Clinicopathological Considerations of Late Death in Cases of Treated Congenital Hydrocephalus

Hiroshi ITO, Hirokazu KAWAKAMI,
Hirobumi MARUYAMA and Tetsuro MIWA

Department of Neurosurgery, Tokyo Medical College, Tokyo

Abstract

Although it is well known that the prognosis of child hydrocephalus has been improved by shunting operations, little is known about the question of why these children die after a long period. Between 1966 and 1980 at the Tokyo Medical College, the shunting procedure was carried out in 95 cases of infantile hydrocephalus. In 14 cases, death occurred more than three years after the operation for hydrocephalus. In eight cases an autopsy was performed. Six out of 14 cases died suddenly in spite of the shunt functioning well and these cases presented the so-called "hydrocephalic crisis". On the other hand, eight cases deteriorated gradually and died despite the most intensive and ingenious neurosurgical care. The 14 cases were classified into two types as follows:

- 1) Sudden death type; most of the cases were combined with myelomeningoceles and the Arnold-Chiari malformation or dysgenetic changes of the brain.
- 2) Gradual deterioration to death type; these cases were combined with intracranial inflammation such as severe meningo-ventriculitis.

In the radiological examination of the skull, the posterior cranial fossa was studied. The index value was less than the lower limit (0.25) of the normal level by Schey's method, and a significant difference was observed. Sudden death cases always showed the picture of "hydrocephalic crisis" without any increase in the supratentorial pressure. Autopsy cases showed the posterior cranial fossa to be small and the clival slope to be steep, abnormal deformation of the condiloid, and a concentric osseous ring around the foramen magnum. Such an anatomically abnormal structure of the posterior fossa plays a role in producing an abortive form of craniocerebral dysproportion, together with the cerebral dysplasia such as the Arnold-Chiari malformation. The state of craniocerebral dysproportion is easily affected by elevation of the supratentorial cerebrospinal fluid pressure. These structural changes are the major factors causing acute compression of the brain stem.

On the other hand, gradual deterioration to death cases presented cerebral dysplasia and hydrocephalic change with severe meningoventriculitis. Inflammatory changes in the brain were observed extending even to the basal ganglia. Gradually irreversible changes of the brain structures are considered to be caused by repeated chronic intracranial inflammation itself or by maldevelopment of the central nervous system and a decrease of immunological resistance of the host. The inflammatory changes of the basal cisterns and brain stem region may play a vital role in the gradually fatal outcome. Thus, the gradually deteriorating condition may be produced.

Key words: hydrocephalus, craniocerebral disproportion, small posterior fossa, cerebral dysplasia, craniometry, inflammation

Introduction

Recently, the prognosis of infantile hydrocephalus has been improved by shunting operations. However, some cases die suddenly and some others gradually deteriorate during the postoperative course, in spite of the shunt functioning well. The cases in which the patient had survived for a long time after the shunting opera-

tion are called¹⁵⁾ "late death" in treated congenital hydrocephalus.

Such late death cases have recently been the focus of great interest by numerous investigators.^{9,10,13)} Actually, such cases may well be more numerous than our experience because many patients may die at home without medical attention.

The etiology of sudden death or gradually deteriorating general condition has not yet been clarified. Our present study is an attempt to approach this problem.

Materials and Methods

For the past 15 years, from 1966 to 1980, ninety-five children with congenital or acquired hydrocephalus were seen and treated at our Department of Neurological Surgery, Tokyo Medical College. Among them, there were 58 long-term surviving children and 28 death cases. Among the latter, 14 cases died after more than three years postoperatively (Table I).

Among these 14 cases, 11 were diagnosed to have

Table 1 Cases ('66-Apr. '80)

Total No. of cases	95
Survival	58
Death	28
Unknown (not traced)	9
*Dead cases more than 3yrs. after op.	14

*Subject cases in this study

Table 2 Clinical findings in each of the three groups
Group I Cases accompanied by spina bifida

	Age	Type	Combined CNS-malf.	Preoperative			Shunting op. (revision)
				VFP.	CM.	Neurological	
I.N	3M	non-comm.	myelomeningocele	200 mm. H ₂ O	1.5 cm	hemiparesis convulsion	V-P (0)
Y.H	2M	non-comm.	"	200↑	1.5	convulsion (L) eye sign	V-P (2)
T.H	2M	non-comm.	"	200↑	2.0	fever. convulsion	V-P (4)
I.M	4M	non-comm.	Arnold-Chiari malf.	200	1.3	hypoactivity mental retardation	V-A V-P (3)

Group II Cases showing cerebral dysplasia

	Type	CNS-malf.	Infection	Preoperative			OP (revision)
				VFP	CM	Symp.	
Y.M.	comm.	hemidysplasia.	slight	150↓	3.0	convulsion visual dist.	V-P (2)
N.T.	non-comm.	hemidysplasia. corpus call. agenesis	(-)	180	?	severe convulsion	V-P
E.A.	?	ext. hydrocephalus porencephaly	(-)	120↓	1.3	mal-develop. convulsion	S-P
I.M.	"	ext. hydrocephalus	(-)	120↓	?	convulsion	S-P (2)
T.K.	non-comm.	hemidysplasia encephalocele	(-)	180	1.0	mal-develop. motor dist.	V-P (2)

Group III Cases accompanied by meningo-ventriculitis

	Type	Combined CNS-malf.	CSF infection	Preoperative			OP (revision)
				VFP	CM	Symp.	
I.H.	non-comm.	(-)	severe	170	1.5	mental retard.	V-P (4)
F.T.	"	(-)	slight	150↓	1.5	visual dist. convulsion	V-P (3)
T.H.	"	(-)	slight	200↑	2.0	mental-motor retard.	V-A (2) V-P
O.I.	"	corpus call. agenesis	severe	250↑	2.5	visual dist. mental retard.	V-A (3)
K.J.	comm.	(-)	severe	200↑	2.0	mental retard.	V-P (5)

obstructive hydrocephalus by neuroradiological investigations. Fourteen cases were classified into three groups as follows (Table 2):

Group I (four cases) consisted of hydrocephalus complicated with myelomeningocele or Arnold-Chiari malformation. Each case showed non-communicating hydrocephalus. The ventricular CSF pressure was more than 200 mmH₂O in continuous monitoring and showed severe thinning of the cerebral mantle in radiological examinations.

Group II (five cases) consisted of hydrocephalus accompanied by encephalodysplasia. Three cases showed non-communicating hydrocephalus but in the remaining two cases the type of hydrocephalus was not determined. The ventricular CSF pressure was shown in this group to be much lower than that in the other groups. In the morphological evaluation, malformations or dysgenetic changes of the cerebrum were identified. These included extensive cerebral hemispheric dysplasia, asymmetric ventricles and dysgenesis of midline structures. These cases presented many abnormalities in the neurological examinations.

Group III (five cases) consisted of hydrocephalus complicated by cerebral inflammation such as ventriculitis and meningitis. Four cases were non-communicating hydrocephalus while one case was not clear. They showed elevated CSF pressure and thin cerebral mantles, but with no specific changes. This group was repeatedly treated with shunting operations.

Ventricular CSF pressure was measured by means of a pressure transducer made by the Stetham Co., model p-37. The width of the cerebral mantle was measured on pneumoencephalograms by the distance between the frontal horn and the inner table of the cranium (Fig. 1). As a result, ventricular CSF pressure was

Fig. 1 CSF pressure and the cerebral mantle. The ventricular CSF pressure was measured by a Stetham. model p-37. pressure transducer and width of the cerebral mantle was determined by air ventriculography.

Table 3 Shunting operation and outcome in each of the three groups

		I	II	III
	Age	av. 2.7 M	2.0	4.0
	No. of revisions	av. 2	2	3.4
	Shunt function (before death)	well-functioning	well-functioning	malfunction. 3.
	Sudden onset type	3	2	1
	Gradual course type	1	3	4

reduced to the normal range by shunting operations in all except one case and the width of the cerebral mantle increased slightly in all except two cases with cerebral dysplasia.

In comparing the treatment and prognosis among the three groups, there was no difference in the clinical picture between Groups I and II (Table 3). Group III demonstrated gradual deterioration of the neurological and general state with shunt malfunction before death. Only one case presented a sudden clinical picture of "hydrocephalic crises."

A distinct feature was noted in the mode of death, i.e. the mode of death can be divided into two types. One is the "sudden onset or acute" type and the other is the "gradually deteriorating or chronic" type. The former demonstrated a so-called "hydrocephalic crisis" with convulsions or primary respiratory paralysis, and tended to die within two to three days after the onset. On the other hand, the latter showed gradual deterioration in neurological and general conditions. These cases presented slight fever, discomfort and inactivity over a long period until death. This type showed no hydrocephalic crisis before death (Table 4).

In the radiological examination of the skull, the

Table 4 Symptoms and mode of death

Physical condition \ Mode of death	Sudden onset 6 cases	Gradual course 8 cases
1. Hydrocephalic crises		
recurrent convulsion	4	
acute respiratory arrest	4	1 (pneumonia)
2. General condition		
displeasure-inactive	2	6
inanition	1	3
fever	2	5

Fig. 2 Posterior fossa index (Schey, 1973). Measurement of the posterior fossa index in fourteen cases (left corner) and 40 normal children (right corner).

measurement values of the posterior cranial fossa were compared between normal subjects and hydrocephalic infants in each age group. The posterior fossa index (P.F.I.) was obtained by Schey's Method (1973)¹²⁾ as GI over CD as shown in Fig. 2. CD was obtained by drawing a line parallel to AB (the greatest antero-posterior measurement of the skull) from the posterior clinoid to the posterior surface of the skull. GI is a line drawn perpendicular to CD at a point which allows GI the longest measurement between a point on line CD

(G) and the floor of the posterior fossa. According to the Schey's study, no patient over 18 months of age fell into the "below 0.25" zone. In this series, the dead cases with non-communicating hydrocephalus and fourteen cases, except two cases were noted fell into the 0.1 to 0.25 zone. Whereas the zone was less than 0.25 in four out of 40 control subjects.

In eight of the 14 dead cases autopsies were obtained with neuropathological examinations. The findings are listed in Table 5. It contains three cases of sudden death and five cases of gradual death. In all, sudden death cases non-communicating hydrocephalus with a highly or asymmetrically enlarged ventricular system, multicysts or porencephalic changes in the cerebral hemispheres were demonstrated. Generally, a marked thinning of the cerebral parenchyma due to extreme dilatation of ventricular systems was observed, whereas the paraventricular walls had become thick (gliosis). Among the pathological changes of the infratentorial space or posterior fossa, the so-called small posterior cranial fossa was noted, and its cranial volume was markedly reduced compared to normal subjects. In the cerebellum, Arnold-Chiari malformations of Type II were shown to be accompanied by obstruction of the aqueduct and severe adhesive arachnoiditis of the basal cistern and the peri-brain stem region.

Figure 3 shows autopsy findings of two cases of sudden death (I. M. and I. N.). In these cases the pos-

Table 5 Autopsy findings of eight cases

	Name	Cerebral hemisphere	Post. fossa, cerebellum brain stem	Spinal cord
Sudden death	I.N.	moderate hy. asymmet. ventricle	small post. fossa	mmc (ls)
	I.M.	severe hy. multiple septation of ventricle paraventricular thinning (gliosis)	Arnold Chiari malf. atresia of aqueduct tonsillar herniation	
	Y.H.	moderate hy. multiple porencephaly paraventricular gliosis	upward bulging of cerebellum small post. fossa	mmc (ls)
Gradual death	N.T.	hemidysgenesis glial scars of thalamus. microgyri. dysgenesis of corp. callosum.	basal adhesive arachnoiditis	
	O.I.	severe hy. severe ventriculitis dysgenesis of corp. callosum	stretched brain stem arachnoid cyst	
	T.N.	severe hy. severe ventriculitis multifoci malacia	basal adhesive arachnoiditis fibrous adhesion of vermis	mmc (l)
	F.T.	moderate hy. glial scars of basal ganglia	small post. fossa tonsillar herniation	
	K.J.	severe hy. ventriculitis	basal adhesive arachnoiditis. compressed brain stem.	

Three of the sudden onset type and five of the gradual progressive type

Fig. 3 Cranial base findings in the autopsy cases.

Left: I.M. 3-year-4-month-old girl with non-comm. hydrocephalus and Arnold-Chiari malformation.

Right: I.N. 4-year-3-month-old girl with non-comm. hydrocephalus and myelomeningocele.

af: anterior fossa, mf: middle fossa, pf: posterior fossa, cl: clivus, cb: circular bone formation, te: tentorial edge, fm: foramen magnum, confl: confluence sinus. Large arrows indicate the sharp angle of the clivus. Small arrows indicate the cerebellar tentorial edge and shallow posterior fossa.

terior cranial fossa was generally small and shallow and conical deformation of the foramen magnum caused by circular bony rings was found. The clival sloop was steep

and the attachment of the cerebellar tentorial edge was in a lower position than in the normal structure. Morphological examinations of cerebellum and brain stem showed similar changes, i.e., the sagittal section of the cerebellum and brain stem were elongated, the fourth ventricle was displaced caudally and the elongated cerebellar tonsil looked as if it compressed the medulla and high cervical spinal cord (Fig. 4).

These changes may be caused by Arnold-Chiari malformations combined with the narrow posterior fossa. It might be due to compression of the brain stem in the posterior fossa.

Gradually deteriorating cases in the mode of death presented cerebral dysplasia and hydrocephalic changes with severe meningoventriculitis and partial cerebritis. The inflammatory changes in the brain were observed extending as far as the basal ganglia.

Adhesive arachnoiditis of the brain stem and cerebellum was fairly fibrous and thickened. These changes formed a vicious circle in causing slowly progressive hydrocephalus.

Macroscopic changes in one case of the gradually deteriorated type are presented in Fig. 5. This case demonstrated irregular shaped multiple encephalomalasia in the cerebral hemisphere and revealed histologically severe cerebritis and distinct periventricular changes.

Fig. 4 Case I.M. Female. Sagittal section of Arnold Chiari malf. Type II. The picture showed brain stem compression and cerebellar tonsillar herniation.

Fig. 5 Autopsy finding in a case of meningo-ventriculitis. The coronal section showing marked paraventricular multiple cyst and encephalomalacia. c: cyst formation, v: ventricle.

Discussion

It is well known that in infantile hydrocephalus the survival time has been extended for a long time by the shunting operation. However, there has not been any extensive analysis of the causes of death, or any attempt to explain the mechanism underlying late complications of surgically treated hydrocephalus. Generally, the mortality rate of congenital hydrocephalus is nearly 25% according to various reports.^{7,9,15)}

The authors have experienced four cases which died suddenly in spite of the shunt functioning well after the operation. Therefore, we are greatly interested in the problem of why such a sudden death occurs without any extracranial cause, presenting "hydrocephalic crises." These cases demonstrated pathologically relevant brain herniation. On the other hand, eight late death cases showed a gradually deteriorating condition, and they should be distinguished from those of sudden death in view of the mechanism of late death.

Jones⁷⁾ described the causes of death in his patients, who died only from complicating diseases, and numerous investigators^{9,14,15)} including Jones have not yet reviewed completely the mechanism of the late death in detail.

Out of 14 cases of late death all occurring more than three years after the operation, postmortem studies were carried out in only eight cases. However, it is well known that postmortem studies did not explain the mechanism of late death. Thus, it is necessary to review the morphological changes of the brain and skull, before and after treatment.

In Group I, hydrocephalus associated with Arnold-Chiari malformation or spina bifida was demonstrated to be of the non-communicating type with marked enlargement of the lateral ventricles, the ventricular

C.S.F. pressure being more than 200 mmH₂O. They all depended on the shunt system. Group II with hydrocephalus and cerebral dysplasia, presented maldeveloped psychomotor functions to some degree. The ventricular C.S.F. pressure was not so high before death. All these cases, except one, revealed the non-communicating type of hydrocephalus.

In Group III, the hydrocephalus associated with intracranial inflammation showed different features from those of the other two groups, i.e. there was no complication of the anomaly of the central nervous system, but operative procedures had to be performed repeatedly. The ventricular CSF pressure was returned to the normal range by shunt operations, but one case continued to show a higher level until just before death. It was expected that the thickness of the cerebral mantle would increase with age, but no increase was observed in Group I or II.

In the post-operative clinical course in all cases, no characteristic signs or symptoms were observed, and yet, the modes of death were different. It was similar to the often reported results on sudden death observed in hydrocephalus accompanied by Arnold-Chiari malformation or spina bifida as reported by Salah *et al.*,¹⁰⁾ Emery,⁵⁾ Doran *et al.*⁴⁾ and Sunder-Plassmann *et al.*¹⁵⁾ They reported that there was no specific basis to explain the signs or symptoms before death when hydrocephalus was not associated with malformations of the central nervous system. Sudden death cases always showed the picture of "hydrocephalic crisis" without any supratentorial elevation of the CSF-pressure. This condition of the hydrocephalic crisis seemed to correspond to brain stem compression.^{5,7,13)} However, intracranial malformation alone does not cause the onset of acute symptoms. We compared the volume of the posterior cranial fossa with that of a normal subject to measure the changes of posterior cranial fossa. As a result, the index values were less than the lower limit (0.25) of the normal level by Schey's method,¹²⁾ and yet, no significant difference was observed compared with 80 other cases of hydrocephalus. Morphological hypoplasia of the posterior fossa in hydrocephalic children has been reported numerously using roentgenological measurements of the cranium^{1,8)} or on a morphological¹¹⁾ and anatomical basis.⁴⁾ Schey¹²⁾ stated in detail that the volume of the posterior fossa in children with hydrocephalus does not increase with age. This suggests that the infratentorial brain structure and the isolated pressure¹⁴⁾ are easily affected by slight elevation of intracranial pressure and shift of the brain¹⁵⁾ structure toward the supratentorial space. According to the reports of the postmortem examination, the posterior cranial fossa in hydrocephalus was distinctly smaller and the attachment of the tentorial edge was lower than in normal

subjects.

In addition, many other reports^{1,8,12,13)} showed the posterior cranial fossa to be small. All these cases showed conical deformation with a concentric osseous ring around the foramen magnum.^{10,15)} We found two autopsy cases with steep clival slope and abnormal deformation of the condiloid. Such abnormal anatomical structures of the posterior cranial fossa play an important role in the development into an abortive form of craniocerebral dysproportion together with the cerebral dysplasia, such as the Arnold-Chiari malformation. The state of craniocerebral dysproportion⁶⁾ is easily affected by elevation of the supratentorial CSF pressure. It is reported that the pressure in the infratentorial space is independent of that in the supratentorial space. In some reports,^{3-5,10,15)} the existence of small, narrow posterior cranial fossa and deformations of the cerebral structures of the infratentorial space are the major factors causing acute compression of the brain stem. To prevent sudden death caused by pons and medulla oblongata compression or displacement with a well-functioning shunt system, Doran *et al.*⁴⁾ and Hoffman *et al.*⁶⁾ have recommended performance of suboccipital decompression. However, they also warn that care must be taken not to cause a dangerous decrease of the supratentorial CSF pressure with the shunt operation.

We could not find any reference to the mechanism of gradually deteriorated death after treatment of hydrocephalus. Generally, this mechanism has been considered to be based on sepsis or pneumonia complicated with hydrocephalus. Moreover, there are many cases of gradually deteriorated death as a result of chronically increased intracranial pressure with obstruction of the cerebrospinal fluid pathway or shunt malfunction. From our study, gradually irreversible changes of brain structures are considered to be caused by repeated chronic intracranial inflammation itself or maldevelopment of the central nervous system and decreased immunological resistance in the host. The inflammatory changes of the basal cistern and peri-brain stem region may play a vital role in the gradual deaths.¹³⁾ The postmortem examination revealed severe adhesive arachnoiditis or cerebral parenchyma inflammation with a moderate degree of hydrocephalus. Two of the autopsy cases showed severe congenital cerebral dysplasia or cerebral malformation.

Infantile hydrocephalus complicated with severe cerebral inflammation or basal arachnopathy seems to have brought about the possibility of inducing chronic lesions of the brain stem and medulla oblongata in the future, i.e., the basal and peri-brain stem adhesion may cause mechanical traction and compression or circulatory disturbance (anemic mechanism) in the brain stem and medulla oblongata. Thus, a gradually deter-

iorated condition may be introduced.

Through a series of these studies, we found two types of late death occurring a long time after the operation. To date, there is no suitable therapeutic procedure for hydrocephalus due to congenital cerebral malformation, and first of all apart from this problem, it is necessary to carefully manage the surviving cases for a long time after the operation.

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Request reprints to: H. Ito, M.D., Department of Neurosurgery, Tokyo Medical College Hospital, 6-7-1, Nishishinjuku, shinjuku-ku Tokyo 160, Japan.