Transfontanelar US: Intracranial Cyst Formations

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Resumen

Introducción. La ecografía transfontanelar es la técnica más usada para evaluar las estructuras intracraneales neonatales, con alta sensibilidad y especificidad para el diagnóstico de lesiones morfológicas.

Objetivo. Mostrar algunos ejemplos de las formaciones quísticas intracraneales en los recién nacidos, incluyendo: Variantes normales, Desarrollo de lesiones quísticas como consecuencia de daño perinatal, Estructuras vasculares que semejan lesiones quísticas y Malformaciones.

Revisión del tema. La ecografía transfontanelar es la técnica de elección para la evaluación intracraneal en recién nacidos y lactantes hasta el cierre de las fontanelas, por la ausencia de radiación ionizante portabilidad, el diagnóstico con bajo costo y en tiempo real. La nueva tecnología y el uso de nuevas ventanas eco-

Abstract

Introduction. Transfontanelar ultrasound is the most frequent technique used to evaluate intracranial structures in neonates due to its high sensitivity and specificity for the diagnosis of morphological lesions.

Objective. To describe some examples of intracranial cyst formations in neonates, including normal variants, development of cystic lesions because of perinatal brain damage, vascular structures resembling cystic lesions and congenital disorders.

Current Importance. Transfontanelar ultrasound is the technique of choice when performing an intracranial evaluation in neonates and breastfed infants up to the closure of the fontanelles, because it is a portable, low-cost, real time procedure that involves no ionizing radiation.

The new technology and the use of new ultrasound win-
transfontanelar ultrasound is the most readily available and repeatable method to obtain images of the brain in neonates. Unlike other neuroimaging tools such as MRI and CT, transfontanelar US can be performed in the infant’s crib with little disturbance.

Transfontanelar US diagnostic sensitivity and specificity have increased with the recognition of more subtle patterns of lesions and the appearance of characteristics suggesting development, metabolic and infectious disorders (1).

US can offer important information about anatomical location, size and shape of lesions, as well as their impact over adjacent structures.

Our purpose is to describe some examples of intracranial cyst formations in neonates, including normal variants, development of cystic lesions because of perinatal brain damage, vascular structures resembling cystic lesions and congenital disorders.

**Materials and method**

This study is based on routine transfontanelar ultrasound exams of premature and full-term infants performed in our neonatal intensive care unit.

**Transfontanelar ultrasound technique**

Cranial ultrasound (CU) is an excellent non-invasive tool to obtain brain images during the neonatal period.

Its quality and diagnostic accuracy depend upon several factors: good ultrasound equipment, use of appropriate transducers, use of adequate exploration protocols, use of several acoustic windows, and the examiner’s experience who needs to be aware of neonate patients’ needs, of the normal anatomy in US, of brain maturation and of frequent anomalies of the neonate brain (1, 3).

**Acoustic windows**

Although traditionally ultrasound is performed through the anterior fontanelle, the use of accessory acoustic windows can improve the quality of the image and diagnostic accuracy. The standard acoustic window used to obtain neonate brain images is the anterior fontanelle. However, the cerebellum, brainstem and subcortical white matter are not well visualized through this window. Exploration through the posterior and mastoid fontanelle can help detect lesions in these areas. The temporal window allows...
a good visualization of the mesencephalon and the brainstem (1) (Figure 1).

It is ideal to have a real time, 2D high resolution ultrasound equipment with a special setting for the neonatal brain and a multifrequency transducer or a lineal and convex transducer of diverse frequencies (5, 7.5 and 10 MHz). The transducer has to be small enough to fit the anterior fontanelle of the premature or full-term newborn.

**Intracranial cystic lesions**

Initially, cystic lesions identified with transcranial ultrasound can seem to have a non-specific appearance. However, it is possible to reach a more accurate diagnosis paying attention to the anatomical location of the cyst and its features (6, 7).

Among the lesions identified, we can mention (a) normal variants, (b) development of cystic lesions because of perinatal brain damage, (c) vascular structures resembling cystic lesions, and (d) congenital disorders (7).

**Normal variants**

**Cavum Septi Pellucidum (CSP)**

The Septum Pellucidum consists of two thin membranes of white matter surrounded by gray matter with a virtual space between them (7). It is located between the anterior horns of the lateral ventricles, forming their medial walls, and extending in a ventral position to the foramen of Monro, from the corpus callosum to the Fornix and anteroposteriorly from the genu to the splenium of the corpus callosum (8, 9).

During intrauterine life, these sheets are separated and start to fusion towards the final stages of gestation (7,8). Some authors agree that closure can be completed in two to six months after birth (5).

The cave of septum pellucidum originates due to the lack of the two laminae fusion, resulting in the preservation of CSP into adulthood. It is a normal variant since it appears in 100% of premature infants and in 85% of full-term infants (8, 9) and in up to 10-15% of adults (9).

It communicates with the cavum vergae, but not with ventricles or with the subarachnoid space. (5). CSP is defined when it is greater or equal to 1 cm and it produces lateral ballooning of the Septum Pellucidum (9) (Figure 2).

**Cavum Vergae**

It is a space full of fluid between the two sheets of the septum pellucidum, located posteriorly to an arbitrary vertical plane constituted by the columns of the fornix up to the splenium (7, 9) (Figure 3).

During the sixth month of gestation, the cavum vergae starts closing from its dorsal position to a ventral position, and the closure is complete generally nearly birth, unlike septum pellucidum, which begins closure at a later stage (5, 8).

It is only visualized in 30% of newborns and in less than 1% of adults (8, 9) and generally together with cavum septum pellucidum. Isolated cavum vergae has been documented exceptionally, which suggests that embryonic events are not always produced in a habitual sequence (7).

**Cavum Veli Interpositi**

Velum Interpositum is an invagination of the pia mater located below the fornices and above the third ventricle and thalamus, creating a cistern that contains the middle cerebral veins and the medial posterior choroidal artery. Anteriorly, it reaches the foramen of Monro and it communicates posteriorly with the quadrideminal cistern and the lateral side of the ambiens cistern. It communicates laterally with the choroidal plexus of the lateral ventricles (7, 9).

The cave and cyst of the velum interpositum produce the progressive increase of the normal size of the cistern (9).

The etiology is unknown and it is not usually associated with other anomalies (8). It is a common finding in infants younger than 18 months and rare in adults (8, 9).

The cavum velum interpositum is separated from cavum vergae by the columns of the fornices (5, 7) (Figure 4).

According to the position, intracranial cystic alterations can be classified into (a) cystic lesions in the posterior fossa, (b) supratentorial cystic lesions in a periventricular location, and (c) non-periventricular supratentorial cystic lesions of intra or extraaxial location (7) (Table 1).
Intracranial Cyst Formations

Figure 1.

Figure 2.
A) Diagram with sagittal and coronal views of the cave of septum pellucidum. (RadioGraphics 2006; 26: 173-196, modified). B) Sagittal view ultrasound at the level of the midline, where CSP is identified as a structure full of fluid extending anteroposteriorly from the genu to the splenium of the corpus callosum. C) Coronal view ultrasound at the level of the frontal horns of the lateral ventricles, where the arrowhead signals a triangular anechoic structure corresponding to CSP. CSP: Cavum Septum Pellucidum.
Figure 3.
A) Diagram with sagittal and coronal views of the cave of the septum vergae. (RadioGraphics 2006; 26: 173-196, modified). B) Sagittal view ultrasound at the level of the midline where CV is identified. C) Coronal view ultrasound at the level of lateral ventricles, where arrows signal a structure filled with fluid corresponding to CV. CV: Cavum Vergae.

Figure 4.
A) Diagram with sagittal and coronal views of the Cavum Veli Interpositi. (RadioGraphics 2006; 26: 173-196, modified). B) Coronal view ultrasound at the level of the lateral ventricles, where CVI is signaled between them. C) Sagittal view ultrasound at the level of the midline showing the posterior location below the fornix of CVI.

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<th>Position</th>
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<td>ST cystic lesions</td>
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<td>ST cystic lesions</td>
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Table 1. Classification of cystic formations according to their position.
ST: supratentorial.
A- Cystic lesions of posterior fossa

Mega Cisterna Magna
Cisterna Magna is an arachnoid structure filled with fluid that is in a caudal position to the vermis and that communicates with the fourth ventricle (5,10). In both sagittal and axial views, it is smaller than 8 mm (it ranges from 3-8 mm) (7).

Mega Cisterna Magna is an enlarged retrocerebellar CSF space measuring more than 8 mm in diameter, with normal vermis and hemispheres. A lineal echo-genic area can be identified in the base of the posterior fossa, consisting of dural folds and probably of the inferior insertion of the tentorium (7) (Figure 5).

It is the most frequent lesion of the posterior fossa (6). It is found in 1% of all postnatal cerebral images (7, 10).

Some authors believe that it is secondary to cerebellum damage and loss of volume (7, 10). It has been associated to infarcts, infections (cytomegalovirus) and with chromosome anomalies (trisomy 18) (7).

Dandy–Walker syndrome
It is an association of combined anomaly of the roof of the fourth ventricle and dysgenesis of the cerebellar vermis, with enlargement of the fourth ventricle and cerebellar hypoplasia (5, 7).

The “classic” syndrome includes three criteria: vermis hypoplasia, cystic dilation of the posterior fossa with communication with the fourth ventricle, and enlarged posterior fossa with abnormal elevation of the tentorium and torcular herophili (7). Hydrocephalus occurs in about 80% of the cases (Figure 6).

Dandy–Walker is a less severe variant, characterized by a posterior fossa of a normal size or slightly enlarged. Hydrocephalus is not habitual.

The prevalence is 1 in 35000 babies born alive, without preference for sex (7, 10).

The most important factor for the prognosis is determined by associated alterations of the nervous system, present in 70% of the cases.

Characteristic ultrasound findings of Dandy–Walker syndrome include a big cyst in the posterior fossa due to the enlargement of the fourth ventricle, a small or absent vermis, small cerebellar hemispheres superiorly displaced and superior elevation of the tentorium. The third ventricle and lateral ventricles can be dilated in different degrees. Ultrasound findings in the Dandy–Walker variant are similar to malformations but less severe (5) (Figure 7).

Arachnoid Cyst

These are congenital intra-arachnoidal lesions expanding with CSF secretion and without ventricular communication (11).

They tend to be unilocular and expansive shaped by adjacent structures (Figure 8).

They represent 1% of intracranial masses. Incidence is slightly greater in men (11).

Most of them are supratentorial, 50% are located in the middle cranial fossa (11), 20% in the retrocerebellar posterior fossa, and less frequently in the fourth ventricle or at the ponto-cerebellar cistern.

Arachnoid cysts in the posterior fossa are associated with the Aicardi syndrome (7).

In ultrasound, they appear as CSF spaces between the cerebellum and occipital bones, the cerebral falx is normal, and the position of the torcular herophili is normal, although it can be elevated if the cyst develops early in the fetal period. Sometimes, there is compression or absence of the inferior vermis (7, 10, 11).

Vein of Galen Aneurysmal Malformation
It is a venous ectasia secondary to an abnormal arteriovenous connection, either directly into the vein of Galen or into an affluent. It is located in the midline, in the quadrigeminal cistern plate.

It is the result of an arterio-venous fistula between the primitive choroidal vessels and the median prosencephalic vein of Markowski. The persistent flow through the connection impedes the expected involution of this umbilical vein and the development of the vein of Galen (7).

It can be associated to other venous anomalies (abnormal dural sinuses and sinus stenosis).

It represents 1% of all vascular cerebral malformations (7, 8).

Clinically it is manifested with congestive heart failure or hydrocephalus by compression of the aqueduct or of the third ventricle (Figure 9).
**Figure 5.**
Sagittal view ultrasound at midline level. Mega Cisterna Magna is seen as an anechoic space filled with fluid (arrows) below the cerebellum.

**Figure 6.**
A) Sagittal view ultrasound at the level of the midline showing retrocerebellar cerebrospinal fluid. B) Coronal view ultrasound showing vermis hypoplasia and communication between the posterior cyst and the fourth ventricle.
Figure 7.
A) Sagittal view at the midline level showing cystic dilation of the posterior fossa. B) Coronal view ultrasound at the level of lateral ventricles showing a parallelism between them. C) Coronal view ultrasound signaling cerebellar vermis hypoplasia and the cyst of the posterior fossa.

Figure 8.
A) Parasagittal view ultrasound and B) Coronal view ultrasound of retrocerebellar arachnoid cyst.

Figure 9. Vein of Galen malformation.
A) Parasagittal view. B) Coronal view. C) Coronal view with color Doppler showing turbulent flow in the aneurysmal and dilated vein.
B- Periventricular location of supratentorial cystic lesions

Connatal Cysts

They are cystic areas adjacent to the superolateral margins of the frontal horns and the body of the lateral ventricle and are mainly anterior to the foramen of Monro (6).

They were considered as sequels of ischemic lesions. Nowadays, many cases represent a normal variant due to approximation towards the walls of the frontal horns of the lateral ventricles. When the ventricular walls are close enough, the most external part of the ventricle takes a round configuration resembling a cyst in ultrasound (7).

They are rare lesions, present in 0.7-1% of premature infants with low weight and they spontaneously disappear within 2-3 months without sequels (7, 12) (Figure 10).

Subependymal Cysts

They are located below the external and posterior angle of the foramen of Monro.

They are delimited lesions, measuring from 2-11 mm in diameter (Figure 11).

We can classify them in two groups: congenital related to germinolysis, and acquired secondary to brain hemorrhage.

They have been reported in association to congenital viral infections (cytomegalovirus and rubella), metabolic disorders (Zellweger syndrome), chromosome anomalies, and cocaine use during pregnancy. However, they can be isolated cases found in healthy newborns (7).

Post-hemorrhage cysts are commonly detected in the caudothalamic groove, where the germinal matrix is present towards the end of gestation (33-35 weeks) and that is why finding them is frequent in premature infants (7).

Choroidal Plexus Cysts

They are located in the body of the choroidal plexus and can eventually protrude towards the ventricular cavity (7).

They are isolated in 1% of all pregnancies and they tend to disappear after 26-28 weeks of gestation.

They are associated to aneuploidy, especially trisomy 18, particularly if the cysts are bigger than 1 cm and bilateral.

Some authors agree that there is a connection between choroidal plexus cysts and heart alterations and hydronephrosis (13).

They do not have clinical importance when they are detected after birth.

In ultrasound, they are rounded with variable size and they can be multiple with a double wall (7) (Figure 12).

Periventricular Leukomalacia

It is white-matter necrosis with a characteristic distribution pattern (dorsal and lateral to external angles of lateral ventricles) (6).

They are frequent in premature infants who have had less than 32 weeks of gestation, since white matter is slightly vascularized and has oligodendrocytes progenitors that are sensitive to consequences of ischemia and infection (7).

Pathogen is still controversial; it is thought to be the result of an ischaemia-reperfusion injury of the white matter.

An ultrasound exam shows hyperechoic periventricular areas, more frequently anterior and lateral to the frontal and peritrigonal horns (5, 7).

Patients who are more affected can develop cysts in these areas, known as cystic PVL. These cysts are bilateral, bigger than 10 mm and are located mainly in the parietooccipital region. They are highly predictive of the development of cerebral palsy and of bad prognosis (Figure 13).

Porencephaly

It is a cortical or subcortical, uni or bilateral intraparenchymatous cystic cavity limited by white matter, which may or may not have ventricular communication, appearing as a dilated ventricle (11).

It is the result of parenchymal destruction (hemorrhage, infection, surgery) and it involves the posterior replacement of the destroyed area with CSF (7, 11).

The prevalence is 2.5% of infants with antecedents of perinatal brain damage.

In ultrasound, it is seen echogenic after initial hemorrhagic event. As clot, retraction takes place, it is replaced by an echogenic area with anechoic center and when retraction is completed, it becomes cystic and anechoic. The walls of these cavities rarely show
discreet calcifications (6, 7) (Figure 14).

Hydranencephaly is considered an extreme example of porencephaly, with little or no brain tissue.

**Figure 10.**

**Figure 11.**
A) Coronal view ultrasound showing a left subependymal cyst. B) Parasagittal view showing a well-defined rounded cyst at the level of the caudothalamic groove.
**Figure 12.**
A) Coronal view ultrasound at the level of lateral ventricles and B) Parasagittal view at the level of the caudothalamic groove showing a left choroidal plexus cyst.

**Figure 13.**
A) Posterior coronal view ultrasound showing small cystic areas adjacent to frontal and occipital horns of ventricles (arrowheads). B) Sagittal view in midline showing frontal small cystic areas and other more superficial and bigger frontal and parietal cystic areas (arrowhead).
**C- Nonperiventricular supratentorial cystic lesions**

**Schizencephaly**

It is an anomaly of the neuronal migration characterized by a cleft surrounded by gray matter heterotopia, extending from lateral ventricle ependymal tissue to the pia matter. There is an abnormal pattern of adjacent gyrus of the cleft.

Schizencephaly can be unilateral or bilateral and it is typically located in the topography of the middle cerebral artery.

The etiology is still controversial; it can be the result of a genetic mutation or to be secondary to immature brain damage before neuronal migration, unlike porencephaly in which there is mature brain damage (Figure 15).

**Holoprosencephaly**

Wide range of anomalies of the intracranial development and of the middle region of the face resulting from a failure in diverticulization of the embryonic prosencephalon occurring between the fourth and eighth week of gestation.

The result of these anomalies is the diverse level of fusion of olfactory tracts, optic tracts and brain hemispheres.

The etiology includes genetic and environment factors.

The incidence is around 1-1.4 in 10,000 births.

There are three types: Alobar, semilobar and lobar, with different levels of structure separation.

The most severe type is alobar holoprosencephaly and is characterized by the absence of cerebral falx, third ventricle and corpus callosum; thalami are fused and there is a large dorsal cyst.

The difference between alobar holoprosencephaly and severe hydranencephaly lies in determining if the thalami are fused or not and to determine the presence or absence of the third ventricle (5, 7, 8) (Figure 16).

**Conclusion**

Transfontanellar ultrasound is the technique of choice when performing an intracranial evaluation in neonates and breastfed infants up to the closure of the fontanelles, because it is a portable, low-cost, real time procedure without ionizing radiation. Improvements in technology and the use of new ultrasound accessory windows, such as posterior and mastoid fontanelles, permit a better evaluation of intracranial structures. Transfontanellar ultrasound is an important method for the study of intracranial structures in premature and full-term infants that helps making a differential diagnosis of intracranial cyst formations in neonates.

**Figure 14.**

Oblique anterior coronal view ultrasound showing a cystic image at the anterior frontal level with irregular edges corresponding to porencephaly.
**Figure 15.**  
A) Coronal view at the level of lateral ventricles and B) Anterior coronal view showing a cleft filled with fluid in left hemisphere, corresponding to unilateral schizencephaly of open lip cleft.

**Figure 16. Alobar holoprosencephaly.**  
A) Coronal view showing only one ventricle in the midline, fusioned thalami, thin layer covering the brain, no separation of the brain hemispheres and absence of cerebral falx. B) Sagittal view at the level of the midline showing a dilated ventricle and posterior cyst.
Bibliography


