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Case Presentation
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Stenosis in the Aqueduct of Sylvius and Hydrocephalus: A Case Report

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Abstract

Hydrocephalus is a disorder triggered by physical or functional block of Cerebrospinal Fluid (CSF) flow that leads to progressive ventricular dilatation. Therefore, hydrocephalus has multiple competing classification systems. Congenital forms of hydrocephalus are complicated, both anatomically and mechanically which makes the prenatal diagnosis of the etiology a challenging task to approach as a clinician.

Here, we review a case report about a stenosis of the aqueduct of Sylvius causing infantile hydrocephalus and we describe the major role of Ultrasound and MRI and their interests as being precocious methods of screening.

Keywords: Hydrocephalus; Aqueduct of Sylvius; Antenatal period; Prenatal ultrasound; Fetal MRI

Introduction

Hydrocephalus generally refers to a disorder of Cerebrospinal Fluid (CSF) physiology resulting in abnormal expansion of the cerebral ventricles [1], typically associated with increased intracranial pressure. Hydrocephalus has many causes and Aqueduct stenosis is one of the most common causes of congenital hydrocephalus. We present a case where the antenatal ultrasound had shown fetal hydrocephalus with an anomaly in the posterior fossa.

Case Presentation

A 30-year-old with no dysgraphia at 21 weeks of gestation was referred to our perinatology department due to hydrocephalus, which was detected on a mid-trimester US examination with no other abnormality reported. The first-trimester screening test had shown an increased risk of Down syndrome (1/50). An amniocentesis was carried out and concluded to a chromosomal formula 46, XX, a female karyotype and absence of chromosomal abnormalities.

Inquiries into the antenatal history revealed no evidence of rubella or other viral infections, and no evidence of toxoplasma infection has been obtained.

The interview revealed no chronic maternal illness neither consanguinity nor history of congenital anomalies. Her previous two children were alive and healthy. Serological status for Hepatitis B, C, Syphilis, and HIV was negative

We repeated the US examination with a Voluson P8 scanner (GE Healthcare), equipped with convex and 4D probes which had revealed a tri-ventricular dilatation with a lateral ventricle at 12 mm and the third ventricle at 8 mm, the fourth thin ventricle without other cerebral abnormalities (Figure 1).

A fetal MRI was performed at 30 weeks of gestational age with a 1.5 T scanner (Siemens MAGNETOM AERA). MRI protocol included sequences in frontal and sagittal planes. This exam revealed a significant tri-ventricular dilation responsible for cerebral parenchymal thinning concerning Sylvius aqueduct stenosis and the vermian height slightly decreased.

A multi-disciplinary staff including radiologists, neonatologists, neurosurgeons, and gynecologists took place and the decision was made to bring the pregnancy to term.

The birth was 39 gestational ages and 5 days vaginally with the birth of a newborn of feminine sex and Apgar score: One minute at 4, 3 min at 3, and 10 min at 3 with death at 30 min of life.

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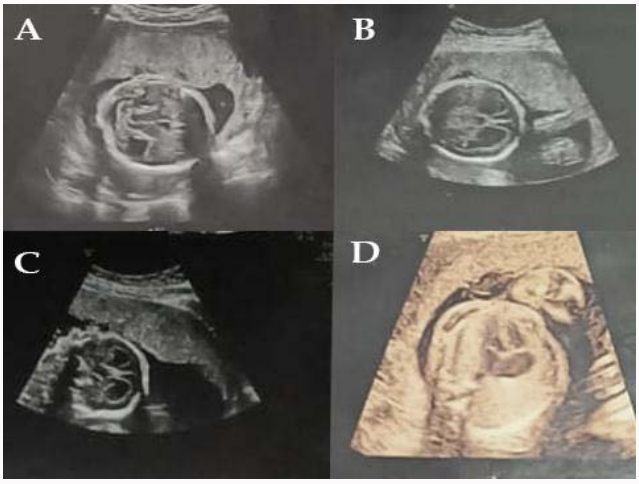

Figure 1A-1D: Ultrasound imaging showing tri-ventricular dilatation without other cerebral abnormalities.

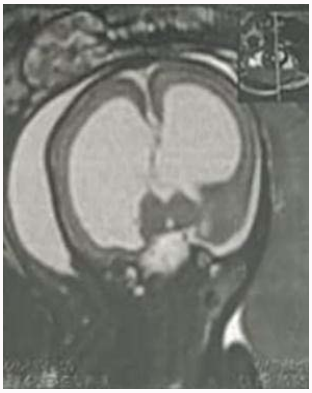

Figure 2: Frontal section of the fetal MRI showing important tri-ventricular dilation with parenchymal thinning.

Foetaphalogic examination confirmed the echography and radiological findings by showing a tri-ventricular hydrocephalus responsible for a significant thinning of the residual cerebral parenchyma caused by tight stenosis of the Sylvius aqueduct.

Discussion

Hydrocephalus could be defined as a disproportion between the production, flow, and resorption of CSF [2].

It may be idiopathic or secondary, “congenital” or “acquired”, “communicating” when there is a free passage of CSF from ventricular cavities to meningeal or "no communicative", that is, blocked by an anatomical obstacle and it can be described according to the ventricular cavities concerned [1].

CSF is secreted mostly at the choroid plexus once produced; it passes through the lateral ventricles to the third ventricle.

It continues through the Monro holes to the fourth ventricle through the aqueduct of Sylvius. This intraventricular area juxtaposes another peri-cerebral and peri-medullary space [1]. Both areas communicate through the medial and lateral openings of the fourth ventricle and its resorption is mainly at the level of arachnoid granulations.

It is a liquid that ensures a nutritional role; it serves as a means

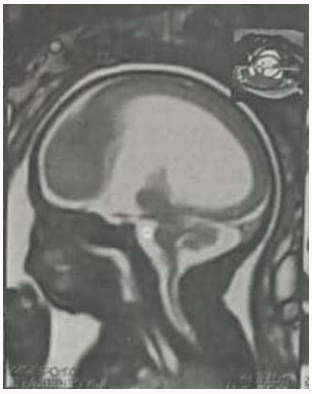

Figure 3: Sagittal section of fetal MRI showing a tri-ventricular hydrocephalus by stenosis of the aqueduct of Sylvius.

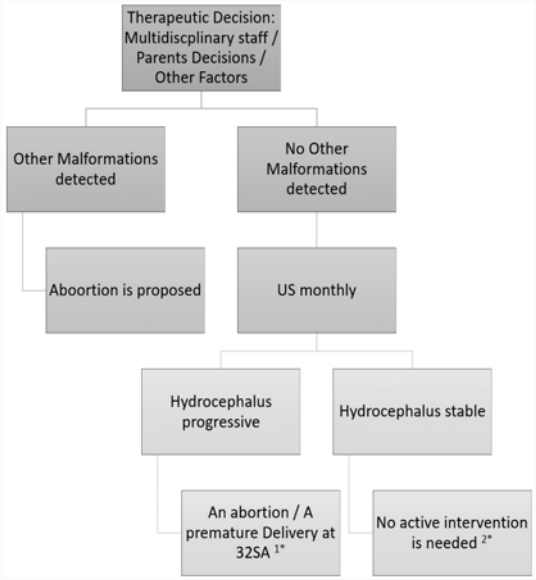

Figure 4: Tree of decision in front of hydrocephalus.
1*: Several attitudes are possible depending on the term of pregnancy and the choice of parents knowing that in these forms, the long-term mental prognosis is unfavorable. An abortion, a premature delivery at 32 SA for rapid placement of a shunt, or ventricular decompression in utero by ventriculo-amniotic shunt.
2*: No active intervention is necessary and the opportunity for derivation will be discussed in the weeks or months following birth.

of transport and has a role in the homeostasis of the central nervous system.

Hydrocephalus is the result of a CSF circulation disorder where three mechanisms could be found [3]: Abnormality of secretion, which exists only as a pathological situation leading to hyperproduction as some tumors of choroid plexus (papilloma’s) [3].

Circulation anomaly, which is the most common mechanism when the blockage most often occurs in ventricular narrowing: Either at the interventricular foramens, at the Sylvius aqueduct, or the posterior cerebral fossa, Abnormalities of resorption may be following an infectious process, hemorrhagic or venous hyper pressure. In antenatal, we have two diagnostic means: Ultrasound [4]: The systematic monitoring of all pregnancies currently makes possible early detection of antenatal hydrocephaly.

This diagnosis, too heavy of consequences, should only be evoked

with caution, on repeated examinations.

MRI allows good visualization of the different structures of the fetal brain. During pregnancy, an MRI is indicated for all ventricular dilations greater than 12 mm as well as in evolutionary forms for the study of cerebral gyration and parenchyma [5] (Figure 2, 3).

It can also be indicated in addition to ultrasound in search of an etiological diagnosis of pathology.

Before a ventricular dilation, we must: First, we confirm the diagnosis by respecting the rules of measurement, assess the importance of dilatation, and investigate if it is a question of bilateral ventriculomegaly, 3rd and/or 4th ventricle involvement, extra-cerebral malformations making the prognosis unfavorable and evaluate its scalability compared to one or more previous studies [6].

The conduct to be treated cannot be systematized [7]; it must be discussed before each case taking into account in particular the quality of the child's life in a multidisciplinary team and the parents are informed of the conclusions and participate in the decision.

This decision depends on several factors: age of the pregnancy, characteristics of hydrocephalus (early the importance of hydrocephalus, evolution, possible association with other Malformations and etiology) (Figure 4).

Their exact cause is most often unknown: Malformation, genetic origin, hereditary, teratogenic, or infectious [8].

From Spina bifida, Arnold Chiari's malformation, and Dandy-Walker's syndrome to Sylvius's aqueduct stenosis, which corresponds to narrowing of the ventricular tract, preventing CSF from circulating normally.

Conclusion

Hydrocephalus is a complex condition with both genetic and environmental causes. In acquired hydrocephalus, an extrinsic cause is often readily by history or based on imaging findings [9], however, in the majority of the cases, this cause is not readily discernible and an anomaly in the intrauterine life may be the cause such as stenosis in the aqueduct of Sylvius.

A careful review of imaging findings is critical to arriving at a diagnosis in the antenatal period such as Ultrasound and Fetal MRI to conduct a decision.

Though we believe that hydrocephalus in many children is a marker of abnormal brain development and the major causes of this pathology remain unknown, therefore ongoing research is imperative using new technologies as new genetic technologies [10] can lead to the discovery of causative genes and the phenotypes associated with them.

References

1. Filis AK, Aghayev K, Vrionis FD. Cerebrospinal fluid and hydrocephalus: Physiology, diagnosis, and treatment. Cancer Control. 2017;24(1):6-8.
2. Estey CM. Congenital hydrocephalus. Vet Clin North Am Small Anim Pract. 2016;46(2):217-29.
3. McAllister JP. Pathophysiology of congenital and neonatal hydrocephalus. Semin Fetal Neonatal Med. 2012;17(5):285-94.
4. D'Addario V. The role of ultrasonography in recognizing the cause of fetal cerebral ventriculomegaly. J Perinat Med. 2004;32(1):5-12.
5. Matys T, Horsburgh A, Kirollos RW, Massoud TF. The aqueduct of Sylvius: Applied 3-T magnetic resonance imaging anatomy and morphometry with neuroendoscopic relevance. Neurosurgery. 2013;73(2 Suppl Operative):132-40.
6. Murphy C, Vassallo G, Burkitt-Wright E, Hupton E, Eelloo J, Lewis L, et al. A retrospective regional study of aqueduct stenosis and fourth ventricle outflow obstruction in the paediatric complex neurofibromatosis type 1 population; Aetiology, clinical presentation and management. Clin Neurol Neurosurg. 2020;193:105791.
7. Hirsch JF, Hirsch E, Sainte Rose C, Renier D, Pierre-Khan A. Stenosis of the aqueduct of Sylvius. Etiology and treatment. J Neurosurg Sci. 1986;30(1-2):29-39.
8. Globus JH, Bergman P. Atresia and stenosis of the aqueduct of Sylvius. J Neuropathol Exp Neurol. 1946;5:342-63.
9. Mitra DK, Srinivas M. Congenital hydrocephalus. Indian J Pediatr. 1997;64(6 Suppl):15-21.
10. Kahle KT, Kulkarni AV, Limbrick DD, Warf BC. Hydrocephalus in children. Lancet Lond Engl. 2016;387(10020):788-99.