Diagnostic Criteria and Assessment of the Condition in Surgically Arrested Hydrocephalus

Hiroshi ITO, Yoshihisa ONODERA, Kunihiko TAKANASHI, Kenichi TAJIMA and Tetsuro MIWA

Department of Neurosurgery, Tokyo Medical College, Tokyo, Japan

Summary

Many investigators differ in opinion regarding the clinical concept and accurate criteria for the definition of surgically arrested hydrocephalus. We have studied the diagnostic criteria and various conditions of arrested hydrocephalus through the investigation of a series of 67 patients treated by shunting operation for progressive hydrocephalus. Of these, shunts were removed in 14 cases and these patients had no need for further treatment. We made investigations using neurological, morphological, neuroradiological and psychological methods, CSF dynamic studies including radioisotope cisternography and intraventricular CSF pressure measurements, and biochemical study of fatty acid in CSF.

On neurological examination, squint, poor vision and motor weakness were noted in the arrested stage in a few cases. Six patients were educable but slightly retarded, while two were imbeciles. Seven patients (50%) presented no obvious physical handicaps, however, five were slightly handicapped. Intelligence developed and the degree of physical disability gradually diminished with increasing age. Physical handicaps and residual neurological signs were correlated with the etiology of hydrocephalus.

After the shunting operation head circumference remained unchanged for a period and then gradually increased within normal developmental range. Thickness of the cerebral mantle increased from 8 to 30 mm in six cases of congenital hydrocephalus. Usually, cases where thickness of the cerebral mantle were less than 14 mm within 3 months after birth, the subject had a tendency to show poor prognosis. In the arrested stage, the CSF pulse waves consisted of multiphasic components, mean intraventricular CSF pressure was found to be constant by long-term monitoring, and various RI-cisternographic patterns were observed. Three cases showed transient ventricular reflux with early clearance and convexity flow. However, in another 11 cases changes in RI-cisternographic patterns were inconsistent. Examination of fatty acid metabolism in CSF revealed that the total value of fatty acid was closest to normal range after the operation.

Arrested hydrocephalus proved to exist in varying degrees as the causes of hydrocephalus vary. Studies from various points of view provided valuable data as to the analysis of arrested hydrocephalus. Diagnosis of arrested hydrocephalus cannot be made on the basis of only simple examination, but should be made by collective judgement from the results of various examinations.

Key words:
arrested hydrocephalus, CSF dynamics, mental-motor development, shunt independency, compensated hydrocephalus

Introduction

In general, surgically arrested hydrocephalus may be defined as a state of cessation of abnormal head growth, total arrest of the progressive neurological symptoms and wasting of the brain tissue. Investigators, however, differ in opinions regarding the clinical concept and accurate criteria for the definition of surgically arrested hydrocephalus. Some investigators state that the presence of arrested hydrocephalus itself is open to controversy.

Arrested hydrocephalus may be defined easily from acute progressive hydrocephalus clinically, but arrested hydrocephalus is sometimes difficult to differentiate from slowly progressive hydrocephalus.

It is necessary to precisely define arrested hydrocephalus as a clinical entity, but recently there has been some confusion in the use of terminology of arrested hydrocephalus.

Therefore, we investigated the diagnostic criteria and various conditions of arrested hydrocephalus through a series of 14 patients from a total of 67 cases.

Materials and Methods

The clinical materials consisted of 67 infantile hydrocephalic patients admitted to the Neurosurgical Department of the Tokyo Medical College Hospital over a period of 10 years since 1965, and this number does not include children with hydrocephalus caused by tumor. All of these 67 infants were treated by ventriculo-atrial shunting or ventriculo-peritoneal shunting. Of the 41 children still alive, 14 cases are no longer shunt dependent. Their shunt systems were removed because the hydrocephalic condition of these 14 patients had been arrested.

A comparative study of pre- and postoperative data was performed for each of these patients. Arrested hydrocephalus was observed in 21 percent of the 67 infants with hydrocephalus. In our clinic, infantile hydrocephalus was subdivided into four major groups (Table 1). Group A consisted of 41 cases with congenital hydrocephalus, Group B of 10 cases with bacterial meningitic hydrocephalus, Group C of 13 hydrocephalic patients accompanied by central nervous system anomalies, and Group D of 3 cases with hydrocephalus of unknown origin. The percentage of mortality in the four main etiological groups differed significantly. In Group A the mortality rate was the highest among four groups and arrested hydrocephalus occured in only four cases. In Group B, arrested hydrocephalus was found in five cases (50%). Eleven of 14 cases of arrested hydrocephalus was of the communicating type.

For the purpose of the evaluation of the state of a arrested hydrocephalus, we investigated not only neurological, psychological and morphological signs, but also cerebrospinal fluid dynamics, which has recently become the focus of attention in the study of hydrocephalus. For the latter we used such tests as ventricular CSF pressure study and isotope cisternography. Then biochemical analysis of fatty acid, one of the significant metabolic factors in the CSF, was done (Table 2).

Results

The preoperative duration of pressure symptoms and thickness of the cerebral mantle at the

Table 1 "Arrested" cases in 67 infantile hydrocephalus according to etiology (Dept. Neurosurg., Tokyo Medical College)

		Total in Series	Arrested	not Arrested	not Traced	Dead
A	Congenital Hydrocephalus	41	4	20	3	14
B	Post-meningitic Hydrocephalus	10	5	3	1	1
C	Hydrocephalus with CNS-malformation	13	4	3	2	4
D	Hydrocephalus (origin unknown)	3	1	1	0	1

(more than 2 years after operation)

Table 2 The diagnostic tests for the evaluation of arrested hydrocephalus

1. Neurological Sign and Symptom	: Setting sun sign. Visual disturbance. Motor disability. Decerebrate sign etc.
2. Physical, Mental Ability	: Physical Handicap (normal ~ incapacitated) Intelligence Assessments (WPPSI. WISC)
3. Morphological Finding	: Head Circumference. Cranial Index. Cerebral Mantle.
4. CSF Dynamics	: Intraventricular fluid mean pressure. Pulse wave form. Infusion test.
5. RI-Cisternography	: RI flow pattern, Ventricular stasis. Convexity flow.
6. CSF Fatty acid	: Total fatty acid composition

time of operation were examined in each group (Fig. 1). Before shunting procedure, Group B received prolonged antibiotic treatment for meningitis. However, all the patients were treated with shunting operation within 2.5 months after birth. The thickness of the cerebral mantle was more than 20 mm in 11 cases. In two cases it was less than 14 mm at the time of operation.

It is well-known that progressive hydrocephalic patients have many neurological signs. On the contrary, only a few residual neurological signs of arrested hydrocephalus were recognized as shown in Table 3. The difference of neurological signs between the progressive and the arrested cases was statistically significant.

These neurological sequelae are variable according to the etiology and grade of hydrocephalus. There appears to be significant association between neurological sequelae and the etiology of the hydrocephalus. For their developmental assessment of infants and children, studies by Tanaka-Binet, Wechsler Intelligence Scale (WISC), Wechsler Pre-school

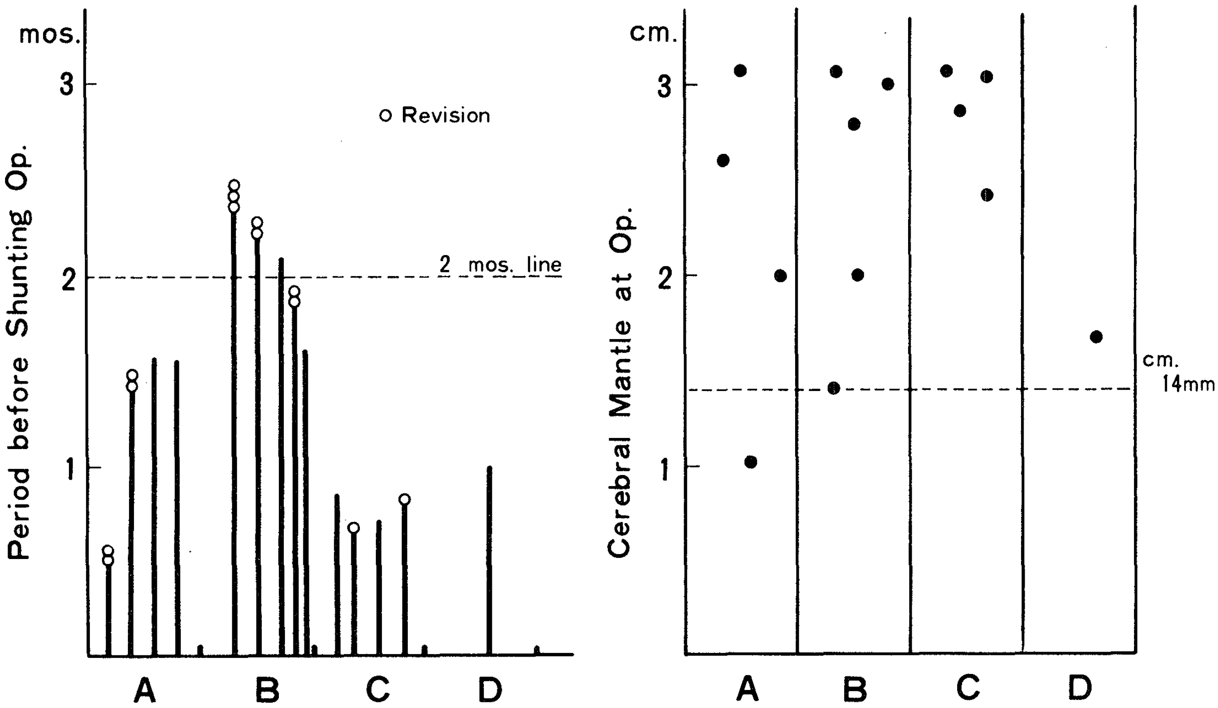

Fig. 1 Period before shunting operation and cerebral mantle. *Left*: Period before shunting operation in the four groups differs significantly. *Right*: Thickness of the cerebral mantle at the shunting operation.

Table 3 Residual neurological signs at the arrested stage in the four groups

	Arrested Stage	Residual Neurological Signs	
A	4	Optic nerve atrophy	1
		Strabismus	2
		Muscle weakness	1
B	5	Convulsion	1
		Strabismus	1
		Muscle weakness	1
C	4	Paraparesis	1
		Strabismus	1
D	1	Strabismus	1

and Primary Scale of Intelligence (WPPSI) were carried out at annual intervals according to the age of the children.

Among the total 14 cases with an average age of 6 years and 5 months, six patients including three of four cases in Group A showed an IQ score of more than 85. The patients in Group B, in general, had low IQ. Causes for the lower IQ were considered to be the influence of severe infection and brain damage rather than hydrocephalus *per se* (Table 4). Judgement of physical handicaps in survivors of hydrocephalus was made using a modified version of Laurence's classification (1969), and physical disability of the 14 cases was divided into three levels according to severity (Table 5).

Many of the children in Group A and B recovered to lead satisfactory and meaningful lives. In Group A, two patients had handicaps that required some nursing.

For the purpose of morphological evaluation of the skull and brain, the thickness of the cerebral mantle was measured by air study, and head circumference was also measured. Head enlargement was confirmed by plotting the head circumference on a Nellhaus's standard chart. Though head enlargement is always present in infancy, abnormal head enlargement was observed before shunting operation in all cases (Fig. 2).

After the shunting operation, the rate of abnormal head growth usually decreased but sometimes continued for a period, and then gradually returned to the standard range. In general, the head which had been abnormally

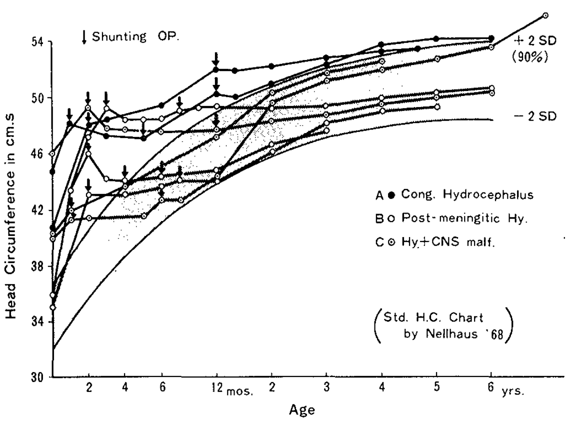

Fig. 2 Head growth curve.
More than three years follow-up of head growth in each group after successful shunting operation. It parallels the normal growth curve. Shaded zone indicates the standard head circumference range by Nellhaus (1968).

Table 4 Intelligence quotient. With an IQ of 85 being the low normal ability level, 6 of 13 children are at least of low normal intelligence.

	No. of Cases	Average age (years)	~100	99~85	84~60	59~
A	4	$6\frac{9}{12}$	2	1		1
B	5	$6\frac{4}{12}$		1	2	1 (Unknown 1)
C	4	$6\frac{7}{12}$	1	1	2	
D	1	$4\frac{1}{12}$			1	
			3	3	5	2

Table 5 Physical handicaps. Degrees of physical handicaps in 14 children with arrested hydrocephalus. Modified Laurence's classification of physical handicaps in survivors of hydrocephalus.

		A	B	C	D	Total
1. Normal:	Slight squint. & brisk reflex only	2	3	1	1	7
2. Slightly Handicapped:	Slight spasticity, impairment vision, unsteady gait, slight imbalance	2	2	1		5
3. Severely Handicapped:	Partial paralysis, flexion deformities, blindness. etc.			2		2

(by M. Laurence 1969)

enlarged for the patients's age and sex, resumed growth at a normal rate within 1 year after the shunting operation and no abnormal enlargement was seen except when malfunction of the shunt occurred.

The cerebral mantle increased from 8 to 30 mm in thickness in six cases (Fig. 3). The average increase rate was 28 mm in Group A, and 7 mm in Group B and C. Two cases in Group B, however, showed an increase of only 5 mm. The postoperative increase in thickness of the cerebral mantle was the largest in congenital hydrocephalus cases and the smallest in post-menigitic hydrocephalus cases.

Fig. 4 shows the clinical course from birth of a 9-year-old girl having arrested hydrocephalus (a patient in Group A). She received a ventriculoatrial shunt at 6 weeks of age. Her hydrocephalic condition was thought to have become arrested in three and a half years after the operation and the shunt was removed. She showed good development of intelligence, was able to attend an ordinary primary school and is presently leading a useful life.

Cerebrospinal fluid pressure study and constant infusion saline manometry are relatively safe procedures which can offer useful information regarding the condition of the CSF circulation and absorption. The authors already reported the study of intraventricular CSF pressure in infantile hydrocephalus. In the present study we compared progressive hydrocephalus with arrested hydrocephalus by measurements of CSF mean pressure and intraventricular CSF pulse waves, loading saline manometry, and various other tests. In arrested hydrocephalus, the CSF pulse waves consisted of multiphasic

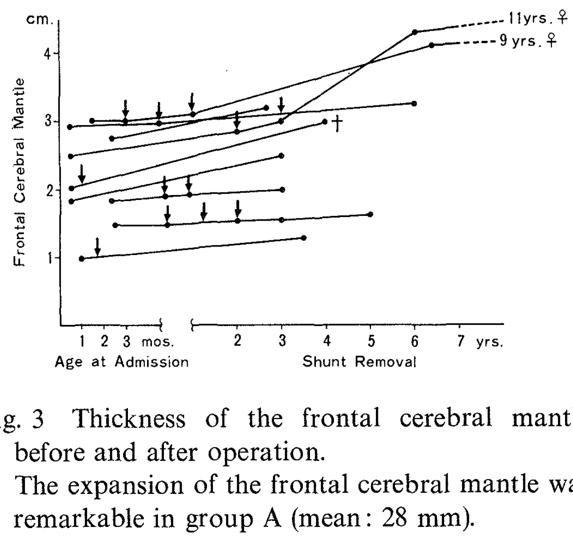

Fig. 3 Thickness of the frontal cerebral mantle before and after operation.
The expansion of the frontal cerebral mantle was remarkable in group A (mean: 28 mm).

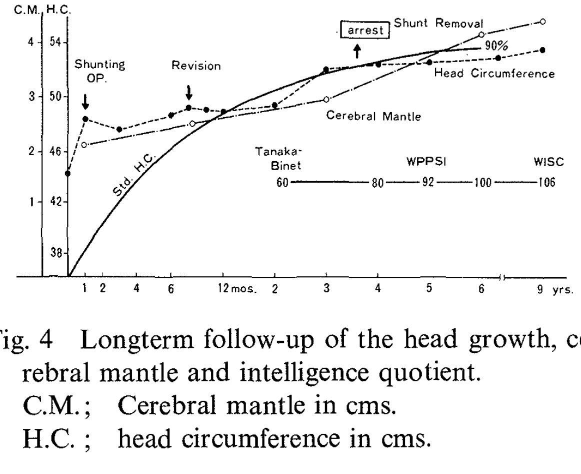

Fig. 4 Longterm follow-up of the head growth, cerebral mantle and intelligence quotient.
C.M.; Cerebral mantle in cms.
H.C. ; head circumference in cms.

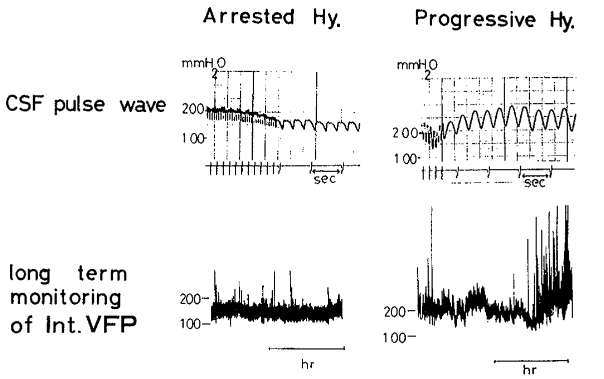

Fig. 5 In progressive hydrocephalus cases intraventricular CSF pulse waves are monophasic and high in amplitude. While in arrested cases, the pulse waves consist of multiphasic components.

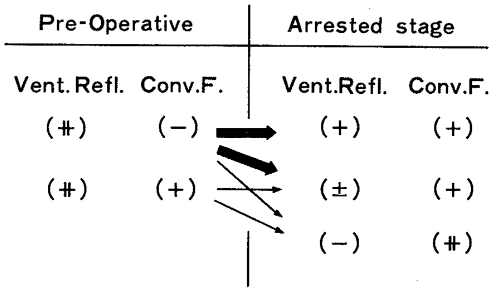

Fig. 7 Changes of RI-cisternographic patterns in arrested hydrocephalus. Left: Preoperative stage. Right: The arrested stage. Vent. reflux. (++): ventricular reflux and delayed clearance. (+): Ventricular reflux with early clearance. (±): difficult to judge. Conv. F. (++): convexity early clearance (normal). (+): convexity slightly delayed clearance. Heavy arrows indicate the general tendency in the preoperative and arrested stages.

components while in progressive hydrocephalus with large ventricle the pulse waves were composed of monophasic components and tended to be slower in elevation and higher in amplitude. The CSF mean pressure was found to be constant in arrested hydrocephalus during continuous long-term monitoring of the intraventricular CSF pressure. On the contrary, progressive hydrocephalus often manifested abnormal rises in CSF mean pressure and its elevation became remarkable (Fig. 5).

In addition, intraventricular saline loading methods (Holtz's modification 1970) were also examined. These techniques are helpful in the evaluation of the need for shunting procedure in patients with enlarged ventricular systems and this procedure can also be applied in the study of arrested hydrocephalus. The results are shown in Fig. 6. In arrested cases pressure curve responds with a rapid rise and then resumes its previous level within 5 minutes after saline injection. The findings in progressive cases were

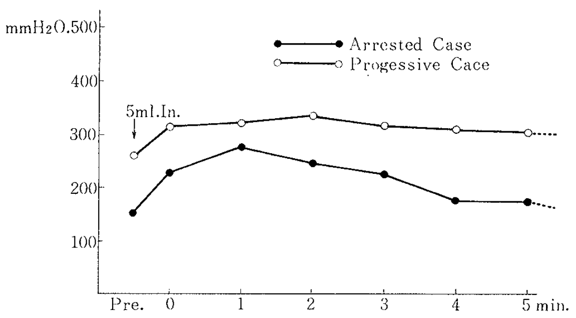

Fig. 6 Constant saline injection and loading test. CSF pressure rapidly rises and returns to its previous level within about five minutes after saline injection.

quite different in responce to saline infusion from those obtained in arrested cases.

It is well-known that radioisotope cisternography is very useful for obtaining information about the functional status of CSF circulatory dynamics. Changes in cisternographic pattern of the preoperative and arrested state were generally as follows (Fig. 7). In the pre-operative stage ventricular stasis was observed in every case. In communicating hydrocephalus a scanty convexity flow of radioisotope was seen only in three patients and was not noted in most of the other cases. In the arrested stage various changes of RI-pattern were observed. Only in a few cases, ventricular reflux with early RI-clearance and convexity RI-flow which indicate improvement of CSF circulation and absorption were observed (Fig. 8). However, at present, complete understanding of changes of RI-cisternographic figures is difficult. In the near future, it is hoped that these analyses will become possible.

Changes of metabolism in CSF are sometimes useful to indirectly estimate the process of brain disease. Therefore, studies of lipid or fatty acid, which is one of the main components of brain tissue, are significant and must be given the same attention as studies of protein and nucleic acid metabolisms. The authors have reported significant results about changes of fatty acid composition in CSF in infantile hydrocephalus. After shunting operation, the total value of fatty acid in CSF decreases to the normal range. The

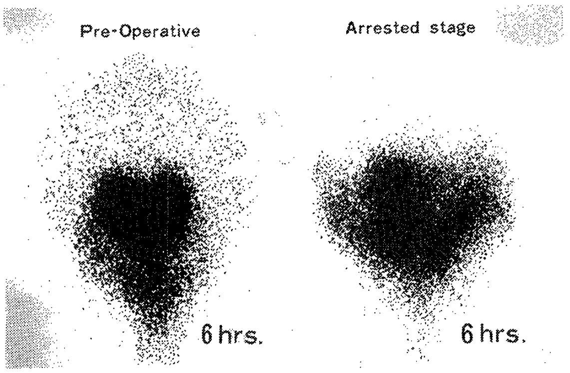

Fig. 8 RI cisternograms in Case S.K., 6 years old girl. Left: RI cisternogram when the patient was 3 months old. Ventricular reflux and no convexity activity are shown. Right: RI cisternogram after 6 years at the arrested stage. Transient ventricular reflux and convexity flow are observed.

total value of fatty acid in CSF was measured at 12 months and 2 years after operation, respectively. The value decreased or was within the normal range in cases showing good prognosis while it was very high or showed remarkable fluctuations in the dead and poor cases. In eight of the arrested cases long-term follow-up study proved that the total value of fatty acids decreased to normal range and showed stability after shunting operation (Fig. 9). When the total value of fatty acids was high or showed fluctuations as in progressive cases, the condition of the patient did not improve to an arrested hydrocephalic state.

Discussion

Recent advances in studies of CSF dynamics[1,2,4,19] have contributed to the evaluation of

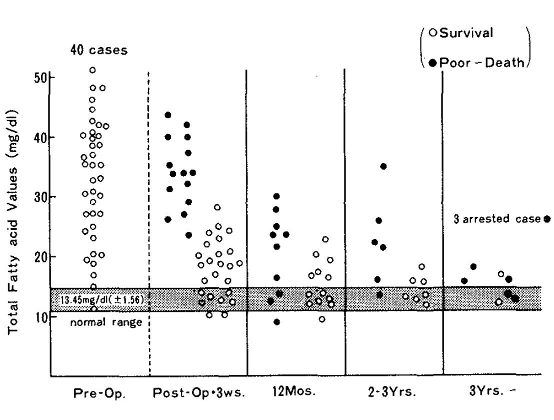

Fig. 9 Total fatty acid values in CSF. Mean value: 13.45 mg/dl ± 1.56.

hydrocephalus, but many unsolved problems remain in arrested and compensated hydrocephalus. In general, conditions of arrested hydrocephalus are clearly different from those of acute progressive hydrocephalus. The term "arrested hydrocephalus" is used by some authors as a synonym of so-called chronic or compensated hydrocephalus.[6,8] For the assessment of the condition of hydrocephalus, various diagnostic methods and tests have been considered, and recently neuroradiological study, CSF dynamic study [3,19,20,25,29] and biochemical study of CSF[11,32] have proven more helpful than any other procedures in resolving these problems in infants and children with hydrocephalus.

Apart from the problem of whether or not infantile hydrocephalus can be arrested naturally[5,14,16,21], prognosis of hydrocephalus is apt to be influenced by etiological factors. Occasionally, we have experienced hydrocephalic cases which have been considered to be arrested after shunting operation. Differing from Foltz's opinion[5], "once a shunt, always a shunt," many investigators have reported arrested states of hydrocephalus postoperatively, and their shunt systems have been removed and the hydrocephalus remained in an arrested state (Table 6).

As to what the condition of arrested hydrocephalus is, Laurence[16] and Matson[28] have already argued about the clinical definition. The criteria of arrested hydrocephalus have been defined by Matson[21] (1969), and Hageberg and Sjögren[8] (1966) as follows: 1) a large head, usually in or above the 90th percentile in normal head growth, 2) a history of satisfactory development until the age of 6 to 8 years in improved cases, 3) mild to moderate ataxia, 4) mild moderated spastic paraparesis, and 5) mental retardation that borders on educability, etc. This conception consists of morphological and neuropsychological considerations.

Recently, many authors have been discussing the application of various measures of CSF dynamics[9,15,20,25,29] to obtain more accurate criteria for the definition of arrested hydrocephalus. The combined use of these procedures such as RI cisternography[7,9,22], intraventricular fluid pressure wave[24] and pulse wave measurements[15], and loading test[3,15,25] can be applied in the study of arrested hydrocephalus. The results may be valu-

Table 6 Percentage of arrested hydrocephalus cases (all types of infantile hydrocephalus)

Reportors	Year Published	Follow up or Post op. (year)	Percentage of arrested cases
1 Hagberg & Sjögren	1962, 1966	6～12	35%
2 Foltz & Schurtleff	1963, 1966	3	77%
3 Laurence & Coates	1958, 1969	2～3	47%
4 Matson & Schick	1961, 1969	4～5	rare
5 Lober & Zachary	1968	7½	83%
6 Milhorat	1972	6 mos～2	
7 Holtzer	1973	1～10½	16.8%
8 Our clinic	1975	2～6	21%

able adjuncts in assessing whether or not the hydrocephalic process is arrested. Thus, arrested hydrocephalus may best be defined in terms of normal CSF dynamics, total arrest of neurological sign and clinical evidence of continued mental-motor development with increasing age. It seems, however, that such cases are very rarely observed. As physiological and anatomical development is characteristic in children, the diagnosis of "arrested" should not be made unless evidence of continuing mental-motor development is shown with increasing age. Matson[21], Milhorat[23] and Matsumoto[27] emphasized these facts. Especially, Matson and Milhorat emphasized that such continuing mental-motor development could be recognized only in cases treated surgically in an early stage and was never seen in naturally arrested cases. The distinction is made on clinical grounds alone and requires a prolonged follow up for adequate documentation. Matson[21] called the attention of physicians to the importance of the correct use of the term "arrested."

Progressive hydrocephalus is usually considered as a state in which unless the patient is surgically treated, irreversible brain damage will occur[16,25,30] and may become compensated hydrocephalus which may be called chronic hydrocephalus.[18] Infant hydrocephalus and adult chronic hydrocephalus (so-called "Normal pressure hydrocephalus") are not exactly the same because of differences in anatomical and physiological conditions. In infants, the two forces produced by the CSF and venous systems can not oppose each other completely because the cranial walls yield. As the ventricle enlarges, the skull also enlarges against atmospheric pressure.[12] For the two forces to oppose each other completely, they require a rigid support to react against. Therefore, the stress within the brain parenchyma is smaller in children. But, brain dysfunction from extreme hydrocephalus in infants and children is seldom superimposed on a wide range of cerebral dysfunction already existing, and the effect of hydrocephalus produces lack of development rather than a loss of acquired capability. We think, therefore, that there is a compensatory mechanism in the human brain to adapt the pressure gradient between the brain tissue and the ventricle. Such a hypothesis has been supported by Milhorat,[23] Hagberg[8] and Holtzer.[13,14]

The arrested state may appear to be normal by 2 years of age.[16] However, as the causes of hydrocephalus are various, clinical evaluation and prognosis are difficult to summarize because of case selection and varying periods of follow up.

In general, post-menigitic hydrocephalus arrives at an arrested state in approximately 3 to 4 years. This clinical tendency was also observed in this study, but the reason for it is not clear. It is probable that a complete surgical arrested state may be due to the relative increase of efficiency of CSF circulation, absorption and improvement in function of arachnoid villi.

Laurence[16] has not only reported on the morbidity and mortality of an original 182 hydrocephalus infants, but on the neurological and intellectual development of the untreated survivors. In a report, published in 1962, he emphasized that no correlation was found between intelligence, head size, cerebral mantle thickness or duration of progressive disease. Many investigators stated that the head circumference, thickness of the cerebral mantle and intelligence could not be correlated. However, recent clinical surveys by many authors[10,26,31] have revealed that infants with hydrocephalus were more likely to have a good IQ when they were treated before the age of 2 to 6 months. Lorber[18] and others[21] have also emphasized the correlation with age, but they found no correlation between the preoperative thickness of the cerebral mantle and ultimate intellectual development. These findings suggest that as the young infant's brain is in some way more vulnerable than that of the older child, hydrocephalus should be managed in the early stage. Histological studies of infantile hydrocephalus[23,30] revealed progressive gliosis in chronic hydrocephalic state together with finding of cerebral atrophy. We[10] have previously found that adequate treatment of congenital hydrocephalus with a cerebral mantle thickness of more than 25 mm within three months after birth will nearly always result in a child with the potential for normal or relatively unimpaired intelligence.

The characteristics of cranial enlargement in infantile hydrocephalus have been previously described.[9,10] An abnormally enlarged head may decrease in size after shunting operation, and then continuous head growth usually is resumed at normal rate within 1 year after the operation.

In Laurence's series,[16,17] "arrest" took place between the age of 9 months and 2 years. However, he has not accurately presented his criteria for arrested hydrocephalus.

On the other hand, Matson[21,28] states that if the head size shows above the 90th percentile of figures taken from standard growth curves and until the child does not completely catch up to his normal place on the curve, the hydrocephalus cannot in fact be said to have reached an arrested state. But, the authors believe that evaluation based on only a morphological conclusion is not sufficient.

Fundamental[9,19,24] and clinical studies[3,9,25] of CSF dynamics expedited progress in the accurate criteria for the definition of arrested hydrocephalus. Recently, many valuable results have been reported by using the pattern of CSF bulk flow and absorption by isotope cisternography,[7,22] indirect measurement of CSF absorptive capacity by constant infusion saline manometry, and analysis of mean CSF pressure measurement.[3,5]

Especially, Foltz[9] investigated the intraventricular wave form and loading of the CSF compartment by saline to demonstrate an effect of pressure in the ventricle as well as in the subdural space in hydrocephalic patients. In the progressive hydrocephalic patient, the ventricular and subdural wave forms are very similar. In arrested hydrocephalus, the ventricular wave form is considerably modified by passing through the overlying brain.

Generally, three factors, that is intracranial CSF pressure, ventricular volume and intracranial blood volume play an important role in the compensatory mechanism in hydrocephalus.

In our study, we used the rapid saline injection and loading test in compensated and arrested hydrocephalus.

We considered that the compensated state was a loss of relationship of three factors.

There were also differences in intraventricular pulse wave form between the progressive hydrocephalus with thin cerebral mantle and arrested hydrocephalic patients with thick cerebral mantle, and the significant difference was considered to have relation to cerebral blood volume/pressure and CSF volume/pressure.

In progressive hydrocephalic patients, the ventricular fluid pressure (VFP) curve often showed a considerable amount of fluctuation of amplitude including episodes of very high pressure such as the plateau waves during long term monitoring.[6,9,20] While, these waves were not observed in patients with arrested hydrocephalus.

Radionuclide cisternography has been demonstrated to be a useful diagnostic procedure in the evaluation of hydrocephalus in children. In progressive hydrocephalus there were uniform demonstration of ventricular reflux or stasis with delayed clearance from the ventricle and subarachnoid space.[6,9,22,23] At the arrested state, transient ventricular filling with convexity flow was seen, but clearance was more rapid than in the progressive cases. In our three cases, it is evident that the decrease of ventricular stasis or transient ventricular filling accurately indicates arrested tendency. Various changes in patterns of RI cisternography in the transitional process to the arrested state are influenced by the enlargement of the ventricle and the pathological change of the subarachnoid space. Therefore, additional information on quantitative RI studies[9] in follow up of longer periods will be required.

The total value of fatty acid in CSF remarkably increased in intracranial hypertension. Fatty acids increase has been also reported[9] in demyelinating diseases of the central nervous system. Authors[32] described the total value of fatty acid and each fatty acid composition in CSF before and after the shunting operation for the treatment of infantile hydrocephalus.

When abnormally high values of total fatty acid continue after the shunting operation, the existence of pathological conditions were estimated as: 1) fatty acid released into the CSF through the destructed blood-brain barrier; and 2) the shunting system is functioning well, but the synthetic processes of the lipid or fatty acid are abnormal in the brain tissue.

From our experience, if the total value of fatty acid decreases to normal range, these patients may be considered as having a surgically arrested hydrocephalus. When intracranial hypertension continued, the total value of fatty acid was abnormally increased, and these results were considered partly due to the possibility of brain edema, brain tissue damage or abnormal metabolic change. In infancy or early childhood, the myelination of the brain tissue is very remarkable, and as a result, destructive processes are apt to cause more considerable damage to the developing brain.

In arrested hydrocephalus, we obtained the following results: 1) abnormally high value of the total fatty acid decreased after the shunting operation; 2) no change in any fatty acid content even in a period of 2 to 3 years after the operation; 3) the values of the total fatty acid related to a state of intracranial pressure; and 4) when a high or unstable value of total fatty acid existed in the course of several months to a year after the shunting operation, some remarkable organic change of the brain, which was different from the arrested hydrocephalus, was considered.

We are confident that the study of fatty acid metabolism in CSF together with protein and amino acid metabolism is significant, and applies to the effect of CSF pressure on the brain tissue in comparison of total fatty acid before and after shunting operation.

In the present study we investigated the clinical significance of neurological findings, isotopic study, roentgenological studies, psycho-motor development, CSF dynamics and fatty acid metabolism in CSF, individually, at the preoperative state and the arrested state after the operation.

It is certain that arrested hydrocephalus exists in varying degrees as causes of hydrocephalus vary. Various-dimensional studies gave us valuable data for the analysis of hydrocephalus. Diagnosis of arrested hydrocephalus should be made by collective judgement of the results of various examinations. It can not be evaluated on the basis of a simple examination.

In conclusion we should like to say that arrested hydrocephalus is difficult to differentiate clinically from compensated hydrocephalus. In compensated hydrocephalus compensatory balance takes place, as reported by Milhorat,[23] to eliminate the pressure gradient between the brain tissue and the ventricle, that is, not only abnormal absorption of CSF but also such compensatory adjustments as ventricular enlargement, cerebral atrophy, and decrease of cerebral blood volume. Accordingly, we considered the compensated state to have bad influence on the brain, and the state cannot be said to be good. Surgical efforts should be made as much as possible in order to improve progressive hydrocephalus into an arrested state.

References

1) Bering, E. A. Jr.: Pathophysiology of Hydrocephalus. Workshop in Hydrocephalus Philadelphia, Univ. Pensylvania Press., 9–28, 1966
2) Bering, E. A. Jr. and Sato, O.: Hydrocephalus; Changes in formation and absorption of cerebrospinal fluid within the cerebral ventricle. *J. Neurosurg.* 20: 1050–1063, 1963
3) Bird, M. T., Ratcheson, R. A. and Seigel, B. A., et al.: The evaluation of arrested communicating hydrocephalus utilizing cerebrospinal fluid dynamics. *Develop. Med. Child Neurol.* 15: 474–482, 1973

4) Epstein, C. M.: The distribution of intracranial forces in acute and chronic hydrocephalus. *J. Neurological Sciences.* 21: 171–180, 1974
5) Foltz, E. L. and Shurtleff, D. B.: Five year comparative study of hydrocephalus in children with and without operation (113 cases). *J. Neurosurg.* 20: 1064–1078, 1963
6) Fox, J. L., McCullough, D. C. and Green, R. C.: Effect of cerebrospinal fluid shunts on intracranial pressure and on cerebrospinal fluid dynamics. *J. Neurol. Neurosurg. Psychiat.* 36: 302–312, 1973
7) Glasauer, F. E., Alker, G. J. Jr. and Leslie, E. V.: Isotope cisternography and ventriculography. *Amer. J. Dis. Child.* 120: 109–114, 1970
8) Hagberg, B. and Sjörgren, I.: The chronic brain syndrome of infantile hydrocephalus. *Amer. J. Dis. Child.* 112: 189–196, 1966
9) Harbert, J. C., *et al.* (eds.): Cisternography and Hydrocephalus (A Symposium). C. C. Thomas, Ill, U.S.A., 1972
10) Ito H., Onodera Y., Miwa T., *et al.*: Some observation on prognosis in congenital hydrocephalus (First report)—with reference to the preoperative evaluation of the hydrocephalic infants—. *Neurol. Surg.* 3: 245–254, 1975
11) Ito H., Onodera Y., Miwa T., *et al.*: Some observations on prognosis in congenital hydrocephalus (Second report)—with reference to the physical handicap in survival cases—. *Neurol. Surg.* 4: 313–322, 1975
12) Hochwald, C. M., Epstein, F., Malhan, C., *et al.*: The relationship of compensated to decompensated hydrocephalus in the cat. *J. Neurosurg.* 39: 694–697, 1973
13) Holtzer, G. J. and de Lange, S. A.: Mental condition and ventricular size in arrested hydrocephalus; An analysis of 29 shunt-independent children. *Develop. Med. Child. Neurol.* Suppl. 25: 39–43, 1971
14) Holtzer, G. J. and de Lange, S. A.: Shunt-independent arrest of hydrocephalus. *J. Neurosurg* 39: 698–701, 1973
15) Tajima K., Ito H., and Miwa T.: The clinical study of ventricular fluid pressure in hydrocephalic patients. *Neurol. Surg.* Part II., in press, 1976
16) Laurence, K. M. and Coates, S.: The natural history of hydrocephalus. Detailed analysis of 182 unoperated cases. *Arch. Dis. Child.*: 345–362, 1962
17) Laurence, K. M.: Neurological and intellectual sequelae of hydrocephalus. *Arch. Neurol.* 20: 73–81, 1969
18) Lorber, J. and Zachary, R. B.: Primary congenital hydrocephalus. Longterm results of controlled therapeutic trial. *Arch. Dis. Child.* 43: 516–527, 1968
19) Lorenzo, A. V., Page, L. K. and Watters, G. V.: Relationship between cerebrospinal fluid formation, absorption and pressure in human hydrocephalus. *Brain* 93: 679–692, 1970
20) Markakin, E.: Continuous ICP monitoring as aid for the indication of shunting procedures in children. In "Progress in Paediatric Neurosurgery," pp. 163–166. Hippokrates Verlag, 1972
21) Matson, D. D.: "Neurosurgery of Infancy and Childhoood," 2nd ed., C. C. Thomas, Springfield, Ill., 1969
22) McCullough, D. C., Harbert, J. C., Miale, A. Jr., *et al.*: Radioisotope cisternography in the evaluation of hydrocephalus in infancy and childhood. *Radiol.* 102: 645–652, 1972
23) Milhorat, T. H.: "Hydrocephalus and The Cerebrospinal Fluid," Baltimore, U.S.A., 1972
24) Obeuchian, T. G.: Continuous pressure monitoring in experimental obstructive hydrocephalus. *Arch. Neurol.* 29: 287–298, 1973
25) Page, L. K., Bresnan, M. J. and Lorenzo, A. V.: Cerebrospinal fluid perfusion studies in childhood hydrocephalus. *Surg. Neurol.* 1: 317–321, 1973
26) Raimondi, A. J. and Soare, P.: Intellectual development in shunted hydrocephalic children. *Am. J. Dis. Child.* 127: 664–671, 1974
27) Matsumoto S.: Pathophsiology and management of compensated hydrocephalus or arrested hydrocephalus. Panel discussion V., 34th Annual Meeting of the Japan Neurosurgical Society, Oct. 24, 1975
28) Schick, Ŕ. W. and Matson, D. D.: What is Arrested Hydrocephalus. *J. Pediat.* 58: 791–799, 1961
29) Trotter, J. L., Luzecky, M. and Siegel, B. A., *et al.*: Cerebrospinal fluid infusion test. *Neurology.* 24: 181–186, 1974
30) Weller, R. O., and Williams, B. N.: Cerebral biopsy and assessment of brain damage in hydrocephalus. *Arch. Dis. Childh.* 50: 763–768, 1975
31) Young, H. F., Nulsen, F. E., Weiss, M. H., *et al.*: The relationship of intelligence and cerebral mantle in treated infantile hydrocephalus. *Pediat.* 52: 38–54, 1973
32) Onodera Y., Ito H., and Miwa T., *et al.*: The quantitative fatty acid composition of cerebrospinal fluid in congenital hydrocephalus. *Brain Develop.* 7: 23–28, 1975