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Discussion to Dr. Pudenz' Speech (1)

Hydrocephalus and its various subtypes

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Mr. President! Colleagues and Guests!

I am greatly impressed with Dr. Pudenz' excellent presentation of his own invention, the ventriculoatrial shunt. Since I am asked by President Mitsuno to discuss his paper, I would like to add our small experiences of the procedure and would then refer to the subtypes of infantile hydrocephalus, namely, hydroencephalodysplasia and Chiari-Arnald syndrome.

Hydrocephalus is a state that the cerebrospinal fluid abnormally accumulates in the cranium to cause intracranial hypertension. There must be many types and many causes. Except those due to tumors, we have experienced 133 cases of hydrocephalus for the last ten years, as shown in Table 1. The veniriculoatrial shunt has been performed on 36 cases, as seen in Table 2. Good results are obtained in 70% of the cases in the follow-up. In spite of the complications listed in the slide, I believe that

Table 1. Type of Hydrocephalus
133 cases

1 non-communicating hydrocephalus..55	
stenosis of Foramen Monroi..... 1	
aqueductal stenosis 5	
Dandy-Walker syndrome12	
Dandy-Walker syndrome with cervical meningocele 1	
secondary basilar impression (chondrodystrophia (foetalis)(4)	
site of lesion unknown24	
post-meningitic 6	
2 communicating hydrocephalus18	
3 dysgenetic hydrocephalus (hydroencephalodysplasia) 9	
4 type unknown 9	
5 Chiari-Arnold syndrome.....42	
with spina bifida (S-type)15	
without spina bifida (C-type).....14	
with basilar impression (B-type)13	

Table 2. Ventriculo-Atriostomy for
Infantile Hydrocephalus

I No. of operations	36 cases
Age distribution	2m-7y
Type of hydrocephalus	
non-communicating	19 cases
communicating	9
dysgenetic	5
Chiari-Arnold	3
II Results of operation	
(A) Follow up studies (the longest follow up: 5 years) improved	30 cases
at present in good condition	25 70%
not improved	6
(B) Postoperative complications in earlier stage	
acute cardiac arrest	1
acute pulmonary edema	1
obstruction of the tube	3
local wound infection	3
in later stage	
infection (meningitis, sepsis)	3
obstruction of the tube (venous thrombosis)	5
distracted shunts due to growth	3
(C) Revision	8

this shunting operation is the most effective procedure for the treatment of hydrocephalus at present and should be performed as early as possible before intracranial hypertension does cause severe cerebral damages.

The most important point of this procedure is to place the distal end of the cardiac tube precisely into the right atrium. As shown in Figure 1 and Figure 2, we are checking the position of the tip by the following three guides:

- 1) measurement of the distance between the inserting point and the right atrium.
- 2) changes of speed of dripping infusion of physiological saline through the cardiac tube, as the tube gradually proceeds in the venous system. (When the distal end of the tube is in the right atrium, the dripping speed is maximum, usually more than 1.5 times as the initial speed.)
- 3) Changes of ECG. (When the end of the tube goes into the cardiac ventricle, ventricular extrasystole appears in the electrocardiographic tracings. If the extrasystole reverts to a normal rhythm, when the tube is withdrawn a little, the tip is in the right atrium. We are using ordinary ECG techniques as Mark and Sweet¹⁾ did. The intracardiac lead as advocated by Robertson and others²⁾ has not

Three Landmarks to Insert
the Tip Properly into
the Right Atrium

Fig. 1. Three landmarks to insert the tip properly into the right atrium

- 1) Measurement of the length between the orifice (the facial vein) and the upper margin of the sternal end of the clavicle and that between the latter and the r. atrium (on X-ray films)
- 2) Changes of speed of dripping infusion of physiological saline solution through the tube, as the tube gradually proceeds in the venous system
- 3) Changes of ECG (ventricular extrasystole)

Fig. 2. Three landmarks to insert the tip properly into the r. atrium

been successful in our hands because of many artifacts.)

In 5 out of 9 cases of hydroencephalodysplasia or dysgenetic hydrocephaly, the subarachnoid-atrial shunt was performed, with good results in 4 cases. The slide (omitted) shows a case with absence of the frontal, temporal and Rolandic regions on the right side accompanied by accumulation of the cerebrospinal fluid in the subarachnoid space.

We prefer the name "Chiari-Arnold syndrome" to Arnold-Chiari malformation, in honor of Hans Chiari who first described the syndrome in 1891^{3/4)}, 3 years before the report of Julius Arnold⁵⁾. As shown in Table 3,

Table 3. Chiari-Arnold Syndrome (Arnold-Chiari Malformation)

Chiari's Classification (1891, 1895)	our Classification (1964)
1. 14 cases — 1) C-type 14 cases	
herniation of the inferior portion of the cerebellum	ptosis of the inferior portion of the cerebellum
with hydrocephalus 14	without spina bifida
with meningocele 1	
(Age 3½ M—68 Y)	(Age 17 Y—40 Y)
2. 7 — 2) S-type 15	
herniation of the inferior portion of the cerebellum, the brain stem and the 4th ventricle	with spina bifida
with hydrocephalus and spina bifida	
(Age 0—6 M)	(Age 0—4 Y)
3. 1 — 3) B-type 13	
herniation of the cerebellum into the cervical meningocele	with basilar impression
(Age 5 M)	(Age 12 Y—39 Y)

Table 4. Chiari-Arnold Syndrome S-Type (with Spina Bifida) 15 cases

Clinical Symptoms	
Enlarged head circumference	15 cases
Mental retardation	3
Convulsion	3
Visual Disturbance	2
Choked disc	2
Vertical nystagmus	1
Spastic paresis of upper extremities	1
Gait disturbance	5
Spastic paresis of lower extremities	2
Flaccid paresis of lowe extremities	6
Deformity of feet	3
Incontinentia urinae et albi	8
Sensory disturbance of lower extremitis	4

Table 5. Chiari-Arnold Syndrome S-Type (with spina bifida)

Results of operation			
	effective	not effective	dead
Suboccipital decompression			
upper cervical laminectomy	3	1	1
Decompression			
ventriculo-pleurostomy	1	0	0
Decompression			
ventriculo-atrionostomy	2	1	0
Decompression			
ventriculo-cisternostomy	0	1	0
Subarachnoid-peritonostomy	0	1	0
Operative findings			
meningocele	1		
meningomyelocele	14		
		lumbar	4
		sacral	4
		lumbosacral	6
		sacral, cervical	1
cerebellar herniation			
down to the heigh of atlas			5
down to the height of axis			5

we divide this syndrome into three types: the C-type with ptosis of the inferior portion of the cerebellum and no spina bifida (14 cases), the S-type with spina bifida (15 cases) and the B-type with basilar impression (13 cases).

The onset of symptoms in the B-and C-types is mostly after 10 years. whereas the S-type exhibits hydrocephalus before 26 months. Therefore, as

Fig. 3. Chiari-Arnold Syndrome, S-Type
(with Spina Bifida)

infantile hydrocephalus, only the S-type becomes a subject of discussion. The age and sex distribution and the onset of hydrocephalus in the S-type is illustrated in Figure 3.

Clinical manifestations of the S-type are tabulated in Table 4. Table 5 shows the results of operation and the operative findings. Suboccipital decompression is often not enough and shunting procedures are necessary. Good results are obtained in 60% in the follow-up. Early discovery and early treatment will improve the results.

In conclusion, I believe with Dr. Pudenz, the ventriculoatrial shunt is the most effective procedure available at present for the treatment of hydrocephalus, not only of its ordinary types, but of some its subtypes.

Thank you for your attention.

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Discussion to Dr. Pudenz' Speech (2)

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We have been most fortunate to have an occasion to hear Dr. Pudenz discusses his work. His results are clear-cut and his analysis of the clinical data is excellent. I think all can agree, that for the present time at least, his procedure is the one of choice from the many procedures available for the treatment of congenital hydrocephalus.

We too have had experience in his method with good results even though the instruments used by us are modified in some respects to his instruments. For example, in our series there were two slits in the cardiac tube and no diaphragm valve for the flushing device. It will be noted that in tables 1 and 2, the clinical results obtained with this method are obviously better than those obtained by previously used methods. In respect to the newer method however, it must unfortunately be pointed out, that in some instances we experienced an obstruction to the cardiac end of the tube. Out of twenty four cases, five had to be subjected to repeated operation two or three times.

On the other hand, in previous series, a thrombosis of the major arteries or veins was a common occurrence; in the newer method such a complication was not encountered in our experience. I wonder if this finding, together with the experience of other workers in the field of general surgery, does not indicate that thrombosis occurs less frequently in the Japanese. In

Table 1. Results of ventriculoatriostomy (1.5 Months to 3 Years)

1) Alive:	
Hydrocephalus arrested	16
Still progressive	2
2) Dead	5
3) Unknown	1
Total	24

Table 2. Surgical results of hydrocephalus under two years of age

	Alive	Dead within one month	Dead over one month	Unknown	Total
V. mastoidost.		2	2		3
Torkildsen's proc.		1			1
V. peritoneost.		1			1
V. salpingost.				1	1
Plect.		1			1
V. atriost.	11	3	2	1	17
Total					24