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Case Report

Fetal and postnatal MRI findings of Blake pouch remnant causing obstructive hydrocephalus [☆]

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ABSTRACT

Blake pouch remnant, also known as Blake pouch cyst or persistent Blake pouch, is a posterior fossa embryologic anomaly that is often seen in isolation with most affected patients being asymptomatic. However, even in isolation, Blake pouch remnant can result in obstructive hydrocephalus requiring early neurosurgical intervention making it an important diagnosis for the fetal radiologist to consider. We present a rare case of a patient with prenatally diagnosed “inferior vermian hypoplasia” on fetal MRI that went on to develop progressive obstructive hydrocephalus in infancy secondary to what was determined to be a Blake pouch remnant.

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Introduction

Blake pouch is a normal embryologic structure that arises from the tele choroidea along the roof of the developing rhombencephalon which is seen in the first trimester fetus, though is typically not visible by imaging by the second trimester [1]. Blake pouch fenestrates to varying degrees in the late first trimester and early second trimester and can persist in the fetal and postnatal period, either in isolation or coexisting with vermian hypoplasia [2]. These posterior fossa anomalies are frequent referrals to many fetal imaging

practices to “rule out Dandy-Walker Malformation.” While classic Dandy-Walker malformation portends a relatively poor prognosis, the clinical implications of prenatally isolated Blake pouch remnant and/or vermian hypoplasia are less clear.

We present a rare case of a patient with Blake pouch remnant that was initially referred for fetal MRI at 36 weeks gestational age (GA) for further evaluation of ventriculomegaly on fetal ultrasound (US). Fetal MRI demonstrated what was described as “inferior vermian hypoplasia” with flattening of the inferior vermis. By the age of 3 months, the patient had severe progressive ventriculomegaly with obstructive hydrocephalus requiring neurosurgical intervention. This case highlights the

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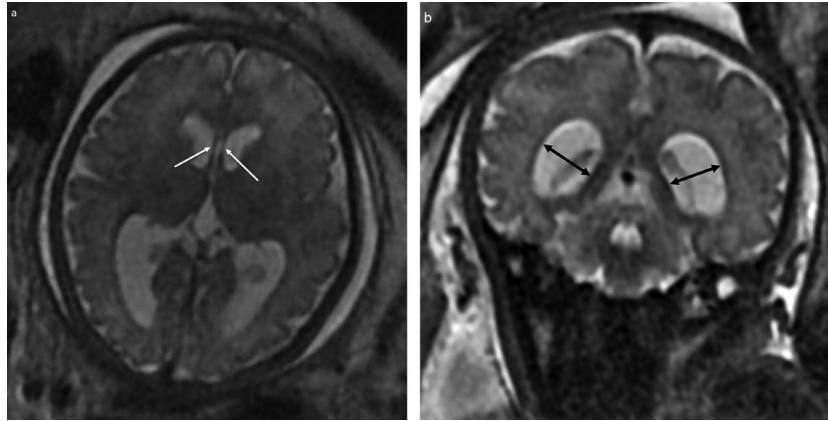


Fig. 1 – Axial (a) and coronal (b) T2-SSFSE images from fetal MRI performed at 36 weeks 2 days GA confirms normal appearance of the cavum septi pellucidum (white arrows), however there is moderate ventriculomegaly of the lateral ventricles. Transverse atrial diameter measured on coronal images (black arrows) were 15 mm bilaterally.

importance of accurate characterization of posterior fossa findings on fetal MRI particularly in the setting of ventriculomegaly and the need for close postnatal follow-up.

Case Report

A 33-year-old woman was referred to the Cincinnati Fetal Center at 36 weeks 2 days GA for further evaluation of fetal ventriculomegaly and nonvisualization of the cavum septum pellucidum identified on prenatal US at 34 weeks 5 days GA. This case report is Health Insurance Portability and Accountability Act compliant and did not require approval from the Institutional Review Board. Signed informed consent to use de-identified medical information for research purposes was obtained from the patient at initial evaluation and retained within the electronic medical records system.

Pregnancy was only remarkable for hyperemesis and intermittent high blood pressure, requiring no medication management. Her past medical history was remarkable for migraine headaches. There was no history of diabetes. The patient denied any illnesses, infections, or known adverse environmental exposures during the pregnancy. Her 10-year-old daughter had a history of attention deficit hyperactivity disorder. There was no consanguinity nor any family history of birth defects, genetic syndromes, other recurrent fetal losses, or neonatal deaths.

Fetal MRI was performed at 36 weeks 2 days GA. The cavum septi pellucidum was clearly present and appeared normal, as did the corpus callosum. There was moderate symmetric ventriculomegaly of the lateral ventricles, with the transverse atrial diameter measuring up to 15 mm on each side (Fig. 1a, b). The cerebral aqueduct appeared patent. The tectum-vermian angle measured 12°. The overall height of the vermis fell within normal range for GA, however it was prospectively noted that the inferior surface of the vermis appeared flattened and was smaller than the superior half of the vermis.

The cisterna magna was not enlarged and the torcular was not elevated. No other abnormalities were identified (Fig. 2a-d).

The baby was born at 37 weeks GA. Head circumference at birth was at the 37th percentile and initial neurologic exam at birth was normal. Head US on the first day of life was performed and was remarkable for a new left grade I germinal matrix hemorrhage. The ventricles were prospectively interpreted as normal in size, though in retrospect appear enlarged with a transverse atrial diameter of approximately 15 mm, similar to the fetal MRI. Transmastoid views of the posterior fossa were normal. At three months of age, the patient presented with increased head circumference, now at the 93rd percentile. A repeat head US demonstrated marked interval increase in ventriculomegaly of the lateral and third ventricles, with the transverse atrial diameter greater than 3 cm. Limited acoustic window precluded a more detailed evaluation (Fig. 3a-d).

The patient underwent brain MRI one week later. Again seen was severe ventriculomegaly of the lateral and third ventricles, without significant periventricular edema. The cerebral aqueduct was widely patent, and a dephasing jet is seen on T2 weighted images confirming its patency. The cerebellar vermis was again rotated, marked by an elevated tectum-vermian angle now 22°, and was flattened along its inferior aspect. The fourth ventricle choroid plexus was displaced along the inferior surface of the vermis. Best seen on 3D FIESTA/TruFISP images is a thin obstructing membrane along the inferior aspect of the posterior fossa dorsal to the cervicomedullary junction. Imaging findings are consistent with Blake pouch remnant resulting in ventricular obstruction. No other abnormalities in the brain were identified (Fig. 4a-e).

The patient was referred to neurosurgery as a result of the imaging findings. She was clinically asymptomatic however an additional repeat MRI at 11 months of age showed further increase in the degree of ventriculomegaly and as a result she underwent suboccipital craniotomy with open cyst fenestration at that time. However, she continued to have excessive enlargement of head circumference and difficulties

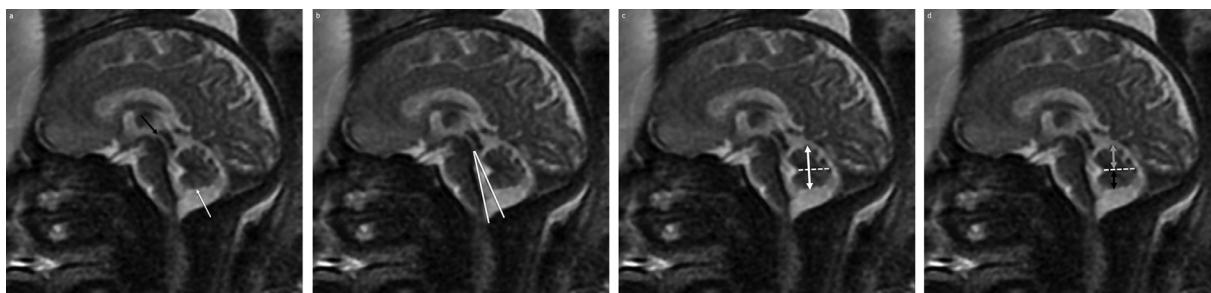


Fig. 2 – Sagittal T2-SSFSE images of the fetal vermis at midline from fetal MRI at 36 weeks 2 days GA. On first inspection (a) is flattening of the inferior aspect of the vermis (white arrow) noted, and the cerebral aqueduct (black arrow) appears patent. The tegmento-vermian angle (b) is mildly elevated at 12 degrees. Overall vermis size (c) demonstrates normal AP (white dashed line) and normal height (white arrow) of the vermis. When evaluating superior-inferior vermian distribution (d) the superior vermis (grey arrow) is larger than the inferior vermis (black arrow).

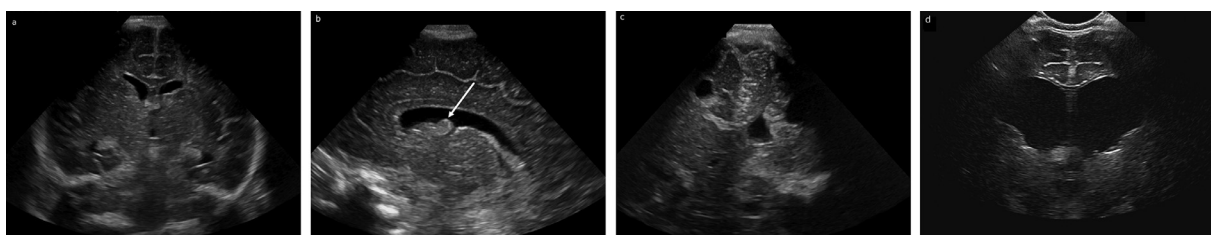


Fig. 3 – Coronal ultrasound image (a) of the brain at day of life 0 demonstrates no apparent progressive ventriculomegaly when compared to the fetal imaging. Best seen on the sagittal image (b) there is a grade I germinal matrix hemorrhage (white arrow) in the left caudothalamic groove. Transmastoid image (c) of the posterior fossa demonstrates no gross abnormalities. Coronal images from repeat head ultrasound performed at 3 months of age (d) demonstrates marked interval worsening in the degree of ventriculomegaly involving the lateral ventricles.

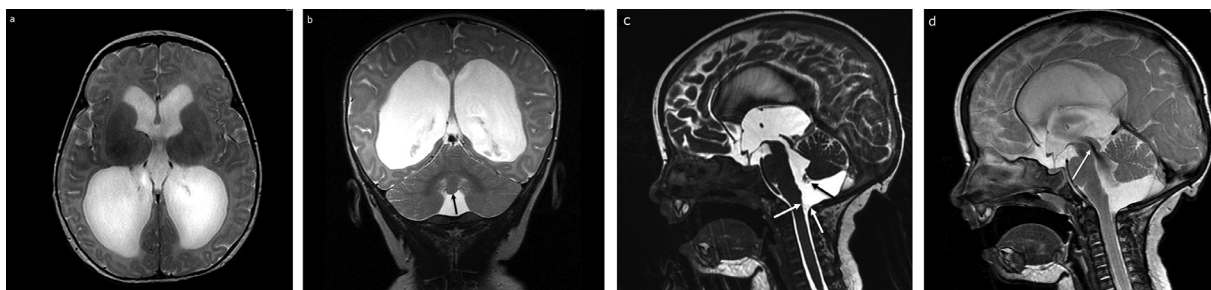


Fig. 4 – Axial (a) and coronal (b) T2-FSE images through the brain at 3 months of age demonstrate severe ventriculomegaly of the lateral and third ventricles. No apparent periventricular edema to suggest acute ventricular obstruction. On the coronal image the inferior vermis (black arrow) appears elevated. Best seen on 3D sagittal FIESTA/TruFISP image through midline (c) is the thin obstructing membrane dorsal to the brainstem at the level of the foramen magnum (white arrows) compatible with Blake pouch remnant. There is also characteristic inferior displacement of the choroid plexus (black arrow). Sagittal T2-FSE image (d) demonstrates a dephasing jet (white arrow) through the cerebral aqueduct confirming its patency.

maintaining balance. As a result a ventriculo-peritoneal shunt was placed at 15 months of age. At age 3, she had no gross motor, cognitive or speech deficits and was discharged from neurology clinic. She continued to receive occupational therapy for mild fine motor deficits.

Discussion

Blake pouch is a normal embryologic structure that arises from the posterior membranous area from the roof of the

primitive rhombencephalon. By around the fourth-fifth week of gestation, the posterior membranous area expands to form Blake pouch, which rather than a true cyst is in direct communication with the fourth ventricle [2]. This pouch then fenestrates to varying degrees to create the foramina of Lushka and Magendie, allowing for normal outflow of the CSF generated within the choroid plexus within the ventricular system.

Blake pouch is not typically visible by imaging in the second trimester on fetal MRI, by which time the fourth ventricle is typically “closed” which is marked by a normal tegmento-vermian angle. The diagnosis of Blake pouch remnant on fetal imaging can be challenging. When evaluating the fetal cerebellar vermis, there are 4 main points to consider: (1) tegmento-vermian angle, (2) overall vermis size, (3) superior-inferior vermis proportions, and (4) foliation pattern. The tegmento-vermian angle is the angle between the dorsal aspect of the brainstem and ventral aspect of the vermis. Recent fetal MRI literature describes an angle of less than 10° by 24 weeks GA as normal, which is smaller than measurements previously described in the US literature of 18° [5]. The vermis size measured by largest height and anterior-posterior (AP) dimensions, parallel and perpendicular to the axis of the ventral vermis, respectively, should also be routinely measured and compared to normal values. Superior and inferior vermis height proportions can also be evaluated on either side of the AP dimension or fastigial point-decline line, with the height of the inferior vermis being about equal to or slightly more than the superior vermis [3]. Finally, vermian lobulation can be assessed with all major lobules visible by 27 weeks GA.

The diagnosis of a Blake pouch remnant on fetal imaging mainly lies in the elevated tegmento-vermian angle with an otherwise normal size and morphology of the fetal vermis [4]. On postnatal imaging an additional characteristic feature is displacement of the choroid plexus along the inferior aspect of the vermis. However, vermian hypoplasia and Blake pouch remnant can coexist along the same pathologic spectrum, which explains rotation of the vermis in cases of vermian hypoplasia [2]. Blake pouch remnant in isolation is typically an incidental finding and greater than 90% of those affected have reportedly normal 5 year neurologic outcomes without intervention [6]. However, obstructive hydrocephalus secondary to Blake pouch remnant can occur with the largest series in the literature describing 6 cases, 4 of which were in children and 5 of which required neurosurgical intervention [7]. While an elevated tegmento-vermian angle is the key to diagnosis, mild to moderate ventriculomegaly is the most common finding on prenatal imaging, which typically resolves or remains stable in about 84% of patients and progresses in about 16% [6].

Prenatal diagnosis of Blake pouch remnant resulting in postnatal obstructive hydrocephalus is extremely rare with only 1 case previously reported in the literature [8]. This reported case, like ours, also showed ventriculomegaly on fetal MRI. Our case is unique because the degree of ventriculomegaly was only moderate, rather than severe. Also, the extent of the posterior fossa abnormality was subtle in our case as the tegmento-vermian angle was only 12°, which would have fallen within normal range based on the existing litera-

ture at the time of the exam. The 1 clue to the fetal imager that allowed for prospective consideration of a causative posterior fossa abnormality was the description of small inferior vermis with associated flattening. Though this is sometimes referred to as “inferior vermian hypoplasia” the existence of this entity has come under question as the vermis is believed to form ventral to dorsal rather than superior to inferior, meaning it is likely that many cases of presumed “inferior vermian hypoplasia” are likely Blake pouch remnants [4]. The other primary differential consideration is an arachnoid cyst, which does not freely communicate with the fourth ventricle and would not typically cause elevation of the tegmento-vermian angle. This case highlights the importance of careful descriptions of vermis abnormalities particularly in the setting of ventriculomegaly. This case also emphasizes the importance of close neurological postnatal follow-up in these patients to allow for timely neurosurgical intervention as needed. Finally, our case has long term follow-up illustrating how a patient with isolated Blake pouch remnant can ultimately have a good clinical outcome even in the setting of obstructive hydrocephalus with timely neurosurgical intervention.

In conclusion, Blake pouch remnant is an important diagnosis that can be made on fetal MRI by careful evaluation of the tegmento-vermian angle, vermis size, superior-inferior vermian proportions, and vermian morphology. Even though in isolation the prognosis is good, close postnatal follow-up is necessary in order to evaluate for developing obstructive hydrocephalus, particularly in the setting of fetal ventriculomegaly. With prompt neurosurgical intervention, patients who develop obstructive hydrocephalus secondary to Blake pouch remnant can have a good outcome.

Patient Consent Statement

Signed informed consent to use de-identified medical information for research purposes was obtained from the patient at initial evaluation and retained within the electronic medical records system.

REFERENCES

- [1] Chapman T, Menashe SJ, Zare M, Alessio AM, Ishak GE. Establishment of normative values for the fetal posterior fossa by magnetic resonance imaging. *Prenat Diagn* 2018;38(13):1035–41.
- [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. *Child's Nerv Syst* 1996;12(6):303–8.
- [3] Robinson AJ, Blaser S, Toi A, Chitayat D, Halliday W, Pantazi S. The fetal cerebellar vermis: Assessment for abnormal development by ultrasonography and magnetic resonance imaging. *Ultrasound Q* 2007;23(3):211–23.
- [4] Robinson AJ, Ederies MA. Diagnostic imaging of posterior fossa anomalies in the fetus. *Semin Fetal Neonatal Med* 2016;21(5):312–20.

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- [5] Volpe P, Contro E, De Musso F, Ghi T, Farina A, Tempesta A. Brainstem-vermis and brainstem-tentorium angles allow accurate categorization of fetal upward rotation of cerebellar vermis. *Ultrasound Obstet Gynecol* 2012;58(3):509–15.
- [6] Gandolfi Colleoni G, Contro E, Carletti A, Ghi T, Campobasso G, Rembouskos G. Prenatal diagnosis and outcome of fetal posterior fossa fluid collections. *Ultrasound Obstet. Gynecol* 2012;39(6):625–31.
- [7] Cornips EMJ, Overvliet GM, Weber JW, Postma AA, Hoeberigs CM, Baldewijns MLL. The clinical spectrum of Blake's pouch cyst: report of six illustrative cases. *Child's Nerv Syst* 2010;26(8):1057–64.
- [8] Vakakmudi U, Rangasami R, Gopinath V. Prenatal Blake pouch cyst with hydrocephalus. *Neurol India* 2016;64(4):830–1.