REVIEW ARTICLE

Interhemispheric Supratentorial Cysts

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Abstract

Interhemispheric supratentorial cystic-like fluid collections may be occasional findings occurring during the routine second trimester ultrasound screening. The correct diagnosis of the origin of the cyst is necessary for an adequate counseling: In most cases, the cyst is a benign finding but sometimes it can be the sign of a serious abnormality with neurological sequelae. They may be located from the anterior to the posterior aspect of the brain in three regions: (1) the area of the anterior complex [cavum septi pellucidi (CSP)]/cavum vergae (CV), frontal horns of the lateral ventricles, genu of the corpus callosum (GCC); (2) the intermediate zone (thalami and third ventricle); (3) the area of the posterior complex [arachnoidal space delimited by the medial walls of the occipital lobes and the splenium of the corpus callosum (CC)]. Cysts of the anterior complex include dilatation of the CSP/CV and agenesis of septum pellucidum (ASP). A cystic structure in the intermediate zone may be due to dilatation of the third ventricle in case of aqueductal stenosis (AS) or agenesis of the corpus callosum (ACC). Interhemispheric cysts located in the area of the posterior complex are arachnoidal cysts originating from the cavum veli interpositi, the quadrigeminal and suprasellar cisterns. In order to define the correct location and consequent clinical significance of the cyst, a complete neurosonogram is needed with the midsagittal view being the most informative plane.

Keywords: Cavum septi pellucidi, Cavum vergae, Corpus callosum, Interhemispheric cyst, Velum interpositum.

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Transventricular, transcerebellar, and transthalamic planes are the axial planes routinely used to screen for supratentorial and infratentorial fetal brain abnormalities and for measure head size.[1] In the transthalamic plane, two physiological hypo-anechoic areas are normally present, located respectively in the area of the so-called anterior and posterior complexes.[2]

The anterior complex includes the rectangular anechoic area of the cavum septi pellucidi (CSP), which is bordered on both sides by the frontal horns of the lateral ventricles, and anteriorly by the genu of the corpus callosum (GCC)[3] (Fig. 1).

The posterior complex is a triangular hypoechoic area with the apex pointing posteriorly to the interhemispheric fissure: It is delimited anteriorly by the splenium of the corpus callosum (CC) and laterally by the medial walls of the occipital lobes (Fig. 1).

In the intermediate zone between the anterior and posterior complexes, the virtual cavity of third ventricle delimited by the thalami can be seen (Fig. 1).

A complete understanding of the anatomical landmarks of the anterior and posterior complexes and intermediate zone may be obtained with the midsagittal view of the brain. This plane shows the complete length of the CC with its components, that from the anterior to the posterior aspect are namely: The rostrum, the genu, the body, and the splenium (Fig. 2). The rostrum and genu delineate anteriorly the CSP; this cavity continues posteriorly in the CV. The body is above the third ventricle; the splenium is above the quadrigeminal cistern.

Interhemispheric Cysts in the Area of the Anterior Complex

The anterior complex includes the rectangular cystic area of the CSP. This is a closed fluid-filled cavity between the two leaves of septum pellucidum that separate the frontal horns of the lateral ventricles. It also borders with the inferior surface of the CC and superior surface of the fornix. It is visualized from around 17 weeks' gestation and disappear upon fusion of the two leaves postnatally.

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Visualization of the CSP is an important marker of normal fetal brain development. Cavum septi pellucidi communicates posteriorly

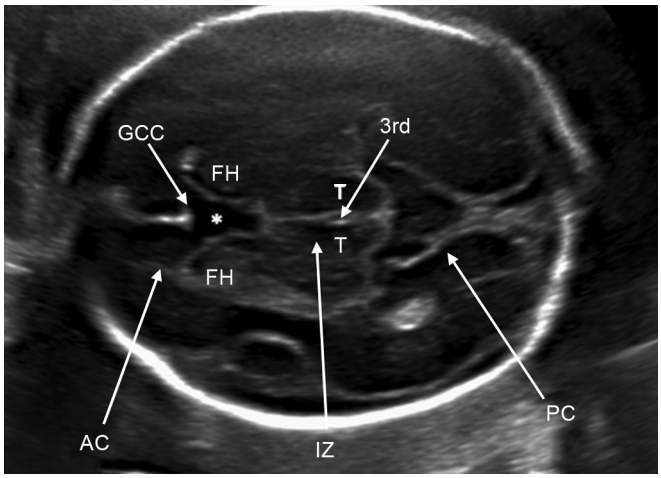


Fig. 1: Transthalamic plane showing the anterior and posterior complexes with intermediate zone. The anterior complex (AC) includes the rectangular anechoic area of the CSP (*), which is bordered on both sides by the frontal horns of the lateral ventricles (FH), and anteriorly by the genu of the corpus callosum (GCC). The posterior complex (PC) is the triangular hypoechoic area with the apex pointing posteriorly. In the intermediate zone between the virtual cavity of third ventricle (3rd) delimited by the thalami (T) can be seen

with the CV extending between the body of CC and fornix to the splenium of the CC.[4]

Abnormal cystic appearance of the anterior complex may be due to (1) dilatation of the CSP/CV; (2) agenesis of the septum pellucidum (ASP) with communication between the frontal horns of the lateral ventricles.

- Dilatation of CSP/CV: The CSP may be larger than normal and may show an abnormal elongation in the anterior–posterior aspect, creating the false impression of a rectangular cyst located between the two normally shaped frontal horns of the lateral ventricles (Fig. 3A). The sagittal view shows the C-shaped cystic structure located below a normal CC (Fig. 3B). In the vast majority of cases, it is a normal variant without any clinical consequence. Some authors report cases of enlarged CSP in fetuses with abnormal karyotype, mainly trisomy 18, but in most cases further brain abnormalities are associated including subtle anomalies in any of the structures that surround CSP/CV and that cannot be recognized by ultrasound.[5] Huge cysts of CSP have been reported in the adults with headache, schizophrenia, and other psychiatric disorders[6,7] but no case of evolution from a simply mild dilatation to a true cyst has been reported.
- Agenesis of the septum pellucidum: Septal agenesis may be associated with various congenital brain malformations, namely septo-optic dysplasia (SOD), holoprosencephaly (HPE), schizencephaly, or agenesis of the corpus callosum (ACC). In cases of isolated ASP, the lateral leaves of the CSP are missing and the CSP directly communicates with the frontal horns of the lateral ventricles creating a crescent-shaped cystic structure (Fig. 4A). The midsagittal and coronal views show the normal development of the CC (Figs 4B and C). Isolated ASP is usually asymptomatic[8]; however, 18% of the fetuses with ASP are affected by SOD, complicated by visual impairment/blindness, hypothalamic-pituitary insufficiency (retarded growth, diabetes insipidus), seizures, and motorial handicap in case of SOD/plus schizencephaly. The differential diagnosis between ASP and SOD may rely on growth hormone (GH) and adrenocortico tropic hormone (ACTH) assays on the fetal blood or the visualization of the optic chiasm; different techniques for visualization and measurement of the optic chiasm have been reported: The first one using a coronal plane,[9] the second one using a tilted axial plane on the orbits,[10] and the third one using an axial plane below the thalamic plane.[11] However, both abnormal hormonal assays and chiasm hypoplasia may develop late in pregnancy or even after delivery. For this reason, the prenatal diagnosis of SOD associated with ASP is still a diagnostic challenge and a counseling dilemma.

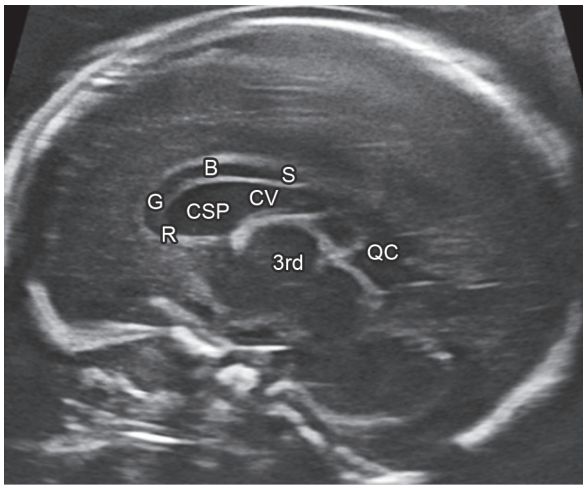
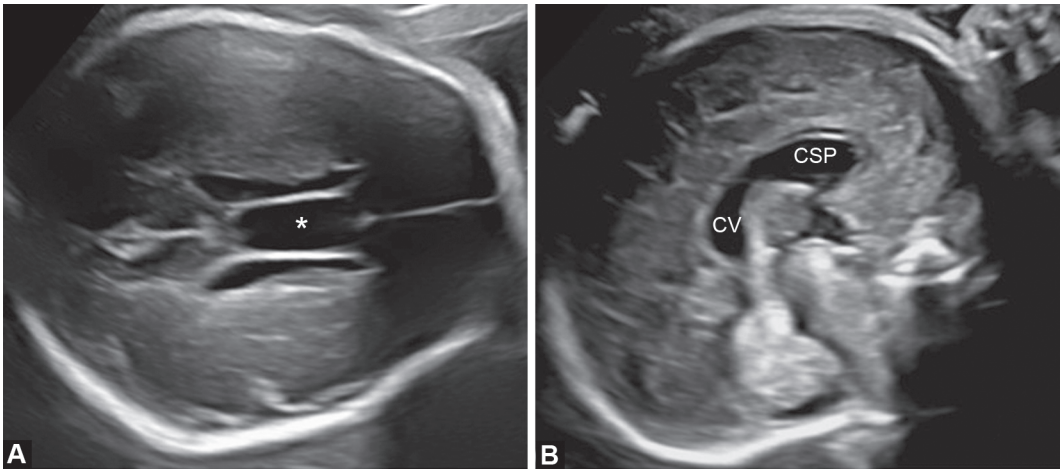
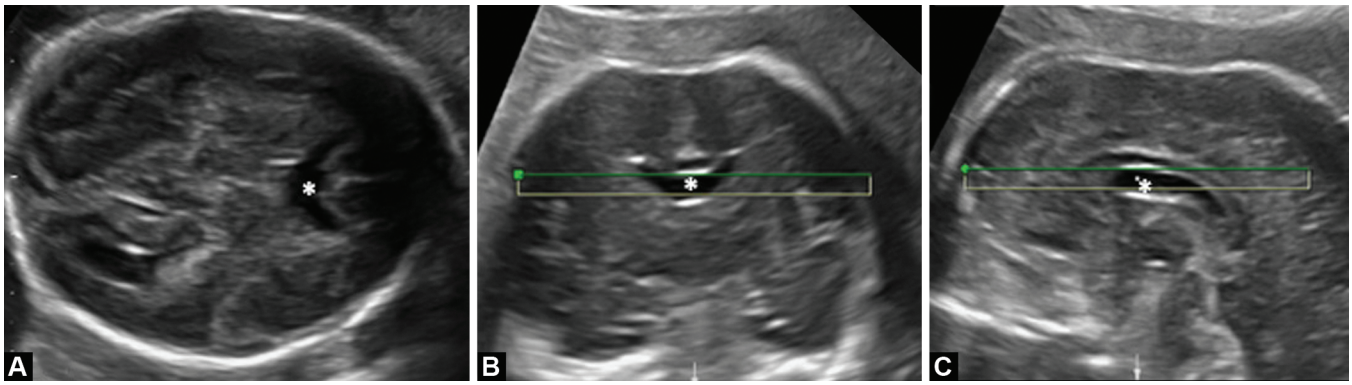


Fig. 2: Midsagittal view of the brain showing the complete length of the CC with its components that from the anterior to the posterior aspect are namely: The rostrum (R), the genu (G), the body (B), and the splenium (S). The rostrum and genu delineate anteriorly the cavum septi pellucidi (CSP); this cavity continues posteriorly in the cavum vergae (CV). The body is above the third ventricle (3rd); the splenium is above the quadrigeminal cistern (QC)

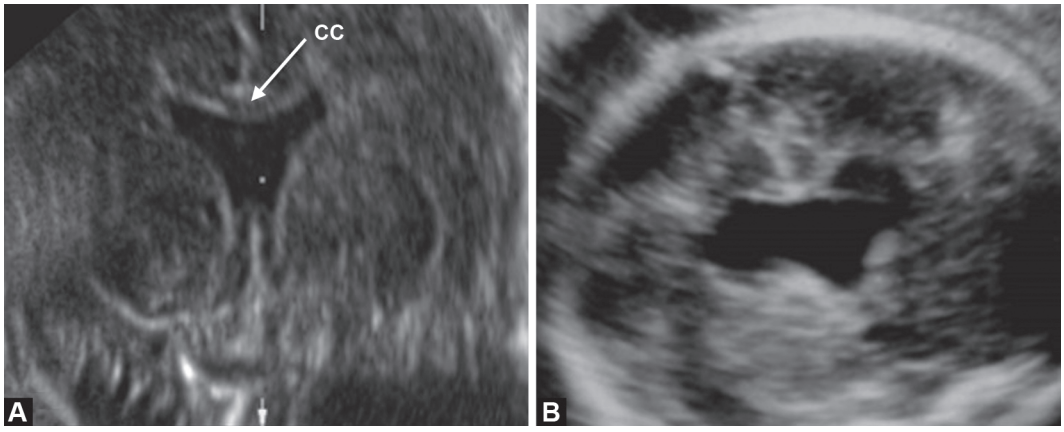
Agenesis of septum pellucidum/SOD should also be differentiated from lobar HPE. In coronal section on the anterior complex, ASP/SOD is characterized by the presence of the CC above the fused frontal horns; in lobar HPE, the fused frontal horns have a typical squared shape and the CC above them is absent or hypoplastic[12] (Fig. 5).



Figs 3A and B: Enlarged cavum septi pellucidi/cavum vergae. (A) The axial view shows a rectangular cyst (*) located between the two lateral ventricles; (B) The midsagittal view shows the C-shaped cystic structure generated by dilated cavum septi pellucidi (CSP) and cavum vergae (CV) located below a normal corpus callosum



Figs 4A to C: Isolated agenesis of septum pellucidum. The lateral leaves of the CSP (*) are missing and the CSP directly communicates with the frontal horns of the lateral ventricles creating a crescent-shaped cystic structure (A). The midsagittal (B) and coronal (C) view show the normal development of the corpus callosum



Figs 5A and B: Coronal views on the anterior complex in case of agenesis of septum pellucidum (A) and lobar holoprosencephaly (B). The CC (arrow) is normally present above the fused frontal horns in agenesis of septum pellucidum; in lobar holoprosencephaly the fused frontal horns have a typical squared shape and the CC above them is absent

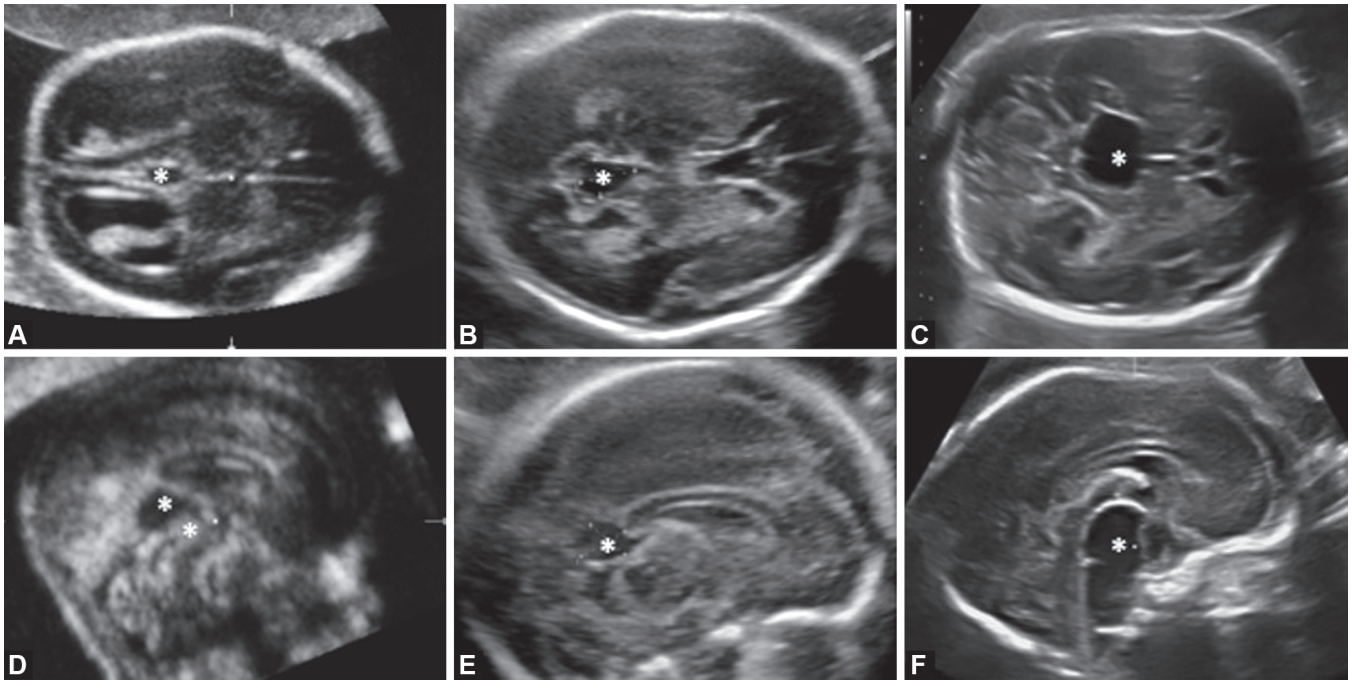
Interhemispheric Cysts in the Area of the Posterior Complex

The posterior complex is the triangular cystic area delimited anteriorly by the splenium of the CC, laterally by the medial walls of the occipital lobes, pointing posteriorly to the interhemispheric fissure. Interhemispheric cysts located in the area of the posterior complex may originate from different regions of the arachnoidal spaces at the base of the skull: Cavum velum interpositum, quadrigeminal cistern, and suprasellar cistern.[13] In the axial plane, the sonographic appearance is the same, but a correct differential diagnosis can be achieved by the midsagittal view (Fig. 6).

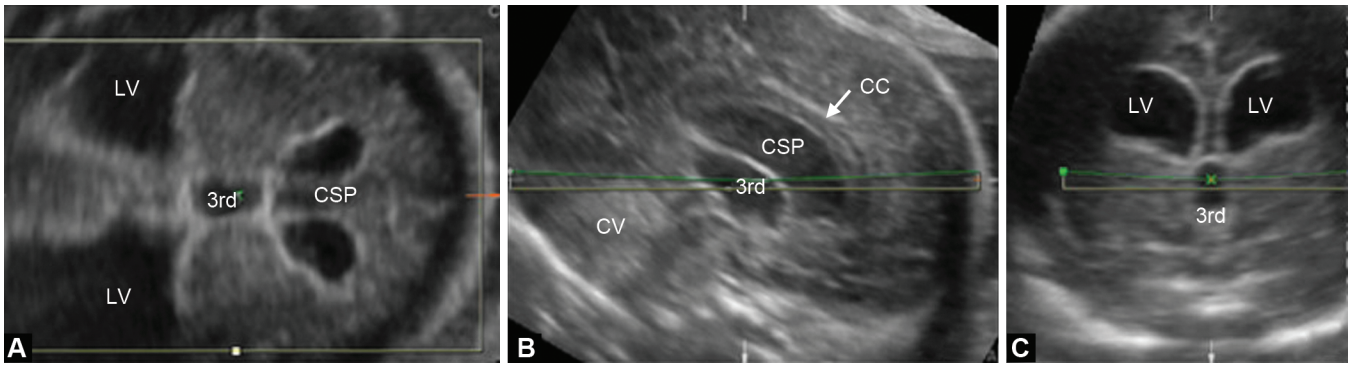
- Cavum veli interpositi (CVI) cyst is located below the splenium of the CC. It originates from a cystic dilatation of the CVI which is a virtual space within the double-layered tela choroidea of the third ventricle. It is situated in anteroinferior part of the splenium of the CC; the column of the fornix separates it from the CSP and the CV. Occasionally, this space is fluid-filled and sonographically visible as an interhemispheric anechoic cyst.[14,15]
- Quadrigeminal cyst is located posterior to the thalami in the area of the lamina quadrigemina.[16,17]
- Suprasellar cyst is located between the clivus and the brainstem.[18]

Arachnoid cysts have thin walls, do not communicate with the ventricular cavities, may cause compression but not disruption of the cerebral tissues, and may cause ventriculomegaly independently from the cyst size. The prognosis mainly depends on the presence of associated anomalies. In most of isolated cases, the neurological outcome is normal. In a meta-analysis of 10 studies involving 47 fetuses, arachnoid cysts had associated central nervous system (CNS) and extra-CNS anomalies in 73% and 14% of the cases, respectively. The most common associated anomalies were ventriculomegaly and callosal abnormalities. Chromosomal abnormalities were present in 6%, but fetuses with isolated cysts were always euploid. Cavum veli interpositi cysts had associated CNS and extra-CNS anomalies in 31% and 6%, respectively. No chromosomal or callosal anomalies were found in these cases. Intrauterine regression occurred in 23% of CVI cysts and in none of the arachnoid cysts. In children with arachnoid cyst, the occurrence of ventriculomegaly and mass effect on the adjacent structures were observed in 23.9% and 26.8%, respectively. None of the cases included had abnormal motor outcome or intelligence. The rate of surgery was 34.7%. None of the children with a prenatal diagnosis of isolated CVI cyst experienced any of the adverse outcomes explored in this review.[19]

In another recent review of 123 cases from the literature, 68% of patients had a normal outcome, 63% underwent surgical intervention which was not associated with abnormal outcome.



Figs 6A to C: Cysts (*) located in the area of the posterior complex visualized in both axial and midsagittal view. (A to D) Cavum veli interpositi cyst is located below the splenium of the corpus callosum; (B to E) Quadrigeminal cyst is located posterior to the thalami in the area of the lamina quadrigemina; (C to F) Suprasellar cyst is located between the clivus and the brainstem



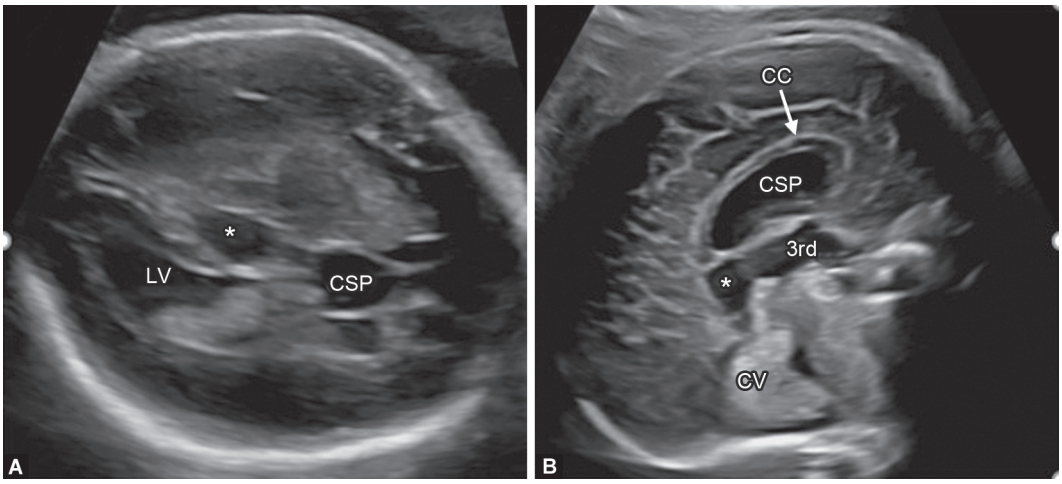
Figs 7A to C: Severe ventriculomegaly in case of aqueductal stenosis in axial (A), midsagittal (B), and coronal (C) views. The dilated third ventricle (3rd) appears as a cystic area in the intermediate zone between the dilated lateral ventricles (LV). In the midsagittal view, the "cystic" third ventricle is seen below the cavum septi pellucidi (CSP) and corpus callosum (CC)

The presence of other intracranial anatomical abnormalities were significant predictors of abnormal outcome.[20]

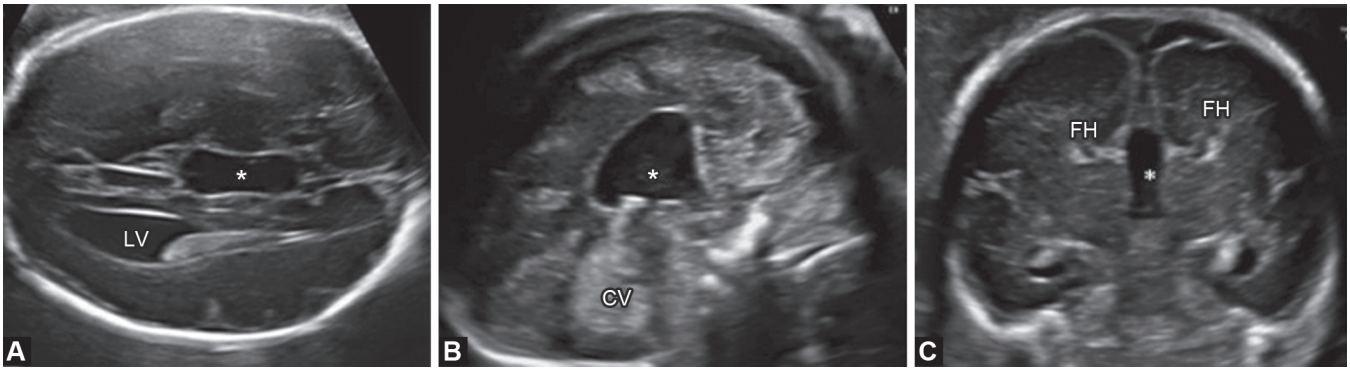
Interhemispheric Cysts in the Intermediate Zone

On the transthalamic plane, the intermediate zone is the area located between the anterior and posterior complexes. Normally in this area, there are no cystic structures; the third ventricle appears as a linear echo between the thalami. A cystic lesion in this area represents dilatation of the third ventricle, which can be due to aqueductal stenosis (AS) or ACC.

- Aqueductal stenosis is the most common cause of prenatal obstructive ventriculomegaly.[21–23] The etiology is multifactorial, including both genetic and acquired forms. Of the genetic causes, chromosomal abnormalities, X-linked and autosomal recessive disorders, copy number variant, and several single-gene disorders have been described.[24] Acquired causes can be intrinsic (obstruction of the aqueduct lumen) or extrinsic (external compression). Prenatally, acquired causes are most commonly intrinsic, resulting from infection (aqueduct gliosis) or intraventricular hemorrhage. The dilatation of the third ventricle is usually a late event appearing after the dilatation of the lateral ventricles.[25] For this reason, in the same axial view where the dilated third ventricle is seen also the dilated lateral ventricles are visible. In the midsagittal view, the "cystic" third ventricle is seen below the CSP and CC (Fig. 7). However, in some cases, the dilatation of the suprapineal recess of the third ventricle may be an early clue for obstructive ventriculomegaly downstream of the third ventricle.[26] It appears as a cystic structure in the intermediate zone in the presence of mild ventriculomegaly (Fig. 8).



Figs 8A and B: Dilatation of the suprapineal recess (*) of the third ventricle in a case of aqueductal stenosis (A = axial view; B = midsagittal view). It appears as a cystic structure in the intermediate zone in the presence of mild ventriculomegaly (3rd = third ventricle; LV = lateral ventricle; CSP = cavum septi pellucidi; CC = corpus callosum)



Figs 9A to C: Interhemispheric cyst (*) associated with ACC. The axial view (A) shows an interhemispheric cyst in the intermediate zone. The midsagittal (B) and coronal (C) views show the dilated third ventricle protruding upward as an interhemispheric cyst. The corpus callosum is absent (LV = lateral ventricles; FH = frontal horns; CV = cerebellar vermis)

- Agenesis of the corpus callosum is one of the most common causes of non-obstructive ventriculomegaly. In some cases of ACC, the third ventricle may be dilated and protrudes upward as an interhemispheric cyst. Three types of cysts have been described:[27] Type a associated with communicating ventriculomegaly but no other cerebral malformation; type b associated with ventriculomegaly secondary to diencephalic malformations (fused thalami) that prohibits the egress of cerebrospinal fluid from the third ventricle into the aqueduct; and type c associated with a small head size and apparent cerebral hemispheric dysplasia or hypoplasia. The differential diagnosis with AS is based on the absence of the CC above the dilated third ventricle and protrusion of the cyst between the two "bull's head" shaped frontal horns in the coronal view of the anterior complex (Fig. 9).

Conclusion

Interhemispheric supratentorial fluid collections may be related to enlargement of physiological median structures (CSP/CV, CVI) or may be related to the presence of true cysts, such as arachnoid cysts, or of cystic-like lesions, such as ASP/SOD, lobar HPE, dilatation of the third ventricle as a consequence of AS or ACC. Even though the interhemispheric location of both paraphysiological and true pathological conditions is similar, their clinical significance and postnatal outcome are quite different: the former are almost constantly normal variants without clinical relevance, whereas the latter are pathological processes that may carry a poor prognosis. Interhemispheric cyst-like lesions related to physiological structures can be prenatally distinguished from pathological fluid collections on the basis of location, cyst size, change in size with time, and absence of associated anomalies. Ultrasonography can be used to perform prenatal differential diagnosis of midline interhemispheric cystic lesions with the midsagittal view being the most informative plane to obtain the correct diagnosis in order to dispensing the proper counseling to the parents.

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