

Early echographic signs of the acrania-exencephaly-anencephaly sequence: case report

Sinais ecográficos precoces da sequência acrania-exencefalia-anencefalia: relato de caso

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ABSTRACT

The acrania-exencephaly-anencephaly sequence is one of the most common brain anomalies and presents well-established ultrasound signs between 11 and 14 weeks, as described in the literature. With advances in ultrasound equipment, signs of this malformation are now being described before 11 weeks. This report aims to describe a case of acrania-exencephaly-anencephaly sequence that was monitored from 4 weeks of gestation with early signs suggestive of this malformation. Changing echogenicity of the amniotic fluid and absence of the rhombencephalic cavity were detected at 8 weeks and 6 days. Acrania, undulating membrane covering amorphous neural tissue with fluid inside the cephalic cavity and elongated cephalic pole, detected at 12 weeks and 4 days, regression of fluid within the cephalic cavity and alteration of the anomalous shape of the cranial pole to a protruding type at 13 weeks and 4 days, and anencephaly at 21 weeks and 5 days. The pregnancy progressed to premature delivery at 27 weeks of gestation, with the fetus born alive but dying shortly thereafter.

Keywords: Neural Tube Defects; Anencephaly; Early Diagnosis; Case Report.

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RESUMO

A sequência acrania-exencefalia-anencefalia é uma das anomalias cerebrais mais frequentes e apresenta sinais ultrassonográficos bem estabelecidos entre 11 e 14 semanas, conforme descrito na literatura. Com a evolução dos aparelhos de ultrassonografia, já começam a se descrever sinais dessa malformação antes de 11 semanas. Assim, este relato objetiva descrever um caso da sequência acrania-exencefalia-anencefalia que foi acompanhado desde 4 semanas de gestação com sinais precoces sugestivos dessa malformação. Alterações na ecogenicidade do líquido amniótico e ausência de cavidade rombencefálica foram detectadas já com 8 semanas e 6 dias. Acrania, membrana ondulante recobrimdo tecido neural amorfo com presença de líquido dentro da cavidade cefálica e polo cefálico de formato alongado, detectados com 12 semanas e 4 dias, regressão do líquido dentro da cavidade cefálica e alteração do formato anômalo do polo craniano para o tipo protuberante com 13 semanas e 4 dias e anencefalia com 21 semanas e 5 dias. A gestação evoluiu para parto prematuro com 27 semanas de gestação, com feto nascido vivo, evoluindo para óbito logo em seguida.

Palavras-chave: Defeitos Do Tubo neural; Anencefalia; Diagnóstico Precoce; Relato De Caso.

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The authors declare no conflicts of interest

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INTRODUCTION

Anencephaly is a malformation of the central nervous system (CNS) resulting from defects in the closure of the rostral neurospore in which the calvaria is not properly formed. It has a reported incidence of 1 to 5 per 1,000 births and a mortality rate of 100% in intrauterine life or after birth^{1,2}. This neural tube defect is also described as the acrania-exencephaly-anencephaly (AEA) sequence. Acrania, which occurs 18 to 20 days after fertilization, represents the first stage of this sequence of maldevelopment. Disorganized, vasculomembranous brain tissue is then seen above the orbits, characterizing exencephaly. In evolution, due to the failure of the prosencephalon and mesencephalon to close with normal fusion at the level of the rhombencephalon and cervical spinal cord, anencephaly is seen, with the absence of the brain. The brainstem, cerebellum, and diencephalon may be present but are generally hypoplastic³.

Ultrasound is one of the most important tests for detecting anencephaly and is considered an “always detectable” malformation in the first trimester of pregnancy, usually between 11 weeks and 13 weeks and 6 days^{2,4-6}. Many studies report detection rates of 100% in the same age group¹, which means that it must be recognizable at this gestational age through systematic examination of the fetus’ head and brain, including the formation of the skull bones, choroid plexuses and ventricles, according to the guidelines of the International Society of Ultrasound in Obstetrics and Gynecology (ISUOG)^{2,3,5}. These same guidelines state that skull bone

mineralization should be demonstrated in axial and coronal sections from 11 weeks of gestation onwards, when distortion or rupture defects should be excluded¹. Most ossification points are found in the lateral parts of the frontal and parietal bones, while ossification of the calvaria may not be visible at 11 weeks in the median sagittal plane, so in order to avoid an incorrect diagnosis, it is essential to assess frontal bone ossification in axial and coronal planes^{4,7,8}. Multiplanar 3D ultrasound can be applied as a complementary diagnostic tool and the implementation of additional transvaginal access can improve detection rates at early gestational ages^{1,3,9,10}.

The main ultrasound findings include, in the coronal plane of the face, the absence of the calvaria above the orbits, a “Mickey-Mouse” bilobular face (brain tissue/cerebellar hemispheres visible in the coronal plane with the appearance of two semicircular structures hovering over the surface of the fetus, falling to the side of the head, similar to the rounded ears of “Mickey Mouse”), increased echogenicity of the amniotic fluid (seen in up to 89% of cases with gestational age above 11 weeks), the frog face, or “frog eyes”, cystic morphology of the head, which can be elongated, irregular and protruding^{1-3,8,11}.

A retrospective, descriptive, multicenter cohort study, which included 88 cases of the AEA sequence, aimed to classify different phenotypic appearances of this malformation in the first trimester of pregnancy and assess the feasibility of this classification with the aim of facilitating and increasing detection rates in early pregnancy and thus making management timely, safer, and less traumatic. The abdominal approach was complemented by the transvaginal

approach in 95.5% of cases. This sequence was classified by two independent observers as bulging/salient, elongated, bilobular, cystic, shortened, and irregular. Interobserver reliability had an interclass correlation coefficient of 0.903, which was considered good. The most common subtype was overhanging (31%), followed by elongated (25%), bilobular (19%), cystic (11%), shortened (8%), and irregular (6%)¹.

Most undiagnosed cases of anencephaly are at gestations of less than 11 weeks, where detection rates can vary between 69 and 89% among non-specialists³. Thus, there is currently a search for an even earlier diagnosis, with a crown-rump length (CRL) of less than 45mm, with this study describing, in addition to the classic signs described between 11-13 weeks of gestation, of the AEA sequence, ultrasound findings before 11 weeks, consistent with those reported by other authors¹²⁻¹⁶, contributing to the description of the early findings of the AEA sequence, as well as the phenotypic changes that this pathology can present throughout its evolution.

METHODS

The information contained in this case report was obtained by reviewing the medical records, photographic records of the 2D and 3D ultrasound diagnostic imaging methods, and laboratory data to which the patient was submitted. The Case Reports guidelines (CARE) were used to write this report. This project complies with Resolution No. 466/2012 of the *Conselho Nacional de Saúde* and the determinations of the Declaration of Helsinki and was submitted to the Research Ethics Committee of the *Universidade Federal do Delta do Parnaíba* (UFDPa) (CAAE 74859223.6.0000.0192) and approved in accordance with substantiated opinion 6.566.527, with the data collected after the patient had signed the Informed Consent Form (ICF).

CASE REPORT

Patient, 23 years old, nulliparous, brown, supermarket cashier operator, from Parnaíba (PI). The patient had a history of polycystic ovary syndrome, with complaints of oligomenorrhea and acne. She reported that she had been using contraceptives for a year and had stopped about 8 months before the medical appointment due to her desire to become pregnant. She also had a fasting blood glucose of 108.8mg/dl in the month of conception, with a previous diagnosis of pre-diabetes and insulin resistance.

Transvaginal ultrasound, performed approximately two weeks before conception, showed enlarged ovaries with peripheral follicles (consistent with polycystic ovaries), central hyperechogenicity, and endometrial thickness of 8.4mm. Due to persistent menstrual delay, a new ultrasound examination was performed 55 days after the first one, whose findings were a gestational sac with an average diameter of 0.69cm, consistent with 5 weeks and 3 days. However, based on the date of the last menstrual period (LMP), it would be 7 weeks and 6 days, with a yolk sac visualized, but without an embryo.

Following up 25 days after the previous exam, a new ultrasonography revealed a gestational sac with an average

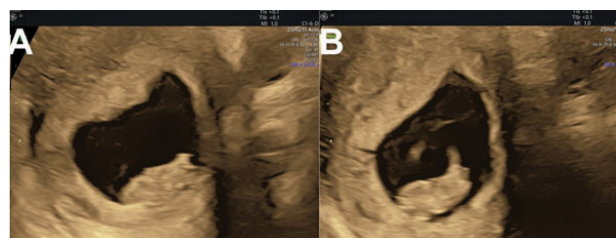


Figure 1. Second transvaginal ultrasound scan at 8 weeks and 6 days, dated by ultrasound. **A.** Echogenic changes in the amniotic cavity, with debris; **B.** Absence of rhombencephalic cavity in the cephalic pole.

Source: Author's archives (2023).

diameter of 3.3cm, now with the embryo present and a crown-rump length (CRL) of 21.5mm, consistent with 8 weeks and 6 days (according to the LMP, it would be 11 weeks and 3 days). In this examination, as described in Figure 1, low amplitude echoes were noted in the amniotic fluid and the cephalic pole was abnormally shaped due to the absence of the rhombencephalic cavity, a structure that should have been visible at 8 weeks' gestation (Figure 2).

During first trimester morphology, the fetus had a crown-rump length of 60.6mm, compatible with 12 weeks and 4 days (15 weeks and 1 day by LMP dating). In addition, the fetus had a nuchal translucency (NT) of 1.7mm, a nasal bone present and a tricuspid valve without regurgitation. However, the cephalic pole was without the skull cap and with an undulating membrane covering amorphous neural tissue, as shown in Figure 3.

In a new examination of the fetus a week later, therefore, at 13 weeks and 4 days, dated by the CRL, the cephalic pole showed a change in shape and a reduction in the anechoic area around the neural tissue (Figure 4), with three-dimensional ultrasound (Figure 5) allowing better visualization of the cephalic pole malformation and the "beret sign".



Figure 2. Normal embryo showing the rhombencephalic cavity as a cystic structure in the cephalic pole, as expected for the gestational age.

Source: Author's archives (2023).

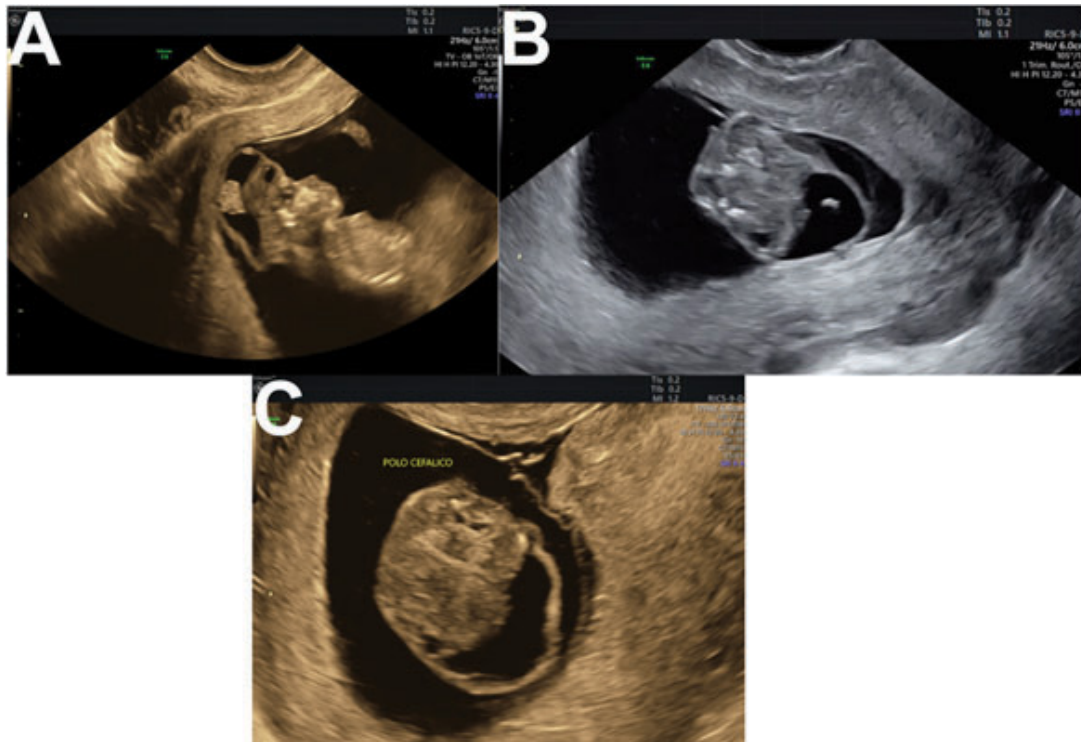


Figure 3. Fetus at 12 weeks and 4 days, showing absence of the cranial vault, undulating membrane covering neural tissue surrounded by anechoic area (cerebrospinal fluid). Sections: **A.** Sagittal; **B.** Coronal; **C.** Transverse.

Source: Author's archives (2023).

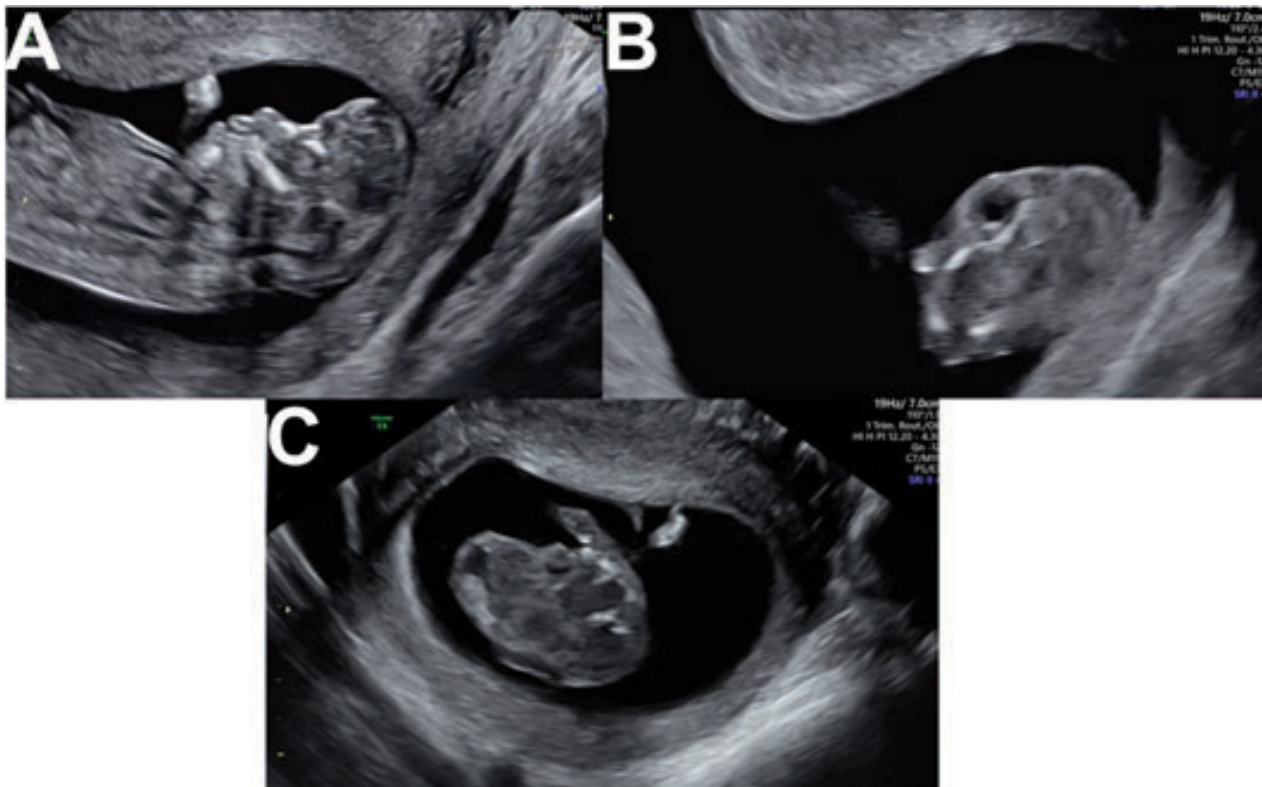


Figure 4. Fetus at 13 weeks and 4 days, showing absence of the cranial vault, with an undulating membrane covering the neural tissue. Note the disappearance of the anechoic area (cerebrospinal fluid, which surrounded the neural tissue in the previous exam). Sections: **A.** Sagittal; **B.** Coronal; **C.** Transverse.

Source: Author's archives (2023).

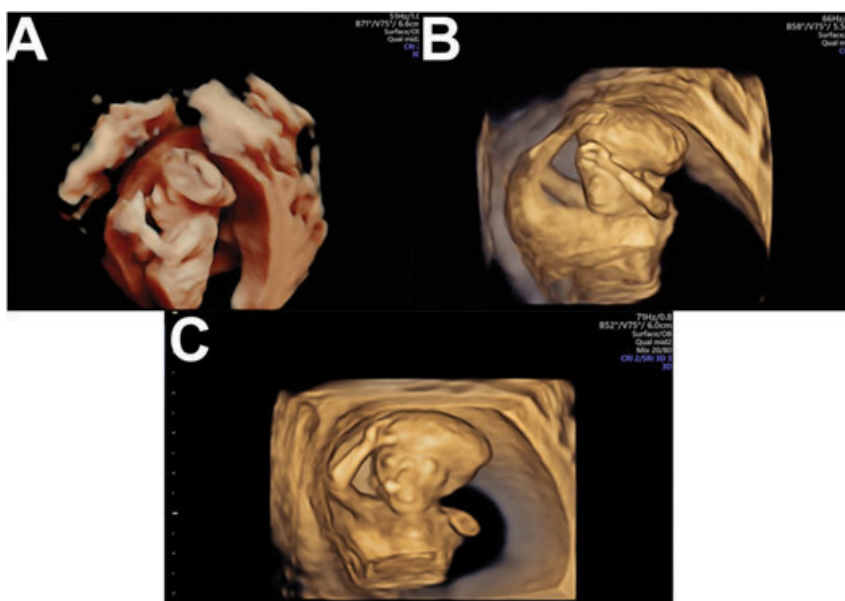


Figure 5. Three-dimensional ultrasound images of the fetus at 13 weeks showing the abnormal morphology of the cephalic segment, with the appearance of the “beret sign”. This examination can help to exclude differential diagnoses such as holoprosencephaly and encephalocele.

Source: Author’s archives (2023).

With the diagnosis confirmed, the patient decided to continue with the pregnancy. The 2nd trimester morphology scan showed the malformation already in the anencephalic state, with the absence of the calvaria and brain parenchyma, as shown in Figure 6. Labor was premature at 27 weeks’ gestation. The newborn weighed 570 grams, was born alive, but died within a few minutes.

DISCUSSION

The diagnosis of the AEA sequence between 11 and 13 weeks of gestation is already well established in the literature, but attempts are currently being made to establish even earlier echographic signs of this pathology, such as

increased echogenicity of the amniotic fluid, a finding present and visualized in this case, reported at 8 weeks and 6 days (Figure 2). Although this alteration is not a specific finding for anencephaly, its presence is usually high in cases of the acrania-exencephaly-anencephaly sequence and has been reported by several authors, who describe, in addition to abnormalities of the cephalic pole with the “Mickey Mouse” sign, hyperechogenicity of the amniotic fluid^{3,9,10,12,13}.

Increased echogenicity of the amniotic fluid is a warning sign of neural tube malformations and therefore a possible marker for identifying the AEA sequence in the first trimester assessment. Furthermore, the presence of echogenic changes in the amniotic fluid reinforces the hypothesis that anencephaly is the final pathway of acrania, with the changes

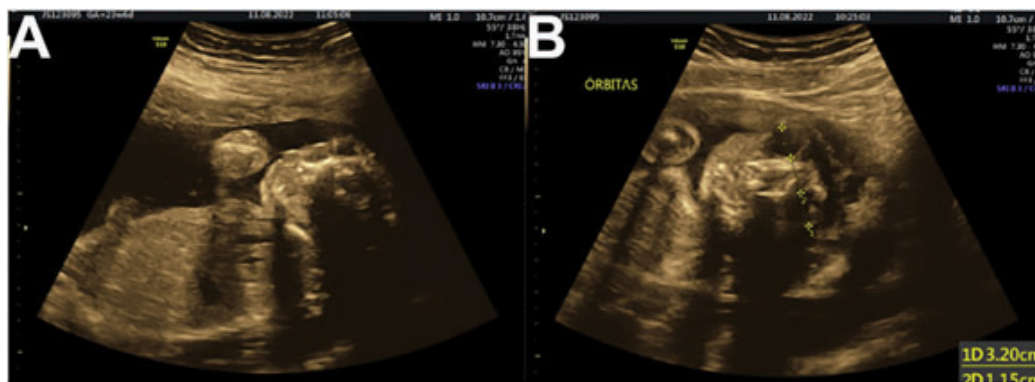


Figure 6. Morphology of the second trimester (gestational age 21 weeks and 5 days), showing acrania and anencephaly. Sections: **A.** Sagittal; **B.** Coronal.

Source: Author’s archives (2023).

in the amniotic fluid probably due to debris of neural tissue caused by the physical and chemical aggression inflicted on the unprotected cephalic pole¹³. Although we can't close the diagnosis at this gestational age with this sign, it is a warning and an indication for ultrasound control to define the case.

In the case described, at week 8 of gestation (21.5mm CRL), when the embryo's cephalic region was analyzed, no anechoic areas were visualized (Figures 2A and 2B), as expected for this gestational age, such as the rhombencephalic cavity (Figure 1). The failure to visualize cystic brain structures at this gestational age has not yet been described as an echographic sign of the AEA sequence; however, sonoembryology describes the need to visualize these structures, as they are part of the normal development of the embryo. This agrees with what Rolnik et al. (2020)¹⁰ says, that abnormalities in the central nervous system can be suspected at 8 weeks of gestation when analyzing the brain cavities, telencephalon, diencephalon, mesencephalon, and rhombencephalon.

It was also possible to see the classic signs of the AEA sequence and different phenotypic patterns, such as the absence of the calvaria (acrania) and an amorphous neural mass (exencephaly) covered by an undulating membrane with fluid around it (Figure 3). These findings agree with those reported by Szkodziak et al. (2020)⁸, according to whom, in these cases, the brain is covered by a thin layer of ectoderm, leaving the nervous tissue vulnerable to destruction. Wertaschnigg et al. (2020)¹ add that the abnormal cephalic pole, at this gestational age, can take on different phenotypes, such as overhanging, elongated, bilobular, cystic, shortened, and irregular. During follow-ups, these subtypes can vary, although he was unable to demonstrate this in his study, given the interruptions in pregnancies after diagnosis. This

hypothesis, however, was confirmed in the present report, since at 12 weeks, the ultrasound features characterized the elongated phenotypic type of the AEA sequence (Figures 7A and 7B), the second most frequent variation. However, one week later, when the ultrasound was checked, there was a regression of the fluid around the neural mass and a change in the phenotypic pattern, with the fetus presenting the overhanging type (Figures 7C and 7D). Thus, in this case, it was documented that the phenotypic pattern of the acrania-exencephaly-anencephaly sequence can change over the weeks, and therefore, knowing all the possible types can help in making an accurate diagnosis.

A 3D ultrasound can prevent diagnostic errors by complementing the analysis of the cranial vault, helping to exclude differential diagnoses with early-stage AEA, such as holoprosencephaly and encephalocele, substantially improving accuracy and patients' understanding of the malformation^{3,9,10,17}. In this report, the 3D images showed the "beret sign" (Figure 5). According to Szkodziak et al. (2020)⁸, in a retrospective observational study of two American centers (including 10 cases of the AEA sequence out of a total of 4,060, with pregnant women ranging from 20 and 37 years of age between the 12th and 16th week of gestation), this finding is defined as brain structures surrounded by a thin undulating membrane at the site of the cranial vault, an anechoic space between this membrane and the remaining brain structures, corresponding to the cerebrospinal fluid, with a non-visible cerebral sickle. These ultrasound signs were described in the sagittal and coronal planes.

It has been described that, in cases of anencephaly, there may be a difference of two weeks less in the dating of gestational age when the crown-rump length is used as a parameter, due to alterations in the cephalic region².

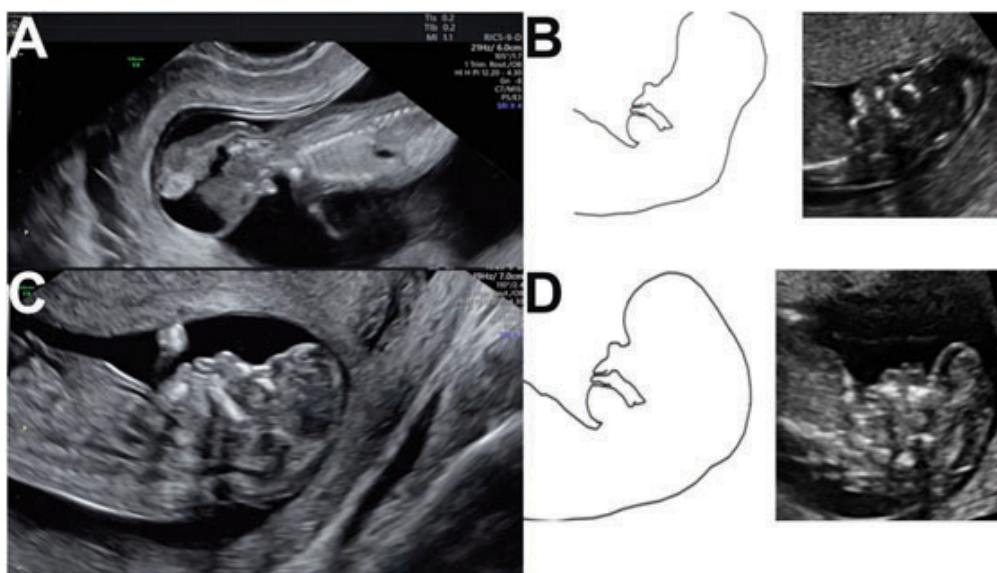


Figure 7. A. Evolution of the phenotypic pattern of the acrania-exencephaly-anencephaly sequence from elongated at 12 weeks, observed by the author and in B. By Wertaschnigg et al. (2020)¹; C. To overhanging at 13 weeks, observed by the author and in D. By Wertaschnigg et al. (2020)¹.

Source: Author's archives (2023) and Wertaschnigg et al. (2020)¹.

Although this was observed in the case described, where the CRL showed the patient to be two weeks and three days younger than the LMP, the patient had a preconception history of polycystic ovaries on ultrasound and oligomenorrhea, which makes it difficult to use her last menstrual period as a parameter for dating gestational age. It should also be reiterated that, in the early stages, the crown-rump length may be normal in fetuses that have developed anencephaly, given the still small degree of destruction of the cephalic pole¹².

Although she chose not to terminate the pregnancy, this was her right. In 2012, the *Suprema Corte Federal* (Federal Supreme Court) ruled that aborting an anencephalic fetus is not a crime. With this demand, the *Conselho Federal de Medicina* (Federal Council of Medicine) approved resolution 1,989/2012 defining guidelines for the diagnosis of this malformation. Thus, for the diagnosis of anencephaly and subsequent legal claim for abortion, an ultrasound examination must be carried out after the twelfth week of pregnancy, and the report must be signed by two qualified doctors. The pregnant woman must be provided with all the information and clarification necessary to make her decision, always guaranteeing her right to freely decide on the course of action to be adopted¹⁸.

Patients who choose not to terminate the pregnancy, as in this case, should be referred to high-risk prenatal care and monitored regularly. Psychological support for the mother and her family should also be offered. The risk of recurrence in future pregnancies should be informed to the mother and is around fifty times higher. Pre-conceptual use of folic acid can halve this risk and should be provided to the patient¹⁶. The World Health Organization recommends daily supplementation with 0.4 mg of folic acid before pregnancy in all women of childbearing age to prevent neural tube defects, but in cases where the woman has a previous history of anencephalic pregnancy or has a first-degree relative affected by the disease, anencephaly prevention should be carried out with oral supplementation of 4.0 mg of folic acid daily three months before conception and maintained during the first trimester of pregnancy¹⁹.

CONCLUSION

This case report followed the echographic evolution of the acrania-exencephaly-anencephaly sequence in the 1st and 2nd trimesters, in addition to the classic findings between 11 and 13 weeks of gestation such as absence of the skull, amorphous neural mass, it was possible to analyze even earlier echographic signs of an 8-week embryo, such as increased echogenicity of the amniotic fluid, discordance in gestational age by dating the CRL and LMP and abnormal morphology of the cephalic segment, with no visualization of the rhombencephalic cavity. The different possible presentations and phenotypic evolutions of this sequence can also be documented in the same case, including three-dimensional rendered images.

AUTHORS' CONTRIBUTION:

We describe contributions to the paper using the taxonomy (CRediT) provided below: Conceptualization, Data curation, *Formal analysis, Methodology, Supervision, Writing – review and editing*: LC Oliveira. *Formal analysis, Research, Methodology, Validation, Visualization, Writing – original draft, Writing – review and editing*: LCR da Silva; *Research, Writing – review and editing*: LRR da Silva; G de A Carvalho; AS Dourado; EB Viana.

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