

## Case Report

# Combined neuroendoscopic cyst wall fenestration and cyst-peritoneal shunt in an infant with gliependymal cyst

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## ABSTRACT

**Background:** Gliependymal cysts (GECs) are rare, benign congenital intracranial cysts that account for 1% of all intracranial cysts. Surgical interventions are required for patients with symptomatic GECs. However, the optimal treatment remains controversial, especially in infants. Here, we report a male infant case of GECs that successfully underwent minimally invasive combined neuroendoscopic cyst wall fenestration and cyst-peritoneal (CP) shunt.

**Case Description:** The boy was delivered transvaginally at 38 weeks and 6 days of gestation with no neurological deficits. Magnetic resonance imaging (MRI) at birth revealed multiple cysts with smooth and rounded borders and a non-enhancing wall in the right parieto-occipital region. The size of the cyst had increased rapidly compared to that of the prenatal MRI, which was performed at 37 weeks and 2 days. On the day of birth, Ommaya cerebrospinal fluid (CSF) reservoir was placed into the largest outer cyst. The patient underwent intermittent CSF drainage; however, he experienced occasional vomiting. At 2 months, he underwent combined neuroendoscopic cyst wall fenestration and CP shunt through a small hole. The patient's postoperative course was uneventful and there was no recurrence of the cyst. The pathological diagnosis was GEC.

**Conclusion:** Combined neuroendoscopic cyst wall fenestration and CP shunt are a minimally invasive and effective treatment for infants with GECs.

**Keywords:** Cyst-peritoneal shunt, Ependymal cyst, Gliependymal cyst, Neuroendoscopic cyst wall fenestration

## INTRODUCTION

Gliependymal cysts (GECs) are rare and benign congenital intracranial cysts that account for 1% of all intracranial cysts.<sup>[7]</sup> It is postulated that GECs originate from ectopic ependymal cells during the embryonic period because they have a capsule lined with ependymal epithelium, glial layer, and connective tissue<sup>[8]</sup> To date, only 32 cases of GECs have been reported, with the most cases in adults and a few cases in children, especially infants.<sup>[8]</sup> Most adult patients with GECs are asymptomatic; however, infants with GECs tend to be symptomatic.<sup>[8]</sup> Surgical interventions are needed for infant patients with symptomatic GECs. However, the optimal treatment remains controversial.<sup>[4,8]</sup> Here, we report an infant case of GECs that successfully underwent minimally invasive combined neuroendoscopic cyst wall fenestration and cyst-peritoneal (CP) shunt and achieved good outcomes.

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Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

## CASE DESCRIPTION

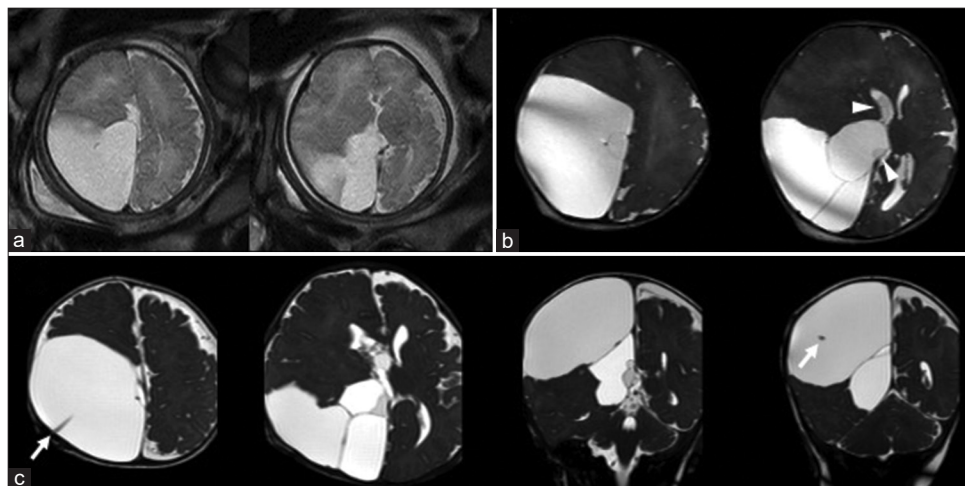
A 33-year-old, healthy, gravida 3, and para 2 woman underwent routine examination by transabdominal ultrasound at 35 weeks and 5 days during a seemingly uneventful pregnancy. Ultrasonography revealed an intracranial cyst in the baby. Prenatal half-Fourier acquisition single-shot turbo spin-echo sequence (HASTE) magnetic resonance imaging (MRI) performed at 37 weeks and 2 days showed a large cyst measuring 70 mm in maximal length in the right parieto-occipital region [Figure 1a].

A boy weighing 3204 g was delivered transvaginally at 38 weeks and 6 days gestation. The patient had Apgar scores of 8 and 9, respectively. The tension in the fontanel was soft. The head circumference was 34 cm (+0.62SD). The patient showed no neurological deficits. At birth, the initial MRI, including 3D-heavily T2-weighted MRI (3D-hT2WI), demonstrated multiple cysts with smooth and rounded borders and a nonenhancing wall in the right parieto-occipital region. The cyst contents were nearly isointense to the cerebrospinal fluid (CSF) but varied in intensity in each lesion [Figure 1b]. The size of the cyst increased rapidly, measuring 90 mm in maximum length, resulting in a mild midline shift. The cysts were not connected to the subarachnoid or right lateral ventricle. Another remarkable finding was that the posterior part of the corpus callosum was partially absent. On the day of birth, Ommaya CSF reservoir was placed into the largest outer cyst. The cyst contained clear

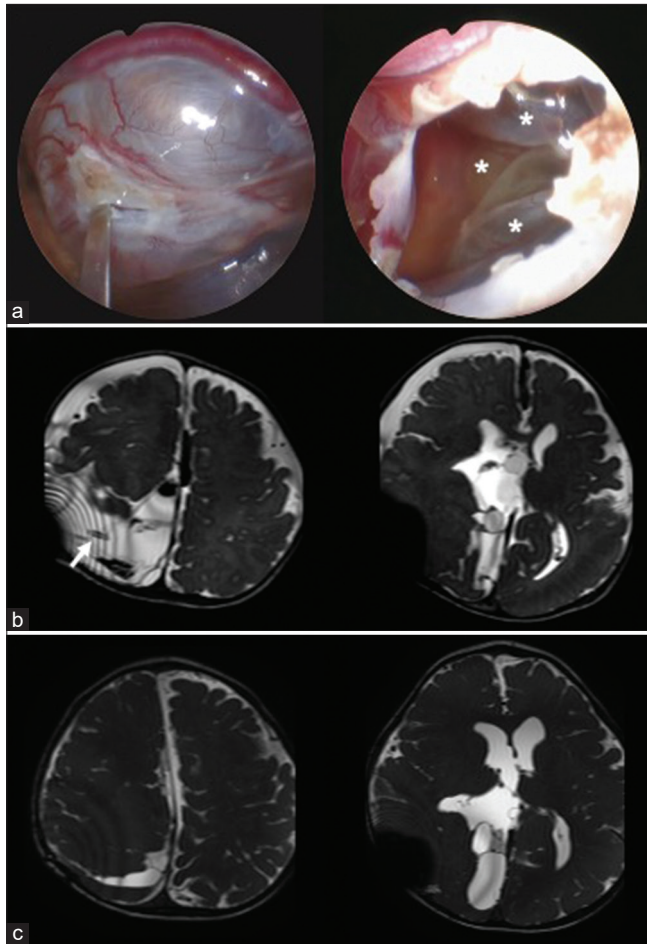
CSF-like fluid. The patient was discharged on postoperative day (POD) 17 and he underwent intermittent CSF drainage through the reservoir. However, he experienced occasional vomiting, and the tension of his fontanel gradually worsened 44 days after the first surgery. 3D-hT2WI at 2 months revealed that the cyst remained large with a mild midline shift even with intermittent drainage [Figure 1c]. We decided on surgical intervention. The patient underwent surgery at 57 days of age. After removing the Ommaya CSF reservoir, the endoscope was inserted into the largest outer cyst wall through the small hole created during the first surgery. The GECs consisted of a thin translucent membrane and veins were observed running over the cyst surface [Figure 2a]. We performed cyst wall fenestration of the largest outer and inner cysts [Figure 2a]. CSF-like clear fluid was found in the inner cyst. Finally, a CP shunt was placed into the large cyst.

Postoperatively, the tension in his fontanel became soft, and vomiting disappeared. 3D-hT2WI performed 4 days after the second surgery revealed that the size of the cyst had decreased [Figure 2b]. The patient's postoperative course was uneventful and he was discharged on POD 15. At the 2-year follow-up, there was no cyst recurrence [Figure 2c], his neurological findings did not deteriorate, and his development was normal.

Pathological examination revealed that the cyst wall was mainly composed of fibroconnective tissue and was partially lined by cuboidal or columnar epithelial cells. A ciliated epithelium was also observed, but goblet cells were not observed. The lining cells were immunopositive for cytokeratin (AE1/AE3) and podoplanin [Figure 3]. In conjunction with radiological



**Figure 1:** (a) Prenatal half-Fourier acquisition single-shot turbo spin-echo sequence (HASTE) image at 37 weeks and 2 days of gestation showing a huge cyst in the right parieto-occipital region. (b) 3D-heavily T2-weighted magnetic resonance imaging (3D-hT2WI) at birth (at 38 weeks and 6 days) revealed multiple cysts with various intensities in the right parieto-occipital region (white arrowheads). These cysts were not connected to the right lateral ventricle. The posterior part of the corpus callosum was partially absent. (c) Axial and coronal 3D-hT2WI at 2 months revealed that the cyst remained large even after intermittent drainage. The Ommaya reservoir is placed in the largest cyst (white arrows).

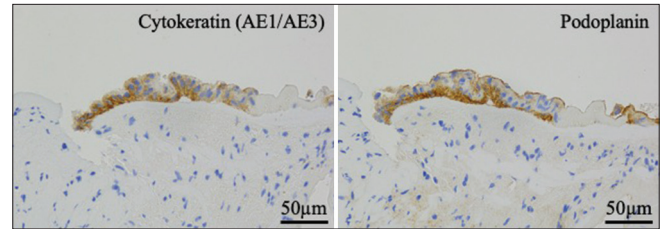


**Figure 2:** (a) Intraoperative photographs of neuroendoscopic cyst wall fenestration. Multiple cysts are shown inside a huge cyst performed wall fenestration (asterisks). (b) 3D-hT2WI 4 days after the operation, revealing the volume of the huge cyst decreased, and cyst-peritoneal shunt was placed into the huge cyst (white arrow). (c) 3D-hT2WI 1 year after operation depicting the volume of the cyst more decreased.

findings of multiple intracerebral cysts and partial defects of the corpus callosum, the pathological diagnosis was GEC.

## DISCUSSION

GECs have several anatomical and clinical characteristics. First, they can originate from different intracranial locations, including the intraparenchymal, intraventricular, and subarachnoid spaces.<sup>[2,6,8-11]</sup> During the embryonic period, in the wall of the neural tube corresponding to the tela choroidea, a transition exists between ciliated ependyma on glial tissue and nonciliated epithelium on connective tissue. For some reason, a short segment of the wall starts ballooning into the cerebral tissue or the subarachnoid space, followed by the formation of a cyst.<sup>[4]</sup> For these embryological reasons, GECs are thought to exist at different intracranial locations. In this



**Figure 3:** Immunohistochemical findings of the epithelium of the cyst. The epithelial cells lining the fibrous wall are immunopositive for cytokeratin (AE1/AE3) and podoplanin.

case, GECs were located in paraventricular lesions, especially around the right lateral ventricle [Figure 1c]. Considering that a previous review reported that GECs located in the lateral ventricle were present in only 1 case (3.7%).<sup>[8]</sup> Our case is extremely rare. Furthermore, GECs show progressive growth in size, especially in fetal or infant cases, resulting in neurological symptoms.<sup>[11]</sup> GECs contain ependymal cells positioned on glia and the secretory activity of ependymal cells may cause progressive growth of the cyst.<sup>[8]</sup> In this case, GECs showed progressive growth in size during only 2 weeks of the fetal period [Figures 1a and b]. Further, infant cases under 1 year of age are rare, and only five cases have been reported [Table 1]. The characteristic of infant GECs cases is symptomatic cysts.<sup>[11]</sup> On the other hand, almost all GECs in adult patients are found incidentally.<sup>[8]</sup> In children, macrocephaly is one of the most common symptoms, followed by failure to thrive.<sup>[8]</sup> Our patient also presented with macrocephalus and high intracranial pressure. In addition, in some cases of GECs, radiological imaging shows developmental anomalies, and the most common anomaly is corpus callosum agenesis.<sup>[10]</sup> The posterior part of the corpus callosum was partially absent in the present patient [Table 1].

For asymptomatic GECs, surgical treatment is typically not necessary. However, symptomatic patients tend to present with symptoms related to increased intracranial pressure, and local mass effect surgical treatment is needed. Common surgical approaches for GECs include burr hole opening for evacuation of the fluid component, open craniotomy for total extirpation, open cysto-ventricular fenestration, cystosubarachnoid shunting, partial cyst excision, cystoperitoneal shunt, or a combination of these approaches.<sup>[8]</sup> Recently, Alvarado *et al.* reported that endoscopic cystoventricular fenestration is a safe and effective surgical option.<sup>[1]</sup> Because of the rarity of GECs and the various locations of cysts, the optimal treatment for symptomatic GECs remains unclear. Especially, in infants, surgical invasion should be considered in selecting the surgical method. In the previous reports of infant GECs, the surgical treatment of choice was craniotomy in all cases.<sup>[2,6,9-11]</sup> In one case, a CP shunt was selected first; however, craniotomy was selected secondarily after recurrence recurrence.<sup>[6]</sup> The outcome was good in four cases and the cyst recurred at the 1-year follow-up in one case. The previous reports recommend



**Table 1:** Infants presenting with gliependymal cysts.

No.	Reference	Age	Sex	Symptoms	Cyst location	Associated lesions	Treatment	Outcome
1	Takehita <i>et al.</i> , 1982	1 month	M	Macrocephalus	Paraventricular cyst	None	Craniotomy	Good
2	Tange <i>et al.</i> , 2000	1 month	M	Macrocephalus	Interhemispheric cyst	Corpus callosum agenesis	Craniotomy	Good
3	Obaldo <i>et al.</i> , 2007	1 day	F	Proptosis	Middle fossa-orbital cyst	None	CP shunt (after 14 days) → Craniotomy	Good (after 2 <sup>nd</sup> operation)
4	Zheng <i>et al.</i> , 2013	3 months	NS	Macrocephalus	Paraventricular cyst	None	Craniotomy	Recurrence (1 year)
5	Cho <i>et al.</i> , 2014	4 months	F	Proptosis Limited EOM	Midline anterior cyst	None	Craniotomy	Good
6	Present case	2 months	M	Vomit Macrocephalus	Paraventricular cyst	Corpus callosum partial absence	Endoscopic cyst opening CP shunt	Good

M: Male, F: Female, EOM: External ocular movement, CP: Cystoperitoneal

complete resection of the cyst wall of GECs through open craniotomy, considering the recurrent nature of the cyst and additional surgical intervention in the future.<sup>[3]</sup> However, open craniotomy for complete or partial cyst wall resection presents a more invasive approach. Excessively invasive surgical methods should be avoided for infant patients with low body weights. However, the urgent treatment is required considering the development of the patient. Successful treatment of giant GECs at the supracerebellar cistern in a 35-month-old girl with endoscopic fenestration of the cyst wall through a burr hole was published.<sup>[5]</sup> At the 5-year follow-up, no evidence of recurrence was observed. In this case, we successfully treated a symptomatic infant patient with GECs using combined neuroendoscopic cyst wall fenestration through burr hole access and CP shunt; we achieved good outcomes at the 2-year follow-up.

There is a concern about shunt malfunction or obstruction for patients who underwent CP shunting, and we believe that this combined neuroendoscopic cyst wall fenestration and CP shunt is a minimally invasive and considerable treatment for infant patients with GECs. To the best of our knowledge, this is the first reported case of an infant treated using this minimally invasive combined surgical procedure.

## CONCLUSION

Combined neuroendoscopic cyst wall fenestration and CP shunt is a minimally invasive and effective treatment for infants with GECs.

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## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

## Conflicts of interest

There are no conflicts of interest.

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