

# Blake's Pouch Cyst: A Case Report

Saurabh Kumar Verma<sup>1</sup> M. S. Sridhar<sup>1</sup> S. Shashivadhanan<sup>1</sup> Manish Garg<sup>1</sup>

<sup>1</sup>Department of Neurosurgery, Army Hospital (R&R), New Delhi, India

Address for correspondence Col (Dr.) Saurabh Kumar Verma, Department of Neurosurgery, Army Hospital (R&R), New Delhi, Delhi Cantt, India (e-mail: eskay6362@yahoo.co.in).

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## Abstract

It is a rare and underdiagnosed entity. The adagium “one only sees what one knows” is certainly true in cases of Blake's pouch cyst, as all types of posterior fossa cysts and cyst-like malformations may present nearly identical on initial imaging studies. Only one case of Blake's pouch cyst has been reported from this country, except for a case in utero, in which a diagnosis of Blake's pouch cyst was made on prenatal ultrasound and later confirmed by MRI. In this report we describe a case of Blake's pouch cyst in a 9-month-old male child along with the principles of diagnosis of Blake's pouch cyst, in combination with literature review. Differentiating Blake's pouch cyst from other posterior fossa cysts and cyst-like malformations and recognizing the accompanying hydrocephalus that are essentially noncommunicating have important implications not only on clinical management but also on genetic counseling, which is unnecessary in case of Blake's pouch cyst.

## Keywords

- ▶ Blake's pouch cyst
- ▶ posterior fossa
- ▶ inferior medullary velum

## Introduction

Blake's pouch is a normal, transient embryologic cystic-appearing structure that represents posterior ballooning of the inferior medullary velum into the cisterna magna, below and posterior to the vermis that communicates with open fourth ventricle through foramen of Magendie.

Blake's pouch cyst is caused by a failure of regression of Blake's pouch secondary to the nonperforation of the foramen of Magendie.<sup>1–3</sup> Importantly, during embryologic development, the foramina of Luschka open later<sup>4</sup> than the foramen of Magendie.<sup>5–7</sup> Therefore, in case of nonperforation of the foramen of Magendie, the fourth ventricle will enlarge together with the supratentorial ventricles until the foramina of Luschka open and establish an equilibrium of cerebrospinal fluid (CSF) outflow from the ventricles into the cisterns.<sup>8</sup>

However, as the larger foramen of Magendie is permanently missing, the ventricles may stay enlarged (▶**Fig. 1**). According to this theory, in case of Blake's pouch cyst, the cerebellar hemispheres and vermis would rather be compressed (to a certain degree) (▶**Fig. 2**) than underdeveloped, and would therefore reexpand in case of

ventricular shunting.<sup>9</sup> Only one case Blake's pouch cyst has been reported from India by Vakakmudi et al, except for a case in utero, in which a diagnosis of Blake's pouch cyst was made on prenatal ultrasound and later confirmed by magnetic resonance imaging (MRI). Differentiating Blake's pouch cyst from other posterior fossa cysts and cyst-like malformations and recognizing the accompanying hydrocephalus that are essentially noncommunicating have important implications not only on clinical management but also on genetic counseling, which is unnecessary in case of Blake's pouch cyst.

## Case Summary

This was a case of a 9-month-old male child, who presented to our center on September 20, 2015, with 2 months history of the parents having noticed rapidly increasing head size. There was no history of any delayed or reversed milestone. Clinically the patient had bulging anterior fontanelle and exhibited no other signs of increase in intracranial pressure. Head circumference at the time of presentation was 47 cm. MRI of the head showed typical features with hydrocephalus.

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**Fig. 1** MRI of the brain showing patent “aqueduct of Sylvius” along with supratentorial ventriculomegaly in sagittal, axial, and coronal sections (white arrow).

Endoscopic third ventriculostomy (ETV) was done on September 23, 2015, with uneventful postoperative period and gratifying follow-up at 4 months (►Figs. 1, 2).

## Discussion

The adage “one only sees what one knows” is certainly true for Blake’s pouch cyst, as all types of posterior fossa cysts and cyst-like malformations may present nearly identical on initial imaging studies. Blake’s pouch cyst is named after Joseph A. Blake (1864–1937), an American physician who studied the physiology related to the foramen of Magendie.<sup>10</sup> It was first described as an independent entity within the Dandy-Walker complex in 1996 by Tortori-Donati et al.<sup>1,10</sup>

The range of clinical presentation varies from antenatal incidental detection with later spontaneous resolution of Blake’s pouch cyst<sup>11</sup> that, according to the authors, was probably due to delayed perforation of foramina of Magendie, fatal hydrocephalus, with the intermediate range of presentations varying from asymptomatic incidental detection, arrested hydrocephalus, increasing head circumference with or without impaired neurologic development in infants, and decompensating hydrocephalus

at an advanced age. The age of detection has been reported from in utero to as late as 69 years of age.

Our case presented with features of rapidly increasing head size with abnormal head circumference.<sup>1,11</sup>

Typical radiologic features of Blake’s pouch cyst include tetraventricular hydrocephalus, infra- or retrocerebellar cyst, a relatively well-developed, nonrotated cerebellar vermis (as opposed to a Dandy-Walker), cystic dilation of the fourth ventricle without cisternal communication, and some degree of compression on the medial cerebellar hemispheres. Ideally, one may see the fourth ventricular choroid plexus continuing in the roof of the cyst on sagittal magnetic resonance images.<sup>1</sup> The position of the fourth ventricular choroid plexus (not to be misinterpreted as a prominent inferior vermian vein) is different in Blake’s pouch cyst, posterior fossa arachnoid cyst, and Dandy-Walker malformation. After intravenous contrast administration, the choroid plexus is readily seen bending below the vermis and continuing into the pouch in Blake’s pouch cyst, which is essentially a fourth ventricular diverticulum, whereas it is in a normal position in a posterior fossa arachnoid cyst and nonexistent in Dandy-Walker malformation. Blake’s pouch needs to be distinguished from other causes of enlarged retrocerebellar “CSF” space.



**Fig. 2** MRI of the brain showing communication of Blake's pouch cyst with fourth ventricle and membranous wall separating cyst from cisterna magna.

- Mega cisterna magna: communicates freely with both fourth ventricle and subarachnoid space
- Other entities of the Dandy-Walker continuum: hypoplasia of the vermis, high position of the torcular
- Absence of the falx cerebelli arachnoid cyst: no hydrocephalus<sup>1</sup>

Most authors agree that Blake's pouch cyst and mega cisterna magna originate from a defect of the posterior membranous area, whereas Dandy-Walker malformation and Dandy-Walker variant originate from a defect of the anterior membranous area.<sup>3,5</sup>

## Management

ETV is a safe and effective treatment option in patients presenting with a symptomatic Blake's pouch cyst, avoiding the risks and added morbidity of open surgery, as well as shunt dependency with obstruction, overdrainage, and other shunt-related problems.<sup>1,12</sup> We managed our case with ETV with gratifying result at follow-up after 4 months.

## Other Treatment Options

As hydrocephalus in patients with Blake's pouch cyst is caused by nonperforation of the foramen of Magendie, at least theoretically one might consider marsupializing the cystic fourth ventricle, trying to reconstruct a foramen of Magendie and reestablish the normal CSF pathway.<sup>1</sup>

According to Tortori-Donati et al, shunting the lateral ventricles (or pouch) restores normal ventricular size, including collapse of the cystic fourth ventricle.<sup>3</sup>

## Conclusion

The aim of presentation is to sensitize the readers to the fact that all types of posterior fossa cysts and cyst-like malformations may present nearly identical on initial imaging studies.

Differentiating Blake's pouch cyst from other posterior fossa cysts and cyst-like malformations and recognizing the accompanying hydrocephalus that are essentially noncommunicating have important implications not only on clinical management but also on genetic counseling, which is unnecessary in case of Blake's pouch cyst.

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