

Cephalothoracoomphalopagus Tetrabrachius Tetrapus Conjoined Twins: An Extremely Rare Case Report

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Case Report

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Abstract

Background

Conjoined twins are a rare subset of monozygotic twins that usually have a poor prognosis, which has become a significant cause of perinatal morbidity and mortality. This case highlights the importance of routine antenatal care for earlier prenatal diagnosis and well-informed family counseling concerning appropriate intervention for conjoined twins.

Case presentation:

A 31-year-old G2P1 female in 29 4/7 weeks of gestation was referred from a primary health care center to our hospital with conjoined twins. Two-dimensional (2D) and three-dimensional (3D) ultrasonography examinations at 29 4/7 weeks of gestation revealed one fetal head accompanied by an orbital hypotelorism with an interocular diameter of 0.99 cm. The liver and heart were found to be the fusion organs involved. Polyhydramnios was observed with a maximum vertical pocket of 12.07 cm. Diagnosis of cephalo-thoraco-omphalopagus conjoined twins was made right after the first scan and consistently confirmed thereafter by six more repeated ultrasound examinations. Clinical examination after scheduled surgical delivery in 33 5/7 weeks old neonates revealed male conjoined twins with a fused head accompanied by two pairs of ears, along with a single trunk together with four neurologically independent upper limbs and lower limbs, as well as two complements of genitalia and two anuses. The twin showed signs of respiratory distress and died within one hour of birth.

Conclusion

Routine antenatal care and advanced ultrasound scanning are crucial for earlier prenatal diagnosis and well-informed family counseling concerning appropriate intervention for conjoined twins.

INTRODUCTION

Conjoined twins are a rare subset of monozygotic twins with an incidence of 1:50,000 to 1:100,000 live births [1]. This congenital anomaly has a poor prognosis, with approximately 60% dying in utero, and around 35% are stillborn or die within the first few hours of delivery [2]. Female predominance was reported, with a ratio of 3:1 compared to males [3].

Conjoined twins are broadly classified into two major groups based on the body parts interconnected: non-dorsal and dorsal [4]. Twins with non-dorsal union include lateral union (side or parapagus) and ventral union, which consists of head (cephalopagus), thorax (thoracopagus), abdomen (omphalopagus), and pelvis (ischiopagus). Twins with a dorsal union include the sacrum (pygopagus), spine (rachipagus), and cranium (craniopagus) [4]. It is also either symmetrical or asymmetrical [5].

We present an extremely rare case of male 33 5/7 weeks cephalo-thoraco-omphalopagus tetrabrachius tetrapus conjoined twins detected with ultrasound scanning in the third trimester and confirmed postnatally in a 31-year-old secundigravida woman. This case highlights the importance of routine antenatal care for earlier prenatal diagnosis and well-informed family counseling concerning appropriate intervention for conjoined twins.

CASE PRESENTATION

A 31-year-old G2P1 female with no history of spontaneous abortion was referred from a primary health care center to our hospital at 29 4/7 weeks of gestation. Since the beginning of her pregnancy, the patient has undergone routine antenatal care at the primary health care center. The earlier ultrasound scan did not indicate any anomalies, and neither the patient nor the fetus experienced any significant complaints during the first trimester. In the second trimester, a conventional two-dimensional (2D) ultrasonography (USG) examination revealed suspected conjoined twins. She had no previous birth history with congenital anomalies, nor did her family history reveal any such issues.

The 2D USG examination revealed one large fetal head with two obvious thalami (Figs. 3A and 3B), accompanied by orbital hypotelorism with an interocular diameter of 0.99 cm (Fig. 1A). It also showed a medial fusion of the left and right sides of the two auricles, forming a “love” symbol (Fig. 2A). A confirmatory examination by three-dimensional (3D) USG is shown in Fig. 1B and Fig. 2B. A single liver and heart were noted as the structural fusion organs of the twins. Polyhydramnios with a maximum vertical pocket (MVP) of 12.07 cm was also noted. The diagnosis of cephalo-thoraco-omphalopagus conjoined twins was made right after the first scan and consistently confirmed thereafter by six more repeated 2D and 3D USG examinations.

Scheduled surgical delivery was performed at 33 5/7 weeks of gestation. Clinical examination after delivery revealed male conjoined twins with one head accompanied by two pairs of ears, along with a single trunk together with four neurologically independent upper limbs and lower limbs, as well as two sets of genitalia and two anuses (Figs. 1C and 2C). The twin showed signs of respiratory distress and died within one hour of birth.

DISCUSSION

Conjoined twins are a rare developmental anomaly that are usually monochorionic and monoamniotic. This condition affects 1 in 200 monozygotic twins with an overall survival rate of around 25% [6]. Approximately 75% to 90% of conjoined twins are female, with a 3:1 ratio compared to males [3]. The pathophysiology of conjoined twins can be explained by the two known theories: partial fission and fusion [7]. The partial fission theory holds that conjoined twinning results from abnormal cleavage of a single fertilized egg between days 13 and 17 of embryogenesis. In contrast, the fusion theory posits that similarities in stem cell characteristics can lead to secondary fusion of two dizygotic or monozygotic twins [7, 8].

Around 50% to 76% of conjoined twin cases developed polyhydramnios [9]. Polyhydramnios, defined as an amniotic fluid index (AFI) greater than or equal to 24 cm and/or a maximum vertical pocket (MVP) or single deepest pocket (SDP) greater than or equal to 8 cm, is referred to as a condition in which there is excessive amniotic fluid surrounding the fetus due to an imbalance in the production and removal mechanisms [10]. Pregnancies with conjoined twins commonly develop polyhydramnios more often than ordinary twins because they often have structural abnormalities in the upper gastrointestinal tract, cardiovascular circulation, and/or brain swallowing reflex [10, 11].

Early prenatal evaluation and diagnosis help obstetricians explain the maternal-fetal condition as soon as possible. It may offer parents a window of time to make an informed decision about prenatal and postnatal care planning. A careful advanced ultrasound examination is crucial to detect the twins' anatomical details, the degree to which the organs are fused, as well as to prepare for potential surgical separation [12]. Separation of the twins requires multidisciplinary collaboration among neonatologists, radiologists, anesthesiologists, pediatric surgeons, and cardiac surgeons [13].

The survival rate of conjoined twins is considered low, and the prognosis largely depends on the site of fusion and the organs involved [14]. Successful twin separations have been reported in a few studies. Clare *et al.* in 2024 performed an emergency separation of a pair of one alive and one stillborn thoracopagus conjoined twin after 18 hours of delivery [15]. In 2019, Grizelj *et al.* performed a separation of fused liver parenchyma in a xypho-omphalopagus conjoined twins with an extremely rare complication of extrauterine twin-to-twin transfusion syndrome [16]. Another twin separation was performed in a resource-constrained setting by Nazir *et al.* in 2018, a thoraco-omphalopagus conjoined twin with a fused liver was successfully separated using the cotton tourniquet technique under endotracheal anesthesia [17].

This case may represent the first reported case of a rare form of conjoined twinning in Indonesia, featuring twins with a fused head, fused thoraco-umbilical body, four upper limbs, four lower limbs, two genitalia, two anuses, as well as fused organs involving the liver and heart. However, this report has several limitations. We could not provide the captured ultrasound images of the fused visceral organs in this case, such as the liver and heart. Further examinations of the other fused organ involved through a magnetic resonance imaging (MRI) modality were also not observed.

CONCLUSION

Routine obstetric care and advanced ultrasound scanning are crucial for earlier prenatal diagnosis. A detailed anatomical survey is necessary, particularly in women with unremarkable earlier antenatal ultrasound scans. Well-informed family counseling will allow for appropriate intervention and delivery options for the management of conjoined twins.

Abbreviations

Two-dimensional
3D
Three-dimensional
AFI
Amniotic Fluid Index
MRI
Magnetic Resonance Imaging
MVP
Maximum Vertical Pocket
SDP
Single Deepest Pocket
USG
Ultrasonography

Declarations

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

Ethical approval was not required for this anonymized case report in accordance with local

CONSENT FOR PUBLICATION

Detailed informed consent was obtained from the patient's parents for publishing this article, with the condition that no personal details or patient names be published.

COMPETING OF INTERESTS

The authors had reported no conflict of interest.

DATA AVAILABILITY

The datasets supporting the conclusions of this article are included within the article

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None.

Author Contribution

References

1. Ahmadi F, Keramat N, Haghighi H. Conjoined twin. *Int J Fertil Steril*. 2012;6(2):135–6.
2. Mathew RP, Francis S, Basti RS, Suresh HB, Rajarathnam A, Cunha PD, et al. Conjoined twins - role of imaging and recent advances. *J Ultrason*. 2017;17(71):259–66.
3. Kusmayadi DD, Emiliana L, Trifosa SC. Characteristics and management of conjoined twins: A single-centre retrospective descriptive study. *Med J Malaysia*. 2024;79(Suppl 4):17–22.
4. Kattel P, Conjoined Twins, *Nepal Med A*. 2018 Mar-Apr;56(211):708–10.
5. Jabari S, Carbon R, Besendörfer M, Hartmann A, Rompel O, Hoerning A, et al. Asymmetric Omphalopagus in a Triplet after In Vitro Fertilization: A Rare Case of Conjoined Twinning. *Case Rep Pediatr*. 2018;2018:9349606.
6. Métneki J, Varjassy P. Conjoined twins. In: Tarnoki AD, Tarnoki DL, Harris JR, Segal NL, editors. *Twin Research for Everyone: From Biology to Health, Epigenetics, and Psychology*. Academic; 2022. pp. 85–112.
7. Boer LL, Schepens-Franke AN, Oostra RJ. Two is a crowd: On the enigmatic etiopathogenesis of conjoined twinning. *Clin Anat*. 2019;32(5):722–A741.
8. Bindlish A, Sawal A. A detailed description and discussion on conjoined twins. *Cureus*. 2022;14(9):e29526.
9. Kalchbrenner M, Weiner S, Templeton J, Losure TA. Prenatal ultrasound diagnosis of thoracopagus conjoined twins. *J Clin Ultrasound*. 1987;15(1):59–63.
10. Society for Maternal-Fetal Medicine (SMFM), Dashe JS, Pressman EK, Hibbard JU. SMFM Consult Series #46: Evaluation and management of polyhydramnios. *Am J Obstet Gynecol*. 2018;219(4):B2–8.
11. van Gemert MJC, Nikkels PGJ, Ross MG, van den Wijngaard JPHM. Acardiac twin pregnancies part V: Why does an acardiac twin with renal tissue produce polyhydramnios? *Birth Defects Res*. 2021;113(6):500–10.
12. Arnold J, Luton A, Davies J, Introduction. Unique challenges in the care of conjoined twins. *Semin Perinatol*. 2018;42(6):319–20.
13. Heuer GG, Madsen PJ, Flanders TM, et al. Separation of Craniopagus Twins by a Multidisciplinary Team. *N Engl J Med*. 2019;380(4):358–64.
14. Behera MK, Jain S. Conjoined Twins. *Med J Armed Forces India*. 2002;58(2):163–4.
15. Clare N, Margaret AM, Lugobe HM, Egwau AFO, Joseph E. Successful separation of conjoined twins, one healthy and one stillborn: A case report. *J Pediatr Surg Case Rep*. 2024;105:102823.
16. Grizelj R, Sindičić Dessardo N, Bulić K, Luetić T, Mikulić D, Antabak A, et al. Successful separation of xypho-omphalopagus conjoined twins with extrauterine twin-twin transfusion syndrome: a case report. *Croat Med J*. 2019;60(4):301–8.

17. Nazir Z, Mateen Khan MA, Faruque AV, Dilawar B. Separation of conjoined twins in a resource constraint setting – Lessons learned and implications for global surgery initiatives. J Pediatr Surg Case Rep. 2018;39:31–4.

Figures

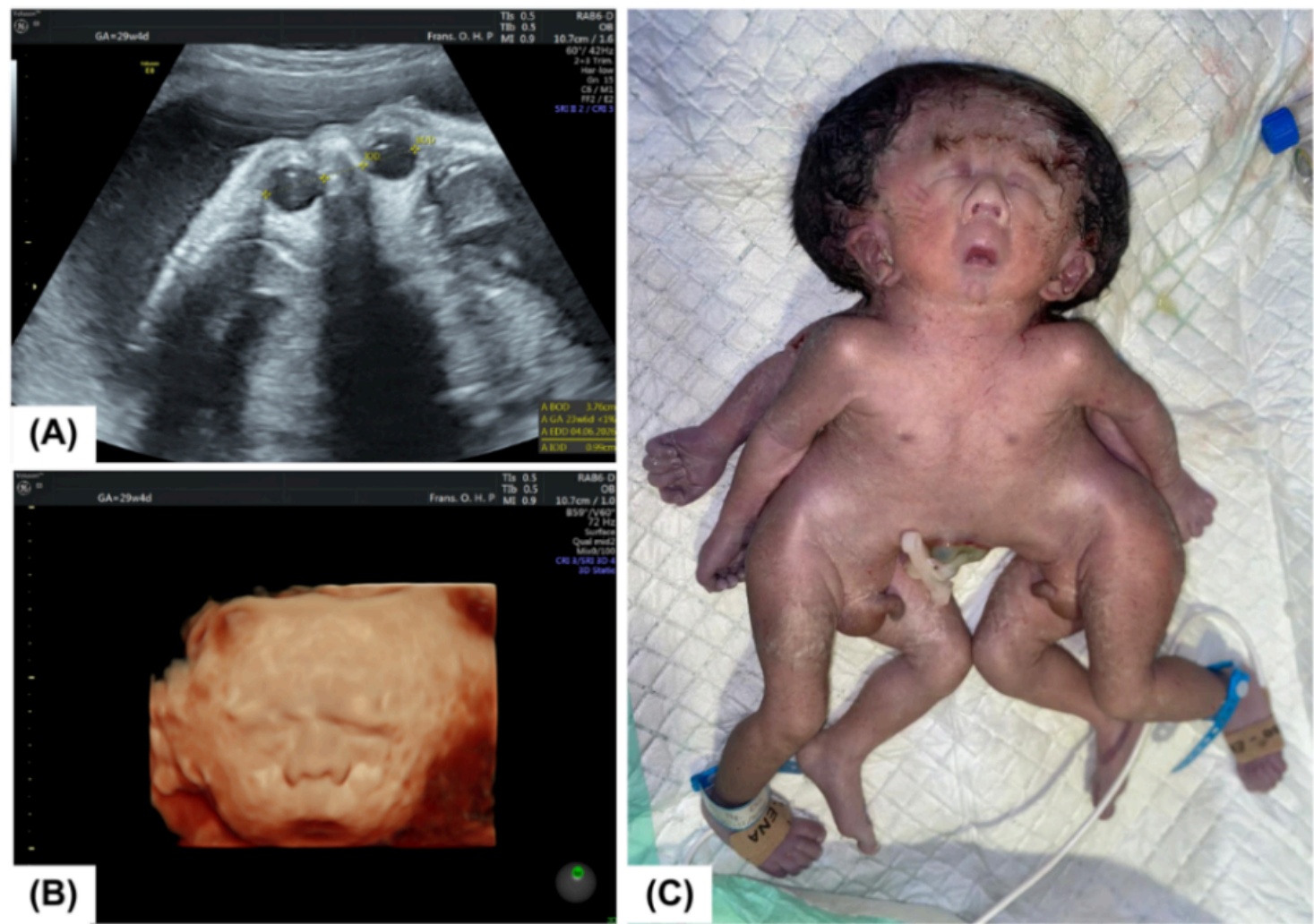


Figure 1

Orbital hypotelorism of fetal face was observed in (A) two-dimensional ultrasound images at 29 4/7 weeks of gestation with 0.99 cm interocular diameter and (B) three-dimensional ultrasound images at 29 4/7 weeks of gestation. (C) Clinical post-delivery examinations at 33 5/7 weeks old showed conjoined twins with one head, one trunk, four upper limbs, two genitalia, two anuses, and four lower limbs

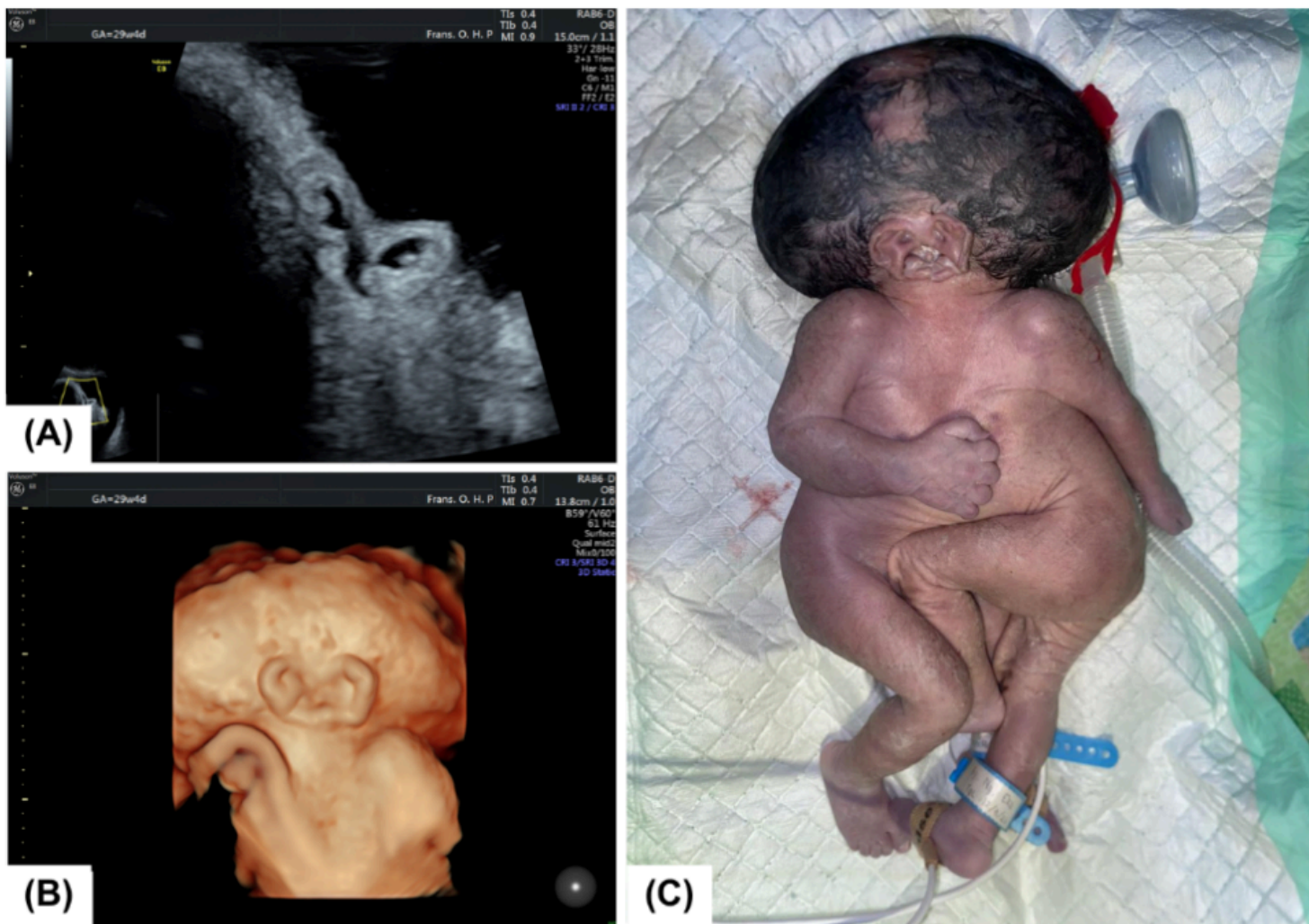


Figure 2

Two auricles of both left and right sides that are fused in the medial, mimicking the "love" symbol was observed in the (A) two-dimensional ultrasound images at 29 4/7 weeks of gestation and (B) three-dimensional ultrasound images at 29 4/7 weeks of gestation, in conjunction with the (C) clinical post-delivery examinations at 33 5/7 weeks old

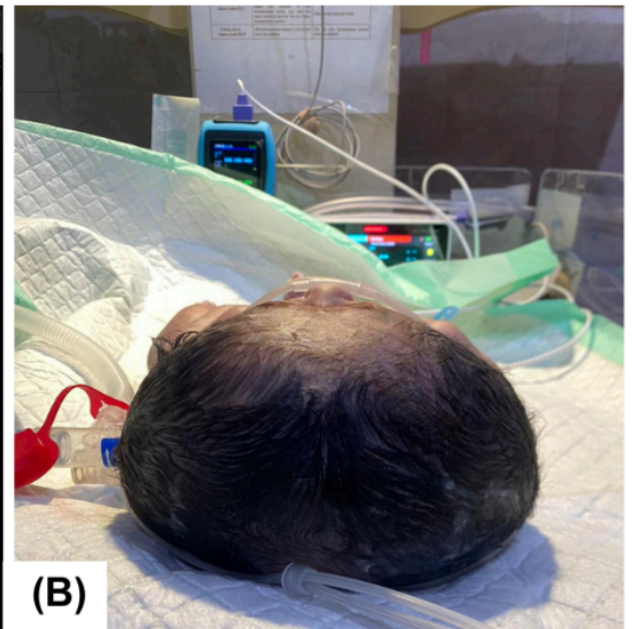
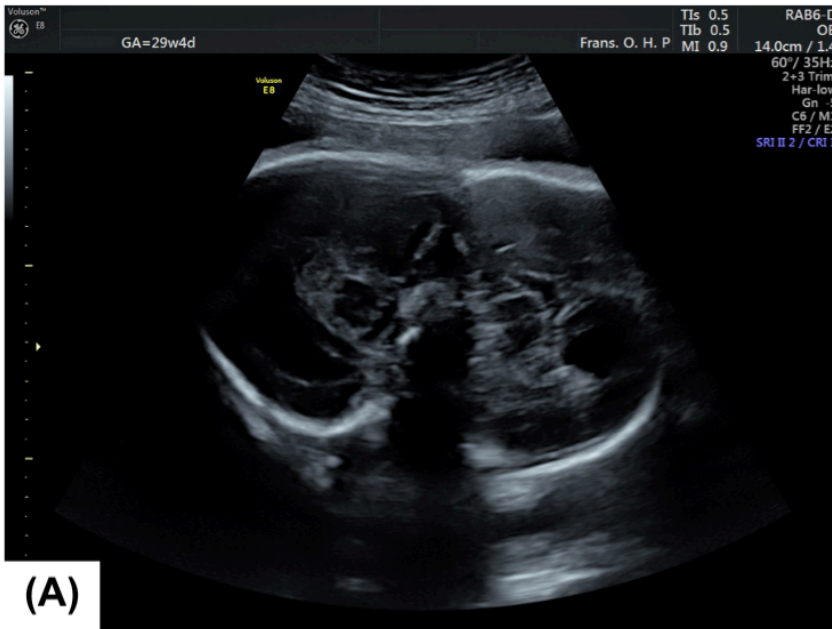


Figure 3

A seemingly single large head with two obvious thalami was observed in the (A) two-dimensional ultrasound images at 29 4/7 weeks of gestation, confirmed by the (B) clinical post-delivery examinations at 33 5/7 weeks old