

Cavum veli interpositi cyst: prenatal diagnosis and postnatal outcome

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ABSTRACT

Objective The cavum veli interpositi (CVI) is a space within the double-layered tela choroidea of the third ventricle. Occasionally, this space is fluid-filled and sonographically visible as an interhemispheric anechoic cyst. Because of its rarity, the incidence of CVI cyst is undetermined and the outcome of affected individuals has been found to be variable. The aim of this study was to report our experience of the sonographic findings and outcome of fetuses affected by CVI cysts.

Methods In five fetuses with a CVI cyst, we performed targeted prenatal ultrasound scans of intracranial structures and a detailed anatomical survey to rule out associated malformations. Follow-up consisted of neurological examination and neurosonography.

Results The CVI cyst appeared as a well-defined anechoic lesion without adjacent mass effect. In all fetuses the cyst was single and in two cases it enlarged slightly during pregnancy. The cyst was isolated in three fetuses and associated with borderline ventriculomegaly in two. A single umbilical artery was the only associated extracranial anomaly and this was detected in only one fetus. Neurosonography confirmed the presence of CVI cysts in all cases after delivery. During postnatal follow-up (range 10–48 months), the cyst regressed in one case within 1 month after delivery while the size of the others remained stable. No infant developed psychomotor disorders.

Conclusions Prenatal sonographic diagnosis of CVI cysts is feasible. Its finding in isolation is consistent with favorable postnatal outcome. Copyright © 2009 ISUOG. Published by John Wiley & Sons, Ltd.

INTRODUCTION

The velum interpositum is the double-layered tela choroidea of the third ventricle. It is located anteroinferior to the splenium of the corpus callosum and below the

columns of the fornix¹. It is bordered anteriorly by the foramen of Monro and posteriorly by the pineal body. The cavum veli interpositi (CVI) is the space within the double-layered tela choroidea and occasionally it is fluid-filled and sonographically visible as an interhemispheric anechoic cyst with regular borders. It is not certain whether CVI cysts represent a simple variant of the norm or are developmental malformations. Because of its rarity, the distinction between a dilated cavum and a true cyst is arbitrary. Furthermore, the prognosis associated with CVI cysts is uncertain and may be related to the size of the cyst when detected postnatally. Psychotic disorders², low-pressure hydrocephalus³, psychomotor retardation and epilepsy⁴ have all been reported in pediatric and adult patients with large CVI cysts but normal outcomes have been reported in prenatally diagnosed cases^{5,6}.

The aim of this study was to report our experience of the prenatal ultrasound findings and outcome of fetuses affected by CVI cysts.

METHODS

This was a retrospective study of five fetuses diagnosed in our hospital as being affected with CVI cyst in the period between March 2004 and January 2007. All five cases had been referred for sonographic investigation, due to the suspicion of either isolated borderline ventriculomegaly ($n = 2$) or isolated interhemispheric cyst ($n = 3$). The gestational age at the time of first examination in our unit ranged from 19 to 30 weeks and the maternal age ranged from 20 to 41 years. One patient had a monochorionic diamniotic twin pregnancy with only one fetus affected with the CVI cyst.

All ultrasound examinations were performed by one experienced operator (V.D.). Transabdominal ultrasound using an Aloka Prosound 5500A (Aloka Co., Tokyo, Japan) ultrasound machine equipped with a 5-MHz curvilinear probe was performed in all cases and in three cases with cephalic presentation a transvaginal examination

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with a 7.5-MHz convex probe was also performed. By the time the last three cases were examined, three-dimensional evaluation had become available in our unit (GE Voluson Pro, GE Healthcare Technologies, Milwaukee, WI, USA). Axial, sagittal and coronal planes of the fetal brain were used to evaluate the shape, size (mean of the anteroposterior, vertical and transverse diameters) and anatomical relationships of the cyst. All the fetuses also underwent a detailed anatomical survey to rule out associated anomalies. Serial scans were performed every 4 weeks until delivery in order to monitor the shape and size of the cyst. Amniocentesis for fetal karyotyping was performed in two cases because of advanced maternal age. Clinicians were informed of the results of investigations, but no treatment was undertaken as a result.

Information on gestational age at delivery and mode of delivery were collected for all the pregnancies. Neonates underwent neurosonography at birth. Neurological evaluation included observation of general movements⁷ (writhing movements at birth and at the first month, then fidgity movements quarterly up to the 18th month).

RESULTS

In all five cases the midsagittal view allowed visualization of the cyst below the splenium of the corpus callosum and posterior to the cavum septi pellucidi and cavum vergae (Figure 1). Results are summarized in Table 1. The mean diameter was 12.4 (range, 9–20) mm and the cyst size remained stable throughout pregnancy in three cases. It showed a slight increase in size until the end of pregnancy in the other two cases. The finding was isolated in three cases, and associated with bilateral borderline ventriculomegaly in the other two cases, with atrial widths of 11 and 11.6 mm. A single umbilical artery was the only extracranial anomaly and this was detected in the fetus with borderline ventriculomegaly that had a 40-year-old mother. Because of advanced maternal age, fetal karyotyping was performed in these two patients. The fetus with a single umbilical artery had trisomy 21 and the other was normal.

Gestational age at delivery ranged from 36 to 40 weeks. Cesarean section was performed in two cases (Cases 3



Figure 1 Cavum veli interpositi cyst: the midsagittal view allows correct visualization of the cyst (C), which is located below the splenium of the corpus callosum (large arrow); the column of the fornix (small arrow) separates it from the cavum septi pellucidi and the cavum vergae.

and 4) for indications not associated with the prenatal diagnosis (previous Cesarean section, twin pregnancy). Birth weight ranged from 2800 to 3450 g. No neonate needed intensive care.

Duration of neonatal follow-up ranged from 10 to 48 months; neonatal neurosonography confirmed the presence of CVI cyst in all five fetuses. The cyst regressed in one case (Case 4) within 1 month after delivery. In the others it remained stable in size during the follow-up period.

DISCUSSION

Embryologically, the CVI is a true cistern originating from the roof plate of the diencephalon by an extension of the pia mater, which protrudes into the primitive neural tube at about the third month of gestation. The pathophysiological mechanism leading to cystic dilatation is unclear: it could represent a progressive enlargement of the normal cistern or it could be the result of abnormal separation of the crura of the fornices⁸.

This study shows that prenatal diagnosis of CVI cyst is feasible by ultrasound examination of the fetal brain. It appears as a well-defined interhemispheric anechoic

Table 1 Prenatal findings and postnatal outcome of five cases of cavum veli interpositi (CVI) cyst

Case	MA (years)	GA (weeks)	CVI cyst mean diameter (mm)*		Associated anomalies	Fetal karyotype	Confirmed final diagnosis	Follow-up	
			Initial	Final				Age at final check (months)	Outcome
1	33	19	10	16	No	Unknown	Stable	48	Normal
2	20	30	20	20	No	Unknown	Stable	16	Normal
3	40	21	9	12	BV, SUA	47,XY+21	Stable	10	Normal
4†	41	30	9	9	BV	46,XY	Regression in 1 month	18	Normal
5	32	23	14	14	No	Unknown	Stable	12	Normal

*Mean of the anteroposterior, vertical and transverse diameters at first and at final measurement. †Monochorionic diamniotic twin pregnancy with only one fetus affected with CVI cyst. BV, borderline ventriculomegaly; GA, gestational age; MA, maternal age; SUA, single umbilical artery.

lesion which, in the axial view of the brain, is located posterior to the thalami. The midsagittal view allows visualization of the cyst, which is located anteroinferior to the splenium of the corpus callosum, superior and posterior to the thalami; the columns of the fornix form the superolateral walls of the CVI cyst and separate it from the cavum vergae and the cavum septi pellucidi. The internal cerebral veins, directed posteriorly under the splenium of the corpus callosum to join the vein of Galen, are displaced inferiorly and laterally by the CVI cyst. The differential diagnosis includes: dilated cavum septi pellucidi and cavum vergae, and arachnoid cysts of the quadrigeminal cistern. The midsagittal view is useful in the differential diagnosis: dilated cavum septi pellucidi and cavum vergae are superior and anterior to the columns of the fornix, located below the body of the corpus callosum; arachnoid cysts of the quadrigeminal cistern are located below the internal cerebral veins and may compress the tectum of the midbrain.

The clinical relevance of CVI cyst is unclear and there is some discrepancy between results of reported postnatal and prenatal series. Raimondi *et al.*³ analyzed 29 children with CVI cyst and proposed a clinical syndrome including macrocephaly, sunken fontanelle, developmental delay, failure to thrive, seizures and hydrocephalus. Gangemi *et al.*⁴ reported that a 9-year-old boy with psychomotor retardation and epileptic seizures had a large CVI cyst diagnosed by computed tomography and magnetic resonance imaging (MRI). The patient was treated by endoscopic fenestration. The surgery resulted in a decrease in the size of the cyst and a reduction in number and intensity of epileptic seizures. Supprian *et al.*² reported a case of monozygotic twins in which one had a psychotic disorder and a large CVI cyst. On the other hand, Yokota⁹ showed that when the cistern of the velum interpositum is dilated, and therefore radiologically visible, it can be asymptomatic, as in the case of a 71-year-old woman studied for a thalamic hemorrhage who had a true asymptomatic CVI cyst.

With respect to prenatal studies, Hertzberg *et al.*¹⁰ reported the case of one fetus, scanned at 29 weeks of gestation, with a CVI cyst obstructing the foramen of Monro resulting in hydrocephalus which was resolved by a ventriculoperitoneal shunt. Chen *et al.*¹¹ hypothesized that hydrocephalus could result from an arachnoid cyst of the velum interpositum, which sometimes requires surgical fenestration. However, because of its anatomical position, it is very difficult to visualize a CVI cyst associated with hydrocephalus; it is more probable that one will verify only borderline ventriculomegaly because of the partial obstruction to the outflow of the cerebrospinal fluid from the third ventricle⁶. Vergani *et al.*⁶ suggested that, because of its anatomical position, CVI cyst is not associated with compression and damage to the surrounding brain tissue. Over a period of 7 years the authors analyzed seven CVI cysts. Six persisted and only one resolved. Two fetuses showed ventriculomegaly while no extracranial abnormalities were detected. Pediatric follow-up (range, 9–79 months)

showed normal development in all cases. Eisenberg *et al.*¹² and Shah *et al.*⁵ reported two cases with CVI cyst detected prenatally and confirmed postnatally. In both studies, one case showed a relative reduction of the cyst's size during the follow-up with MRI. Their neonatal and infant follow-up (range, 6–48 months) revealed normal growth and development. Our prenatal observations are consistent with those reported by others^{5,6,12}: the CVI cyst may undergo spontaneous resolution after delivery, and the postnatal outcome is good even in cases of persisting cyst.

In conclusion, CVI cyst is an uncommon fetal ultrasound finding, which seems to be benign. However, careful fetal neurosonography is important to rule out associated anomalies which can influence postnatal outcome. When the diagnosis of isolated CVI cyst is certain, no additional prenatal investigations are necessary and an expectant approach, with monthly sonographic examinations until delivery, can be adopted. Due to the reported association of large CVI cysts and adverse pediatric outcome, postnatal sonographic follow-up with serial examinations at delivery and at 6 and 12 months is recommended in order to rule out the rare possibility of postnatal enlargement of the CVI cyst.

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