Fontanelle Pressure (ICP) in Infantile Hydrocephalus —Investigation of ICP in 6 Cases of Hydrocephalus with Normal Head Circumference and without Shunting Operation—

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Summary: A follow up study on ICP was carried out in 6 cases of hydrocephalus within normal head circumference during the period 8-9 months after birth. There were no shunting operations. An abnormal plateau like wave recorded in 2 cases with myelomeningocele during the follow-up period. In other cases, almost normal baseline pressure was recorded without abnormal pressure wave and all these cases showed favorable morbidity (function). These results indicate that the importance of a follow-up study on ICP for months after birth in such cases as hydrocephalus without large head circumference; Furthermore, shunting operation should be recommended in cases showing abnormal pressure wave during the follow-up period.

Key words: fontanelle pressure — infantile hydrocephalus — normal head circumference — REM (Rapid Eye Movement) period, pressure wave — baseline pressure

Introduction

Infantile hydrocephalus shows a variety of spinal fluid circulation and ventricular configurations. A much debated problem, such as a differentiation of the types of hydrocephalus (compensated, arrested or slowly progressive), is not yet clearly solved. In this report, we studied the correlation between the prognosis and the fluctuation of intracranial pressure or ventricular configuration, in particular, we analyzed the 6 cases follow-up for three to five years without shunting operation because the head circumference corresponded to the upper limit of the normal range.

Methods

The psychomotor development of the

infants was estimated according to the Enjoji's method, while the Tsumori's method was employed additionally in the patients older than four years and 8 months. The head circumference and size of the cerebral ventricle were simultaneously measured at sequential intervals. The CT scan made it possible to calculate ventricle; brain-volume ratio (VBVR) (Honda et al. 1979) on mini-computer by comparing the ventricular cross-section area to brain cross-sectional area by using the following equation; $R = (\sum_{\lambda=2}^n V_i / \sum_{\lambda=2}^n B_i) \times 100$.

Intracranial pressure (ICP) was estimated continuously for over five hours through the anterior fontanelle by using APT-16 transducer (Shojima 1980) and F.P. sensor (Honda et al. 1982). Being measured only during sleep, the baseline pressure in non-REM period and the pres-

sure wave pattern, as well as the amplitude in REM period, were estimated. RI cisternography was studied in some patients.

Summary of Cases

Case 1

Five years and four months old, female, postmeningitic hydrocephalus.

Septic meningitis occurred in the second month after birth with signs of increased ICP such as vomiting, irritability and enlargement of head circumference ($<+2SD$). The baseline pressure of ICP was as high as 170 mmH₂O and the ventricle showed a rapid expansion up to the greatest VBVR of 50 % within 2 months. In the 4th month, ICP became stable, with the baseline pressure being 120-130 mmH₂O, and the fluctuation of the pressure in the REM period seemed to be smaller than normal. psychomotor development remained in the normal range until three years old, although a slight retardation was recognized after 5 years of age (Fig. 1).

Case 2

Five years and five months old, female, postmeningitic hydrocephalus.

In the third month after birth, she suffered from meningitis with only a transient rise of ICP. In the fifth month, the baseline pressure became 130 mmH₂O with the highest ICP in REM period being around 220 to 240 mmH₂O and only a slight increase of amplitude. The VBVR had shown a constant value of 24 % for three years and the psychomotor development was quite normal (Fig. 2).

Case 4

Three years and six months old, female, postmeningitic hydrocephalus.

In the two and half months after birth, aseptic meningitis developed with a transient increase of ICP. The baseline pressure became stable being around 120 mm-

H₂O. On continuous measurement of ICP, a rise of amplitude in REM period twice as much as that in non-REM period was observed only once in the fifth month but became normal in the sixth month. Although the VBVR was 31 % in the third month, it tended to reduce to 22 % in three years, compared with the development of head circumference. The follow-up study for three and half years showed that psychomotor development was completely normal (Fig. 3).

Case 5

Three years old, male, hydrocephalus following the operation of meningocele.

Sacro-lumbar meningocele was operated upon shortly after birth. Thereafter, ventricular enlargement occurred slowly, the VBVR being from 23 to 25 %. Neither a abrupt manifestation of clinical symptoms nor a rapid expansion of the cerebral ventricle has been observed. The amplitude of ICP in REM period increased from the 16th day on compared with the normal average, which seemed to indicate the reduction of capacity in the buffer system, whereas he was followed up without any manipulation. The ICP became stable after one month, the baseline pressure (100 mmH₂O) was almost within normal limits. However, a plateau like wave around 250 mmH₂O appeared in REM period from the latter half of the sixth month. On the other hand, an enlargement of the subdural space became manifest in the CT scan. The psychomotor development after three years seemed to be within normal limits (Fig. 4).

Case 6

Five years old, female, postoperative hydrocephalus after meningocele.

Radical operation of lumbar myelomeningocele was carried out shortly after birth. The cerebral ventricle has enlarged progressively during a few postoperative months, the greatest VBVR being 38 %.

ICP Via Fontanelle During Sleep

Fig. 1. Case 1 (postmeningitic hydrocephalus)

ICP at 2 months after birth showed a relatively high baseline pressure of (170 mmH₂O). At 4 months, ICP remained normal (120–130 mmH₂O) without elevation and oscillation even in REM period. Serial CT scan demonstrated a marked ventricular dilatation (VBVR 50%) without progressive hydrocephalus. This stage is considered to be so-called hydrocephalus disproportion.

However, the circumference of head remained within the range from +1SD to +2SD. The baseline pressure of ICP after one month from birth showed 120 mmH₂O, just on the border line of normality. From the fifth month after birth, a plateau like

wave appeared in REM period, of which the amplitude attained a value more than twice as great as that in non-REM period. The psychomotor development was almost within normal limits up to one to two years of age, but a moderate disturbance and a

ICP Via Fontanelle During REM Period

T.K (F)

Fig. 2. Case 2 (postmeningitic hydrocephalus)

ICP at 5 months after birth showed baseline pressures of 130 mmH₂O and 220–240 mmH₂O during REM period. CT scan demonstrated a mild dilatation of the ventricular system (VBVR 24%).

retardation of intelligence were observed at 5 years (Fig. 5).

Results

The developmental curve of the head circumference remained within the limits from +1SD to +2SD in all six cases. Of four cases with normal psychomotor development, three (except for case 5) pre-

sented no pressure wave in the follow-up period of eight to ten months after birth. The enlargement of the cerebral ventricle was moderate, the VBVR being about 25%. The VBVR of 12 normal infants ranged from 2.4% to 7.8%. In cases 1 and 6 with disturbed psychomotor development, the cerebral ventricle enlarged rapidly in a short time, the VBVR being about 40% or more. The baseline pressure of ICP in case 1 was 170 mmH₂O within two months

CASE 4

ICP Via Fontanelle During Sleep

Fig. 3. Case 4 (postmeningitic hydrocephalus)

Although ICP showed base line pressure of 120 mmH₂O at 2 weeks after birth, a transient plateau-like wave during REM period was observed at 5 months, which returned to normal range one month later. Serial CT scan demonstrated gradual decrease in the ventricular size.

after birth and case 6 presented with a plateau-like wave, which was observed in case 5 as well, necessitating frequent follow-up examinations though the pressure is normal now in their third year. The RI cisternography was carried out in cases 1

to 4. The ventricular reflux and RI retention in 48 hours were observed in all cases. when reexamined after 1.2 years in case 4, the spinal fluid circulation was demonstrated to be normal (Table 1).

ICP Via Fontanelle During Sleep

Fig. 4. Case 5 (hydrocephalus with lumbo-sacral myelomeningocele)

Despite a slowly progressive ventricular dilatation after surgery, ICP remained within normal baseline pressure of 100 mmH₂O at one month after birth and a plateau-like wave (250 mmH₂O) was recorded during REM period at 6 months. Serial CT scan showed a dilatation of the subdural space and of the longitudinal fissure.

Y.I (F)

ICP Via Fontanelle During Sleep

Fig. 5. A case of hydrocephalus with lumbar myelomeningocele ICP at one month after birth remained within normal, baseline pressure of 120 mmH₂O during REM and non-REM period. However, a plateau wave of high amplitude was recorded during REM period at 5 months after birth. The amplitude was 2 times higher than that of non-REM period. Serial CT scan showed a progressive enlargement of the ventricular systems with dilatation of the subdural space and the longitudinal fissure.

TABLE 1 (1)

Case No.	Etiology	I C P			Head Circumference
		Highest base-line pressure mmH ₂ O	Wave Pattern	Ratio Amplitude (REM/non-REM)	
Case 1	Meningitis	170 (2.5M)	Normal	≪ 2	+1SD~+2SD
Case 2	Meningitis	130 (5M)	Normal	≪ 2	+1SD~+2SD
Case 3	Meningitis	150 (7M)	Normal	≪ 2	+1SD~+2SD
Case 4	Meningitis	150 (7M)	Normal	≪ 2	+1SD~+2SD
Case 5	Meningocele	130 (1M)	Plateau like Wave	2 <	+1SD~+2SD
Case 6	Meningocele	160 (7M)	Plateau like Wave	2 <	+1SD~+2SD

Discussion

Laurence (1962) reported that the infantile hydrocephalus does not progress further after two years, the majority becoming stable within 9 months. Though the differentiation of the cases into arrested (Dirocco et al. 1977), compensated or slowly progressive types (Gordon et al. 1977) is still an unsolved problem, it may be of value in estimating the progress of hydrocephalus to study the changes of ICP and ventricular configuration while the anterior fontanelle is still open about nine months after birth.

From the etiological point of view, it must be noted that the hydrocephalus in three cases out of four was a sequel to meningitis, when the primary brain damage was slight. In secondary hydrocephalus after meningitis, the intraventricular absorption (Sahar, 1969) seems to play an important role in the balance of production and absorption of spinal fluid. However, according to the follow-up data of CT in the above mentioned three cases, the subependymal area became more clearly demarcated against the cerebral ventricle with age, suggesting that another route

of absorption of the spinal fluid might exist. The ventricle of these cases enlarged moderately, the VBVR being about 25 %. Whether they were of compensated or arrested type, the enlargement was rather limited compared with the findings of Osaka et al. (1977).

The remaining two cases were those of hydrocephalus following the meningocele operation. Previous reports stated that hydrocephalus with myelomeningocele tends to become arrested (Hammock, 1976; Shurtleff, 1975) and the head circumference particularly in the elderly infants fell within the normal range, with which the findings of these two cases were completely in accord.

When the ICP was estimated with F.P. sensor or APT-16 transducer, the baseline pressure of 15 neonates within one month of birth was 65 ± 27 mmH₂O and that of three infants aged between one and two months was 81 ± 24 mmH₂O. Neonates of less than one month demonstrated only a slight rise in the value of ICP during REM period (Fig. 6). It has been explained in the following way; the cavity of the skull is rather open without a fusion of sutures; the contractions of the heart

TABLE 1 (2)

Ventricles			Motor and Mental development (Age in last examination)	RI Cisternography
Maximum VBVR	VBVR at Last CT	(Age)		
50 %	88 %	4 y	Slight Deterioration (5 y)	RI Ventricular Reflux and Retention (48 hr) (3M)
31 %	24 %	4 y	Normal (5 y)	RI Ventricular Reflux and Retention (48 hr) (5M)
28 %	28 %	3 y	Normal (3 y)	RI Ventricular Reflux and Retention (48 hr) (4M)
31 %	22 %	3 y	Normal (4 y)	RI Ventricular Reflux and Retention (48 hr) (3M)
24 %	26 %	3 y	Normal (3 y)	not performed
38 %	36 %	3 y	Middle Retardation (5 y)	not performed

ICP Via Fontanelle During Sleep

Fig. 6. ICP recording at 15 days after birth.

- (1) ICP in a normal infant showed baseline pressure of 70 mmH₂O without elevation of ICP during REM period.
- (2) ICP in infant with marked hydrocephalus showed extremely high ICP above 200 mmH₂O associated with a relatively low amplitude wave, even in REM period.
- (3) In a moderately hydrocephalic infant, the baseline pressure was 170 mmH₂O and 250 mmH₂O in REM period with high amplitude wave.

and blood vessels are weak; the transmission of pulse is weakened through the high water content of the white matter of newborn infants (Volpe et al. 1977). We postulated, however, that it was caused by the relatively large buffering effects of the venous and spinal fluid space of the newborn.

The baseline pressure within two months after birth of 18 cases in which progressive hydrocephalus was developed (including those operated upon by shunt) was 148 ± 29 mmH₂O. When compared with normal values, 120 mmH₂O may be regarded as a borderline between normal and abnormal values, which has also been called a critical pressure. The value in these cases increased in REM period probably due to the diminution of the buffer system, whereas it disappeared both in non-REM and REM periods in those cases with extremely advanced hydrocephalus (Fig. 7). The observation was accord with the Wald et al.'s (1974) report in whose the maximum skull wrapping with silicone caused a rise in ICP and the disappearance of amplitude. A collapse of vascular space is considered to occur and to cause an extensive brain damage.

As a normal baseline pressure in infants after three months from birth is often about 120 to 130 mmH₂O for the reasons given above, the pressure in REM period is of rather more value than the baseline pressure. We have considered that a fluctuation of ICP in REM period is important as a natural burden in the intracranial system. The rise of pressure in REM period and the plateau wave are often observed in adult cases with normal pressure hydrocephalus as well as in infants with hydrocephalus (Cooper, 1966; Hayden, 1970; Pierre Karn 1976). Various opinions have been presented on the mechanism of a rise of pressure in REM period; 1. an increase of blood flow induced by catecholamines (Reivich, 1976) 2. an increase of the vascular bed by the

Fig. 7. Type of abnormal pressure wave in hydrocephalic infant

- (1) This case is one of progressive hydrocephalus with myelomeningocele. Lundberg's plateau wave was demonstrated at 4 months after birth, although this type of wave was rarely seen in a hydrocephalic infant.
- (2) B-like wave: A rhythmical pressure wave showing 100 mmH₂O lower than the baseline pressure which simulated Lundberg's B wave.
- (3) Plateau-like wave: REM period ICP rose up to 270–400 mmH₂O with a high amplitude twice as great as that of non-REM period. Most cases of infantile hydrocephalus showed this ICP pattern.
- (4) ICP recording in normal infant at 6 months after birth.

lowered cerebral vasomotor tone through a hypothalamic action (Risberg, 1969) (Shima et al. 1980). 3. disturbed venous return (Shutleff, 1975). All invariably point to an

increase in cerebral blood volume, although the exact value during REM period is not clearly established, due to individual differences. Since the ICP in REM period of normal infants after three to six months from birth is about 200 mmH₂O, it may be calculated to be at least 10 ml according to Shulman and Marmarou's (1971) pressure volume considerations in infants.

Fig. 7 shows the pressure wave in cases with progressive hydrocephalus operated upon by shunt in three to four months after birth with good prognosis. Plateau waves were observed in only one of 18 cases. As this case had an Arnold-Chiari's malformation, the disturbance of flow in the straight sinus might have played some role (Shurtleff, 1975). It has been reported by Lundberg (1961) that the plateau wave rages from 500 to 1000 mmH₂O, although it is infrequently encountered in infantile hydrocephalus (Liguori et al. 1979) (Paraicz, 1975). The second abnormal wave, B-like wave, appears in the sleep stage before REM period. Martin (1971) concluded that sporadic B wave was probably physiological, whereas the rhythmic one, which indicates some abnormalities, is not due to respiration. Our findings are in accordance with this observation; Whereas some secondary respiratory changes can be observed. This type of wave appeared for only a short time, about 10 to 15 minutes. The third abnormal wave was a plateau one, the maximum pressure being 250 to 400 mmH₂O. A greater part of waves in infantile hydrocephalus belonged to this type. Another salient feature of this type is the increasing rate of amplitude in REM period. When compared with normal infants of the same age, the amplitude of hydrocephalus cases in REM period increased more than twice as much as that in non-REM period (Fig. 7). Gücer et al. (1979) reported that experiment studies of ICP in normal monkeys revealed a definite increase of pres-

sure in REM period although the increase was less than twice as much as that in non-REM period. Plateau and plateau like waves were also observed in other types disease such as subdural effusion (Honda et al. 1978, 1979) or tumor with slow progress (Liguori et al. 1979). On the correlation curve of volume and pressure, the appearance of a plateau wave indicates a right-ward deviation of the intracranial circumstances, that is the buffer system around the venous and spinal fluid space approaches the break point (Granholm, 1976). A rapid rise of ICP and amplitude is caused by an increase of the cerebral blood volume in REM period. Symon et al. (1972) reported that B wave was characteristic of low pressure hydrocephalus. Venes (1979) also discussed that fluctuations on the side of the capillary bed might arise with an increase of the cerebral blood volume in the those cases of low compliance, but the reason is not yet established.

Analyzing six cases without shunt in this report based on the above data, Case 1 to 4 showed the fluctuation similar to normal ICP after six months from birth, in cases with a shunt operation. The baseline pressure of ICP in case 1 attained 170 mmH₂O from the second month after birth. In accordance with the report of Gordon et al. (1977) which describes that the cerebral ventricle starts to enlarge as ICP approaches 170 mmH₂O, the cerebral ventricle enlarged rapidly after four months from birth (VBVR=50 %); in contrary, the baseline pressure of ICP decreased to 120 mmH₂O, with little pressure fluctuation. Shulman (1971) defined a state of craniocerebral disproportion as a single cause of enlargement of the cerebral ventricle without a proportionate change of the head circumference. Our case 1 may correspond to this, missing the chance of shunt operation.

Sixty-five percent of cases with myelomeningocele developed hydrocephalus,

while the enlargement of the head circumference remained mostly around the upper limit of normal range. Case 6 presented with a pressure wave of 300 mmH₂O, with progressive enlargement of the ventricle. Case 5 presented a plateau like wave of 250 to 270 mmH₂O without progressive enlargement of the ventricle. Case 5 must be followed attentively and Case 6 should have been operated upon.

Cases of hydrocephalus with stationary head circumference or that around the upper limit of normal range do not always indicate the state of craniocerebral disproportion, as in Case 1. The reason why the head circumference remained unchanged may be explained differently in the following cases; in three cases with hydrocephalus after meningitis, it was mainly due to the fact the baseline pressure remained under 170 to 180 mmH₂O during a transient increase of pressure; secondly, due to a slow progress of pressure compensation and an improved CSF circulation with the development of the head circumference. On the other hand, in cases with hydrocephalus after myelomeningocele surgery, it may be due to an enlargement of the subdural space including the hemispheric fissure.

The parameters on psychomotor development are rather unreliable during the period from birth to three years of age, and only after five years, does it become reliable (Dirocco, 1977) (Hagberg, 1962). Therefore, a continuous measurement of ICP in infants may be of importance for an early and appropriate therapy. In future prospect, it is desirable that a continuous, non-surgical method of measuring ICP in those cases with closed fontanelle, for which the transducer employed in this study is of no use, should be developed and, in the cases of open fontanelle the data concerning ICP should be obtained.

Conclusion

Continuous measurement of ICP has been carried out for an average of 9 months after birth in cases with enlarged cerebral ventricles. Since many cases presented with abnormal ICP fluctuation, measurement may be of great importance in demonstrating the need for proper treatment as early as possible.

We followed six cases of hydrocephalus with normal head circumference for three to five years. Two cases with meningocele presented with a plateau-like wave in ICP and one case at least had an indication for shunt operation. One of four cases with hydrocephalus after meningitis proceeded to the state of craniocerebral disproportion and the other three with normal psychomotor development showed a normal fluctuation of ICP, so there was no indication for a shunt operation.

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