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Congenital Hydrocephalus: A Rare Case Report and Literature Review

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ABSTRACT
Hydrocephalus is a condition characterized by the abnormal accumulation of cerebrospinal fluid (CSF) within the brain's ventricles and/or subarachnoid space, leading to ventricular enlargement and elevated intracranial pressure.
This anomaly is sometimes detected during a routine fetal scan between 18 and 22 weeks of gestation, although early signs may be visible by the end of the first trimester. Ultrasound and magnetic resonance imaging data play a crucial role in guiding patient counseling and pregnancy management. It is essential to inform the couple about the prognosis of these anomalies to ensure optimal care and decision-making.
We report the case of severe hydrocephalus diagnosed prenatally at 23 weeks of gestation. Through this clinical case, we discuss the ultrasound signs supporting a positive diagnosis of hydrocephalus as well as the management approach.

Keywords: Fetal hydrocephalus, Ventriculomegaly, Prenatal diagnosis, morphology ultrasound.

INTRODUCTION:

Hydrocephalus is a brain abnormality marked by an excessive buildup of cerebrospinal fluid (CSF) in the cerebral ventricles[1].
Although hydrocephalus can develop at any age, congenital hydrocephalus is distinguished by its early beginning during the intrauterine phase, high treatment difficulty, and frequently unfavorable neurological results[1].
Congenital hydrocephalus can be caused by different developmental
Abnormalities.
Genetics, infection, intraventricular hemorrhage, and brain structural abnormalities such neural tube malformations, agenesis of the corpus callosum, and cerebral aqueduct stenosis are among the causes of hydrocephalus. Congenital hydrocephalus is most commonly caused by aberrant brain development, cerebral aqueduct blockage, Chiari malformations, and Dandy-Walker deformities[2].
Worldwide, congenital hydrocephalus incidence varies. Nonetheless, it ranges from 2.2 to 18 per 10,000 live births[2].
This anomaly can occasionally be found at the end of the first trimester, or it can occasionally be seen during a routine fetal ultrasound at 18 to 22 weeks gestation.
The prognosis of congenital defects must be communicated to the couple for better care; ultrasound and magnetic resonance imaging data is crucial for counseling patients and managing pregnancies.
Here, we present a case of congenital hydrocephalus . Furthermore, the essential literary works required

for comprehending and tackling this persistent phenomenon are supplied.

Case rapport:

Mrs B.M is 24 years old with no medical or surgical antecedents. She is primigravida; the result of a non-consanguineous marriage; free of dysgravidia , pregnant at 23 weeks of amenorrhea and is on martial therapy with no folic acid. The patient missed the first trimester screening and the second trimester prenatal screening test. A morphology scan confirms the triventricular hydrocephalus **(Figure 1).**

The ultrasonography showed the biparietal diameter of the fetus was 6.2 cm (>97, the head circumference was 22.4 cm and the cerebral cortex became thinner **(Figure 2; 3)**.

Severe ventriculomegaly, with atrial width >15 mm. The fetus has no other anomalies through the scan.

Our patient refuse to undergo a second trimester prenatal screening tests a karyotype exam and magnetic resonance imaging for financial reasons.

The TORCH infection screen revealed no abnormalities.

Following a multidisciplinary prenatal consultation and after receiving informed consent, the parents chose to terminate the pregnancy.

The findings indicate that prenatal counseling and ongoing follow-up can help prevent further complications.

Consequently, sharing our clinical case and discussing the ultrasound features of these defects, along with their epidemiology, prenatal diagnosis, and outcomes, is essential.

DISCUSSION:

An aberrant buildup of cerebrospinal fluid (CSF) in the brain is the hallmark of hydrocephalus. The primary clinical sign in a fetus is enlargement of the head, though cerebral ventriculomegaly can occasionally cause a head circumference that is within normal bounds. There could be pressure on the CSF, compressing it and harming the brain[3] .One newborn out of every 1,000 has congenital hydrocephalus[4].

It is a serious and common malformation, linked to significant fetal and neonatal morbidity and mortality. Prognostic factors include associated anomalies (both intracranial and extra cranial), aneuploidy, underlying etiology, and the degree of ventricular enlargement.

Hydrocephalus can be divided into two types:

Communicating hydrocephalus occurs when the flow of cerebrospinal fluid (CSF) is obstructed after it leaves the ventricles, but CSF can still circulate between the ventricles, which remain connected[5].

Non-communicating hydrocephalus, also known as obstructive hydrocephalus, arises when the flow of CSF is blocked within one or more of the narrow pathways linking the ventricles. A common cause is aqueductal stenosis, a narrowing of the aqueduct of Sylvius, the small channel connecting the third and fourth ventricles in the brain[5].

A routine fetal examination includes a prenatal sonographic assessment of the cerebral ventricles, which can be detected as early as 15 weeks of gestation and specially the morphology scan. Measuring these ventricles is a key part of evaluating the central nervous system (CNS) [6].

Hydrocephaly was diagnosed when the ventricle-to-hemisphere ratio exceeded the upper limits of the reference ranges. Later studies tracked the in utero growth of various parts of the ventricular system, leading to the establishment of reference values for ventricular width throughout pregnancy.

The accepted standard today is the measurement of atrial width at the level of the choroid plexus.

An atrial diameter more than or equal to 10 mm is referred to be VM (ISUOG 2007). VM is classified as mild if the atrial diameter is between 10 and 15 mm and as severe if it is greater than 15 mm; however, other writers employ the classifications of mild (10–12 mm), moderate (13–15 mm), and severe (20 mm). (1991's Bromley et al).[7].

When the ventricles are slightly enlarged, the word "ventriculomegaly" is typically used, and when they measure more than 15 mm, the term "hydrocephalus" is employed.[7]

In our case the morphology scan showed a biparietal diameter of the fetus was 6.2 cm (>97) **(Figure 3)**, the head circumference was 22.4 cm and the cerebral cortex became thinner **(Figure 2)**.

There are various etiologies and classifications, but all differentiate between "communicating" (where CSF flows from the ventricular system to the subarachnoid spaces) and "obstructive" (where CSF flow is blocked at some point within the ventricular system)[1].

Conventionally, chromosomal analysis, magnetic resonance imaging (MRI), and screening for infections(usually TORCH) are the methods used to investigate the etiology of fetal hydrocephalus .

Targeted ultrasound exams and fetal echocardiography are essential components of the diagnostic work-up when ventriculomegaly is discovered, as the possibility of concomitant malformations is high and the presence of central nervous system or extra neural abnormalities is possible specially when ventricular width >=15 mm [6].

From a diagnostic perspective, the extent of ventriculomegaly (whether bi-, tri-, or tetraventricular) can provide clues about the location of the obstruction. For instance, Dandy-Walker malformation is typically linked to tetraventricular hydrocephalus, while hemorrhage or aqueductal stenosis may result in triventricular hydrocephalus[7]. Magnetic resonance imaging plays a very important role in detailing abnormalities.

Our patient refuse to undergo a second trimester prenatal screening tests a karyotype exam and magnetic resonance imaging for financial reasons.

The TORCH infection screen revealed no abnormalities.

The identification of additional anomalies allows for a more accurate adjustment of the current therapeutic approach and also provides information about the future prognosis of the fetus.

The progression of ventriculomegaly in utero appears to be linked to the prognosis, with a more favorable outcome when the condition improves or resolves, regardless of the initial severity (whether mild, moderate, or severe)[6]

Severe ventriculomegaly (greater than 15 mm) is uncommon and has a low probability of resolving spontaneously[8].

While numerous studies focus on the clinical outcomes of congenital hydrocephalus following postnatal treatment, there is a limited number of articles that provide long-term follow-up data[9].

Overall, the 10-year survival rate is approximately 60%, with only half of these individuals achieving normal cognitive function sufficient for independent living, though they may experience learning difficulties in school. Intellectual disability, cerebral palsy, and epilepsy are notably common in children with congenital hydrocephalus[1].

A new chapter in invasive fetal therapy commenced in the early 1980s when multiple independent groups pioneered shunting procedures for the treatment of hydrocephalus[10].

Precautions and criteria for case selection should be standardized. While there are various intervention methods, there is no clear evidence that one method is superior to the others[11]

At that time, the understanding of the natural history, pathophysiology, and criteria for patient selection was rudimentary and incomplete. However, experimental studies by various researchers using suitable animal models helped clarify the pathophysiology of these conditions and laid the theoretical groundwork for intervention[12].

When the hydrocephaly is severe with a poor prognosis and detected earlier, some patients prefer to terminate the pregnancy; the same in our first case.

Conclusion:

Hydrocephalus is one of the most frequently identified cerebral abnormalities through ultrasonography, yet it remains challenging to diagnose with precision. Prenatal counseling and family support are crucial, but accurate diagnoses are essential for effective guidance. The detection of isolated hydrocephalus can present a dilemma due to the wide variability in outcomes. Therefore, it is important to develop clear guidelines for the diagnosis and management of fetal hydrocephalus.

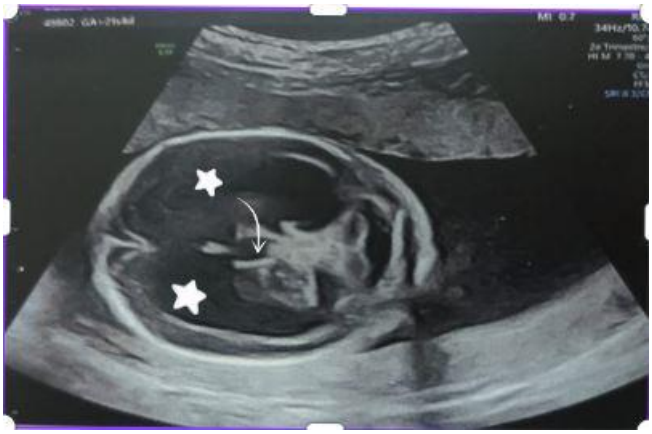

Figure 1 : Tri-ventricular Hydrocephalus : star: lateral ventricles arrow : Third ventricles

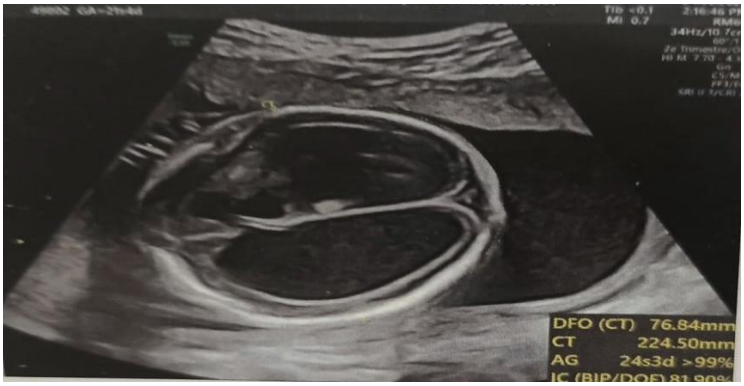

Figure 2 : Hydrocephalus : the head circumference was 22.4 cm and the cerebral cortex became thinner

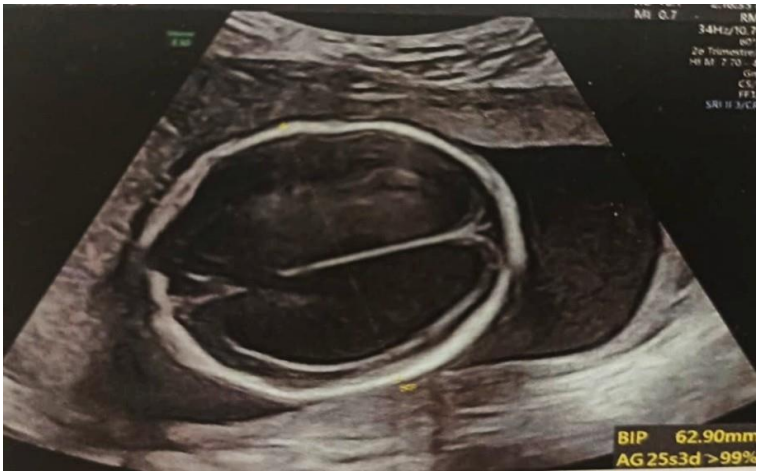

Figure 3 : hydrocephalus : a biparietal diameter of the fetus was 6.2 cm (>97).

Referneces

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