primary shunt within 6 months of insertion. It can be seen, however, that after the first postoperative year the percentage of children with unrevised primary shunts remained steady at 20 to 25%. Again, this method of examining results lends itself to possible distortion as the numbers of children diminish with increase in the follow-up period. Only 2 children have been followed up for 5 years, both having had revision of their primary shunt, giving a revision rate of 100%.

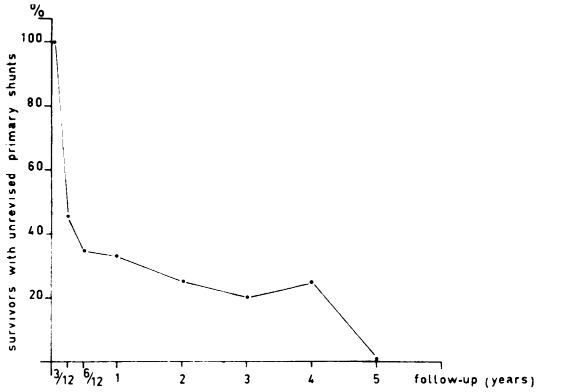

FIG. 1.—*Percentage of patients with unrevised primary ventriculo-peritoneal shunt in consecutive follow-up periods.*

(b) Frequency of revisions. By the word 'revision' is meant surgical intervention involving replacement of some part of the shunting system (upper and/or lower catheter) but leaving the patient with a functioning ventriculo-peritoneal shunt. The frequency of such revisions is again looked at in consecutive follow-up periods. In considering the results at the end of each of these periods the following children were excluded: those whose survival or follow-up did not extend to the end of the period and those in whom the ventriculo-peritoneal shunt had been replaced by a ventriculo-atrial system during the period. Table III shows the number of children who required from 0 to 3 revisions (no child required more than 3 revisions) within each of the follow-up periods considered. From the total number of revisions a mean monthly revision rate was calculated. This is shown graphically in Fig. 2. It will be seen that the mean monthly revision rate is highest in the first 3 months after insertion of the primary shunt and falls rapidly thereafter. After 2 years no further revisions were required.

(c) Conversion to ventriculo-atrial shunt. In 11 cases (22%) the ventriculo-peritoneal system was replaced by a ventriculo-atrial shunt, 73% within 6 months of insertion of the primary shunt. It is noteworthy also that 82% of these conversions were carried out before 1968, though only 52% of primary shunts were inserted in this period, suggesting a decreasing tendency to resort to conversion to ventriculo-atrial shunt as experience with the ventriculo-peritoneal system grew.

In each of the 11 cases the indication for conversion to ventriculo-atrial shunt was obstruction of the peritoneal catheter. As indicated in Table IV, no specific number of distal blocks was taken as an

TABLE III

Number of ventriculo-peritoneal shunt revisions and mean monthly revision rate in children surviving to the end of consecutive follow-up periods

Follow-up period (yr)	No. alive throughout period with V-P shunt	Number of revisions of V-P shunt					Mean monthly revision rate
		0	1	2	3	Total	
0–3/12	40	21	15	3	1	24	0·20
3–6/12	37	28	6	1	0	8	0·072
6/12–1	29	26	1	2	0	5	0·058
1–2	21	18	3	0	0	3	0·048
2–3	10	10	0	0	0	0	0
3–4	3	3	0	0	0	0	0
4–5	1	1	0	0	0	0	0

V-P, ventriculo-peritoneal.

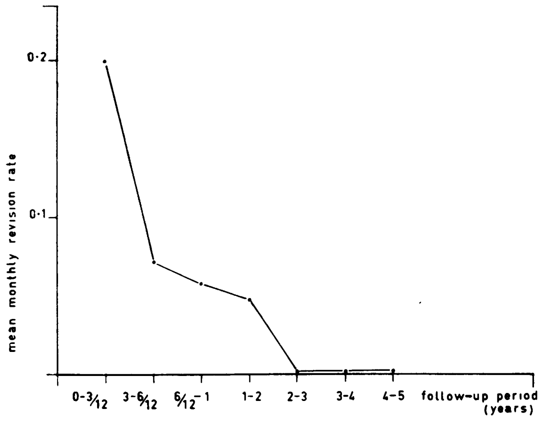

FIG. 2.—*Mean monthly ventriculo-peritoneal shunt revision rate for consecutive follow-up periods (data from Table III).*

TABLE IV

Frequency of peritoneal catheter obstruction preceding conversion to ventriculo-atrial shunt

No. of cases	No. of distal blocks				
	1	2	3	4	Total
11	3	4	3	1	24

indication for conversion, the number in individual cases ranging from 1 to 4.

Degree of control of hydrocephalus. There have been many attempts to find a reliable method of assessing degree of control of hydrocephalus. Measurements depending on clinical examination alone have particular limitations. The occipitofrontal circumference, however, which has been shown to be related to ventricular size (Lorber, 1961), has been widely used for this purpose. In considering the distribution of occipitofrontal circumference before operation and on follow-up in this series, the grading defined by Forrest, Hole, and Wynne (1966) has been adopted (Fig. 3). This allows direct comparison with their results from ventriculo-atrial shunting (Table V).

Complications. The frequency of ventriculo-peritoneal shunt revision and conversion has been discussed. The complications necessitating such intervention are outlined below.

A total of 66 complications were recognized, all occurring in the first 2 years after insertion of the primary shunt. 29 (44%) of these were related to the ventricular catheter and 37 (56%) to the peritoneal catheter.

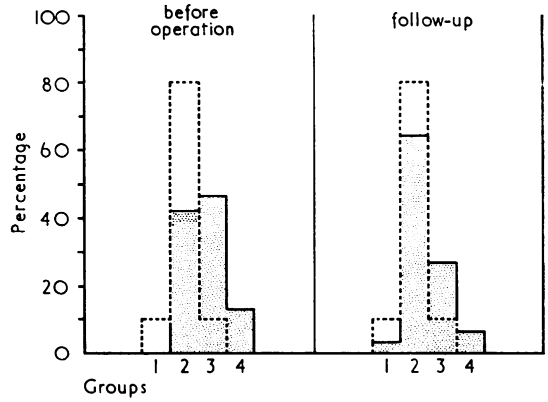

FIG. 3.—*Distribution of occipitofrontal circumference before shunting and at follow-up, compared with a normal population (broken lines). Grouping as defined by Forrest* et al. *(1966). 1, below 10th centile; 2, between 10th and 90th centile; 3, less than 4 cm above 90th centile; 4, more than 4 cm above 90th centile.*

TABLE V

Comparison of occipitofrontal circumference before shunting and at latest follow-up in patients treated by ventriculo-peritoneal and ventriculo-atrial shunts. (Data for the latter and grading of OFC taken from Forrest et al., *1966)*

OFC Group	Before operation				Latest follow-up			
	V-P		V-A (Forrest *et al.*)		V-P		V-A (Forrest *et al.*)	
	No.	%	No.	%	No.	%	No.	%
1	0	0	3	3	1	3	3	3
2	14	42	32	35	21	64	59	65
3	15	46	41	45	9	27	25	27
4	4	12	15	17	2	6	4	4

OFC, occipitofrontal circumference; V-P, ventriculo-peritoneal shunt; V-A, ventriculo-atrial shunt.

Ventricular catheter complications. There were 10 episodes of ventriculitis and 19 of ventricular catheter obstruction, 34 and 66%, respectively. The causes of obstruction of the ventricular catheter were no different from those reported in ventriculo-atrial shunts (Cast and Schurr, 1969).

Peritoneal catheter complications. Infection, i.e. peritonitis, did not occur as a complication of the peritoneal catheter, all cases being due to obstruction. The most common mechanisms of obstruction were kinking, fibrous tissue encapsulation, and displacement of the catheter, usually into the abdominal wall. In three instances the catheter had retracted up over the rib cage and in one had penetrated the diaphragm and caused a large effusion of CSF in the extrapleural space.

Predisposing factors. As reported by other authors (Lorber, 1969), early insertion of the primary shunt was associated with a relatively high complication rate. For example, of patients who required no shunt revision in the first 3 months after insertion 80% were over 1 month of age at the time of primary shunting. By contrast, 75% of those requiring three revisions in this period had undergone primary shunting at less than 1 month of age.

It could be argued that the high complication rate of the children who had early insertion might reflect a more severe degree of hydrocephalus. There was, however, no significant difference in initial mean cortical thickness between patients with revised and unrevised primary shunts at both the 1- and 2-year follow-up periods (Table VI).

TABLE VI

Need for shunt revision related to initial severity of hydrocephalus, measured by thickness of cerebral mantle at vertex

		Mean initial cerebral mantle (mm)
At 1 yr	Primary shunt still *in situ*	22·3 + SD 4·0
	Primary shunt no longer *in situ*	21·5 + SD 4·4
At 2 yr	Primary shunt still *in situ*	21·0 + SD 3·8
	Primary shunt no longer *in situ*	21·2 + SD 4·6

Discussion

Both ventriculo-atrial and ventriculo-peritoneal shunts are capable of controlling hydrocephalus successfully though, in exceptional cases, either system may fail completely and have to be replaced by the other. Comparison of the two must, therefore, take account of the cost at which control of hydrocephalus is achieved. This cost can be measured in terms of the frequency and severity of complications.

Frequency of complications. Nearly two-thirds of patients in the present series required revision of some part of the ventriculo-peritoneal shunt within 6 months of its insertion. The long distal catheter proved particularly liable to obstruction. Though comparison with other series is difficult in view of variations in methods of presenting the data, obstruction of the distal catheter appears to be less frequent in most series of ventriculo-atrial shunts. Obstruction and other complications of such shunts are, however, by no means rare. For example, in Hemmer's (1967) series of 181 patients, 62% required revision at some stage and 65% of revisions were carried out within the first 6 months. Zachary and Sharrard (1967) reported that only 9 out of 43 of their 7-year survivors had required no revision; the remaining 34 had had no fewer than 160 revisions. Noble *et al.* (1970) reported on a series of 143 patients whose follow-up varied from only 3 months to 5½ years: 67 patients (47%) had required 118 revisions. Lorber (1969), in describing a controlled trial of early and delayed ventriculo-atrial shunting, quoted figures for revision rates which allow more direct comparison with the present series. Depending on whether the primary shunt was inserted in the first 14 days or later, between 0·058 and 0·038 revisions per patient per month were required in children whose mean follow-up was 12 to 18 months. In the present series, the revision rate was much higher (0·20) in the first 3 months but fell to 0·048 in the second year.

In brief, there is little doubt that the ventriculo-peritoneal shunt is more likely to become obstructed in the first 6 months after insertion than the ventriculo-atrial. After the first year, however, complications are very infrequent.

Severity of complications. It is, however, when the nature of complications is considered that the advantages of the ventriculo-peritoneal shunt become apparent. Apart from simple obstruction, the only important complication encountered was ventriculitis, which occurred in 18% of patients in the first 6 months but in none thereafter. Ventriculitis is, of course, not peculiar to the ventriculo-peritoneal shunt. As shown in Table II, no deaths in this series could be attributed directly to the ventriculo-peritoneal shunt. Though in the past ventriculo-peritoneal shunts have usually been resorted to in extreme circumstances, e.g. after repeated failure of ventriculo-atrial shunting, reports of fatal complications are extremely rare (Rubin, Ghatak, and Visudhipan, 1972).

The ventriculo-atrial shunt is liable to other and more serious complications. 3 out of 4 children who died with ventriculo-atrial shunts *in situ* succumbed to shunt complications (Table II). It is conceded that the infants in this series who were given ventriculo-atrial shunts were unfavourably selected. Nevertheless, Forrest and Cooper (1968) considered that 10% of their patients died as a direct result of a ventriculo-atrial shunt inserted as a primary procedure, and in the series of Guthkelch (1967) valve complications were responsible for 69% of deaths. Septicaemia from valve colonization occurred in 13·5% of 1540 patients with ventriculo-atrial shunts culled from published reports by Luthardt (1970). Massive pulmonary embolism, endocarditis, thrombosis of the superior vena cava and right atrium, and even cardiac perforation are well recognized dangers (Crome and Erdohazi, 1966; Noble *et al.*, 1970; Tsingoglou and Eckstein, 1970). The long-term effects of ventriculo-atrial shunting are unknown, but the necropsy finding of some degree of pulmonary embolization in 93% (Emery and Hilton, 1961) and more recent reports of 'shunt nephritis' (Strauffer, 1970) are disquieting.

In addition to freedom from serious hazards, the ventriculo-peritoneal shunt offers further advantages. The operative technique is simple and the placing of the distal catheter is not critical. Even if revision is more often required in the first year, it is a much easier procedure. Migration of the peritoneal catheter with growth is not a problem and prophylactic lengthening is unnecessary. The operation can, moreover, be carried out with less trepidation after treatment of ventriculitis. Neither in this series nor in that of Attai *et al.* (1972) did ileal loop diversion lead to particular dangers in children with ventriculo-peritoneal shunts.

In conclusion, it is suggested that the ventriculo-peritoneal shunt merits more widespread use as the primary measure for control of hydrocephalus associated with myelomeningocele.

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