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Hydrocephalus Research Update
—Controversies in Definition and Classification of Hydrocephalus—

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Abstract

Classification of hydrocephalus is the most crucial but the most complicated academic challenge within the hydrocephalus research field. The major difficulty in this challenge arises from the fact that the classification is based on almost all subjects in hydrocephalus research, i.e., definition and terminology of hydrocephalus, pathophysiology, hydrocephalus chronology, specific forms of hydrocephalus, associated congenital anomalies/syndrome and underlying conditions, diagnostic procedures for hydrocephalus, and treatment modalities in hydrocephalus. The current status of the classification of hydrocephalus in individual subgroups was reviewed and summarized from publications in the last 60 years (1950–2010), and discussed focusing on the variety of characteristics in hydrocephalus, with more and more new aspects recently disclosed not only in fetal and pediatric but also in adult hydrocephalus. A recently-reported classification of hydrocephalus, ''Multi-categorical Hydrocephalus Classification'' provides comprehensive coverage of the entire aspects of hydrocephalus with current important classification categories and subtypes.

Key words: hydrocephalus, neuroendoscopy, classification and definition, fetal hydrocephalus, adult hydrocephalus, cerebrospinal fluid dynamics

Introduction

''*The basic facts permit an understanding of the dynamics of cerebrospinal fluid (CSF) circulation and of the various types of hydrocephalus*'' (Anthony J. Raimondi, 1972).

Since hydrocephalus is not a single pathological disease but a pathophysiological condition of disturbed cerebrospinal fluid (CSF) dynamics with or without underlying disease, the classification is often confused and complex. There is a great variety of classification categories, items, and criteria. Since hydrocephalus in the individual patient can be classified in various individual categories and items, the combinations of the classified subtypes of hydrocephalus will be almost limitless, i.e., ''congenital/neonatal/primary/syndromic with holoprosencephaly/macrocephalic/non-communicating/internal/high-pressure/progressive'' hydrocephalus etc. in children, and ''adult/idiopathic/normocephalic / dementia / communicating / internal / normal-pressure/progressive'' hydrocephalus, etc. in adults.

Hydrocephalus Research is de facto recognized by neurosurgeons, by *all* other medical and surgical specialties, and by societies. The principles are being established, limits extended and defined, and practitioners identified, but in the one hundred year history of *Hydrocephalus Research*,[1–42] various specific entities, pathophysiology, treatment modalities/indication, and fundamental basic science remain controversial.

It is hoped that a critical review of the epoch-making publications in hydrocephalus may serve as an update on *Hydrocephalus Research*, and simultaneously stimulate present and future hydrocephalus researchers to advance the present knowledge and treatment modalities in hydrocephalus, to more effectively treat the sick children suffering from this malady. In the field of *Hydrocephalus Research* in adults, there is growing new consensus of entities other than classical ''normal pressure hydrocephalus'' (NPH), which have been always debatable in the concept of this entity.[12,30]

Based on such various characteristics in hydrocephalus, with more and more new aspects recently disclosed not only in fetal and pediatric but also in adult hydrocephalus, the current status of classification of hydrocephalus in the individual subgroup was reviewed focusing on the critical points of the individual category of hydrocephalus.

The purpose of this paper is to review the previously proposed or published classifications of hydrocephalus from the fetal to the adult types, and to discuss the various aspects in hydrocephalus based on the current considerable variations in classification categories and subtypes.

Materials and Methods

Previous publications related to *Hydrocephalus* and *CSF Research* were identified using ''PubMed advanced search'' during the period of 1950 to April 20, 2010 with the key words, ''Hydrocephalus'' and/or ''CSF'' either in the ''key words'' or the title from all English publications.

Previous proposals and publications related to classification of hydrocephalus were identified using ''PubMed advanced search'' during the period of 1950 to April 20, 2010 with the key words, ''Hydrocephalus Classification'' either in the ''key words'' or the title from all English publications. Based on the entire review of these publications dealing with classification of hydrocephalus, the historical trends of *Hydrocephalus Research* in Japan are summarized. Critical discussion considers these historical trends to present major proposals for the classification of hydrocephalus in Japan.

Results

Results of the ''PubMed advanced search'' during the period of 1950 to April 20, 2010 for previous publications related to *Hydrocephalus Research* and *CSF Research* are summarized in Fig. 1 and Fig. 2, respectively. The annual numbers of publications related to *Hydrocephalus Research* have been increasing steadily (Fig. 1). The number of publications related to *Hydrocephalus Research* listed in 1950 was just 12, but reached to over 300 within following 60 years. Compared to *Hydrocephalus Research*, the numbers of publications related to *CSF Research* suggested that *CSF Research* was more active than *Hydrocephalus Research* in the 1950s and early 1960s, and has been slowly increasing or unchanged (Fig. 2).

The ''PubMed advanced search'' during the period of 1950 to April 20 identified 2010 previous proposals and publications related to classification of hydrocephalus listed in Table 1. Based on the review of all these publications dealing with classification of hydrocephalus, the historical trend of *Hydrocephalus Research* in Japan was marked down with one recent publication on this topic published in the Journal of Hydrocephalus[13] (Table 2).

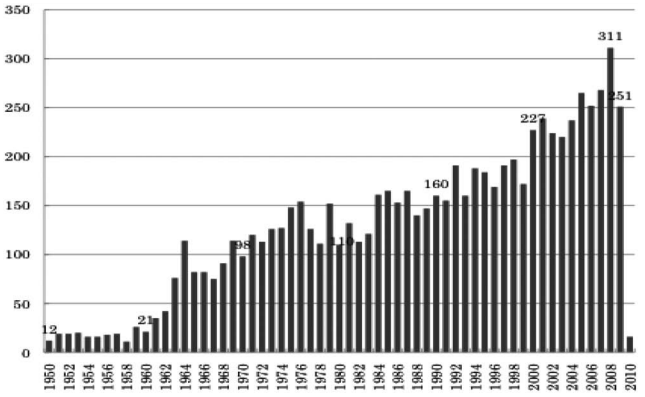

Fig. 1 PubMed advanced search results during the period of 1950 to April 20, 2010 for previous publications related to *Hydrocephalus Research* (accessed on February 17, 2010). The annual numbers of the publication related to *Hydrocephalus Research* have been increasing steadily. The number of publications related to *Hydrocephalus Research* listed in 1950 was just 12, but reached to over 300 within the following 60 years.

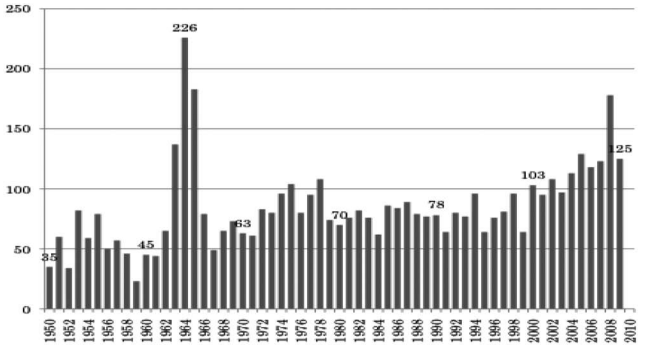

Fig. 2 PubMed advanced search results during the period of 1950 to April 20, 2010 for previous publications related to cerebrospinal fluid (*CSF*) *Research*. The numbers of publication related to *CSF Research* suggested that *CSF Research* was already active in the 1950s and early 1960s, and has been slowly increasing or unchanged.

Discussion

I. Classification of hydrocephalus in CSF dynamics

Raimondi (1972) summarized the basic concept of hydrocephalus in relation to the CSF dynamics as ''*The basic facts permit an understanding of the dynamics of cerebrospinal fluid (CSF) circulation and of the various types of hydrocephalus*.'' Classically, there were two major definitions of disturbed CSF dynamics or circulation in the major pathways, ''*communicating vs. non-communicating*'' by Dandy (1919),[2] and ''non-obstructive vs. obstructive'' by

Table 1 List of publications related to classification of hydrocephalus 1950–2010

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2: Hamilton MG. Treatment of hydrocephalus in adults. Semin Pediatr Neurol. 2009 Mar; 16 (1): 34-41. Review. PubMed PMID: 19410156.
3: Rekate HL. A contemporary definition and classification of hydrocephalus. Semin Pediatr Neurol. 2009 Mar; 16 (1): 9-15. Review. PubMed PMID: 19410151.
4: Newman M, Tucker B, Nornes S, Ward A, Lardelli M. Altering presenilin gene activity in zebrafish embryos causes changes in expression of genes with potential involvement in Alzheimer's disease pathogenesis. J Alzheimers Dis. 2009 Jan; 16 (1): 133-47. PubMed PMID: 19158429.
5: Rekate HL. Shunt-related headaches: the slit ventricle syndromes. Childs Nerv Syst. 2008 Apr; 24 (4): 423-30. Epub 2008 Feb 8. Review. PubMed PMID: 18259760.
6: Czosnyka Z, Keong N, Kim DJ, Radolovich D, Smielewski P, Lavinio A, Schmidt EA, Momjian S, Owler B, Pickard JD, Czosnyka M. Pulse amplitude of intracranial pressure waveform in hydrocephalus. Acta Neurochir Suppl. 2008; 102: 137-40. PubMed PMID: 19388305.
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Contd.

Russel (1949).[39]

It has been generally accepted that the major CSF pathway starts from the bilateral lateral ventricles with the choroid plexus as the significant CSF production source merging with CSF produced in the third and fourth ventricles, and CSF passes outside the ventricular system into the cisterns or subarachnoid space. An appreciable volume of CSF comes from sources other than the choroid plexus in animals (Bering and Sato, 1963),[1] (Sato et al, 1975).[40] The major absorption site is the arachnoid granulation (Pacchionian body) or villi which soaks CSF up into the sinuses, mainly the superior sagittal sinus (Weed, 1914, 1916).[41,42] With the bi-directional volume movement of CSF in the major pathway, the CSF dynamics create the bulk flow (Dandy and Blackfan, 1914).[3] The rate of CSF production is approximately 500 ml over 24 hours in humans and the CSF major pathway contains some 130–140 ml, so there may be physiologic turnover of the CSF three or four times every day.

Based on these conventional concepts of CSF dynamics, the hydrocephalus has been defined as a state of ''disturbed CSF circulation'' and classified classically into two types, communicating and non-communicating (Dandy, 1919).[2] In the definition of Dandy's communicating/non-communicating hydrocephalus, the communication of the CSF pathway is between the lateral ventricle and the lumbar subarachnoid space (confirmed by injection of dye

Table 1 Contd.

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into the lateral ventricle and detection by lumbar puncture). However, the terminology of obstructive and non-obstructive hydrocephalus (Russel, 1949),[39] is defined as a condition of disturbed CSF circulation due to a blockage at any region in the major CSF pathway including the ventricular system and cistern/subarachnoid apace, so that the causes for non-obstructive hydrocephalus are limited to either CSF overproduction by choroid plexus papilloma or CSF malabsorption due to sinus thrombosis or fetal/neonatal hydrocephalus during the period of immature development of the Pacchionian bodies, etc. These two classifications are based on a concept to classify the type of hydrocephalus considering the disturbed CSF dynamics only in the major CSF pathway: ''Major Pathway Hydrocephalus'' (Oi and Di Rocco, 2006).[14]

Since the CSF dynamics during the fetal and neonatal/early infantile periods are mainly maintained in the ''minor CSF pathway,'' hydrocephalus occurring during these periods can be defined as disturbed CSF circulation in the ''minor CSF pathway'' (''Minor CSF Pathway Hydrocephalus'': Oi and Di Rocco, 2006).[14] Therefore, the mechanism or pathogenesis of hydrocephalus may be different from the ''Major CSF Pathway Hydrocephalus.'' The classical

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classification of ''communicating vs. non-communicating'' (Dandy)[2] and ''obstructive vs. non-obstructive'' (Russell)[39] do not correctly reflect the disturbed CSF circulation in the fetal and neonatal/early infantile periods when the major CSF pathway is not yet mature (Fig. 3).

In the CSF circulation, as in the immature form, CSF absorption may possibly be disturbed at various absorption sites including the subpial space→perivascular space→subarachnoid space→neuroepithelium intracellular space, choroid plexus epithelium→venous fenestrated capillary→galenic system, and/or perineural space→lymphatic channel. Our computed tomography (CT) ventriculo-cisternography data demonstrated remarkable intraparenchymal CSF passage and delayed clearance of the contrast not only in the ventriculo-cisternal space (''major CSF pathway'') but also from the cerebral parenchyma as in the ''minor CSF pathway'' (''Minor CSF Pathway Hydrocephalus''). The condition of ''*Minor CSF Pathway Hydrocephalus*,''[14] however, may improve later with the development of the Pacchionian body in late infancy which will increase the CSF absorption function in the major CSF pathway. The high success rate of neuroendoscopic surgery, or even spontaneous arrested

Table 2 Historical trend of Hydrocephalus Research for ''Classification of Hydrocephalus'' in Japan

Year	Author(s): Concept	Source
2010	Oi: ''Multi-categorical Hydrocephalus Classification'' (McHC)	*Journal of Hydrocephalus* 2: 1–21, 2010[13]
2006	Oi and Di Rocco: Proposal of ''Evolution Theory in CSF Dynamics'' and ''Minor Pathway Hydrocephalus''	*Childs Nerv Syst* 22: 662–669, 2006[14]
2003	Oi: Prenatal Diagnosis and Management of ''Fetal Hydrocephalus''	*Childs Nerv Syst* 19: 508–516, 2003
2000	Ishizaki et al.: ''Acute and Subacute Hydrocephalus'' Rat Model.	*Pediatr Neurosurg* 33: 298–305, 2000
2000	Mori: Actualities in Hydrocephalus Classification and Management Possibilities.	*Neurol Res* 22: 127–130, 2000
1998	Oi et al.: ''Perspective Classification of Congenital Hydrocephalus'' (PCCH).	*J Neurosurg* 88: 685–694, 1998[18]
1996	Hirai et al.: Clinical Grading in Post-SAH hydrocephalus.	*Neurosurgery* 39: 441–447, 1996
1995	Mori et al.: Classification of Hydrocephalus and Outcome of Treatment	*Brain Dev* 17: 338–348, 1995
1995	Mori: Evolution of New Classifications.	*Childs Nerv Syst* 11: 523–532, 1995
1990	Mori: ''Intractable Infantile Hydrocephalus''	*Childs Nerv Syst* 6: 198–204, 1990
1964	Sano: Hydrocephalus and Its Various Subtypes	*Neurol Med Chir (Tokyo)* 6: 12–17, 1964

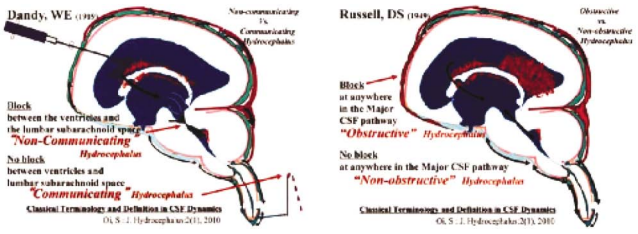

Fig. 3 Schematic drawings of two classical types of hydrocephalus classified in the conventional concept of cerebrospinal fluid (CSF) dynamics. Reprinted with permission from Oi, S.[13] *Left*: Communicating and non-communicating hydrocephalus.[2] *Right*: Obstructive and non-obstructive hydrocephalus.[39] In the definition of Dandy's communicating/non-communicating hydrocephalus, the communication of the CSF pathway is between the lateral ventricle and the lumbar subarachnoid space (confirmed by injection of dye into the lateral ventricle and detection by lumbar puncture). Obstructive and non-obstructive hydrocephalus is defined as a condition of disturbed CSF circulation due to a blockage at any region in the major CSF pathway including the ventricular system and cistern/subarachnoid space.

hydrocephalus and disappearance of external hydrocephalus, are all expected after this period, when the ''major CSF pathway'' is completed and functionally matured.[14]

II. Classification of fetal and congenital hydrocephalus

The underlying lesions or causative factors may differ completely in the fetal or perinatal period from childhood and adulthood. These two categories are most essential in the classification of hydrocephalus, and the underlying lesion, if any, is the most important factor for the cause of hydrocephalus.

Up to the late 1990s, no available classification systems took into account the chronological changes of the hydrocephalic state from the fetal to neonatal and infantile periods, and reflected the underlying developmental or embryological stages of the brain, especially the neuronal maturation process. It was essential to have some definitive method with which to estimate postnatal prognosis in fetal hydrocephalus. The prognosis may also depend on the progression of the fetal hydrocephalus and the affected brain, and on the degree of damage to the neuronal maturation process.

Therefore, a new classification system was developed for congenital hydrocephalus, ''Perspective Classification of Congenital Hydrocephalus'' (PCCH).[18,28] This classification is based on the stage, type, and clinical category of congenital hydrocephalus. Regarding the clinical-embryological stages, each stage reflects both clinical and embryological developmental aspects of the neuronal maturation process in the hydrocephalic fetus or in-

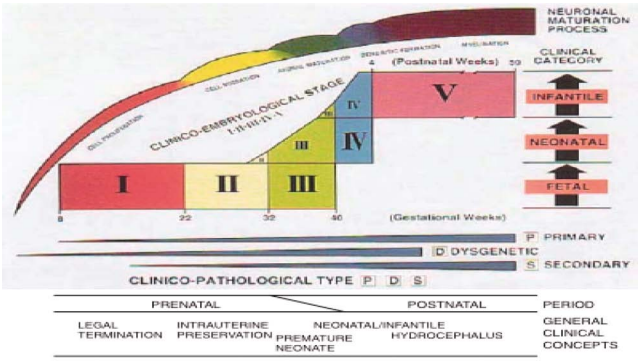

Fig. 4 Schematic drawings of the concept of ''Perspective Classification of Congenital Hydrocephalus'' (PCCH Stages I–V). Reprinted with permission from Oi, S.[13] *Stage I occurs between 8 and 21 weeks of gestation, which is the period of legally, except for fetus disease, permissible termination of pregnancy in Japan. Cell proliferation is the main process in neuronal maturation. **Stage II extends from 22 to 31 weeks of gestation, the period of intrauterine preservation of the fetus before pulmonary maturation is completed. Cell differentiation and migration are the main processes in neuronal maturation. ***Stage III extends from 32 to 40 weeks of gestation, a period of possible premature/preterm neonatal hydrocephalus, if delivery occurs. Axonal maturation is the main process in neuronal maturation. ****Stage IV occurs between 0 and 4 weeks of postnatal age, the period of neonatal hydrocephalus. Dendritic maturation is the main process in neuronal maturation. *****Stage V extends from 5 to 50 weeks of postnatal age, the period of infantile hydrocephalus. Myelination is the main process in neuronal maturation.

fant, as summarized in Fig. 4.

The clinicoembryological stages (PCCH Stages I–V) are as follows.

Stage I occurs between 8 and 21 weeks of gestation, which is the period of legally, except for fetal disease, permissible termination of pregnancy in Japan. Cell proliferation is the main process in neuronal maturation.

Stage II extends from 22 to 31 weeks of gestation, the period of intrauterine preservation of the fetus before pulmonary maturation is completed. Cell differentiation and migration are the main processes in neuronal maturation.

Stage III extends from 32 to 40 weeks of gestation, a period of possible premature/preterm neonatal hydrocephalus, if delivery occurs. Axonal maturation is the main process in neuronal maturation.

Stage IV occurs between 0 and 4 weeks of postnatal age, the period of neonatal hydrocephalus. Dendritic maturation is the main process in neuronal maturation.

Stage V extends from 5 to 50 weeks of postnatal age, the period of infantile hydrocephalus. Myelination is the main process in neuronal maturation.

In each stage, individual conditions with differing features of hydrocephalus can be classified along with the embryological or developmental background of the affected brain and the CSF circulation in each pathological type with subtypes. The three clinicopathological subtypes are: primary hydrocephalus, including communicating or non-complicated hydrocephalus, aqueductal stenosis, foramen atresia, and others; dysgenetic hydrocephalus, including hydrocephalus with spina bifida, bifid cranium, Dandy-Walker cyst, holoprosencephaly, hydranencephaly, lissencephaly, congenital cyst, and others; and secondary hydrocephalus, hydrocephalus due to brain tumor, hemorrhagic or other vascular disease(s), infection, trauma, subdural fluid collection, and others. These conditions should be considered in the standard clinical categories of fetal, neonatal, and infantile hydrocephalus, based on the essential differences in their pathophysiological appearance, including the dynamics of intracranial pressure and CSF circulation.

Further analysis performed using the new classification, PCCH, suggests that postnatal outcomes differ, depending on the time of onset of the hydrocephalus, even within the same category or sub types. The intelligence quotient (IQ) or developmental quotient (DQ) of patients whose hydrocephalus was diagnosed at PCCH Stage III was higher compared with those diagnosed at Stage II in cases of primary hydrocephalus, and compared with those cases with some types of dysgenetic hydrocephalus such as myeloschisis.[18]

III. Classification of adult hydrocephalus

It is generally recognized that the intracranial pressure (ICP) dynamics of hydrocephalus change chronologically. The chronological changes in hydrocephalic ICP dynamics and symptomatology were reported in adult patients, namely, ''Hydrocephalus Chronology in Adults'' (HCA), and the Staging of I–V from the acute to chronic and post-shunt periods.[29] The triad of symptoms, dementia, gait disturbance and urinary incontinence is observed even in cases revealing high ICP dynamics and non-communicating hydrocephalus before the truly chronic stage, such as HCA Stage III.[12] Hydrocephalus in this stage can be treated with a medium-pressure shunt system. There is some phase discrepancy between the symptomatology of ''NPH'' (Hakim and Adams)[7] and the chronological change in ICP dynamics in hydrocephalus. In the truly chronic phase of hydrocephalus, it may be normal-

ized in the ICP dynamics. ICP dynamics is associated with biomechanical changes in the hydrocephalic brain in the chronic stage (Fig. 5).

The author proposed a unique category of hydrocephalus in adults, namely, a long-term type of hydrocephalus, ''long-standing overt ventriculomegaly in adult'' (LOVA).[30] Although its mechanism still remains unclear, patients with LOVA often suffer from a progressive course of hydrocephalus that continues into adulthood. We also reported that the hydrocephalic state in LOVA is extremely difficult to treat with a shunt because of lost intracranial compliance. Patients with LOVA in whom significant progressive symptoms of hydrocephalus had developed were first diagnosed as hydrocephalic during adulthood.[30] In all patients, ventriculomegaly was prominent, involving the lateral and third ventricles as demonstrated on CT and/or magnetic resonance images. None of the patients had any known underlying disease or symptoms or signs, indicating that the hydrocephalus had first occurred at birth or during infancy in accordance with neuroimaging findings of long-standing hydrocephalus. The specific diagnostic criteria for LOVA includes macrocephaly greater than two standard deviations in head circumference, 57 cm in female and 58 cm in male patients, and/or neuroradiological evidence of a significantly expanded or destroyed sella turcica in addition to the noncommunicating overt ventriculomegaly.[30]

LOVA is a chronological concept of hydrocephalus. As described here, LOVA may be summarized as a complex entity with the following compatible subtypes: 1) onset may be congenital in origin but becomes manifest during adulthood; 2) the underlying lesion is aqueductal stenosis; 3) symptoms include macrocephaly, increased ICP symptoms, dementia and subnormal IQ, but occasionally high or even super-high IQ; 4) pathophysiological characteristics include noncommunicating CSF circulation and ICP dynamics that mainly consists of high ICP; 5) the chronology is long term and progressive; and 6) the hydrocephalus becomes arrested again after shunt placement or neuroendoscopic ventriculostomy.[30]

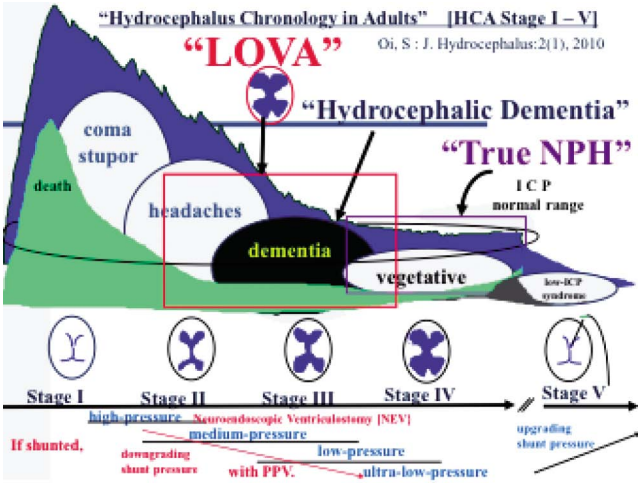

Fig. 5 Schematic drawings of the concept of ''Hydrocephalus Chronology in Adults'' (HCA Stages I–V).[12] Reprinted with permission from Oi, S.[13] Staging of I–V from the acute to chronic and post-shunt periods. The triad of symptoms, dementia, gait disturbance, and urinary incontinence, is observed even in patients with high intracranial pressure (ICP) dynamics and non-communicating hydrocephalus before the truly chronic stage, such as HCA Stage III. Hydrocephalus in this stage can be treated with a medium-pressure shunt system. There is some phase discrepancy between the symptomatology of ''normal pressure hydrocephalus (NPH)''[7] and the chronological change in ICP dynamics in hydrocephalus. In the truly chronic phase of hydrocephalus, it may be normalized in the ICP dynamics. ICP dynamics is associated with biomechanical changes in the hydrocephalic brain in the chronic stage.

IV. Classification of post-shunt isolated compartments

The authors have previously reported that excess drainage of CSF via a ventricular shunt system will cause morphological changes in the CSF path-

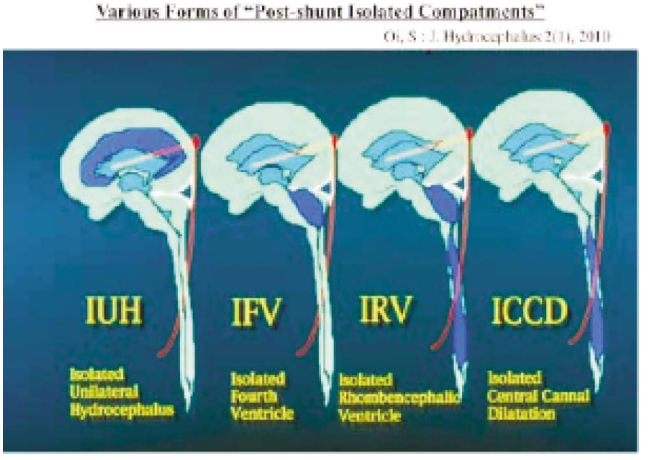

Fig. 6 Schematic drawings of the concept of ''Post-shunt Isolated Compartments'' (Types I–IV).[19] Reprinted with permission from Oi, S.[13] Isolated unilateral hydrocephalus (IUH), isolated fourth ventricle (IFV), disproportionately large fourth ventricle, isolated rhombencephalic ventricle (IRV), isolated central canal dilation (ICCD), isolated quarto-ventriculomegaly, dorsal sac in holoprosencephaly, and loculated ventricle.[19]

ways and possibly lead to isolation of compartments.[19–24,26] These phenomena produce a slit-like ventricle most commonly seen in young infants[19,21–23,25] and occasionally lead to the slit ventricle syndrome.[25] The mechanism of development of an isolated ventricle after shunting is closely related to the presence of a slit-like ventricle.[23] The mechanism of obstruction at the foramen of Monro in isolated unilateral hydrocephalus (IUH)[23] and that of aqueductal obstruction in isolated fourth ventricles (IFV)[21,22] occurring after shunt placement are essentially the same. Both occur in a previously communicating ventricular system, and in both cases reduction of the size of all ventricles is initially seen after shunting.[8] Isolation then gradually develops and relargement of the isolated compartment is ob-

Table 3 ''Multi-categorical Hydrocephalus Classification'': [McHC] to cover the entire aspects of hydrocephalus with all considerable classification items and factors (Oi, S)[13]

Mc HC: **Multi-categorical Hydrocephalus Classification [Oi, S: Journal of hydrocephalus Vol.2 No.1, 2010]**

Subjects	Categories	Subtypes		**/***[reference]
Patient	I. Onset (Pre-/Post-natal)	1 Congenital	2 Acquired	
	Onset (Age)	1 Fetal [PCCH Stage ( )]	2 Neonatal	3 Infantile
		4 Child	5 Adult	**[Oi, S]
	Others	( )		
	II. Cause	1 Primary	2 Secondary	3 Idiopathic
	III. Underlying Lesion	1 Dysgenetic	Syndromic ( )	
		2 Post hemorrhagic	3 Post meningitic	4 Post traumatic
		5 With Tumor / Cyst / Mass Lesions		others ( )
	IV. Symptomatology (Head)	1 Macrocephalic	2 Normocephalic	3 Microcephalic
	(Symptom)	1 Occult (asymptomatic)	2 Symptomatic	3 Overt
	(Conciousness & Mentality)	1 Comatose	2 Stuporous	3 Dementia
		4 Retarded	others ( )	***[Hakim, S]
	(Syndrome:)	1 Hydrocephalus / Parkinsonism Complex		**[Oi, S]
CSF	V. Pathophysiology:	1 Communicating	2 Noncommunicating	***[Dandy, WE]
	CSF Circulation (Occlusion)	3 Non-obstructive	4 Obstructive	***[Russell, DS]
	(Accumulation)	1 External	2 Internal	3 Interstitial ***[Raimondi, AJ]
		4 Localized	5 Minor Pathway	***[Sato, O], **[Oi, S & Di Rocco, C]
	(Isolated Compartment)	Isolated Compartment: [1 UH 2 IFV 3 IRV 4 ICCD 5 DCH		
		6 DLFV 7 HMH others ( )]		***[Rekate, H]**[Oi, S]
	VI. Pathophysiology (ICP Dynamics)	1 High Pressure	2 Normal Pressure	***[Hakim, S] ***[Di Rocco, C]
	VII. Chronology (Phase)	1 Acute	2 Chronic	3 Long standing
	(Progression)	1 Progressive	2 Spontaneously Arrested	**[Oi, S]
Treatment	VIII. Post-shunt	1 Shunt dependent	2 Shunt independent	
	IX. Post-neurondoscopic	1 Slit ventricle syndrome	2 Postoperative Subdural Hematoma	
	Ventriculostomy [NEV]	1 NEV dependent	2 NEV independent	
	X. Others	Others ( )		

* ICP = Intracranial Pressure, CSF = Cerebrospinal Fluid, NEV = Neuroendscopic Ventriculostomy, UH = Unilateral Hydrocephalus, IFV = Isolated Fourth Ventricle, IRV = Isolated Rhombencephalic Ventricle, ICCD = Isolated Central Canal Dilatation, DCH = Double Compartment Hydrocephalus, DLFV = Disproportionately Large Fourth Ventricle, HMH = Hydromyelic Hydrocephalus

**Reference: 1. Hydromyelic Hydrocephalus: Oi, S et al: Journal of Neurosurgery 74: 371-379, 1991
2. Experimental Models of Congenital Hydrocephalus and Compatible Clinical Types: Oi, S et al: Child's Nerv Syst 12: 292-302, 1996.
3. Perspective Classification of Congenital Hydrocephalus[PCCH Stage I-V]: Oi, S et al: Journal of Neurosurgery 88: 685, 1998
4. Long-standing Overt Ventriculomegaly in Adults [LOVA]: Oi, S et al: Journal of Neurosurgery 92: 933-940, 2000
5. Hydrocephalus-Parkinsonism Complex: Oi, S et al: Child's Nerv Syst 20: 37-40, 2004
6. Evolution Theory of CSF Dynamics and Minor Pathway Hydrocephalus: Oi, S and Di Rocco, C: Child's Nerv Syst 22: 662-669, 2006
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***Reference: 8. Communicating/ Non-communicating Hydrocephalus: Dandy, WE: Ann Surg 70: 129-142, 1919 and Dandy, WE & Blackfan, KD: Am J Dis Child 8: 406-482,1914
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Considerable items in classification for hydrocephalus among 3 subjects: patient, cerebrospinal fluid (CSF), and treatment are divided into 10 categories. McHC Category I: Onset (age, phase); II: Cause; III: Underlying lesion; IV: Symptomatology; V: Pathophysiology 1, CSF circulation; VI: Pathophysiology 2, intracranial pressure (ICP) dynamics; VII: Chronology; VIII: Post-shunt; IX: Post-endoscopic ventriculostomy (EVS); and X: Others. There were 54 subtypes of hydrocephalus listed up, and these were divided into the 10 McHC Categories, 2–7 in each, respectively.

served. Dynamic studies of the CSF using metrizamide CT ventriculography have confirmed the presence of a one-way valve at either the foramen of Monro or the aqueduct,[21,22] and pressure gradients between the compartments have also been recorded. We suggested that similar isolation may occur after placement of a shunt in the lateral ventricle in cases of communicating holoneural canal dilation. Various types of isolation (Types I to IV) may then develop, depending upon the site of occlusion[19] (Fig. 6).

Neuroendoscopic surgery was used to treat patients with various forms of isolated compartments with specific pathophysiology[16] including IUH, IFV, disproportionately large fourth ventricle (DLFV), isolated rhombencephalic ventricle (IRV), isolated central canal dilatation (ICCD), isolated quarto-ventriculomegaly (IQV), dorsal sac in holoprosencephaly (DS), and loculated ventricle (LV).[16]

V. Proposal for comprehensive ''Multi-categorical Hydrocephalus Classification'' (McHC)

Various items for classification of hydrocephalus were previously proposed, such as associated anomalies/underlying lesions, CSF circulation/intracranial pressure (ICP) patterns, clinical features, and other categories. No definitive classification has been published comprehensively to cover the variety of these aspects.

The authors designed and developed a new classification of hydrocephalus, McHC (Oi, 2010)[13] to cover the entire aspects of hydrocephalus with all important classification items and factors. Important items in the classification for hydrocephalus cover 3 subjects, patient, CSF and treatment, and are divided into 10 categories, ''McHC'' Category I: Onset (age, phase); II: Cause; III: Underlying lesion; IV: Symptomatology; V: Pathophysiology 1, CSF circulation; VI: Pathophysiology 2, ICP dynamics; VII: Chronology; VIII: Post-shunt; IX: Post-endoscopic ventriculostomy (EVS); and X: Others. A total of 54 subtypes of hydrocephalus are listed, and these are divided into 10 ''McHC'' Categories, 2–7 in each (Table 3). There could be theoretically 72,576,000 patterns of hydrocephalus classified with all these combinations. ''McHC'' has been applied to analyze the clinical data prospectively collected from the most experienced centers in Japan as a part of the nationwide cooperative study for fetal and congenital hydrocephalus in the Center of Excellence (COE)—Fetal Hydrocephalus Top 10 Japan nominated from the retrospective survey for 2008.

Conclusions

Classification of hydrocephalus is the most crucial but the most complicated academic challenge within hydrocephalus research. The major difficulty in this challenge arises from the fact that the classifications are based on almost all subjects in hydrocephalus research. The current status of classification of hydrocephalus in the individual subgroups was reviewed and summarized from publications in the last 60 years (1950–2010), and discussed focusing on the variety of characteristics in hydrocephalus, with more and more new aspects recently disclosed not only in fetal and pediatric but also in adult hydrocephalus. A recently reported classification of hydrocephalus, McHC provides comprehensive coverage of the entire aspects of hydrocephalus with current important classification categories and subtypes.

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