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Prenatal Ultrasound Diagnosis of Agenesis of the Corpus Callosum: A Case Report and Review of the Literature.

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Abstract*: Agenesis of the corpus callosum is a frequent malformation, affecting 0.05% of births. The diagnosis can be made prenatally, by performing a prenatal ultrasound. The prognosis of children with this condition is better known today and depends on the presence or absence of associated malformations. The purpose of this work is to review the ultrasound criteria for prenatal screening of this condition.*

Keywords: Agenesis of the corpus callosum, ultra-sound diagnosis, prenatal diagnosis.

1. Introduction

The corpus callosum is the main inter-hemispheric commissure that connects the two cerebral hemispheres. Its agenesis is the most frequent cerebral anomaly. Its incidence is 0.05% in the general population [1].
This anomaly corresponds to a total or partial absence of the corpus callosum. Its diagnosis is accessible at prenatal diagnosis via morphological ultrasound. The latter must look for other malformations that may be associated, notably cerebral, cardiac and facial. These etiologies can be genetic and are mostly seen in syndromic forms, or metabolic, infectious, notably CMV and rubella, or environmental and toxic, such as the consumption of cocaine or anti-epileptics [2].
The search for corpus callosum in prenatal ultrasound is not part of the standard ultrasound workup to be performed in screening according to the recommendations of the French College of Fetal Ultrasound, however, one must be attentive to the presence of indirect signs that may suspect a corpus callosum anomaly, and will impose a complementary diagnosis [1].

2. Clinical case:

It is about Mrs B.Y. 36 years old, primiparous with no notable pathological history, no notion of consanguineous marriage, no notion of known genetic abnormality in the family, the course of the current pregnancy is marked by the discovery of gestational diabetes based on a fasting blood sugar level done at the end of T1, Consulted in our training for the follow-up of her pregnancy, the patient

benefited from a biological check-up of the 1st trimester which showed a gestational diabetes diagnosed on a positive fasting glycaemia, and from a T1 ultrasound without particularity notably a normal nuchal translucency.

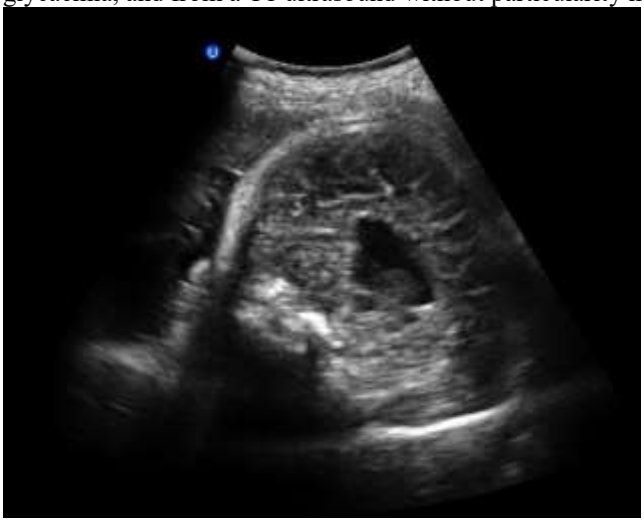

Figure 1: absence of the corpus callosum with dilatation of the 3rd ventricle and disappearance of the septal cavity, note the radial aspect of the cingulate gyrus.

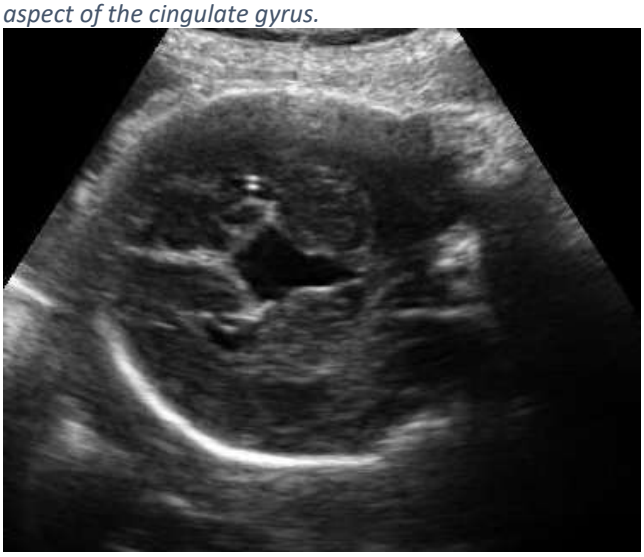

Figure 2: Bull's horn appearance: frontal horns of the lateral ventricles with a dilated V3 in the center, note the interhemispheric scissure that descends to V3 and disappearance of the cingulate gyrus.

During the 2nd trimester ultrasound performed at 26 weeks of amenorrhea, the diagnosis of agenesis of the corpus callosum was suspected in view of several direct and indirect signs: absence of visualization of the cavity of the septum pullicidum, with a 3rd

ventricle that projects forward and takes the place of the cavum of the septum pullicidum, a colpocephaly, and an abnormal course of the pericalleus artery that takes on a radial aspect.
The rest of the ultrasound does not have another morphological abnormality or intrauterine growth retardation (Figure 1,2,3,4).
During the course of the pregnancy, the patient was put on insulin therapy with hygionic dietary measures, and delivered a male infant by vaginal delivery at 38 weeks of amenorrhea.

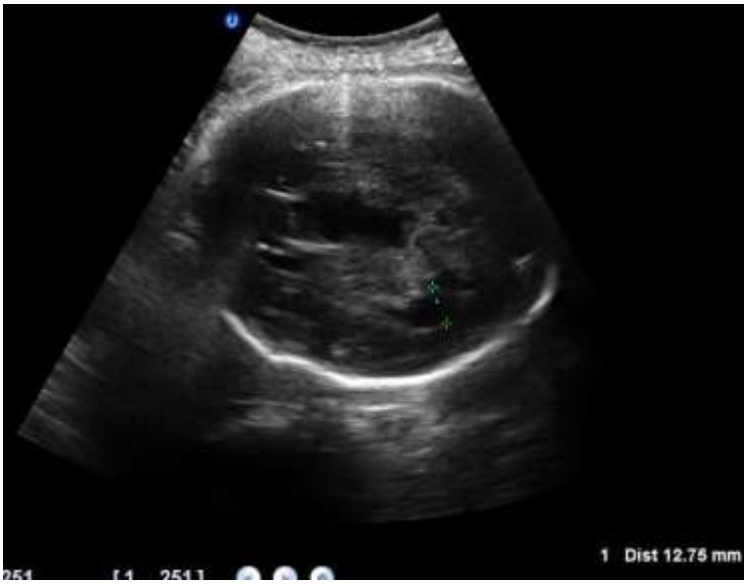

Figure 3: cephalic perimeter view: the V3 is dilated and takes the place of the septal cavity in front, and dilation of the occipital horn of the lateral ventricle: colpocephaly. Note the interhemispheric scissure that comes in contact with V3.

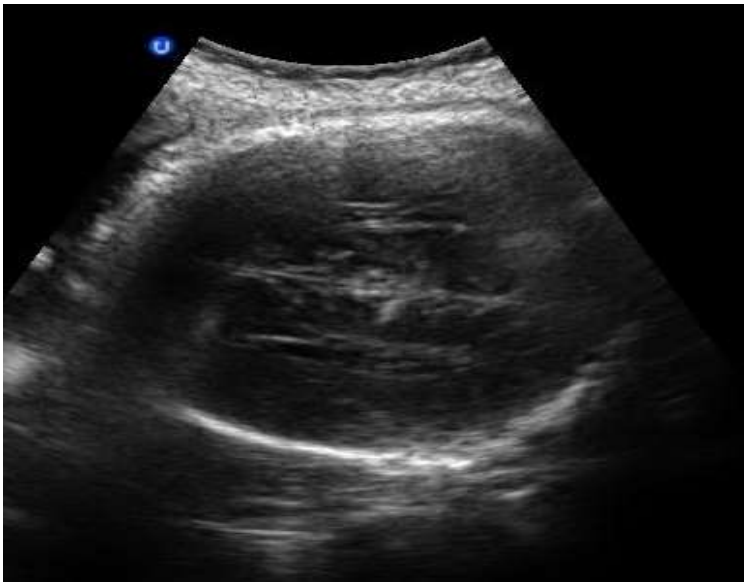

Figure 4: Triple sheet appearance on a high axial cephalic section.

3. Discussion:

The corpus callosum develops from the lamina reuniens, which corresponds to the thickening of the lamina terminalis that joins the telencephalic vesicles. The lamina reuniens appears around the 6th week and is at the origin of the corpus callosum but also of the anterior and posterior white commissures. A groove appears on the upper surface of the lamina reuniens and cells develop in this groove to form the commissural mass; nerve fibers (axons) from hemispheric neurons penetrate the commissural mass around the 11th week; fibers from the cingulate gyrus are the first to cross the midline, and serve as guides for the axons of the corpus callosum

in crossing the midline. It is conventionally accepted that the corpus callosum develops from front to back, with first the formation of the knee around the 13th week, then the body and the splenium, but the rostrum develops last during the 18th week.
Agenesis results from a defect in the formation or guidance of axons towards or beyond the midline; genetic, toxic (alcohol) or infectious (cytomegalovirus) factors can alter the development of the corpus callosum. Primitive meninges are located in a ventral groove developed at the level of the lamina reuniens [3];[4].

The current recommendations of the French College of Fetal Ultrasound for antenatal screening do not recommend the systematic realization of a sagittal section of the corpus callosum [8], however the visualization of indirect signs on routine sections, in particular that of the cranial perimeter, should raise the alarm in order to proceed with the diagnosis. On an axial slice, the absence of the septal cavity in the total form can be noticed, this will be replaced by the cavity of the 3rd ventricle which projects anteriorly and comes in contact with the interhemispheric scissure. Posteriorly and on the same section we note a dilatation of the occipital horns of the lateral ventricles or what is called colpocephaly or teardrop ventricle, while the frontal horns remain thin. If we go up a little higher in ultrasound section of the cranial perimeter, we note a triple sheet aspect which corresponds to the inter-hemispheric scissure with the scythe of the brain, this one goes down more than the normal and takes the place of the corpus callosum. On the diagnostic ultrasound, the realization of other sections to complete the exploration is necessary. On a sagittal slice we note the absence of visualization of the corpus callosum in its complete form, see a part that normally should be distal in the partial form. On the same section we note the absence of the cingulate gyrus and the pericallear artery, both of which take on a radial appearance. Finally on this section we note that the septal cavity is absent and a 3rd ventricle which is dilated. On a coronal section a typical bull's horn appearance is seen, corresponding to the frontal horns with the 3rd ventricle in the middle. The occipital ventricular horns of the lateral ventricles appear dilated. The interhemispheric fissure descends to the 3rd ventricle without interposition of the septal cavity [5];[9].
In our case, all the ultrasound signs were present and the diagnosis of agenesis of the corpus callosum was made. Then we proceeded to the search for other malformations. Syndromic associations are possible, especially at the encephalic level, such as anomalies of gyration or of the posterior cerebral fossa. The search for facial, cardiac and digestive anomalies must be done with caution [7] [8].
If there is any doubt about the ultrasound diagnosis of agenesis of the corpus callosum, which is mostly seen in partial agenesis, fetal MRI should be used. The radiological signs are often the same as those found in ultrasound but with more precision. It also allows a more detailed morphological exploration [6].
The prognosis of agenesis of the corpus callosum is better known today. In isolated primary forms, learning difficulties are encountered during childhood, and these difficulties are sometimes late and require long-term support.
In secondary forms, such as the case of interhemispheric cysts or lipomas which are responsible for a secondary destruction of the corpus callosum, the prognosis is better. They may be associated with sensory or motor disorders, more attributable to the impact of the cyst on the cortical bone than to the agenesis of the corpus callosum [10].

4. Conclusion:

Agenesis of the corpus callosum is a malformative pathology quite frequent in the fetus in comparison with other cerebral anomalies. Its diagnosis is possible in prenatal ultrasound. It is based on a careful screening ultrasound, leading to a detailed diagnostic ultrasound.
This prenatal diagnosis is of interest in order to provide the best conditions for the reception of the newborn, adapted to the presumed prognosis.

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