NMC Case Report Journal 2020; 7: 141–145 DOI: 10.2176/nmccrj.cr.2019-0263

Case Report

Progressive Hydrocephalus after Endoscopic Third Ventriculostomy in Pediatric Patients with Blake's Pouch Cyst

Youhei Takeuchi,[1] Tomomi Kimiwada,[1] Reizo Shirane,[1] and Teiji Tominaga[2]

The pathophysiology and optimal treatment for hydrocephalus with Blake's pouch cyst (BPC) remain controversial. The authors present two pediatric cases of hydrocephalus associated with BPC, in which the patients' hydrocephalus progressed after endoscopic third ventriculostomy (ETV), despite a patent stoma of the third ventricular floor.
Case 1: A 4-year-old girl with delayed gait development was diagnosed with BPC-associated hydrocephalus and received ETV. Postoperatively, the patient presented headaches and nausea. Computed tomography (CT) scans demonstrated larger ventricles than those observed on the preoperative images. Because phase-contrast cine magnetic resonance imaging (MRI) and constructive interference in steady state (CISS) MRI revealed patent cerebrospinal fluid (CSF) flow at the third ventricular floor level, a ventriculoperitoneal shunt (VPS) was placed using a programmable pressure valve to treat the hydrocephalus.
Case 2: A 6-year-old girl with newly developed repeated convulsive seizures was diagnosed with BPC-associated hydrocephalus and received ETV. Phase-contrast cine MRI on the 5th postoperative day showed hyperdynamic CSF flow at the third ventricular floor level. She also developed vomiting and headache 6 weeks after ETV. CT scans demonstrated much larger tetraventricular hydrocephalus than that observed on the preoperative images. VPS placement improved her hydrocephalus.
Referencing the previous literature, we discuss the CSF dynamics and the mechanism of BPC-associated hydrocephalus, focusing on the third ventricular floor bulging. We hope our experience will help elucidate the pathophysiology and treatment strategies for BPC-associated hydrocephalus.

Keywords: Blake's pouch cyst, hydrocephalus, endoscopic third ventriculostomy, children

Introduction

Cystic malformations in the posterior fossa are classified into two categories embryologically. In one category, the cyst represents an expansion of the roof plate of the brain vesicle and includes the Dandy–Walker (DW) cyst, or Blake's pouch cyst (BPC). Cysts in the other category consist of extra-axial structures such as the arachnoid membrane, which includes arachnoid cysts and the mega-cisterna magna.[1–3)]

BPC is a congenital cystic lesion at the posterior cranial fossa characterized by persistent posterior ballooning of the posterior membranous area (PMA) into the cisterna magna caused by failed spontaneous perforation of the foramen of Magendie during embryogenesis. Radiologically, the choroid plexus bends below the vermis and continues into the pouch in the BPC, which is essentially a fourth ventricular diverticulum. Conversely, the choroid plexus is positioned normally in posterior fossa arachnoid cysts and is non-existent in DW malformations.[2,3)]

Although the pathophysiology and optimal treatment for hydrocephalus with BPC remains controversial, shunt placement, endoscopic third ventriculostomy (ETV), and fenestration of the cyst have been previously reported.[2,4–9)] ETV has been reported to be effective in pediatric patients with BPC-associated hydrocephalus in recent years.[4,6,10)] In line with the current trend of using minimally invasive procedures in neurosurgery, ETV is considered a good alternative for hydrocephalus with BPC.

In this report, we present our experiences with two pediatric patients who showed tetraventricular hydrocephalus associated with BPC. As a first treatment, we performed ETV on both patients. Despite their patent stoma after ETV, their hydrocephalus was aggravated compared with their preoperative hydrocephalus. We next added ventriculoperitoneal shunts (VPS) for these patients. Referencing the previous literature, we discuss the cerebrospinal fluid (CSF) dynamics and the mechanism of BPC-associated hydrocephalus, focusing on the third ventricular floor bulging.

Case Presentation

Case 1

History and examination

A 4-year-old girl with delayed gait development was referred to our department. At 9 months old, her head circumference began increasing, reaching larger than 97% of the normal range. She could walk hand in hand with mother but not walk independently, and neuropsychological testing revealed her IQ score at 88. Magnetic resonance imaging (MRI) demonstrated tetraventricular hydrocephalus with a posterior fossa cyst communicating with the fourth ventricle and located just beneath the vermis as well as a membranous structure in the prepontine cistern (Figs. 1A–1C). These findings were compatible with BPC. Her ETV success score was

[1]Department of Neurosurgery, Miyagi Children's Hospital, Sendai, Miyagi, Japan
[2]Department of Neurosurgery, Tohoku University Graduate School of Medicine, Sendai, Miyagi, Japan

Received: November 17, 2019; Accepted: January 20, 2020
Online June 26, 2020

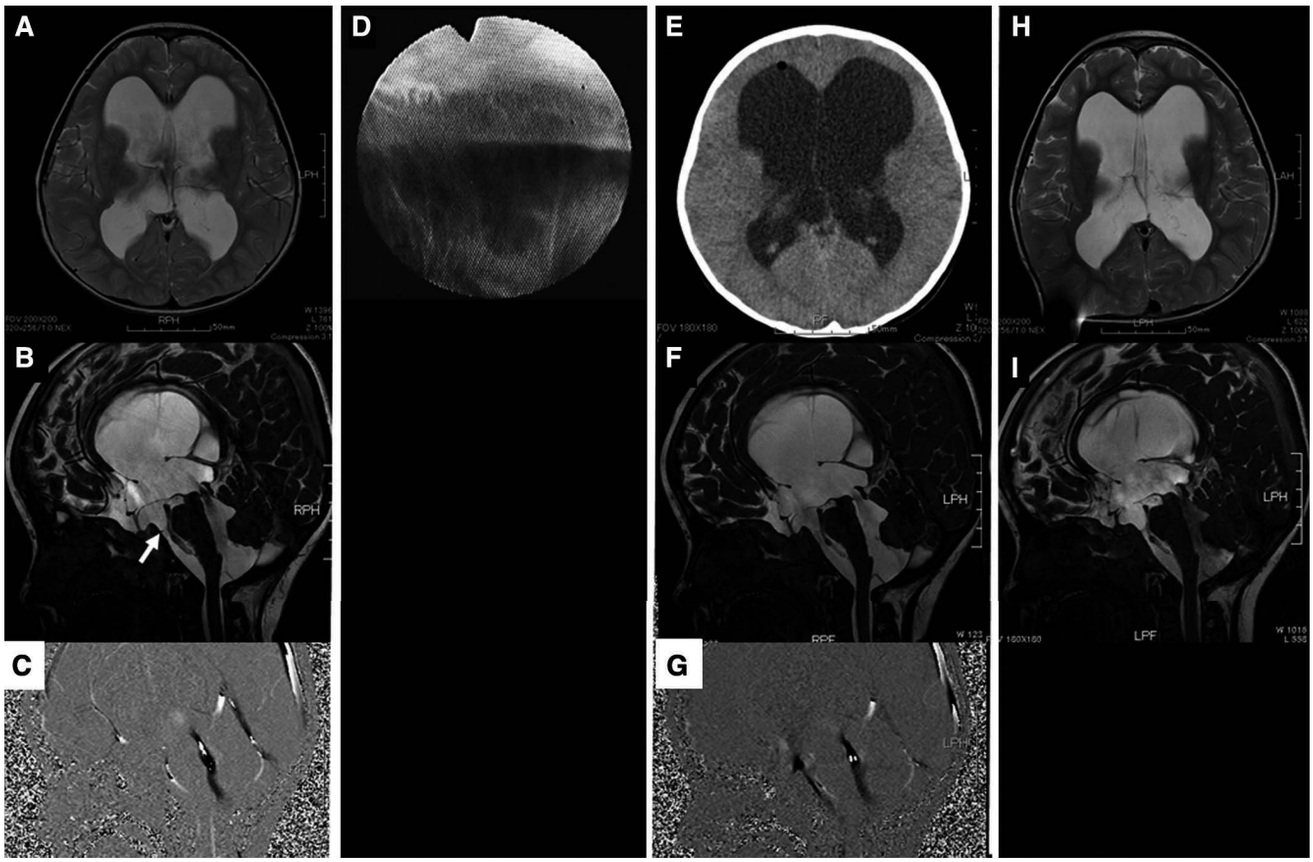

Fig. 1 Radiological images for case 1 obtained preoperatively (**A**–**C**), immediately before VPS after ETV (**E**–**G**), and after VPS (**H, I**). (**A, E, H**) T2-weighted or CT axial images. (**B, F, I**) CISS sagittal MR images. Prepontine membrane is found in **B** (arrow). (**C**, **G**) Phase-contrast cine MR images. (**D**) Intraoperative view. Tough membrane in the prepontine cistern. Note the ventricular size after ETV (**E**) was larger than that before ETV (**A**), and the patent stoma and CSF flow at the third ventricular floor level were apparent when the patient showed symptoms due to increased ICP (**F, G**). CISS: constructive interference in steady state, CSF: cerebrospinal fluid, CT: computed tomography, ETV: endoscopic third ventriculostomy, ICP: intracranial pressure, MR: magnetic resonance, VPS: ventriculoperitoneal shunt.

80 (age: 40, etiology: 30, shunt: 10), and we planned ETV to manage the hydrocephalus as a first treatment.

Endoscopic third ventriculostomy

ETV was performed using a flexible endoscope via the right anterior horn. After making a stoma at the third ventricular floor and Liliequist's membrane, we found another hard membrane in the prepontine cistern via a stoma (Fig. 1D) and perforated the membrane using forceps and a balloon catheter to release CSF flow. Pulsatile movement of the third ventricular floor was observed after the operation.

Postoperative course

After the surgery, the immediate postoperative course was uneventful. To confirm the ETV stoma, routinely performed phase-contrast cine MRI on the 7th postoperative day showed patent CSF flow at the third ventricular floor level. However, the patient presented vomiting and headaches on the 8th postoperative day, and new computed tomography (CT) scans demonstrated larger ventricles than those seen on the preoperative images (Fig. 1E). Because phase-contrast cine and constructive interference in steady state (CISS) MRI revealed patent CSF flow at the third ventricular floor level (Figs. 1F and 1G), a VPS was placed using a programmable pressure valve to treat the hydrocephalus. Following shunt placement, the patient could walk independently in 6 months and run in a year, and follow-up MR images showed improved hydrocephalus (Figs. 1H and 1I).

Case 2

History and examination

A 6-year-old girl with newly developed repeated convulsive seizures was referred to our hospital. Her head circumference had been larger than 97% of the normal range since she was an infant. Neuropsychological testing (WISC-IV) revealed a mildly low IQ score (full-scale IQ 83, verbal comprehension 97, perceptual reasoning 74, working memory 79, and processing speed 94). An electroencephalogram (EEG) showed sharp waves in the right temporal area. MRI demonstrated tetraventricular hydrocephalus with a cyst communicating with the fourth ventricle and projecting to the cisterna magna as well as a membranous structure in prepontine cistern (Figs. 2A–C). These findings were completely compatible

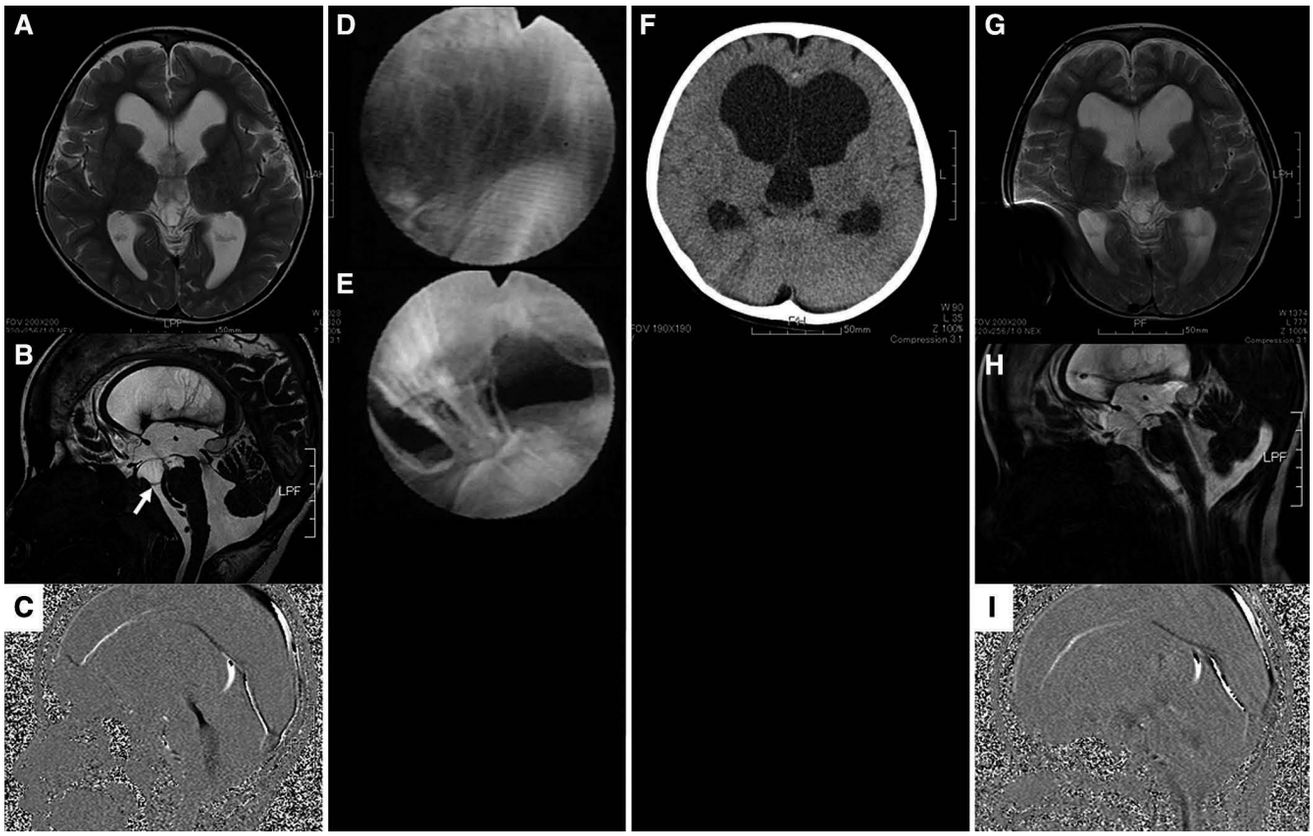

Fig. 2 Radiological images for case 2 obtained preoperatively (**A**–**C**), immediately before VPS after ETV (**F**), and after VPS (**G**–**I**). (**A, F, G**) T2-weighted or CT axial images. (**B, H**) CISS sagittal MR images. Prepontine membrane is found in **B** (arrow). (**C, I**) Phase-contrast cine MR images. (**D, E**) Intraoperative views. Tough membrane in the prepontine cistern (**D**) and perforations in this membrane (**E**). Note the ventricular size after ETV (**F**) was much larger than that before ETV (**A**). Apparent patent stoma and CSF flow at the third ventricular floor level remain confirmed in **H**, **I**. CISS: constructive interference in steady state, CSF: cerebrospinal fluid, CT: computed tomography, ETV: endoscopic third ventriculostomy, MR: magnetic resonance, VPS: ventriculoperitoneal shunt.

with BPC. Her ETV success score was 80 (age: 40, etiology: 30, shunt: 10), and we planned ETV as a first treatment.

Endoscopic third ventriculostomy

ETV was performed using a flexible endoscope as in case 1. We found a hard membrane in the prepontine cistern via a stoma (Fig. 2D) and perforated the membrane to release CSF flow (Fig. 2E). Pulsatile movement of the third ventricular floor was observed postoperatively.

Postoperative course

After the surgery, she transiently developed syndrome of inappropriate secretion of antidiuretic hormone (SIADH) but improved completely within 1 week. Routinely performed follow-up phase-contrast cine MRI on the 5th postoperative day showed hyperdynamic CSF flow at the third ventricular floor level. She was discharged on the 7th postoperative day; however, 6 weeks later, she returned to our hospital with frequent vomiting and headaches. CT scans demonstrated much larger tetraventricular hydrocephalus than that observed on the preoperative images (Fig. 2F). We quickly performed a VPS using a programmable pressure valve. Postoperative CISS MRI showed a clear stoma of the third ventricular floor (Fig. 2H), and CSF flow of the stoma was confirmed by phase-contrast cine MRI (Fig. 2I). Following shunt placement, the patient's clinical course was uneventful, but she continued anti-epileptic drugs because her seizures did not disappear. Follow-up MRI improved hydrocephalus (Fig. 2G).

Discussion

We experienced two pediatric cases with progressive BPC-associated hydrocephalus after ETV. Because BPC is a relatively rare malformation in the posterior cranial fossa, the reported number of case series of BPC remains limited, and few reports are available regarding BPC-associated hydrocephalus managed by ETV (Table 1). To our knowledge, Cornips et al.[4] first reported two pediatric patients with BPC-associated hydrocephalus treated successfully by ETV. One was a 2-year-old boy who presented delayed gait development and macrocephaly, and the patient's gait improved and head circumference stabilized after ETV. The other was a 2-year-old boy who presented persistent open fontanel and macrocephaly. It would have been interesting to

Table 1 Summary of literature cases of BPC related hydrocephalus managed by ETV

Authors (year)	No. of cases	Age at surgery	Bulging of IIIrd ventricle floor	Pulsatile movement at IIIrd ventricle floor	Membrane in basal cistern	No. of cases with ETV benefit	Additional treatment
Cornips et al. (2010)[4]	3	1m, 2y, 2y	Upward (2) Normal (1)	n.d.	n.d.	2	–
Takami et al. (2010)[5]	1	7m	Normal	–	n.d.	0	VPS
Nishiyama et al. (2011)[6]	3	5y, 2y, 14y	Downward (3)	+	Membrane (2) Open (1)	2	–
Brusius et al. (2013)[10]	8	1, 7, 26, 2, 11, 9, 2, 48 months	n.d.	n.d.	n.d.	8	–
Hirono et al. (2014)[7]	1	3y	Normal	n.d.	Tough Liliequst's membrane	0	VPS
Our cases	2	4y, 6y	Upward	+	Membrane (2)	0	VPS

BPC: Blake's pouch cyst, ETV: endoscopic third ventriculostomy, n.d.: not described, VPS: ventriculoperitoneal shunt.

learn whether they found thickened membranes in the prepontine cistern after ETV and whether they perforated the membrane or not, as this was not mentioned in the article. Takami et al.[5] reported a 7-month-old girl with BPC-associated hydrocephalus, who presented tonic–clonic seizures. The patient had three surgeries. The first surgery removed the cyst membrane in the posterior cranial fossa. The patient's hydrocephalus initially improved, but tetraventricular hydrocephalus recurred 3 weeks after the cyst membrane was removed. The second surgery was ETV, but the authors found no pulsatile movement of the third ventricular floor. Finally, the authors placed a VPS, and the patient's hydrocephalus improved. They discussed that the patient's hydrocephalus was caused by the combination of obstructed CSF flow at the fourth ventricular outlets and disequilibrium of the CSF dynamics between production and absorption capacity. Brusius et al.[10] also reported that ETV was a safe and effective procedure for BPC-associated hydrocephalus. They demonstrated eight pediatric patients with BPC-associated hydrocephalus who were treated successfully by ETV. Although detailed information on these eight patients was lacking, the authors concluded that patients with BPC-associated hydrocephalus are eligible candidates for ETV treatment. Nishiyama et al.[6] reported three pediatric patients with BPC-associated hydrocephalus managed by ETV. ETV was effective for two of these three patients. These authors found a hard membrane in the prepontine cistern and confirmed pulsatile movement of the third ventricular floor after perforating this membrane. Hirono et al.[7] described a 3-year-old girl with BPC-associated hydrocephalus suffering from headaches and nausea. ETV was initially performed, but the patient developed bacterial meningitis and secondary hydrocephalus, which necessitated a VPS 1 month after the ETV. These authors also indicated the tough membrane at the basal cistern.

In both our cases, we encountered a fibrous tough membrane in the prepontine cistern after perforating the third ventricular floor (Figs. 1B and 1I and Figs. 2B and 2E) and we confirmed pulsatile movement of the third ventricular floor after perforating this membrane. Nishiyama et al.[6] pointed out the membrane and hypothesized the theory of an immature obstructive CSF pathway in front of the brain stem in patients with BPC-associated hydrocephalus. During development, the enlarged fourth ventricle with persistent BPC pushes the brain stem forward, and the CSF pathway in front of the brain stem can be inhibited. Consequently, a tough membranous structure, such as a prepontine ligament, may develop and obstruct the CSF flow in front of the brain stem. Therefore, perforating this membrane in addition to the third ventricular floor seems essential for successful ETV in patients with BPC-associated hydrocephalus.

In our two cases, we recognized the hard membrane in the prepontine cistern and perforated it; however, the patient's hydrocephalus was aggravated after the ETV. We found it odd that phase-contrast cine and CISS MRI revealed apparent CSF flow at the third ventricular floor level, suggesting a patent stoma, when one patient (case 1) showed symptoms due to increased intracranial pressure (ICP) after ETV. Thus, we concluded that the patient's hydrocephalus had progressed despite the patent stoma. Patent stoma was also confirmed in case 2 after VPS.

Several reasons may explain why the hydrocephalus progressed after ETV despite the patent stoma. One may be the lack of ballooning (downward bulging) of the third ventricular floor, which was not found in our two cases. Ballooning of the third ventricular floor generally suggests a pressure gradient between the third ventricle and cerebral basal cistern, indicating successful ETV. Our patients' third ventricular floor did not exhibit ballooning on their preoperative MRIs, but showed upward bulging, suggesting that the pressure in the basal cistern was higher than that in the third ventricle. By fenestration of the third ventricular floor, the CSF inflow into the third ventricle may have exceeded the outflow from the third ventricle (inflow/outflow mismatch). As it is unknown about the flow through the perforated third ventricular floor and prepontine membrane, upward bulging of the third ventricular floor may be the key for progressive hydrocephalus after ETV. The reasons for upward bulging of the third ventricular floor remain to be elucidated, but one of the possibility is the check-valve mechanism in prepontine

membrane. As Nishiyama et al.[6] suggested above, the CSF flow in front of the brain stem might be obstructed and result in multiple tough membranes with check-valve mechanism. Therefore, inflow/outflow mismatch might occur and result in progressive hydrocephalus after ETV. Another possibility may be that the stoma was occluded only in prepontine cistern and not in the third ventricular floor. As described above, if the membrane in the prepontine cistern must be perforated to improve CSF flow in BPC-associated hydrocephalus, occluding the stoma in the prepontine cistern may deteriorate the hydrocephalus. Unfortunately, we did not observe the stoma endoscopically before the VPS because the VPS was performed via the posterior horn puncture. Another reason is the changes in CSF flow and disequilibrium of CSF dynamics between production and absorption capacity after ETV (production/absorption mismatch).

Considering these findings, we think that ETV cannot be effective for patients with BPC-associated hydrocephalus, especially with upward bulging of third ventricular floor. Though little is known about the mechanism of BPC-associated hydrocephalus, we think BPC-associated hydrocephalus is not a simple non-communicating hydrocephalus due to a simple problem of CSF obstruction at the BPC, but due to multiple problems of CSF pathway such as multiple prepontine membranes with check-valve mechanism and obstructed foramina of Luschka. Therefore, some patients cannot be treated successfully with ETV or BPC wall resection. And we should carefully decide the indication of ETV for patients with BPC-associated hydrocephalus. This report provides important considerations, but includes only two patients, and further studies in a larger population are required. We hope our experience will help elucidate the pathophysiology and treatment strategies for BPC-associated hydrocephalus.

Conclusion

For pediatric patients of BPC-associated hydrocephalus with upward bulging of third ventricular floor, we should carefully decide the indication of ETV because of the risk of progressive hydrocephalus after ETV.

Acknowledgment

We thank Traci Raley, MS, ELS, from Edanz Group (www.edanzediting.com/ac) for editing a draft of this manuscript.

Conflicts of Interest

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

References

1) Utsunomiya H, Yamashita S, Takano K, Ueda Y, Fujii A: Midline cystic malformations of the brain: imaging diagnosis and classification based on embryologic analysis. *Radiat Med* 24: 471–481, 2006
2) Tortori-Donati P, Fondelli MP, Rossi A, Carini S: Cystic malformations of the posterior cranial fossa originating from a defect of the posterior membranous area. Mega cisterna magna and persisting Blake's pouch: two separate entities. *Childs Nerv Syst* 12: 303–308, 1996
3) Nelson MD, Maher K, Gilles FH: A different approach to cysts of the posterior fossa. *Pediatr Radiol* 34: 720–732, 2004
4) Cornips EM, Overvliet GM, Weber JW, Postma AA, Hoeberigs CM, Baldewijns MM, Vles JS: The clinical spectrum of Blake's pouch cyst: report of six illustrative cases. *Childs Nerv Syst* 26: 1057–1064, 2010
5) Takami H, Shin M, Kuroiwa M, Isoo A, Takahashi K, Saito N: Hydrocephalus associated with cystic dilation of the foramina of Magendie and Luschka. *J Neurosurg Pediatr* 5: 415–418, 2010
6) Nishiyama K, Okamoto K, Fujii Y: Pathophisiology and treatment of hydrocephalus with Blake's pouch cyst. *Nerv Syst Childs* 36: 464–466, 2011
7) Hirono S, Ito D, Murai H, et al.: Postnatal development of Blake's pouch cyst: a case report and new insight for its pathogenesis. *Childs Nerv Syst* 30: 1767–1771, 2014
8) Arai H, Sato K: Posterior fossa cysts: clinical, neuroradiological and surgical features. *Childs Nerv Syst* 7: 156–164, 1991
9) Apok V, Constantini S, Roth J: Microsurgical fenestration of retrocerebellar cysts as a treatment for syringomyelia. *Childs Nerv Syst* 28: 653–656, 2012
10) Brusius CV, Cavalheiro S: Endoscopic third ventriculostomy is a safe and effective procedure for the treatment of Blake's pouch cyst. *Arq Neuropsiquiatr* 71: 545–548, 2013

Corresponding author:
Tomomi Kimiwada, MD, PhD, Department of Neurosurgery, Miyagi Children's Hospital, 4-3-37 Ochiai, Aoba-ku, Sendai, Miyagi 989-3126, Japan.
✉kimiwada@miyagi-children.or.jp