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PRENATAL ULTRASOUND FINDINGS IN DANDY WALKER VARIANT

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ABSTRACT

The Dandy-Walker syndrome is a rare congenital malformation of the central nervous system that comprises a spectrum of posterior fossa abnormalities. We are presenting a rare phenotypic variety of Dandy-Walker variant highlighting the importance of the prenatal ultrasound investigation in the diagnosis and specialized care of newborns with severe congenital neurological disorders.

KEYWORDS: Dandy-Walker, cerebellar vermis, ultrasound examination, prenatal diagnosis.

INTRODUCTION

The Dandy-Walker syndrome is a rare congenital malformation of the central nervous system that comprises a spectrum of abnormalities of the posterior fossa which are classified in three versions: classical Dandy-Walker malformation (cystic dilatation of the fourth ventricle, complete or partial agenesis of the cerebellar vermis and an enlarged posterior fossa), Dandy-Walker variant (variable hypoplasia of cerebellar vermis, with or without enlargement of the posterior fossa), and (c) megacisterna magna (enlarged cisterna magna, keeping the integrity of the cerebellar vermis and of the fourth ventricle).[1-4]

The condition is uncommon, the incidence is unknown, and about 50% of the cases of Dandy-Walker malformation are associated with other non-cerebral anomalies.[5,6]

The present study aimed to highlights the benefits of ultrasound examination in the prenatal diagnosis and management of the Dandy-Walker variant.

MATERIALS AND METHODS

The patient AB, aged 32 years, comes to the A.S. Medical Center, from Bucharest, Romania in her 35-week pregnancy for a fetal ultrasound scan.

The ultrasound investigation was conducted by the Division of Maternal-Fetal Medicine and Obstetrics of our medical center, in accordance with the Declaration of Helsinki - Ethical Principles and Good Clinical Practices and after acquiring the patient's informed consent, and performed using a General Electric Voluson E10 ultrasound machine.

RESULTS

The non-invasive prenatal test, Verifi Plus Prenatal Test, made in the first trimester of pregnancy, was negative. Sonographic scan highlighted a singleton fetus 35 weeks old, in evolution, with an estimated fetal weight of 3146 g. Fetal head ultrasound biometry showed biparietal diameter: 89.5 mm, occipitofrontal diameter: 114.3 mm, and head circumference: 321.6 mm (Fig. 1).

Sonographic examination of the fetal brain showed cerebellum: 49.9 mm, with abnormal structure and configuration, vermis hypoplasia: 13 mm with Dandy-Walker variant aspect, symmetrical cerebral hemispheres with normal configuration, normal septum pellucidum, normal corpus callosum, and fissura sylvii, homogeneous choroid plexus, and normal spectrum of the blood flow in the middle cerebral artery: peak systolic velocity 58.28cm/s, end-diastolic velocity 13.87cm/s, systolic/diastolic ratio 4.2, pulsatility index 1.34, and resistivity index 0.76 (Fig. 2 and Fig. 3). No other fetal craniofacial dysmorphisms were found (Fig. 4).

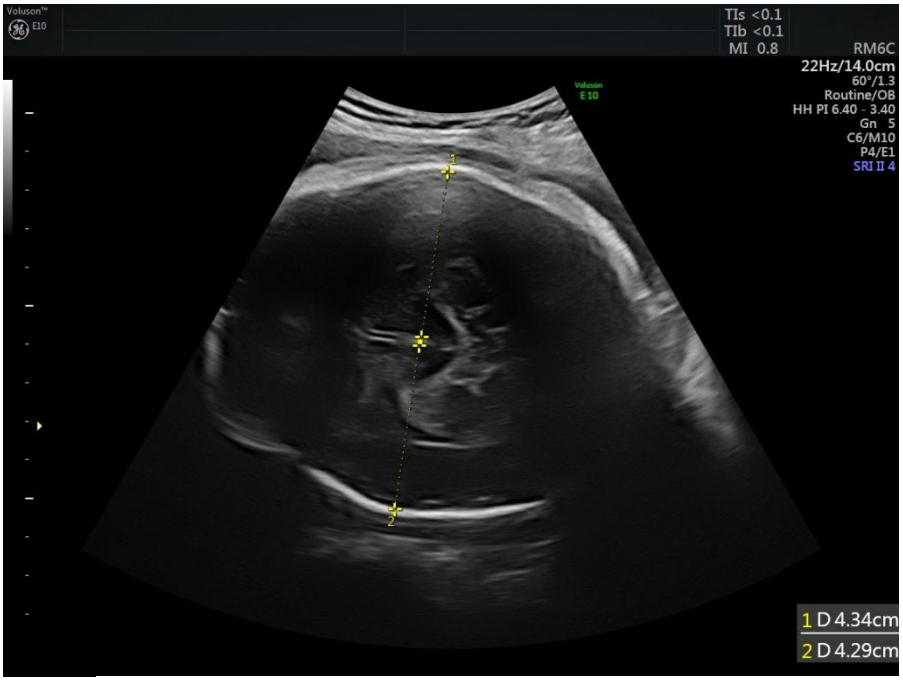

Figure 1: Fetal head biometry: the measurement of the biparietal diameter (BPD).

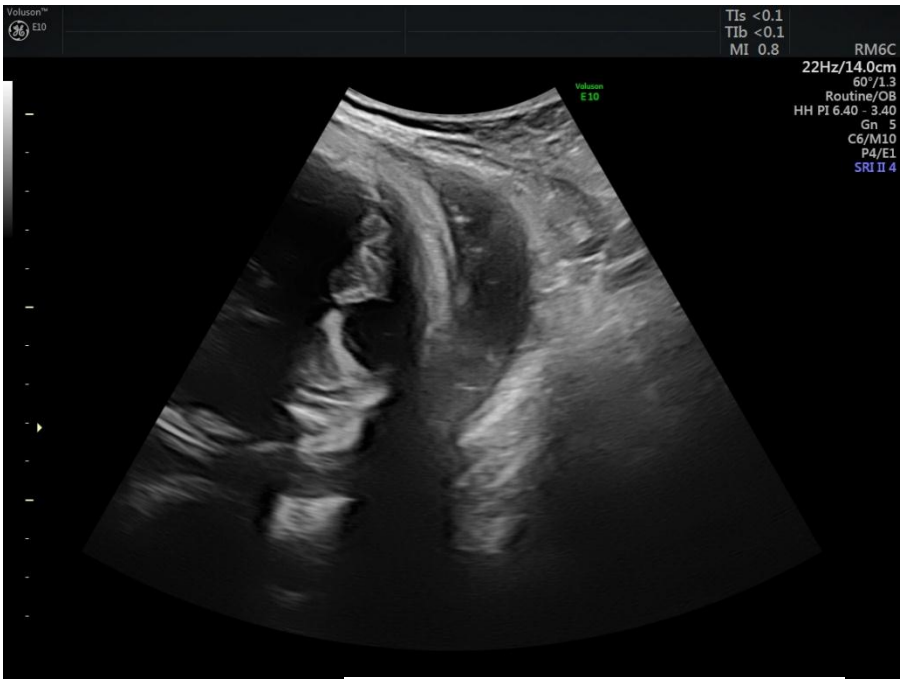

Figure 2: 2D Ultrasound fetal scan showing a Dandy-Walker variant.

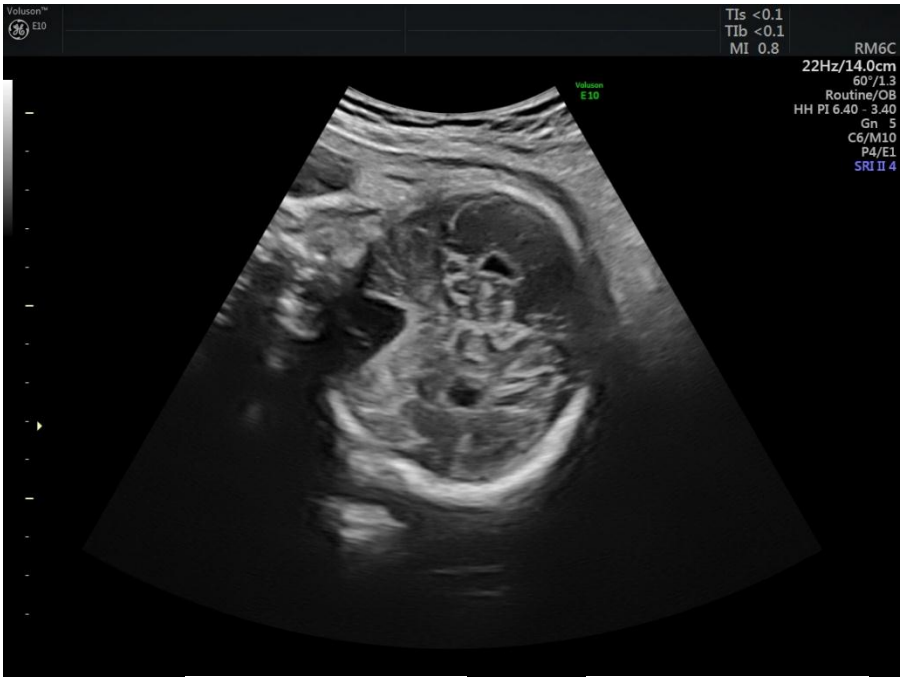

Figure 3: 2D Ultrasound image of the fetal brain: Dandy-Walker variant.

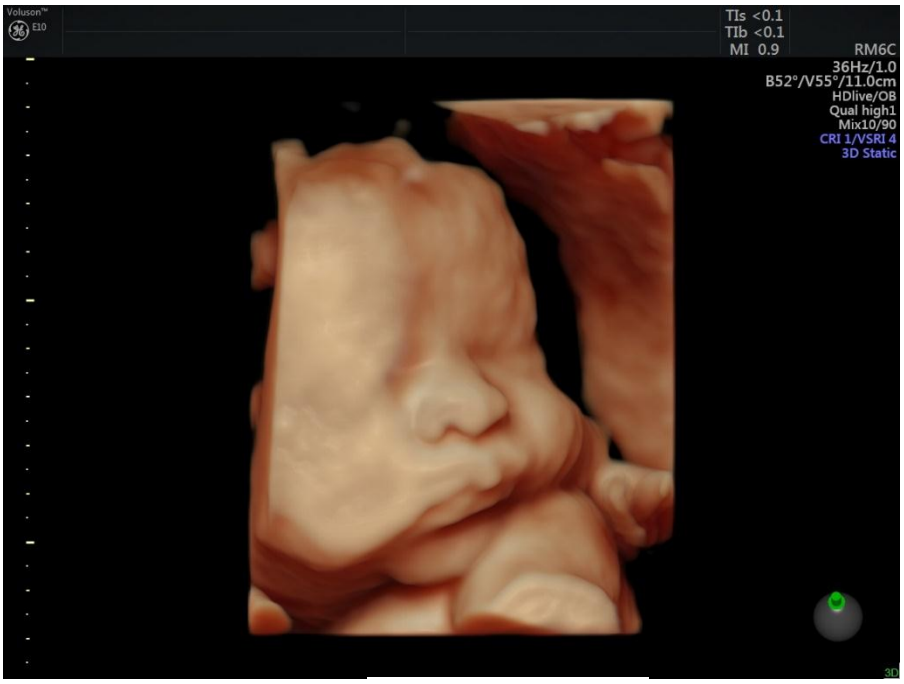

Figure 3: 3D Static Ultrasound image of the fetal face.

The ultrasound scan of the thorax, abdomen, and limbs showed apparently normal shape and structure. No other fetal dysmorphisms over 0.5 cm were found.

On the basis of the fetal morphology scan the following diagnosis was established: Mono-fetal pregnancy 35.2 weeks' of gestation (chronologic) / 36.3 weeks' of gestation (biometric), in evolution with malformed fetus: cerebellar vermis hypoplasia, Dandy-Walker variant.

The parents were informed about the severity of fetal malformations and were referred for a pediatric neurology consultation.

The baby was born at term by cesarean section and was transferred to the neonatal intensive care unit to be monitored and receive specialized care for a newborn with neurological disorders.

DISCUSSION

Dandy–Walker syndrome has been known for more than a century; however, many aspects (aetiology, biology,

types and outcome) are not well understood, and the severity depends on the presence of associated anomalies.[7-9]

The syndromes associated with Dandy–Walker syndrome include posterior fossa brain malformations, hemangiomas, arterial anomalies, coarctation of the aorta and cardiac defects, and eye abnormalities and Ellis–van Creveld syndrome.[10-11]

Early antenatal diagnosis of Dandy-Walker syndrome may improve the prognosis and reduce the mortality, which has been reported to be 30-48%.[12-15]

CONCLUSION

The rare phenotypic variety of serious congenital malformation of the central nervous system, which associates cerebellar vermis hypoplasia, Dandy-Walker variant, highlights the importance of the prenatal ultrasound investigation in the diagnosis and specialized care of newborns with severe congenital neurological disorders.

Authors' contributions

All authors contributed equally with the first-author, in the preparing, review and editing of the article. All authors read and approved the final version of the manuscript.

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