Unilateral Hydrocephalus in Congenital Atresia of the Foramen of Monro

Jong Hyun Kim, M.D., Yong Gu Chung, M.D., Nam Joon Lee, M.D.,* Se Hoon Kim, M.D., Hoon Kap Lee, M.D., Ki Chan Lee, M.D., Jung Keun Suh, M.D.
Department of Neurosurgery, Department of Diagnostic Radiology,* College of Medicine, Korea University, Seoul, Korea

Abstract
Unilateral hydrocephalus is an uncommon disease which may result from obstruction of the foramen of Monro. Congenital or acquired lesions obstructing this foramen have been reported. We present a case of prenatally diagnosed fetal unilateral hydrocephalus. Ventricleoperitoneal shunt operation was performed and following the procedure, ventricular size was decreased and patients neurological status was improved.

Key Words: Unilateral hydrocephalus· Fetal diagnosis· Atresia· Foramen of Monro· Ventricleoperitoneal shunt.

Introduction
Unilateral hydrocephalus means dilatation of one lateral ventricle, which may result from prenatal or postnatal causes. This rarely reported congenital atresia of the foramen of Monro can result in unilateral dilatation of the lateral ventricle. Prenatal ultrasonography can be a useful method to diagnose this entity. There are two options in the surgical management of unilateral hydrocephalus: one is the ventriculoperitoneal shunt and the other is endoscopic fenestration of the occluded foramen of Monro or septum pellucidum. We present a case of prenatally diagnosed unilateral hydrocephalus resulted from atresia of the foramen of Monro which was treated by ventriculoperitoneal shunt.

Case Report
A 2-month-old boy was admitted due to increasing head size. He was diagnosed as having fetal hydrocephalus by prenatal ultrasonography and was born preterm at 32 weeks of gestation by vaginal delivery (Fig. 1). On admission, he was alert without neurological deficit and his developmental milestones were not delayed. His occipitofrontal head circumference was 42 cm above 97th percentile of his age and the anterior fontanelle was bulging. MRI revealed a markedly dilated right lateral ventricle with midline shift to the left and a severely compressed left lateral ventricle. The septum pellucidum and the third ventricle were intact and there were no masses or associated anomalies (Fig. 2). Ventricleoperitoneal shunting was performed using a high pressure-type neonate in-line valve (Heyer-SchulteTM). In the operating room, ventriculography and intraventricular CSF analyses were done. Ventricleoperitoneal shunting revealed no interventricular communication through the right foramen of Monro (Fig. 3). Results of CSF analyses were normal. Shortly after the operation, anterior fontanelle had sunken and overlapping suture lines were palpable. On the 7th postoperative day, occipitofrontal head circumference had decreased to 40 cm. The postoperative course
was uneventful and follow-up CT examination 1 month later revealed a markedly decreased right lateral ventricle with increasing cerebral mantle thickness but a large amount of subdural hygroma had collected (Fig. 4). Because the patient was asymptomatic, this was untreated. One year after, the follow-up CT shows more increased cerebral mantle thickness but still enlarged right lateral ventricle was evident without subdural hygroma (Fig. 5). The patient is now alert and his developmental milestones are not delayed.

Discussion

Previously, Dott has defined the term unilateral hydrocephalus as a condition in which one lateral ventricle has become actively enlarged due to restriction or complete

---

**Fig. 1.** Prenatal ultrasonography of a 32-week-old fetus showing severe unilateral dilatation of the right lateral ventricle (v). Biparietal diameter (arrows) of fetal head was about 9.4 cm.

**Fig. 2.** The preoperative gadolinium-enhanced MRI axial T1 weighted image showing markedly increased right lateral ventricle with thinning of cerebral mantle. The left lateral ventricle is collapsed and deviated to the left. There are no enhancing mass lesions.

**Fig. 3.** CT ventriculography performed during operation demonstrating the right lateral ventricle filled with contrast medium and no interventricular connection.

**Fig. 4.** Follow up CT 1 month after operation showing decreased size of the right lateral ventricle and increasing thickness of the ipsilateral cerebral mantle but a large amount of subdural hygroma (arrows) was collected.
blockage of outflow of the cerebrospinal fluid pathway. Congenital hydrocephalus results from several prenatal causes such as obstruction of aqueduct of Sylvius, congenital malformations of Dandy-Walker or Arnold-Chiari and intrauterine infection involving the central nervous system, especially congenital toxoplasmosis and cytomegalovirus infection. Most of these causes result in bilateral hydrocephalus, but congenital atresia of the foramen of Monro, which is quite rare, is known to cause unilateral hydrocephalus. Unilateral hydrocephalus results more commonly from postnatal causes. Tumors in the area of the foramen of Monro such as craniopharyngioma or hypothalamic astrocytomas may grow in size to occlude one of the foramina although bilateral obstruction occurs more frequently. In the patient with tuberous sclerosis, unilateral obstruction of the foramen of Monro occurs more frequently because they tend to develop subependymal giant cell astrocytoma in this region. Simple choroid plexus cyst and gliomatosis had been reported to result in unilateral hydrocephalus. Infectious processes such as intraventricular mumps infection and intraventricular hemorrhage can also lead to obstruction of the foramen of Monro.

Unilateral hydrocephalus can result not only from morphological obstruction of the foramen of Monro, but also from functional obstruction. Functional obstruction following ventriculo-peritoneal shunt and a patent foramen with unbalanced intracranial compliance has been reported to cause unilateral hydrocephalus. Oi et al. classified unilateral hydrocephalus into four categories. According to his categorization, category 1 is congenital atresia of the foramen of Monro; category 2 is morphological obstruction of the foramen; functional obstruction of the foramen is category 3; unbalanced intracranial compliance or unilateral parenchymal change is category 4.

There have been some reports about prenatal diagnosis of unilateral hydrocephalus. Prenatal ultrasonography is useful in the diagnosis of fetal hydrocephalus. As in our case, unilateral hydrocephalus can be easily diagnosed by ultrasonography.

Most of the previously reported cases of unilateral hydrocephalus have shown good prognosis without neurological deficit or mental retardation following ventriculoperitoneal shunt compared to bilateral congenital hydrocephalus, which may be related to the absence of other associated congenital anomaly. Early detection and treatment may affect good prognosis and should be emphasized.


References

4) Dott NM. A case of left unilateral hydrocephalus in an infant.
Operation-cure. Brain 50: 548-560, 1927

선천성 몬로공 폐쇄증에 의한 일측성 수두증

김종현·정용구·이남준·문수현·이훈갑·이기찬·서중근

= 국문초록 =

선천성 몬로공 폐쇄증에 의한 일측성 수두증이 발생할 수 있는 경우, 이는 그로 인한 일측성 수두증의 발생을 예방하고 치료하기 위해서는 그 원인을 규명하고 치료가 가능해야 한다. 본 연구에서는 선천성 몬로공 폐쇄증에 의한 일측성 수두증의 발생 원인 및 치료 방법에 대해 조사하였고, 이를 바탕으로 임상적 경험을 종합하여 논평하였다.

중심 단어: 선천성, 몬로공 폐쇄증, 일측성 수두증, 치료