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Case Report

Prenatal diagnosis of parapagus diprosopus dibrachius dipus twins with spina bifida in the first trimester using two- and three-dimensional ultrasound

Pei-Yin Yang [a], Ching-Hua Wu [b], Guang-Perng Yeh [c], Charles Tsung-Che Hsieh [c, *]

[a] Department of Obstetrics and Gynecology, Chiayi Christian Hospital, Chiayi, Taiwan
[b] Obstetrics and Gynecology Ultrasound Lab, Changhua Christian Hospital, Changhua, Taiwan
[c] Department of Obstetrics and Gynecology, Changhua Christian Hospital, Changhua, Taiwan

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ABSTRACT

Objective: Here, we report a case of parapagus diprosopus twins with spina bifida diagnosed in the first trimester of pregnancy using two-dimensional (2D) and three-dimensional (3D) ultrasound.
Case report: A 28-year-old Taiwanese woman, gravid 1, para 0, visited our hospital due to an abnormal fetal head shape discovered by 2D ultrasound at 11-weeks gestation. Parapagus diprosopus twins with spina bifida were diagnosed after ultrasound examination. The characteristics of parapagus diprosopus twins are more illustrative in 3D ultrasound than in 2D ultrasound. After counseling, termination of pregnancy was chosen by the couple. Although necropsy was declined, the gross appearance and radiograph of the abortus confirmed our diagnosis.
Conclusion: With the help of 3D ultrasound, we made an early and definitive diagnosis of conjoined twins.

Introduction

Conjoined twins are very uncommon, with an incidence of ~1:50,000 to ~1:100,000 [1–3], occurring in 1% of monochorioic twins [1]. The logical theory of the origin of conjoined twins proposed by an expert would be the "spherical theory", which describes the secondary union of two originally separate embryonic discs in one of two areas: the dorsal neural tube or the ventral yolk sac after the 13th day of conception [4]. Due to significant advances in ultrasound technology, prenatal diagnosis of conjoined twins in the first trimester is feasible [5]. The first case of prenatal diagnosis of conjoined twins in the first trimester was thoracophalipagus at 11-weeks gestation [6].

Early diagnosis and detailed assessments of the shared vital organs are necessary for optimal management of the pregnancy and birth. With the use of three-dimensional (3D) imaging technology, the space orientation and anatomical details are more clearly demonstrative than with traditional two-dimensional (2D) ultrasonography [2,5]. Here, we report a rare case of parapagus diprosopus twins associated with spina bifida diagnosed at 12-weeks gestation by using 2D and 3D ultrasound.

Case presentation

A 28-year-old female, gravida 1, para 0, visited our hospital at 12 weeks of pregnancy. She had received an ultrasound exam in Vietnam, during which an abnormal fetal head shape was recognized. She returned to Taiwan for the subsequent prenatal exam. The couple are not consanguineous. Neither the woman nor her husband had a family member with a history of twinning or congenital anomalies.

We used a high-resolution ultrasound machine (Voluson 730 Expert, General Electric, Zipf, Austria), an abdominal transducer (AB 2–7 convex probe, 2–7 MHz, General Electric), and a 3D transabdominal transducer (RAB 2–5 L4D probe, 2–5 MHz, General Electric). The crown-rump length was 55 mm, compatible with the gestational age. With 2D ultrasonography, we identified a normal-appearing trunk and extremities, one stomach, one heart, and one bladder. However, we also noted widening of the skull like a

* Corresponding author. Department of Obstetrics and Gynecology, Changhua Christian Hospital, Number 135 Nan-hsiao Street, Changhua 500, Taiwan.
E-mail address: 40129@cch.org.tw (C.T.-C. Hsieh).

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helmet. Midline of the brain and bilateral widened central plexuses were traced in the 2D ultrasound (Figure 1). We performed a 3D ultrasound, which indicated a normal body appearance, but a widened thoracic cage and four normal limbs (Figure 2). We then performed a 3D rendering with skeletal mode. Widened cervicothoracic spines were recognized and partial conjunction of spines was suspected (Figure 3). Two posterior fontanelles could be observed with 3D surface rendering. The fusion of the faces was much clearer in the 3D surface rendering. We also used 3D tomographic ultrasound imaging (TUI) to survey the fetal spine and confirmed our suspicion of spina bifida due to an overlying skin defect of the spine (Figure 4). Three orbits were also indicated by the 3D TUI technique (Figure 5). We were not able to define the intracranial structures due to the early gestational age and fetal position.

After discussion with the family, the couples decided to terminate this pregnancy. Although necropsy was declined, the appearance of the abortus was compatible with the results of the 2D and 3D ultrasonography (Figures 6 and 7). There were four limbs and a normal trunk, but a wide thoracic cage, a rounded and wide shared cranial bone, three eyes due to inner-side fusion in two eyes, two noses, and two sets of lips (diprosopus, parapagus, dibrachius, and dipus). The postabortal radiography showed two joined cranial vaults and cervicothoracic spines (Figure 8). All these findings were compatible with our ultrasonographic findings.

Discussion

Different types of fusion exist in the cases of conjoined twins. Spencer [4] proposed that conjoined twins should be classified by the proposed site of the union and be divided into two groups: ventral and dorsal [4]. Ventral unions, such as rostral, caudal, and lateral unions, occur most commonly. Dorsal unions, such as craniopagus, rachipagus, and pygopagus occur less frequently [4,7]. Our case was a ventral lateral union, meaning parapagus. Diprosopus was the subtype of the parapagus, meaning two faces.

The rarest type of conjoined twins is the diprosopus, representing only ~0.4% [8]. This type of conjoined twin has two faces, one head, and one body. In 1992, Fontanarosa et al [9] reported a diprosopus twin diagnosed in the first trimester [9]. Initially, their impression was of a fetus with a large encephalocele. They found this fetus with a bifid head, a broad thorax, a single heart, and four normal limbs using 2D ultrasonography. After termination, the abortus had two faces symmetrically and medially fused, craniorachischisis, and left gastroschisis.

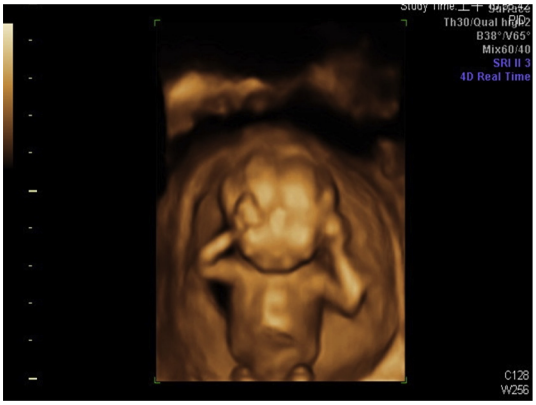

Figure 2. Three-dimensional ultrasound of our case at 12-weeks gestation using the surface mode.

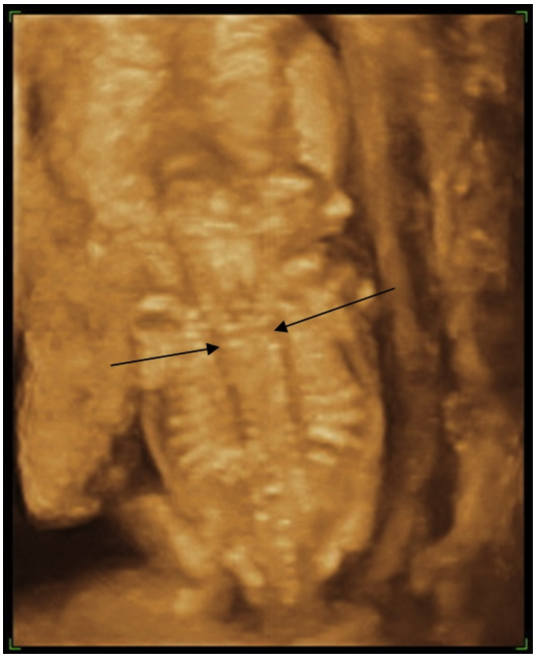

Figure 3. Three-dimensional ultrasound of our case at 12-weeks gestation in the skeletal mode. The black arrows indicate the duplication of cervical spines.

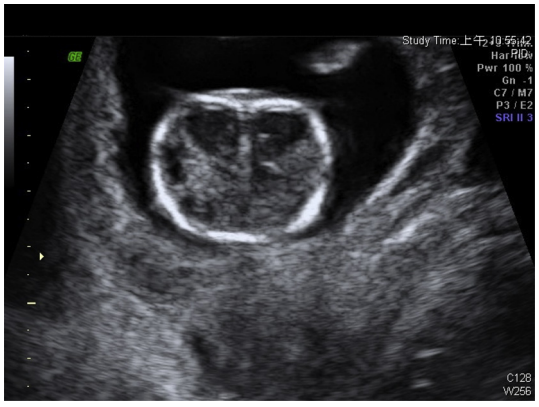

Figure 1. Two-dimensional ultrasound of the brain section of our case at 12-weeks gestation.

Ülker et al [10] presented a case of diprosopus without gastroschisis. Due to the rarity of this condition, the authors were not initially recognized and they had to repeat the exam to ensure an accurate diagnosis. In our case, the diagnosis was easy to make with 3D ultrasound because of its excellent space orientation. We reviewed all of the cases diagnosed as parapagus diprosopus in the first trimester (Table 1). The earliest gestational age at diagnosis was 8-weeks gestation [11]. In that case, obvious bifid cranium poles were noted in early pregnancy, enabling them to make the diagnosis at a very early gestational age. In our case, the gestational age at presenting was 12-weeks gestation, however, our case did not have a bifid cranium. The only clue from the 2D sonography that aided the diagnosis was the abnormal fetal head shape. Our case illustrates the importance of incorporation of fetal skull shape into fetal anatomy screening.

The experts proposed that most cases of parapagus diprosopus have duplicated cervical and upper thoracic spines, and varying

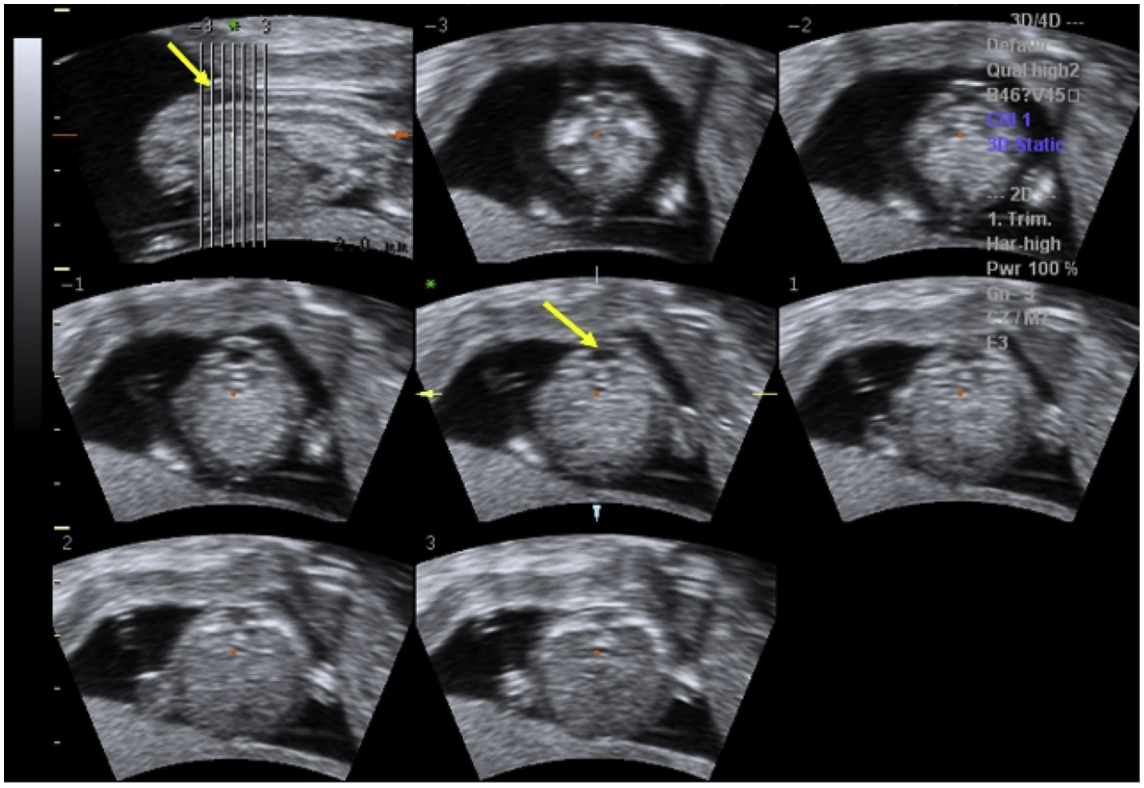

Figure 4. Tomographic ultrasound imaging of the lumbar sacral region in our case at 12-weeks gestation. The yellow arrows indicate the site of spina bifida.

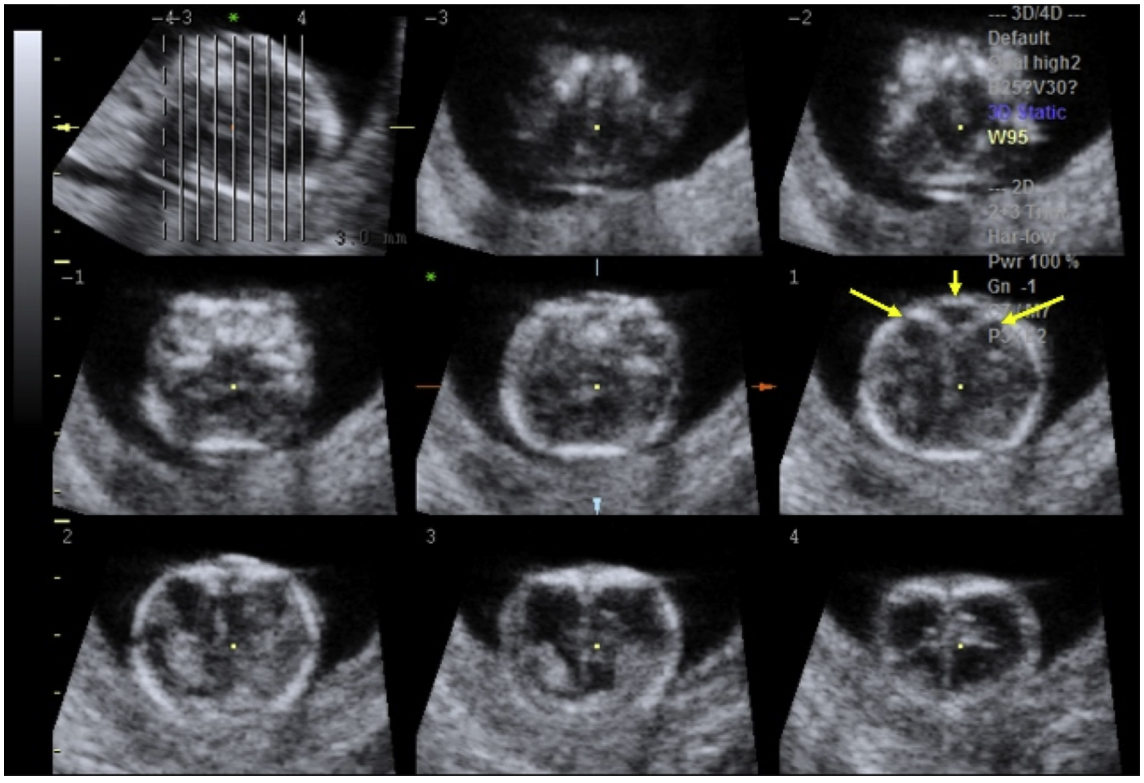

Figure 5. Tomographic ultrasound imaging of the brain region. The yellow arrows indicate three orbits.

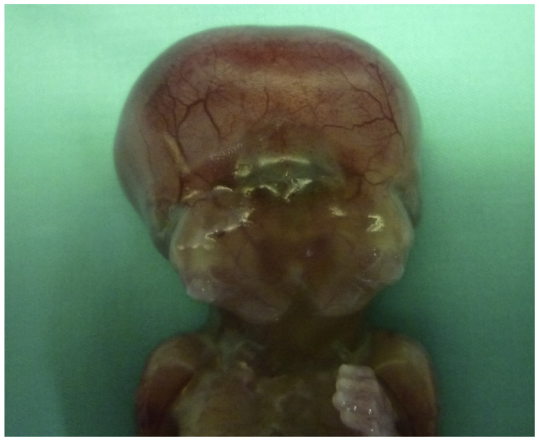

Figure 6. Postabortal image of the fused faces with a single body and upper limbs.

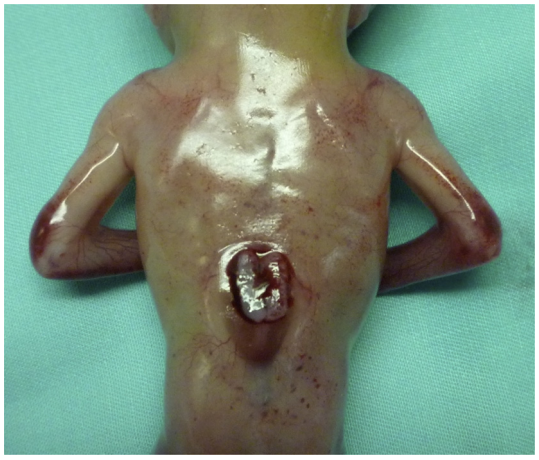

Figure 7. Postabortal image of the fetus with spina bifida and lower limbs.

portions of the shared cranium [4–10]. These findings were confirmed by radiography in our case. An association with other anomalies was also reported. Half of the reported cases had diaphragmatic hernias, and cleft lips and palates were also common.

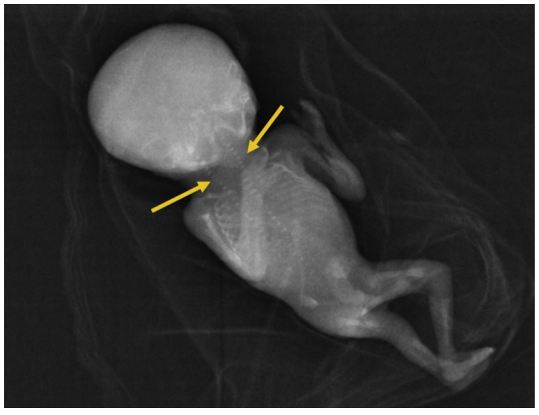

Figure 8. Postabortal X-ray of the fetus showing two joined cervicothoracic spines.

Table 1
Reported cases of parapagus diprosopus diagnosed in the first trimester.

	Age (y)	Diagnosis of age (wks)	Ultrasound finding	2D-/3D-ultrasound diagnosis	Follow-up
Tangsong et al [11] (Case 1)	20	8	—	2D	TOP
Fontanarosa et al [9]	30	11	Craniorachischisis, gastroschisis	2D	TOP
Ülker et al [10]	22	12	Craniorachischisis	2D/3D	TOP
This study	28	12	Spina bifida	2D/3D	TOP

2D = two dimensional; 3D = three dimensional; TOP = termination of pregnancy.

Heart anomalies or gastrointestinal anomalies have also been reported [4,7,8,12]. Because necropsy was declined, the fetal internal structures could not be depicted in our case. There may be a phenotype variant in this region from partial duplication of a few facial structures to complete dicephalus. Isolated duplication of the nose, eyes, or mouth occurs in the mildest forms, as in our case (Figures 6 and 7). In the most severe form of diprosopus, the fetus showed two complete faces [7,8]. Additionally, anencephaly is very frequent in this type of conjoined twins, but was not observed in our case [7,8]. We think liberal use of 3D technologies will improve the accuracy of the diagnosis of conjoined twins and enable early diagnosis.

Conflict of interest

The authors have no conflict of interest relevant to this article.

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