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Cause of Death in Children with Meningomyelocele or Hydrocephalus*

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Meningomyelocele is one of the most important congenital abnormalities, and is often associated with hydrocephalus. A recent survey (Eckstein and Macnab, 1966) showed that the mortality rate with modern treatment had fallen from about 50% in 1958 to about 20% at present. The purpose of this report is to review the causes of death in a combined series of 1091 patients with meningomyelocele or hydrocephalus seen at three children's hospitals in London in recent years. Of these, 206 patients had hydrocephalus only, and 885 had a meningomyelocele with or without hydrocephalus. Table I sets out the distribution of the patients in the various hospitals and indicates the period of review, which varied from one centre to another. The patients were, by and large, an unselected group, except that they had of course all been referred to a paediatric surgical centre for treatment. The basic policy of treatment in the three centres was essentially the same and the Holter valve was used for a ventriculo-atrial shunt in cases of hydrocephalus since 1958 in all three hospitals. While closure of the spinal defect was usually delayed in children seen early in this review, the policy since 1962 has been to close the meningomyelocele as an emergency procedure as soon after birth as possible.

The cause of death was ascertained whenever possible by studying the records of the post-mortem examination or the clinical notes of children in whom necropsy was not performed. In the case of children dying at home or in other hospitals, an effort was made to contact local practitioners to ascertain the cause of death. The necropsy rate on the patients reviewed in the three hospitals was 39%, 77%, and 83%, respectively.

The age at death is shown in Fig. 1. This clearly shows a high peak in the first three months of life tailing off gradually. In fact, 35% of the deaths occurred in the first three months of life and no less than 68% of deaths under the age of 1 year. However, it is significant that 9·4% of the children died after reaching their third birthday, which suggests

TABLE I

Distribution of Patients in the Three Hospitals

Hospital	Years of Review	Total	Alive	Dead
A	1958-1963	396	233	163
B	1958-1965	502	351	151
C	1948-1965	193	134	59
		1091	718	373

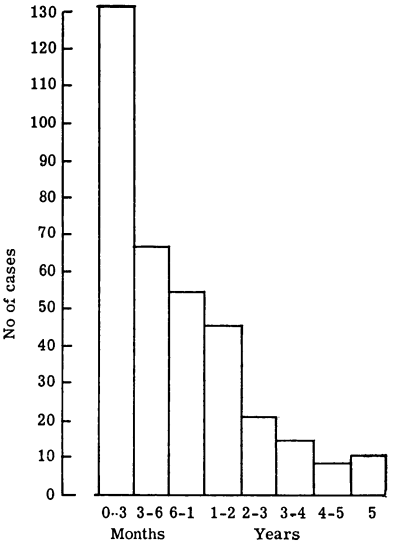

FIG. 1.—*Ages at death in 373 cases of hydrocephalus or meningomyelocele.*

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TABLE II

Causes of Death in 91 Untreated Patients

Cause	No.
Pressure	22
Meningitis	31
Pneumonia	16
Renal failure	3
Septicaemia	1
Pulmonary emboli	0
Not known	18

that there will be a continued small but steady mortality over the years.

Of the patients under review, 100 were not treated for a variety of reasons. Most of these were considered to be too ill or to be suffering from associated severe anomalies. In others there was a fulminating meningitis present when the child was first seen, and yet others had a gross degree of hydrocephalus. A few of the children died while awaiting admission for treatment (all before 1962). Of the untreated patients, 91 died, but these are in an already selected group. The causes of death in the untreated group of cases are shown in Table II. This shows not surprisingly that meningitis was the most important cause, but it must be recalled that these children were adversely selected. The three patients dying of renal failure had gross renal damage at the time of birth. All untreated patients who died did so within a year and usually within a month or two of birth.

The causes of death in the treated children are shown in Table III. In some of the cases it was impossible in retrospect to determine the exact cause of death when there was present, in the same child, a mixture of pneumonia, meningitis, and raised intracranial pressure. The total number of causes of death, therefore, exceeds the number of patients who died. However, the results show once again that meningitis is by far the most important cause of death.

TABLE III

*Causes of Death in 282 Treated Patients**

Cause	No.
Meningitis	127
Pneumonia	62
Pressure	54
Renal failure	31
Septicaemia	11
Pulmonary emboli	4
Post-operative	17
Back closure	9
Primary valve operation	3
Valve revision	5
Others	16
Not known	40

* More than one cause of death in many patients.

It might be thought that there would be a pattern of the cause of death in relation to the age at death, and one might have expected that children would die of meningitis at a younger age and of raised intracranial pressure or shunt failure at a later age. However, Fig. 2 shows that the various causes of death are indeed evenly distributed over the years.

A closer look at the various causes of death shows in fact that a retrospective study of this type is only of limited use. In all, 127 patients died of meningitis and, though this followed direct infection from the spinal lesion in many, it arose following the valve insertion in others. In a small but significant group

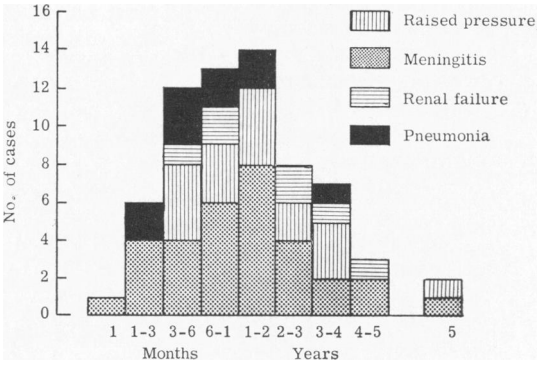

FIG. 2.—*Causes of death (Hospital A only) at various ages in 66 cases.*

of patients, meningitis developed after a few years, either without obvious cause or not infrequently when further surgery was undertaken to improve appearance of the back scar.

Raised intracranial pressure was the cause of death in 54 patients. In most of these, failure of the shunt was not recognized early enough and the children were not referred back to the surgical unit for a revision operation in time. However, it must be recalled that in a certain proportion of these patients it was felt that because the child was totally paralysed and also grossly retarded mentally, no further operation should be attempted and in these particular children the ventriculo-atrial shunt was intentionally not revised when it became blocked. A few patients died with a fluid collection under pressure in the posterior fossa while the ventriculo-atrial shunt was draining the lateral ventricles adequately.

Pneumonia was a contributory or primary cause of death in 62 patients. Again in this group it must be remembered that treatment was intentionally withheld when it was felt that the child was totally

paralysed and grossly retarded. In other children, however, it must be recalled that these had a severe chest deformity in association with meningomyelocele and poor air entry of one or both lungs. Pneumonia also developed not infrequently in children whose shunt became blocked, but even if the shunt system was successfully revised the pneumonia not infrequently continued to progress.

Renal failure was a predominant cause of death in 31 patients. In the large majority the upper urinary tracts were grossly dilated at the time of birth, and it is doubtful whether earlier diversion procedures would have saved these children. There have been no deaths from renal failure in children who have had a urinary diversion with reasonably well-functioning kidneys.

Pulmonary emboli or pulmonary hypertension was present at necropsy as a cause of death in only four cases. This is surprising, since we have previously shown that there is histological evidence of pulmonary embolism in about half the patients coming to necropsy following a ventriculo-atrial shunt (Erdohazi, Eckstein, and Crome, 1966). It may be that in future years the problem of pulmonary hypertension will become more important.

Eleven children died of septicaemia: in most of these there was a fulminating septicaemia associated with infection of the back or meningitis, but in a small number the septicaemia was a result of colonization of the valve, which persisted when the valve was removed or which, in a few cases, was not recognized and treated quickly.

The surgical mortality of children with meningomyelocele and hydrocephalus is surprisingly low. Only 9 patients died very soon after the closure of the spinal defect and these were invariably poor risk patients. Included in this group are two babies with large encephaloceles in whom a large amount of brain substance had to be excised in order to close the skull defect.

In this series of 1091 patients, a total of 643 primary ventriculo-atrial shunts were carried out. In almost all the cases a Holter type of valve was used, but in a small number the Pudenz valve was used, and in an even smaller one the 'D.W.T.' valve. It is noteworthy that only three patients died as a result of the original valve insertion. One of these patients bled to death when the distal catheter was inserted into the transverse sinus, a procedure which has since been abandoned. One further patient died of an inferior vena cava thrombosis as the distal catheter was too long. A third one died with massive pulmonary collapse. It is noteworthy that there was no death resulting from air embolus in this series—a tribute to the various anaesthetists who have helped us with the treatment of these patients.

Revision operations for ventriculo-atrial shunts were required in about 40% of the patients and in a large proportion of these repeated revision procedures were necessary. There were again only 5 deaths which could be directly connected with such revision procedures. Cardiac tamponade occurred in 2 as the result of perforation of the superior vena cava and heart, respectively, and in a third by inserting the distal catheter into the pericardium instead of the pleural cavity. One patient died of a shunt failure as the distal catheter had slipped into the pulmonary artery and a further patient had cardiac arrest at operation.

Summary

The most important causes of death in children with meningomyelocele and hydrocephalus are meningitis, raised intracranial pressure, and pneumonia. Often two or three of these processes are present at the same time and it is difficult to draw any definite conclusions. The primary mortality of closure of the spinal defect is low. Although the morbidity of ventriculo-atrial shunts is considerable, the mortality of shunt procedures and even of revision operations is surprisingly low.

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References

Eckstein, H. B., and Macnab, G. H. (1966). Myelomeningocele and hydrocephalus: the impact of modern treatment. *Lancet*, **1**, 842.

Erdohazi, M., Eckstein, H. B., and Crome, L. (1966). Pulmonary embolization as a complication of ventriculo-atrial shunts for hydrocephalus. *Develop. Med. Child Neurol.*, **8**, Suppl. 2, 360.