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CASE REPORT

SEVERE BILATERAL VENTRICULOMEGALY DIAGNOSED IN A THIRD TRIMESTER FETUS: CASE REPORT AND LITERATURE REVIEW

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ABSTRACT

Fetal ventriculomegaly (VM) is defined as an increase in the diameters of the lateral ventricles greater than 10 mm on a prenatal ultrasound. It has an incidence of 0.3 to 1.5 per 1000 births. The ultrasonographic finding generally occurs during the examination in the second trimester, associated with malformations of the central nervous system (CNS), disruptive events or genetic syndromes. Classification in 1 or 2 ways: mild (10-15 mm) or severe (>15 mm), or mild (10-12 mm), moderate (13-15 mm) or severe (>15 mm). A 26-year-old patient, with a preterm pregnancy, poor prenatal control, was admitted with preterm labor. Stable vital signs, single fetus, alive, obstetric ultrasound with report of severe bilateral VM. It was decided to begin a protocol for resolving the pregnancy via an emergency abdominal route, a male newborn was obtained in cardiorespiratory arrest, neonatal resuscitation maneuvers were not provided. This finding is only a preliminary step to take during the diagnostic approach to recognize the cause of ventricular dilation. When no cause is found, it is defined as "isolated", representing, by definition, provisional discrimination of exclusion.

Key words: Perinatology; Hydrocephalus; Chromosome Aberrations (Source: MeSH NLM)

INTRODUCTION

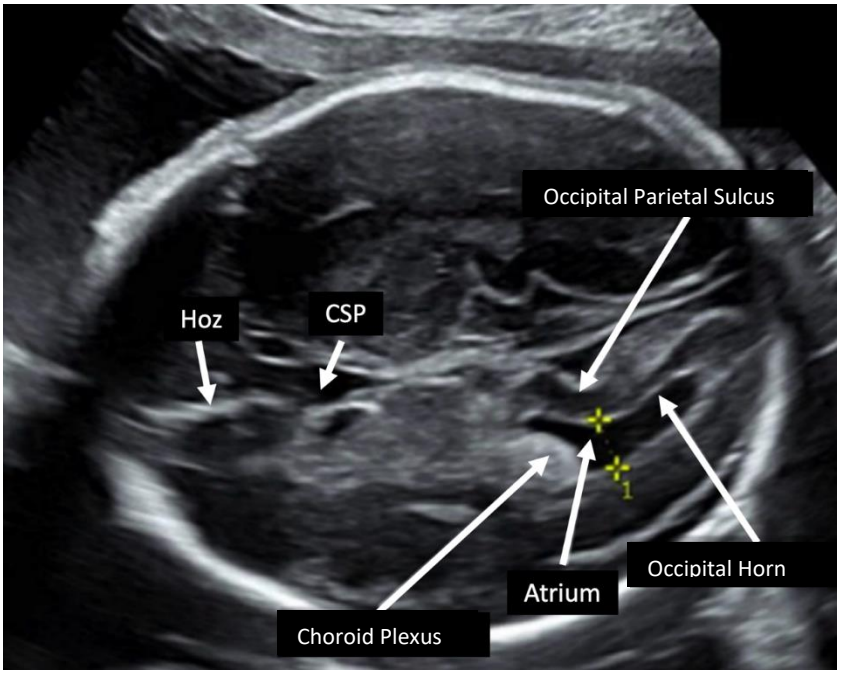
Fetal ventriculomegaly (MV) is defined as an atrial atrial atrial diameter greater than 10 mm on prenatal ultrasound (1). Its incidence ranges from 0.3 to 1.5 per 1,000 births (2). It is not a diagnosis, but an ultrasonic finding, which usually occurs during the examination of fetal structures in the second trimester (3).

It is characterized by dilation of the fetal cerebral lateral ventricles. It is typically classified in 1 or 2 ways: as mild (10 - 15 mm) or severe (>15 mm), or as mild (10 - 12 mm), moderate (13 - 15 mm) or severe (>15 mm) (4). The atrium of the lateral ventricle is the part where the body, posterior horn, and temporal horn converge (**Figure 1**); It has been established that the atrial diameter remains stable between 15 and 40 weeks of gestation, the normal mean diameter of the lateral ventricle ranges between 5.4 and 6 mm, so a measurement of 10 mm represents between 2.5 and 4 standard deviations above the mean (5). Unilateral MV is present in approximately 50 to 60% of cases, and bilateral ventriculomegaly occurs in approximately 40 to 50% (6).

When ventriculomegaly is identified, a thorough evaluation should be performed including a detailed ultrasound evaluation of fetal anatomy, amniocentesis for evaluation of chromosomal abnormalities, as well as genetic testing and PCR examination for fetal infection (7).

A search of the Medline database via PubMed was performed using the following terms: "ventriculomegaly", "dilated cerebral ventricles", "fetal magnetic resonance imaging", "hydrocephalus", "chromosomal abnormalities". The search was limited by the following filters: "Case Reports", "Review", "Systematic Reviews", and "Books and Documents", "Spanish and English", from 1972 to 2021.

A total of 3,847 studies were found, editorials, reviews, duplicate articles were excluded, and 42 studies were selected that included pregnant women diagnosed with ventriculomegaly.



CSP: Cavum del Septum Pellucidum

Figure 1. Correct measurement of the cerebral lateral ventricle

A 26-year-old patient from a municipality in the state of Michoacán, Mexico. Low socioeconomic stratum, dedicated to household chores, lives in a rural type house that has basic services, with an incomplete vaccination schedule for age and sex, does not perform physical activity, positive zoonoses, denied recent infections or during pregnancy, only urinary tract in unknown treatment.

As a relevant obstetric and gynecologic history; Geestas five, cesarean sections two, births one, abortions one. Within the history of prenatal control with onset in the second trimester (22 weeks), she made only 2 prenatal control consultations and 2 ultrasounds in a private clinic, however, she was not given any written report, she did not receive her own vaccination schedule for a pregnant woman, with multiple cases of threatened abortion and preterm delivery that required temporary hospitalizations in a health center in her community.

She was admitted to a second-level General Hospital of referral of her health center with a pregnancy of 32.1 weeks of gestation calculated by LMP, a clinical picture compatible with threatened preterm labor; Colic pain in the hypogastrium with irradiation to the lumbar and sacral area, a sensation of uterine constriction and little transvaginal bleeding, which lasted 26 hours, increasing in intensity, with minimal intervals of improvement. On admission with adequate active fetal kinetics, referring to the same symptomatology already mentioned.

Physical examination revealed blood pressure levels of 108/66 mmHg, heart rate (HR) 62 bpm, respiratory rate (RR) 18 rpm, temperature 36.7C, weight 84.2 kg, height 1.70 m, body mass index (BMI) 29.1. Globose abdomen at the expense of the gestating uterus, fundus height of 29 cm, single fetus, alive, transverse, upper dorsum, cephalic pole in the maternal right hypochondrium, fetal heart rate (FHR) of 131 ppm, uterine activity was perceived at the rate of three contractions in ten minutes lasting approximately 40 seconds. Obstetric ultrasound with a C5-2 convex transducer was performed, and the findings reported in Figure 2 were found. Assessment was limited by sonic window and transverse fetal position. Genital examination with scarce blood remains, without evidence of active bleeding, a vaginal speculum was placed where a central cervix was evidenced, with an approximate dilation of 4 cm, and shortened.

Therefore, the diagnoses of multiple pregnancy with pregnancy of 32.2, fetus with severe bilateral ventriculomegaly in the dorsal superior transverse position, preterm labor, previous uterine scar and satisfied parity were integrated. It was decided to start a protocol for emergency abdominal pregnancy resolution due to maternal history and fetal diagnoses, the laboratory results are shown in **Table 1**.

Table 1. Laboratory Studies Report

	Result
Blood Chemistry	
Glucose	74 mg/dL
Urea	18.3
Blood Urea Nitrogen (BUN)	10.0 mg/dL
Glomerular filtration rate (GFR)	118.41 mL/min
Serum creatinine	0.73 mg/dL
Uric acid	4.5 mg/dL
Blood biometry	
Total leukocytes	7.9 x 103/uL
Hemoglobin	13.1 g/dL
Hematocrit	29.3%
Hemotype	
Or positive	
Coagulation times	
Prothrombin time	12.1 seconds
Partial thromboplastin time	32.9 seconds
Fibrinogen	365

Subsequently, under subarachnoid block and as a result of a cesarean section with a body incision, a male newborn was obtained with a weight of 1,740 g, height of 44 cm, head circumference (CP) of 39.5 cm, thoracic circumference (PT) of 25 cm, foot 6 cm, Capurro test 32 weeks of gestation, newborn in cardiorespiratory arrest, generalized cyanosis, muscle flaccidity, Anatomically normal placenta, no neonatal resuscitation measures were provided, time of death was declared 4 minutes after birth.

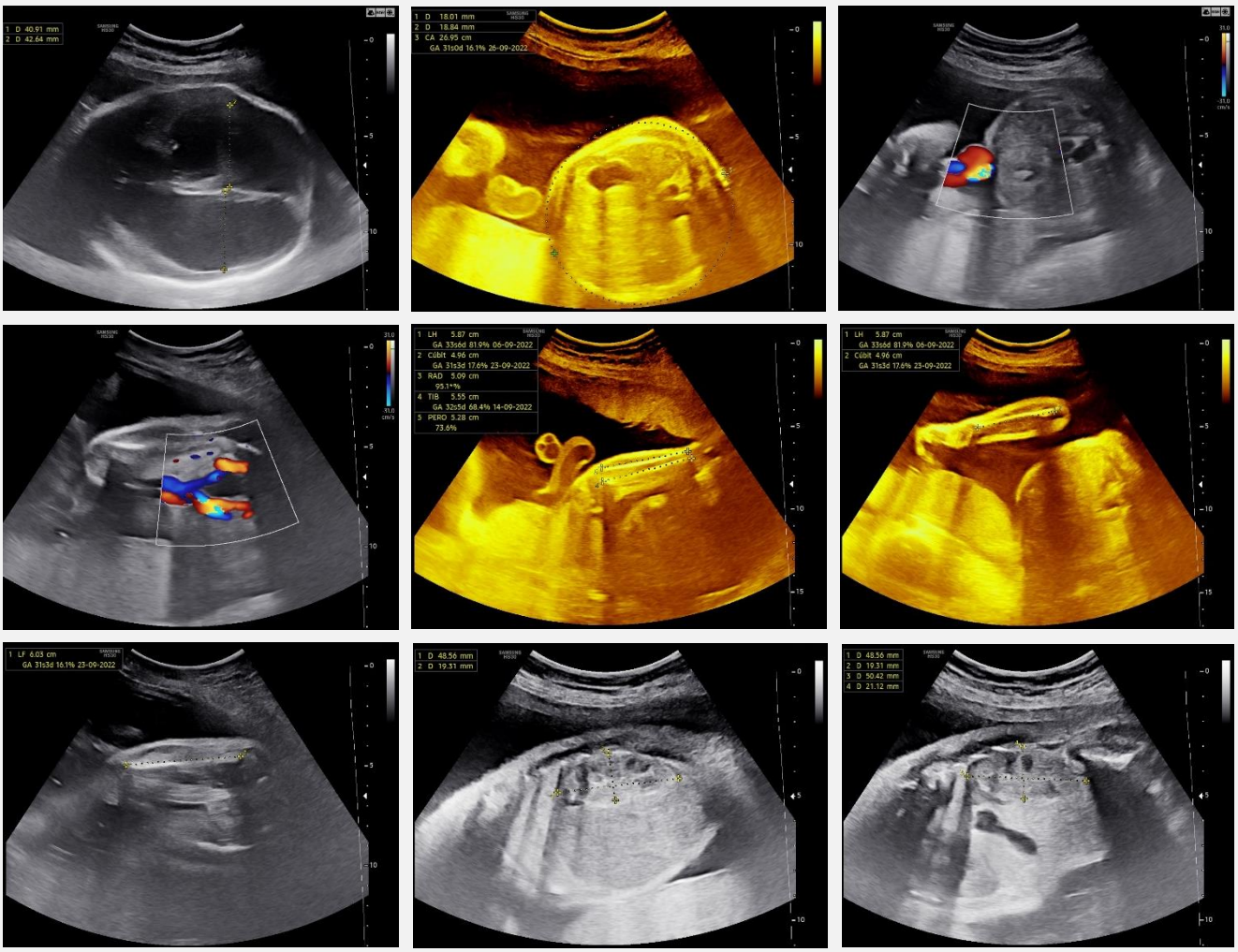


Figure 2. Structural evaluation at the level of the skull showed the presence of a midline, separate thalamus, no dilation of the third ventricle, lateral ventricles with a measurement of the left ventricular atrium of 40.9 mm and right ventricular atrium of 42.64 mm, posterior fossa without alterations with a cisterna magna, at the level of the face integrity of the midline and upper lip was observed. with the presence of orbiting lenses, nasal bone present. Symmetrical chest with four-chamber section with heart enlevocardia, levoapex, AV and VA concordance, perforated mode, integrity of the interventricular septum, unaltered outflow tracts, three-vessel cut-off and three-vessel trachea without alterations. At the level of the abdomen there is abdominal situs solitus, with normal renal pelvicles, two umbilical arteries are observed. Column without alterations. Limbs intact, tubular bones according to gestational age with complete finger count.

DISCUSSION

MV is most often detected by ultrasound performed in the second or third trimester of pregnancy. Ventricular evaluation is an important component of the standard second-trimester exam and includes measurement of the lateral ventricle atrium (VL). The atrium of the LV should be measured in the transventricular (axial) plane at the level demonstrated by the frontal horns and the cavum of the septum pellucidum (CSP), in which the cerebral hemispheres are symmetrical in appearance.

The forceps should be placed at the inner margin of the medial and lateral walls of the atria, at the level of the parieto-occipital groove and the glomus of the choroid plexus, on an axis perpendicular to the long axis of the VL **(Figure 1**, **Table 2**) (5). It should be noted that, due to artifacts in the near-field of the image, caused by shading of the proximal parietal bone, in the standard transventricular plane, only the hemisphere and lateral ventricle distal to the transducer are generally clearly visualized. However, efforts must be made, tilting the probe, to visualize both ventricles (8).

Table 2. Criteria for Proper Measurement of the Lateral Ventricles

Head in the axial plane
The image is enlarged appropriately, so that the fetal head fills most of the image
The focal area is at the appropriate level
The cerebral ventricles have a symmetrical appearance.
An image of the sickle is taken from the midline
The atrium and occipital horn of the lateral ventricle are clear image
The atrium of the lateral ventricle is measured at the level of the parieto-occipital sulcus
Calibrators are placed on the medial and lateral walls of the atrium perpendicular to the longitudinal axis of the ventricle

The third ventricle can be measured on a coronal image at its largest transverse diameter. A measure > 4 mm is considered enlarged. The fourth ventriclecan be measured on a midline sagittal image, a measurement > more than 7 mm is considered abnormal (9).

ETIOLOGIES ASSOCIATED WITH VENTRICULOMEGALY

Approximately 5% of cases of mild to moderate MV have been reported to result from congenital fetal infections, and approximately 5% of fetuses with apparently isolated mild to moderate MV have an abnormal karyotype, most commonly trisomy 21. Another 10% to 15% have abnormal chromosomal microarray (CMA) analysis findings (10). Diagnostic tests (amniocentesis) with CMA should be offered when MV is detected. It is important to perform an initial karyotype analysis or fluorescence in situ hybridization, with CMA reflex if the results of these tests are normal (11).

The most common cause of severe MV is aqueduct stenosis, which results from the narrowing of the cerebral aqueduct of Silvio, located between the third and fourth ventricles, leading to progressive dilation of the lateral and third ventricles (11). Aqueduct stenosis can be congenital (X-linked by L1 CAM mutation) or acquired (e.g., gliosis due to hemorrhage) or due to CNS malformations (rhomboencephalosynapsis) (12).

VENTRICULOMEGALY AND CHROMOSOMAL/GENETIC DISORDERS

Diagnosis of a fetus with MV should also include evaluation of the fetal karyotype. The incidence of chromosomal abnormalities is high (>15%) in both mild/moderate and severe MV in the presence of an associated structural abnormality (13). The incidence of abnormal karyotyping in fetuses with isolated mild/moderate MV is controversial: three meta-analyses report an incidence of abnormal karyoma-type of 2.8% (14), 5% (15), and 4.6% (16), respectively. The variation in results may depend on the prevalence of trisomies in the population studied, which in turn depends on previously implemented screening programs. In the last decade, special attention has been paid to the association of fetal malformations and copy number variations (CNVs) defined as microdeletions or microduplications of segments of the genome, ranging in size from one kilobase (kb) to several megabases (Mb), and identifiable with the use of chromosomal microarrays. Abnormal CNVs can be found in 6.6% of isolated MV fetuses and 24% of non-isolated cases (17).

For this reason, amniocentesis with CMA should be part of the diagnostic study of fetuses with MV, especially in non-isolated cases. In recent years, more than 100 genes associated with fetal MV have been identified, in most cases as part of defined genetic syndromes. One is L1 syndrome, which is caused by mutations in the L1CAM (Cell Adesion Molecule) gene. It affects approximately one in 30,000 men causing X-linked hydrocephalus with aqueductal stenosis (18). Other genetic syndromes have been associated with MV, including RASopathies (Noonan syndrome, Costello syndrome, neurofibromatosis type 1, cardiofaciocutaneous syndrome) and ciliopathies (Meckel syndrome, Joubert syndrome, digital orofacial syndrome) (19), these conditions are often associated with severe MV as well as additional abnormalities. The most common chromosomal and non-chromosomal genetic conditions associated with ventriculomegaly are presented in **Table 3**.

Table 3. Genetic conditions associated with ventriculomegaly

Genetic disorders	Central Nervous System Findings
Chromosomal disorders	
Trisomy 21	Ventriculomegaly, holoprosencephaly
Trisomy 18	Ventriculomegaly, large choroid plexus cyst, cerebellar hypoplasia, mega cisterna magna, holoprosencephaly, dysgenesis of the corpus callosum, spina bifida
Trisomy 13	Ventriculomegaly, cerebellar hypoplasia, mega cisterna magna, holoprosencephaly, agenesis of the corpus callosum, microcephaly
Non-chromosomal disorders	
X-linked hydrocephalus	Ventriculomegaly, agénesia/dysgenesia of the corpus callosum
Ciliopathies (e.g., Meckel-Gruber syndrome*, Joubert syndrome)	Ventriculomegaly, occipital encephalocele, vermian and cerebellar hypoplasia, microcephaly, agenesis of the corpus callosum, holoprosencephaly, anencephaly
Dystroglianopathies (e.g., Walker-Warburg syndrome*)	Ventriculomegaly, malformation of cortical development, cerebellar anomalies, occipital cephalocele, agénesia/dysgenesia of the corpus callosum, brainstem abnormalities (Z-form), ocular anomalies
Tubulinopathies (e.g., TUBA1A)	Ventriculomegaly, malformation of cortical development, basal ganglia dysgenesia, agénesia/dysgenesia of the corpus callosum, cerebellar dysgenesia/hypoplasia, midbrain abnormalities

VENTRICULOMEGALY-RELATED INFECTIONS

MV may be associated in approximately 2% of cases with fetal infections, particularly cytomegalovirus (CMV), toxoplasmosis, parvovirus, Zika virus (20). Sporadic cases of MV associated with other viruses (enteroviruses, parainfluenza viruses) have also been reported, which also cause inflammation of the arachnoid granulations and excess production of cerebrospinal fluid (7).

Many cases of MV due to infectious diseases will have other ultrasound findings, such as brain calcifications, periventricular cysts, or signs external to the CNS (21). Brain imaging findings include cerebral calcification, MV, microcephaly, white matter abnormalities, cortical malformations, cerebellar hypoplasia, subependymal cysts, intraventricular septa, and pathognomonic cystic changes in the occipital or anterior temporal horns (22).

However, in some cases, MV is the only ultrasound feature; therefore, testing for infections should be offered in all cases of isolated MV, and is strongly recommended if the fetus has other features suspicious of an infectious cause. Maternal serology is the first diagnostic test, but it is less sensitive and specific than PCR in amniotic fluid, making amniotic fluid PCR the preferred method of evaluation for fetal infections (21). Cases associated with infection have been reported demonstrating other ultrasound markers, including fetal growth restriction, periventricular, hepatic, and other intra-abdominal calcifications, echogenic fetal bowel, hepatosplenomegaly, ascites, meconium peritonitis, polyhydramnios, and microcephaly. However, these features may not be apparent until later in gestation, and not all infected fetuses will have other ultrasound signs (7).

PCR amniocentesis performed before 21 weeks' gestation has a sensitivity of 45% to 80% for CMV; therefore, a negative result does not exclude CMV infection. PCR performed on amniotic fluid after 21 weeks gestation or 6 to 7 weeks after maternal primary infection has a higher sensitivity and specificity between 97 and 100%. The positive predictive value of the test is close to 100% (23), although false-positive CMV results have been reported by PCR (24). PCR for toxoplasmosis performed in amniotic fluid has a sensitivity of 64%, a negative predictive value of 87%, and a positive predictive value of almost 100% (25).

The woman's history should be reviewed for symptoms resulting from CMV infection, and exposure to potential sources of toxoplasmosis (e.g., outdoor cats, gardening, consumption of

undercooked meat) and Zika virus should be evaluated (26). For women who decline amniocentesis, serum testing for CMV includes IgG and IgM, as does screening for toxoplasmosis. Negative IgG and IgM results for CMV and toxoplasmosis do not suggest prior exposure, excluding these infections as a cause of MV; positive IgG and IgM negative results suggest prior infection and immunity, making congenital infection unlikely as a cause of VM. In women with a positive CMV IgM result, IgG avidity testing is recommended; a low-avidity IgG and a positive IgM indicate an infection in the previous 3 months (27). A positive result for IgG and IgM toxoplasmosis may indicate a recent infection or a false-positive result. A positive IgM toxoplasmosis antibody result should be followed by an IgG avidity test and repeat IgM testing at a reference laboratory. As with CMV, high-avidity IgG suggests that the infection predated pregnancy.

STRUCTURAL ABNORMALITIES ASSOCIATED WITH VENTRICULOMEGALY

MV may be associated with several underlying CNS abnormalities. Some of structural origin, such as holoprosencephaly, hydranencephaly, porencephaly or schizencephaly, and cystic lesions, such as arachnoid cysts, which result in abnormal collections of fluid in the fetal brain that can be misdiagnosed as MV, structural abnormalities that can lead to dilation or enlargement of the VLs have also been described. These include agenesis of the corpus callosum, Dandy-Walker malformation, neural tube defects, cortical defects, and migratory abnormalities or heterotopia (11). The incidence of additional CNS ultrasonographic abnormalities identified in fetuses with mild or moderate MV ranges from 10% to 76%, but appears to be <50% in most studies (28).

VENTRICULOMEGALY RELATED TO THE PRESENCE OF CHOROID PLEXUS CYSTS

Choroid plexus cysts (CPCs) form during normal development, representing neuroepithelial folds that subsequently fill with cerebrospinal fluid (CSF) and cellular debris. They have been identified in about 1% of fetuses during routine second-trimester ultrasound (29). They are defined as unilocular or septate cystic spaces in the choroid larger than 3 mm in diameter (30). The only importance of finding a CPC in the fetus, even an isolated CPC, is the increased risk of trisomy 18 (31). Isolated large CPCs can cause distention of fetal VLs. It has been suggested that this mechanism of MV is benign if other causes of MV and other associated abnormalities have been excluded, and therefore prenatal counseling should be reassuring (32).

DISRUPTIVE EVENTS: FETAL CENTRAL NERVOUS SYSTEM HEMORRHAGE

Mutation of the COL4A1 gene has been described as a feature present in fetuses with severe brain hemorrhagic lesions. These lesions are extensive or multifocal and may be supratentorial (intraparenchymal or intraventricular) and/or infratentorial, and may worsen during pregnancy. These lesions may interfere with the fragility of brain vessels associated with the COL4A1 mutation (33).

Ischemic and hemorrhagic lesions associated with COL4A1 mutations are usually multifocal and often extend to several brain territories (34).

It is suggested to systematically search for mutations in the COL4A1 and COL4A2 genes in cases of severe and/or multifocal hemorrhagic or ischemic-hemorrhagic brain lesions, particularly when these abnormalities are of different ages and are associated with scizencephaly or porencephaly. Exome analysis is likely to be useful in the characterization of cases carrying this phenotype (34).

CONGENITAL AQUEDUCTAL STENOSIS (CAS)

It is a form of obstructive hydrocephalus in which there is a partial or complete obstruction of flow through the cerebral aqueduct leading to enlargement of the VL and third parties in the context of intracranial hypertension. It may be due to genetic or acquired causes (35). Four genes that cause congenital hydrocephalus have been described as the main feature of this disease process, two of which are X-linked (L1CAM and AP1S2) and two autosomal recessive (CCDC88C and MPDZ) (11).

NEURAL TUBE DEFECTS: MYELOMENINGOCELE

Myelomeningocele is the most common open neural tube defect. The persistence of a CSF leak in fetal life at the level of the defect creates a pressure gradient between the brain and the spine that results in significant alterations in brain development (cervical hernia, small posterior fossa,

and distorted ventricular anatomy, among others). This can lead to an alteration of CSF dynamics and the development of MV with progression to communicative and/or non-communicative hydrocephalus (36).

CRANIOSYNOSTOSIS AND VENTROCULOMEGALY

Nonsyndromic craniosynostosis related MV is a disorder that may represent simple MV or shunt-dependent hydrocephalus (37). However, diagnosis can be challenging in these patients and may require invasive methods such as intracranial pressure monitoring (ICP) or lumbar puncture (LP), as both syndromic and non-syndromic disorders can elevate PCI and show excessive symptoms (38). MV is common in syndromic craniosynostosis and has been reported in 30% to 70% of patients with Crouzon and Pfeiffer syndrome (39), and 40% to 90% of patients with Apert syndrome (40). While shunt-dependent hydrocephalus is common in Crouzon and Pfeiffer syndromes, in Apert syndrome, most cases with ventriculomegaly do not require shunt (36).

Once the diagnosis of MV has been confirmed, a complete examination of the fetal anatomy, including a detailed neurosonographic evaluation, preferably using the transvaginal approach, is indicated in order to exclude associated conditions (**Table 4**).

Table 4. Etiologic Classification of Ventriculomegaly and Associated Central Nervous System Abnormalities

MALFORMATIVE
Holoprosencephaly
Agenesis of the corpus callosum
Malformations of cortical development
OBSTRUCTIVE
Intracranial tumor
Aqueduct stenosis
Malformation of the back of the brain
Open Neural Tube Defects: Chiari Malformation Type 2
CLASTIC
Intrauterine infection
Ischemia
Interventricular hemorrhage
ATROPHIC
Metabolic disease
Neurodegenerative disorders
Lesions following feto-fetal transfusion syndrome, and as a consequence of single fetal death in the pregnancy of monochorionic twins

USEFULNESS OF AN ADEQUATE ULTRASOUND FOR THE DETECTION OF VENTRICULOMEGALY

The sensitivity of ultrasound in detecting MV is controversial. The largest multicenter study in Europe (EUROFETUS) (41) reported a sensitivity of 93.5%. However, this apparently good result refers to severe forms of MV and also to cases diagnosed in the third trimester. Before 24 weeks gestation, sensitivity drops to 35%, which likely reflects the natural history of the disease, rather than a measurement error. No data are available regarding false-negative cases of mild MV.

In contrast, mild MV is among the most frequent false positive diagnoses in ultrasound detection of fetal malformations, mainly due to an incorrect measurement technique (21).

ROLE OF NUCELLAR MAGNETIC RESONANCE IMAGING (MRI)

Unlike ultrasound, it is not limited by the ability to penetrate the surrounding fetal skull and maternal soft tissues, and it also demonstrates superior contrast resolution (39).

In order to increase the ability to recognize the underlying causes of MV, MRI has been suggested as an additional diagnostic tool in fetuses with ventricular dilation, particularly mild and moderate fetuses. A meta-analysis by Rossi and Prefumo on the role of MRI in women with CNS malformations reported 10% of the additional information provided by MRI in fetuses with MV regardless of the severity of dilation (42). The MERIDIEN cohort study reported an additional overall diagnostic outcome in 8.8% of cases, with different results according to MV severity: 4.9% in mild MV, 19.4% in moderate MV, and 30.8% in severe MV (43).

A recent multicenter study conducted by the ENSO (European Neurosonography) working group (44) reported additional structural abnormalities detected on prenatal MRI and not performed on ultrasound in only 5.4% of cases of mild and moderate MV (mainly intracranial haemorrhage, polymicrogyny and lissencephaly). The associated abnormalities were detected exclusively at birth and were lost on prenatal imaging in 3.8% of cases. In

severe MV, the same group (45) reported additional structural abnormalities detected exclusively on prenatal MRI in 18.1% of cases. When considering the type of abnormality, cortical developmental malformations were detected on MRI in 32.4% of cases, while midline or acquired lesions (hypoxemic/hemorrhagic) were detected in 26.5% and 14.7% of cases, respectively (7).

TRACKING

When the diagnosis of MV has been established, repeated ultrasound assessments should be planned, scheduled, and performed to assess the course of fetal MV and further evaluate associated abnormalities. According to the natural history of MV, in 57% of cases, stability or cessation of progression occurs, progression in 24%, and resolution in 29% (46). Resolution occurs primarily in the mild VM group.

PROGNOSIS

Ventricular measurements that are closer to 10 mm are more likely to represent a normal variant, fetuses with a ventricular atrial diameter of 10 to 12 mm have a normal postnatal evaluation in more than 90% of cases (47). Approximately 7% to 10% of fetuses with apparently isolated mild MV have been reported to have other structural abnormalities on examination after birth (48). Neurological, motor, and cognitive impairment are more likely when severe MV is present, in these cases, a meta-analysis reported a survival rate of 88%, and only 42% of these children had normal neurodevelopment (49). Neurodevelopmental outcome in people with isolated MV is normal in 74.3% of cases, borderline or moderate disability is present in 14.3% of cases, and severe disability in 11.4%. In addition, 15.1% of cases thought to be prenatally isolated were found to have associated abnormalities after birth (50). It is worth mentioning that the prognosis depends, in addition to the severity of the dilation, on the underlying cause, that is, although they have similar diameters, cases affected by T.21 do not have the same outcome as those of lissencephaly, for example.

PROGNOSIS IN ISOLATED VENTRICULOMEGALY

The incidence of isolated MV has a prevalence of 0.7%, while isolated severe MV has a prevalence of between 0.03% and 0.15% (28). The overall rate of developmental delay in patients with mild to moderate MV has been shown to be 7.9%, which is comparable to that observed in the general population (16). In contrast, only 3.5% of patients survived beyond 2 years when severe MV was involved (51, 52).

CONCLUSIONS

It is important that the VL is measured correctly, as small differences in technique can result in false positive or false negative results. Substantial interobserver variability in interpretation may occur, particularly in borderline ventricular diameters (i.e., around 10 mm). MV should be classified as a sign, it is considered the most frequent anomaly of the CNS, it is non-specific and evolutionary, thus being the most sensitive marker of anomaly of CNS development.

With regard to the different etiologies of MV, the most common is that there is a malformative pathology of the CNS. Only 10% of cases are attributed to infections, and the most frequently isolated pathogens have been CMV, Toxoplasma and Zika, if this etiology is suspected, maternal serology should be performed in all cases, and if the IgG is positive, amniocentesis for PCR is indicated.

The rate of associated anomalies is high, approximately 40%, of the extracranial anomalies, half are cardiac, and are more frequent in the most severe cases (10% vs 55%). As complementary studies, it has been shown that MRI allows a better evaluation of the cortex, mainly in severe and/or progressive MV. The outcome of fetal MV depends on three factors: degree of MV, progression of ventricular dilation, and presence of associated abnormalities. The latter is the most important predictor of outcome and the most useful for providing accurate advice.

We can conclude that the finding of MV is only the first step in recognizing the cause of ventricular dilation. When no cause is found, MV is defined as "isolated," indicating that the etiology is not known. This represents, by definition, a provisional discrimination of exclusion.

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