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Case Report

# Fetal magnetic resonance imaging findings of Dandy-Walker malformation in monochorionic diamniotic twins

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**Abstract.** Fetal magnetic resonance imaging (MRI) findings of Dandy-Walker malformation are presented in twins at 22 weeks of gestation. The mother was 22 years-old. Prenatal ultrasonography at 18 weeks revealed mega cisterna magna in one fetus and a borderline cisterna magna in the other fetus of monochorionic diamniotic twins. Fetal MRI findings were consistent with Dandy-Walker Syndrome in one and Dandy-Walker variant in the other fetus. Dandy-Walker malformation affecting both fetuses of the twins is extremely rare. Although ultrasonography is the primary modality for the diagnosis, fetal MRI is an important tool to confirm the diagnosis, to describe the anatomic features in detail and to identify associated anomalies.

Keywords: Fetal magnetic resonance imaging, Dandy-Walker malformation, monochorionic diamniotic twins

## 1. Introduction

Dandy-Walker syndrome (DWS) is a developmental malformation of the central nervous system characterized by complete or partial absence of the cerebellar vermis, the presence of a posterior fossa cyst, hypoplasia and dysplasia of the cerebellar hemispheres, upward displacement of the tentorium and ventriculomegaly [1–3]. Prenatal diagnosis is possible by ultrasonography (US) and magnetic resonance imaging (MRI). Fetal MRI reveals more information about the anatomy and anomalies [4,5]. In this paper we present our MRI findings of DWS in monochorionic diamniotic twins.

## 2. Case report

A 22 year-old woman (gravida 3, para 0, abortus 2) at 18 weeks of gestation (WG) was referred to our radiology department with a diagnosis of mega cistern magna in both fetuses of monochorionic diamniotic twins. A single placenta was identified; \a double artery and a single vein in the umbilical cord were noted on color Doppler US. At 22 WG, MRI of the fetuses was performed on a 1.5 T MRI equipped with a synergy body coil. Coronal, sagittal and axial

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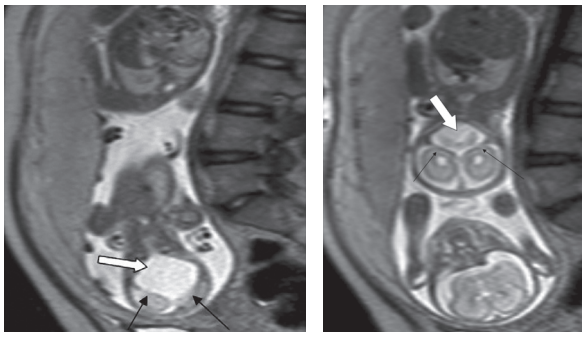
images were obtained using T2-weighted single-shot sequences to establish the diagnosis, to better delineate the anatomic features and to assess for further probable anomalies. Twins were visualized successfully. Both fetuses were male. In one fetus, a huge posterior fossa cyst communicating with 4th ventricle along with elevated tentorium cerebella and enlarged posterior cranial fossa were seen (Fig. 1a). Cerebellar hemispheres were hypoplastic and laterally displaced (Fig. 2). In the other fetus there was a posterior fossa cyst which communicated with the 4th ventricle, the cerebellar hemispheres were hypoplastic but differently from the other tentorium, posterior fossa volumes were normal in this case (Fig. 1b and 3). The Inferior vermis was agenetic in both fetuses. Thus we diagnosed DWS for the one twin, and a Dandy-Walker variant (DWV) for the other fetus.



(a) (b)

Fig. 1. Fetal MRI of twin pregnancy T2 weighted image coronal plane (a) Large posterior fossa cyst in the lower located fetal cranium (white arrow). Note that tentorium cerebella is elevated and posterior cranial fossa is enlarged (black arrows). (b) A posterior fossa cyst is also seen in the upper located fetus (white arrow). Tentorium and posterior fossa volumes are normal in this case (black arrows).

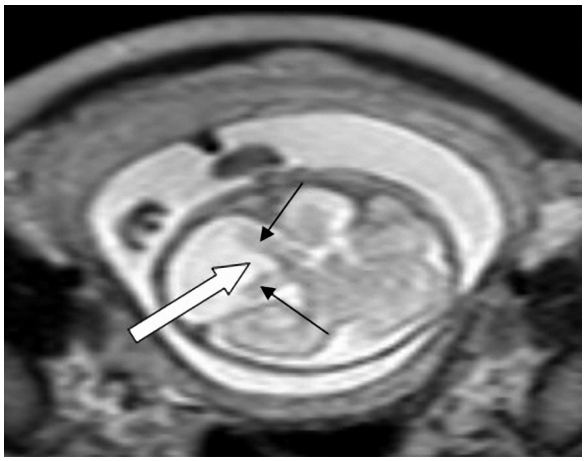


Fig. 2. Axial T2 weighted image from the posterior fossa of the lower located fetus reveals the cyst communicates with the 4th ventricle (white arrow). Cerebellar hemispheres are hypoplastic and laterally displaced.

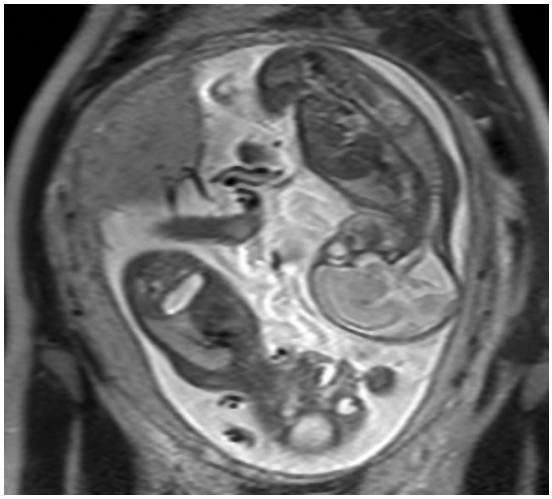


Fig. 3. Sagittal T2 weighted image of the upper located fetus reveals the posterior fossa cyst communicates with 4th ventricle, the inferior vermis is agenetic and cerebellar hemispheres are hypoplastic.

## 3. Discussion

Dandy-Walker malformation (DWM) has been recently described under a new term, the Dandy-Walker complex: a continuum of overlapping developmental anomalies that include DWS, DWV, the mega cisterna magna, and retrocerebellar arachnoid cysts of the posterior cranial fossa, which must be differentiated from each other [1–6]. US is the basic diagnostic modality and MRI is used to further describe the anomalies. Diagnosis is usually made at 18–22 wk. Since the cerebellar vermis is not fully formed until 20 wk, one must care not to over-diagnose this entity [1–7]. The incidence of DWM is reported to range between 1/25 000 to 1/35 000 births [8].

Embryological aspect reveals a disruption of neural tube just closing on the 5th wk of gestation, along with the developmental abnormalities of the cerebellar vermis and the roof of the 4th ventricle [1,4].

US findings of DWS include a large cystic mass of the posterior cranial fossa that communicates with a dilated 4th ventricle as the cardinal finding. There is also

aplasia of the cerebellar vermis, whiich is rotated in the direction of the quadrigeminal plate,and hypoplasia and dysplasia of the cerebellar hemispheres to varying degrees [1]. The tentorium cerebella, straight sinus, confluence of sinuses, and transverse sinus are elevated so the posterior cranial fossa is enlarged and the hypoplastic and dysplastic cerebellar hemispheres are displaced cranially and laterally. The brain stem can be displaced anteriorly and can be hypoplastic. [4,5]. In our case enlarged posterior fossa and a cyst communicating with $4^{th}$ ventricle along with elevated tentorium, hypoplastic and laterally displaced cerebellar hemispheres and agenesis of inferior vermis were seen in one of the fetus, leading to the diagnosis of DWS.

Associated malformations, mostly central nervous system, include agenesis or dysgenesis of the corpus callosum, occasionally extracranial malformations. Seldom, syndromes such as Klippel-Feil syndrome, trisomy 13 and 18 can be seen [1,3]. In our case there were no associated malformations in the twins.

Dandy-Walker variant is a less severe form in which the posterior cranial fossa is not enlarged and the cerebellar vermis hypoplasia and dysplasia are less pronounced, with normally developed cerebellar hemispheres. The $4^{th}$ ventricle is slightly dilated but communicates with the retrocerebellar cyst [4,5].

In the other fetus of our case there was a posterior fossa cyst that was communicating with the $4^{th}$ ventricle, the cerebellar hemispheres were hypoplastic, but differently from the other fetus, tentorium and posterior fossa volumes were normal. Thus diagnosis was as DWV for the second fetus.

Retrocerebellar arachnoid cyst of the posterior cranial fossa is characterized by a large retrocerebellar cystic cavity that compresses the cerebellum and displaces it rostrally. The mega cisterna magna is a normal variant. It is characterized by a fluid-filled cavity located under the cerebellum and behind the medulla oblongata. In both conditions the posterior cranial fossa is not enlarged and there is no communication between the cyst and the $4^{th}$ ventricle [3–5].

Kontopoulo et al. [3] studied the data on 660 patients referred for consultation with complicated monochorionic twin pregnancies. DWS was seen in 10 (1.5%) patients, all cases were isolated DWS and none had a history of DWS in prior pregnancies.

The incidence of DWS of complicated monochorionic twins was reported approximately 200 times higher than expected for the general population [3,9].

DWS is reported to be of sporadic inheritance, although some cases have been associated with congenital infection, genetic syndromes, or with Mendelian and chromosomal disorders [3,8]. Actual pathophysiologic etiology of this complex anomaly is yet unknown.

Cowles et al. [10] reported a DWS diagnosed on 33-WG of a male fetus whose pedigree analysis revealed a family history of isolated DWM in three other males, suggesting an X-linked recessive inheritance pattern.

Ulm et al. [11] reported a family with recurrent DWM. The first offspring had isolated DWM and the subsequent pregnancy revealed isolated DWM in both dizygotic twins. It was reported that chromosome analysis was normal in all three infants, and autopsy confirmed that no other congenital abnormalities. They concluded that evidence suggests that rare families transmit the disorder in an autosomal or X-linked recessive pattern, with a high recurrence risk. Since both of the fetuses in our case were males, one may assume that there might be X-linked recessive pattern. Genetic analysis was not performed since the family refused to do so.

In conclusion DWM is a developmental malformation of the central nervous, which is extremely rare in both fetuses in twins. Diagnoses via US is possible, MRI is required for further assessment.

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