Radiology Case Reports 19 (2024) 4940–4944

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

A rare case report: The value of fetal MRI to detect diprosopus twins ☆

Utami Purbasari, MD, PhD[a,b,*], *Dewi Asih, MD*[c], *Helda H, MD, PhD*[b], *Reza Tigor Manurung, MD*[d], *Puspa Dewi, MD*[a], *Agnes Nina Eureka, MD, MPH*[b]

[a] *Department of Radiology, Fatmawati General Hospital, Jl. RS. Fatmawati Raya, Num4, RT.4/RW.9, West Cilandak, South Jakarta 12430, Indonesia*

[b] *Department of Clinical Epidemiology, Faculty of Public Health, University of Indonesia, Jl. Lingkar Kampus Raya Universitas, Depok, West Java 16424, Indonesia*

[c] *Department of Radiology, University of Prima Indonesia, Jl. Ayahanda No.68a, Sei Putih Tengah, District Medan Petisah, Medan, North Sumatra 20118, Indonesia*

[d] *Department of Obstetrics and Gynecology, Fatmawati General Hospital, Jl. RS. Fatmawati Raya, Num4, RT.4/RW.9, West Cilandak, South Jakarta 12430, Indonesia*

ARTICLE INFO

Article history:
Received 17 June 2024
Revised 9 July 2024
Accepted 11 July 2024

Keywords:
Diprosopus
Fetal MRI
Antenatal diagnosis
Congenital anomalies
Conjoined twins

ABSTRACT

Diprosopus is one of the rarest types of conjoined twins, caused by incomplete zygote separation in early pregnancy. It defines a condition with duplication of facial structures, monocephalic and 1 trunk. Early detection is difficult, but fetal MRI plays an important role in strengthening antenatal diagnosis of conjoined twin pregnancies and other major congenital abnormalities, complementing antenatal ultrasonography. A 28-year-old patient (G2P1A0) was referred from the regional general hospital for suspected malformations, including Dandy-Walker syndrome and a small mandible Antenatal 3-D ultrasound at 35 weeks revealed a single baby with macrosomia, hypoplasia of the vermis, and cleft lip with malformation of facial structures. A 3 Tesla MRI (Signa, GE Healthcare) revealed various developmental brain anomalies, including duplication of the frontotemporal lobes, corpus callosum agenesis, and small posterior fossa. The identification of 4 orbital structures raised suspicions of face duplication. This patient underwent a caesarean section and delivered a diprosopus twin baby. MRI emerged as an indispensable adjunct, complementing ultrasound in detecting congenital malformations. The success of this approach emphasizes collaborative efforts between clinicians and radiologists for accurate identification and management of complex fetal anomalies.

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

* Corresponding author.
E-mail address: upurbasari.1002@gmail.com (U. Purbasari).

https://doi.org/10.1016/j.radcr.2024.07.055

Introduction

Diprosopus is the term used to describe the duplication of facial structures. It is a very rare congenital condition with an incidence rate of 1 case in 180,000 to 15,000,000 births [1]. Diprosopus is characterized by craniofacial duplication, which may involve partial or complete duplication. There are 4 types of diprosopus, as described by Gorlin et al.: a single mouth with duplication of the maxillary arch, a supernumerary mouth laterally placed with rudimentary segments, a single mouth with replication of the mandibular segments, and true facial duplication [1–4].

In the contemporary realm of medical imaging, fetal magnetic resonance imaging (MRI) emerges as a crucial supplementary tool, enhancing prenatal sonography for the evaluation of conjoined twins. Fetal MRI plays a pivotal role, especially in cases such as conjoined twins, where the use of computed tomography (CT) scans is limited due to radiation exposure concerns during pregnancy. MRI offers clear benefits compared to prenatal sonography since it eradicates variability associated with operator dependence and delivers enhanced tissue contrast through increased resolution and a broader field of view (FOV). As a result, MRI plays a crucial role in preparing for complex surgical interventions [5].

Indications to perform fetal magnetic resonance imaging (MRI) can result from insufficient ultrasound assessment, intrinsic ultrasound disadvantages, or suspected congenital malformation. Another indication for fetal MRI could be maternal or family history and screening [5,6].

Clinicians should maintain a high level of suspicion when diagnosing such instances with ultrasound in early pregnancy. In every monochorionic monoamniotic pregnancy (a single placenta with no separate amniotic membranes), the risk of conjoined twins should be considered and can be detected as early as 12 weeks of pregnancy [1,7].

Given the contraindication of computed tomography (CT) scan usage due to increased radiation exposure for both the fetus and the mother, MRI presents distinct advantages over prenatal sonography. Notably, MRI eliminates variability linked to operator dependency and provides superior tissue contrast due to heightened resolution and an expanded FOV. Consequently, MRI assumes a pivotal role in preparing for intricate surgical interventions [5,8].

Case presentation

A 28-year-old female (G2P1A0) was initially admitted at 34 weeks of pregnancy with a suspected Dandy-Walker variant and a small mandible. She didn't have a history of twinning in the family. The first child is a 3.5-year-old girl born healthy with a normal vaginal birth history. There is no history of interfamily marriage. She didn't have a history of diabetes, heart disease, or hypertension. Subsequently, a 3D ultrasound was performed, revealing malformed facial features with cleft palates, a large head, and macrosomia. Several congenital disorders such as VSD and hypoplastic vermis were found. The clinician suspected a possibility of facial duplication because 3 optic discs were seen in cross section, as shown in Figs. 1 and 2.

The patient was then referred to the radiology department for a fetal MRI examination at 34 weeks gestation. On MRI examination, the size of the fetus and the head was large; therefore, initially, we were not aware of the possibility of conjoined twins. Our study utilized a 3 Tesla Signa Pioneer GE Healthcare MRI machine with a body matrix coil (AIR Coil TM) and a Spine Coil attached to the machine table. Encompassing specific sequences like axial coronal sagittal T2 SSFSE (TR 802 ms, TE 124 ms) with coronal FIESTA FS (TR 4 ms, TE 2 ms) and axial FIESTA FS (TR 4 ms, TE 1 ms) with 3 mm slice thickness, 1.5 mm gap for brain and 4 mm slice thickness 2 mm gap for thoracoabdominal region. MRI of the brain was performed at 3 orthogonal planes.

At the axial, sagittal, and coronal view of Fetal MRI, there are multiple developmental abnormalities due to the failure of brain structure separation, such as duplication of the frontotemporal lobes, separated occipital lobes, agenesis of the corpus callosum, agenesis of septum pellucidum, and duplication of lateral ventricles. The posterior fossa was small, and the 2 heads appeared fused at the posterior fossa with

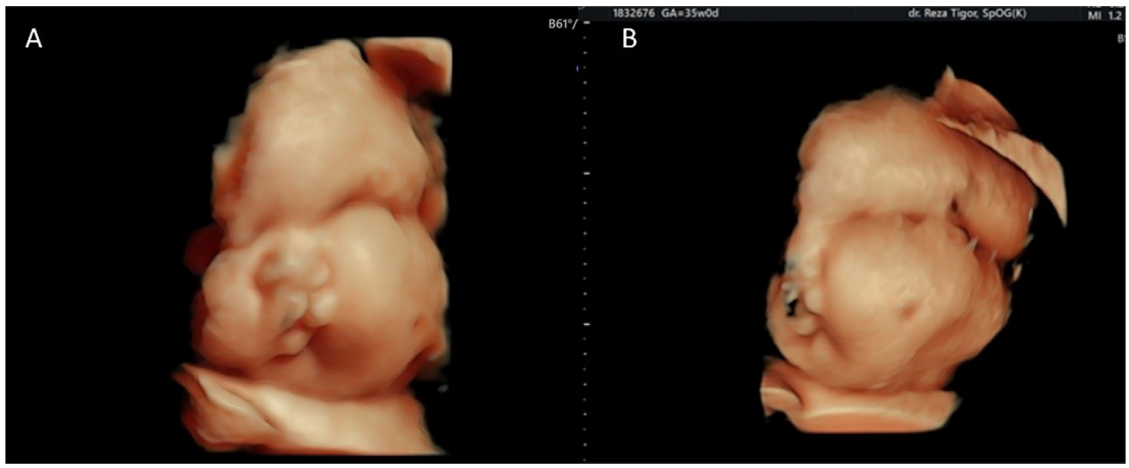

Fig. 1 – (A, B) 3D ultrasound showing a cleft palate, double lips, and malformed facial images.

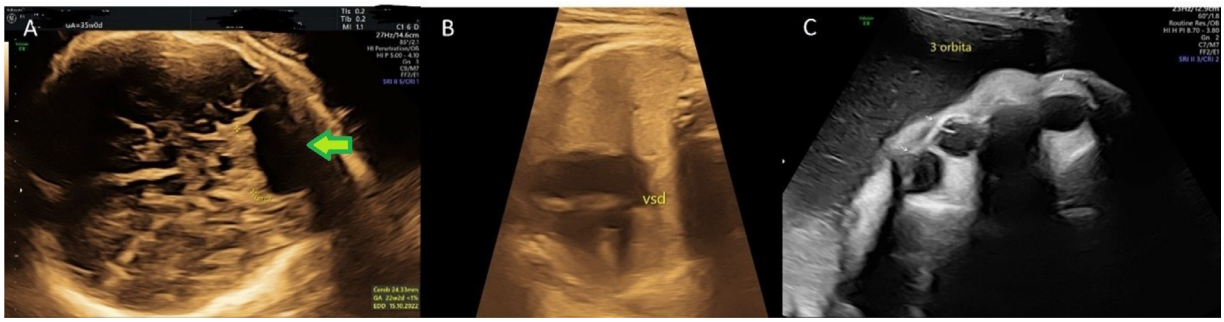

Fig. 2 – (A, B) Axial cut of the head showing cystic structures (marked green arrow) in the posterior fossa, possibly a Dandy-Walker malformation. The heart structure shows a suspicion of VSD (ventricular septal defect). (C) Three optic discs are seen in the axial section.

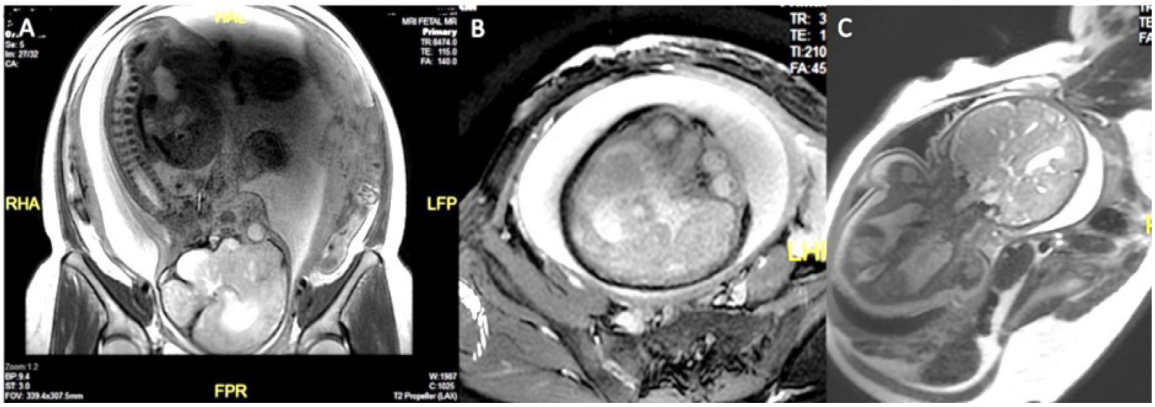

Fig. 3 – (A) Hypoplastic vermis with possible Dandy-Walker Malformation. There are visible malformations of occipital and ventricular structures. (B) There are 3 orbital structures, the fourth orbit is not in line and is somewhat separate. (C) There are 3 orbital structures, the fourth orbit is not in line and is somewhat separate.

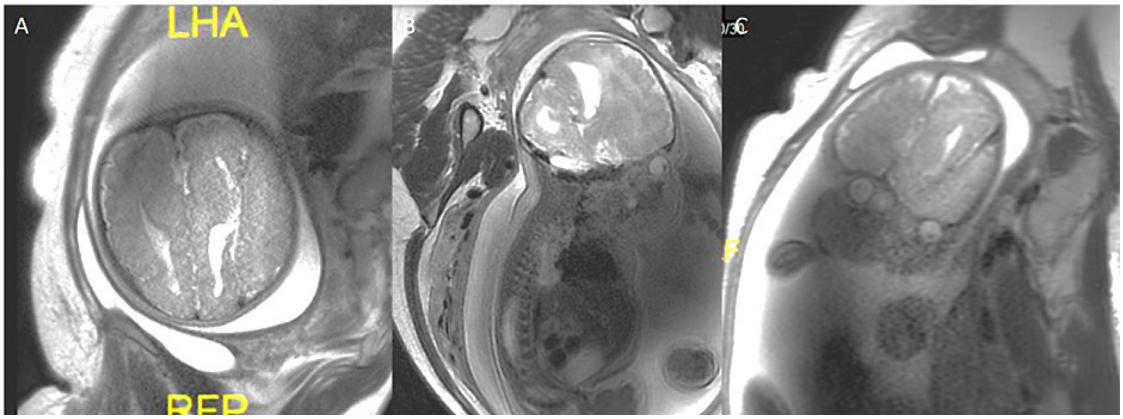

Fig. 4 – (A-C) Agenesis of the corpus callosum and septum pellucidum, malformation of the lateral ventricles, malformation or duplication of the temporal lobe.

small vermis and Dandy-Walker malformation (Figs. 3A and B). There is 1 posterior fossa. Furthermore, posterior fossa fusion and split of the medulla oblongata which fused at the dilated spinal canal were evident (Fig. 3C).

In the facial region, the identification of 4 orbital structures or bulbus oculi (eyeballs) raised suspicion of facial malformation (Figs. 3B and 4C). Although this was a high-risk pregnancy, a caesarean section (c-section) delivery was scheduled. Some

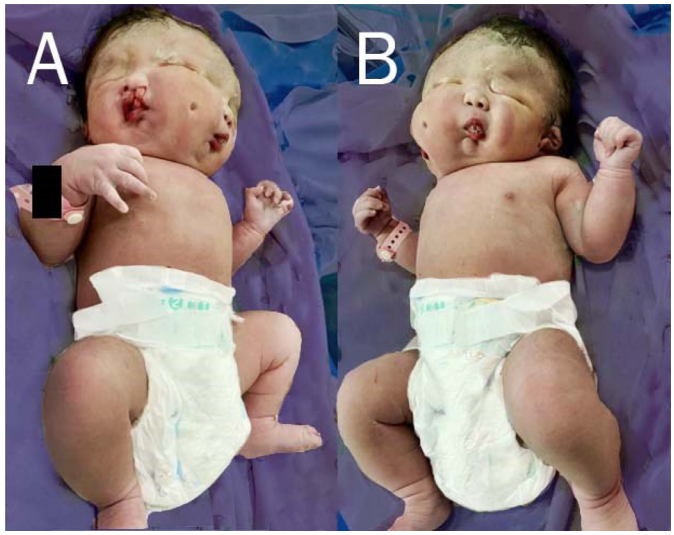

Fig. 5 – (A, B) Baby with diprosopus twins (DT) having 2 faces with 1 head and a trunk.

studies indicate that after 24 weeks of gestation, a c-section is preferred to avoid any birth trauma in conjoined twins, with the patient should be advised and counselled before the delivery process and for future pregnancies [3]. Further planning, including the need for intensive management postdelivery, was communicated to both parents weeks before the delivery. At 38 weeks of gestation, the baby was delivered with a diagnosis of diprosopus twins (DT) (Figs. 5A and B). Both parents refused on further intensive management. The baby was in inpatient care for 3 days before having a cardiac arrest. Resuscitation attempt was refused by both parents and the baby passed away.

Discussion

The presentation of diprosopus twins (DT), a rare and challenging diagnosis characterized by craniofacial duplication, was observed in this case. The fetus was large (macrosomia), noticeably large face and head, as the facial structures might be missing. Hence, collaboration with clinicians and comparative analysis with ultrasonography was essential for an accurate assessment [1]. Notably, the MRI was conducted at 34 weeks, a juncture where cerebral development was nearing completion.

Diprosopus comes from the Greek language, meaning 2-faced. It is a rare congenital anomaly characterized by craniofacial duplication and poses diagnostic challenges that necessitate advanced imaging techniques for accurate assessment [9]. Based on Gorlin's classification, our case corelates with type 2, a supernumerary mouth laterally placed with rudimentary segments [1,3].

In recent years, magnetic resonance imaging (MRI) has emerged as a superior modality, serving as an adjunct to ultrasound in unraveling the complexities of this unique condition [6]. Ultrasonography, a conventional tool in prenatal screening, has limitations in delineating intricate cranial structures in diprosopus cases. Operator-dependent variability and challenges associated with fetal positioning often result in incomplete visualizations, potentially leading to misdiagnosis or incomplete assessment. In contrast, MRI offers distinctive advantages in providing high-resolution and detailed images of fetal anatomy. The elimination of operator-dependent variability ensures a consistent and accurate depiction of duplicated cranial structures. The heightened tissue contrast, superior resolution, and multiplanar capabilities of MRI contribute significantly to a comprehensive understanding of the anomaly [5].

Raafat et al., from their study of CNS anomalies in fetal MRI compared to ultrasound, found that the overall diagnostic accuracy was 98.8% for ultrasound and 99.6% for MRI [10]. However, in cases like morbid maternal obesity, oligohydramnios, ossified skulls, in late-term pregnancy, or cases that show indecisive results, additional Fetal MRI examination is needed to obtain more accurate information [10]. Fetal MRI can indicate the location of the tentorium cerebelli as well as the global volume of the posterior fossa. It allows for morphological and biometric examination of the structures of the posterior fossa. Direct midline sagittal and coronal pictures show the cerebellar vermis best. Nonoblique axial and coronal views are ideal for assessing the cerebellar hemispheres [6].

For the fetal examination, this case was using 3Tesla MRI. The 3T MRI had a higher imaging score than the 1.5T MRI when assessing the different anatomical features of the foetus. There is an overall advantage to undergoing 3T fetal MRI. Improvements in imaging software and methods enable improved visibility and evaluation of prenatal pathology [11].

Research studies, such as those conducted by Salem and Spielmann et al., emphasize the efficacy of MRI in diagnosing diprosopus twins (DT). These studies highlight MRI's ability to visualize intricate details, including the duplication of frontotemporal lobes, distinct occipital lobes, and facial structures. The fetal face is an important part of the antenatal structure to examine. Facial malformation may indicate an underlying chromosome abnormality or syndrome. Routine antena-

tal screening from the early trimester is crucial because if abnormalities are found early, families can receive early counseling to implement more appropriate management. This facial deformity has an unfavorable prognosis because it is often accompanied by abnormalities in brain development. Therefore, when found in late pregnancy, the baby is usually stillborn or does not live long [6,9,5]. In this case, the clinician has provided an explanation of the baby's condition and longtime prognosis for both parents. The baby had Dandy-Walker syndrome and ventricular septal defect (VSD) detected by ultrasound and MRI. Upon consideration, both parents decided against intensive treatment, and the baby died 3 days after birth.

Conclusion

In conclusion, by employing multiplanar techniques, MRI emerged as the optimal adjunctive and complementary modality to ultrasonography for the detection of complex congenital malformations which are difficult to achieve by ultrasound. Successful outcomes hinge upon robust collaboration between clinicians and radiologists, underscoring the necessity for heightened sensitivity in diagnostic approaches [1–4]. The advent of 3D ultrasound technology and faster MRI sequences has facilitated early prenatal diagnosis and proper classification of this type of conjoined twins, allowing for optimal and timely obstetric and postnatal treatment [7].

Patient consent

The authors of this case series titled "*A Rare Case Report: The Value of Fetal MRI to Detect Diprosopus Twins*" state that written informed consent for the publication of this case series was obtained from the legal guardians of the pediatric patients involved. All patient identifying information has been redacted to protect the privacy of the individuals involved. Pictures used in the study have been consented to by the patient's legal guardian.

REFERENCES

[1] Trevisani V, Balestri E, Napoli M, Caraffi SG, Baroni MC, Peluso F, et al. Diprosopus: a rare case of craniofacial duplication and a systematic review of the literature. Genes (Basel) 2023;14(9):1745.
[2] Chikkannaiah P, Prathap N, Venkataramanappa S. Rare case of "diprosopus bicephalous triophthalmus" and review of literature. J Pediatr Neurosci 2018;13(4):465–8.
[3] Nair NM, Swarr DT, Barnes-Davis ME. Preterm infant with diprosopus and holoprosencephaly. Clin Case Rep 2021;9(12):e05163.
[4] Rehder H, Kircher SG, Schoner K, Smogavec M, Behunova J, Ihm U, et al. Brain malformations in diprosopia observed in clinical cases, museum specimens and artistic representations. Orphanet J Rare Dis 2023;18(1):57.
[5] Eshiba SM, Zahran MH, Elnekeidy AM, Abdeldayem TM, Hassan HHM. Added value of fetal MRI as a complementary method to antenatal ultrasound in the assessment of non-CNS fetal congenital anomalies. Egypt J Radiol Nucl Med 2022;53(1):40.
[6] Saleem SN. Fetal MRI: an approach to practice: a review. J Adv Res 2014;5:507–23 Elsevier B.V..
[7] Srinath N, Rajgarhia N, Regmi P, Bajpai A. Prenatal diagnosis of non-janiceps type of cephalopagus conjoined twins: a case report. Egypt J Radiol Nucl Med 2023;54(1):63.
[8] McHugh K KESL. Imaging of conjoined twins. Pediatr Radiol 2006;36(9):899–910 quiz 1002-3.
[9] Bhuyan M HI. Diprosopus a rare craniofacial malformation. Asian J Neurosurg 2018;13(04):1257–9. doi:10.4103/ajns.ajns_202_17.
[10] Raafat M, Alalfy M, Nagy O, Saraya S. Fetal brain MRI: how it added to ultrasound diagnosis of fetal CNS anomalies-1 year experience. Egypt J Radiol Nucl Med 2021;52(1):85.
[11] Victoria T, Johnson AM, Christopher Edgar J, Zarnow DM, et al. Between 1.5-T and 3-T MRI for fetal imaging: is there an advantage to imaging with a higher field strength? AJR 2016;206(1):195–201. doi:10.2214/AJR.14.14205.