APPLIED RADIOLOGY
RADIOLOGICAL CASE

Parapagus dicephalus dibrachius dipus conjoined twin case

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CASE SUMMARY

A 37-year-old $G_4P_2A_2$ Hispanic female presents for her initial obstetrical visit at a gestational age of 12 weeks and four days. She has a history of one therapeutic abortion and one spontaneous abortion (third pregnancy). The patient is blood type O-positive, has an equivocal rubella titer and tests positive for tuberculosis (QuantiFERON).

IMAGING FINDINGS

Grayscale transvaginal ultrasound demonstrates a fetus with duplicated maxillofacial and calvarial structures fused at midline (Figure 1). There is a solitary heart with shared intracranial vasculature, two upper and two lower extremities (Figure 2).

DIAGNOSIS

Conjoined twin pregnancy of the parapagus dicephalus dibrachius dipus (PDDD) type. This type of conjoined twin is defined by lateral fusion with two heads, sharing a single thorax, abdomen and pelvis with two arms and two legs.

DISCUSSION

Conjoined twins occur in approximately 1 percent of monoamniotic monochorionic monozygotic twins.[1] The reported global prevalence ranges from 1:50,000 to 1:100,000 births.[1,2] Of the small number of conjoined twins born alive, there is a 30 percent mortality rate within the first 24 hours of life.[1] Overall, there is a 2:1 female predominance of conjoined twins.

Historically, there have been two opposing embryologic theories for the development of conjoined twins: "fission" and "fusion." The former attributes the development of conjoined twins to incomplete division of the developing embryo, while the latter argues that all eight types occur by secondary fusion of two embryonic discs beyond the 13th day post-conception.[3]

Conjoined twins are divided into two main groups, ventral and dorsal, based on the location of embryonic disc union. They are further classified into eight basic types relative to their site(s) of union or pagus (Greek for joined): Omphalopagus (umbilicus/abdomen), thoracopagus (thorax/upper abdomen), cephalopagus (maxillofacial), craniopagus (skull), ischiopagus (pelvis), rachipagus (spine) and pygopagus (sacrum). Additional terminology includes the suffixes: -parapagus (fused laterally/side-by-side), -brachius (upper limb), -pus (lower limb) and -prosopus (face).[3]

Parapagus conjoined twins can be divided into two types: Parapagus dicephalus (two heads, single trunk/abdomen/pelvis with 4-7 limbs) which was the presented case, and parapagus diprosopus (two faces on a single head, single trunk/abdomen/pelvis and four limbs). The classification of conjoined twins is summarized in Table 1 and the various subtypes of parapagus dicephalus are summarized in Table 2.[3]

The distance between the rostral ends of the embryonic disc increases as the spectrum of parapagus conjoined twins progresses from diprosopus to dicephalus. As this distance increases, there is increasing separation of the conjoined twins; the heads separate first, followed by the thoraces, while the number of limbs increases from four to a maximum of seven.[3]

Conjoined twins often have several associated anomalies in various organ systems, including musculoskeletal, cardiac, gastrointestinal, and the central nervous system. As the number of limbs and degree of separation increases, so does the likelihood of organ duplication. For example, parapagus diprosopus twins (parapagus type with the least degree of physical separation) will share a single heart. However, parapagus dicephalus twins have greater separation that may result in a partially duplicated or two separate hearts. This phenomenon is not limited to the thorax and the same principles apply to the abdomen, pelvis, and CNS.[3]

The earliest published diagnosis of conjoined twins via sonography was seven weeks' gestation (thoracopagus type).[4] A conjoined twin can be ruled out if there is visualization of two placentae or a membrane separating the twins. The following sonographic diagnostic criteria for conjoined twins have been proposed: "absent separating membrane, conjoined body parts, inseparable bodies or heads between the twins despite changes in fetal position

Table 1. Summary of the classification of conjoined twins with illustrations

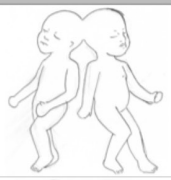
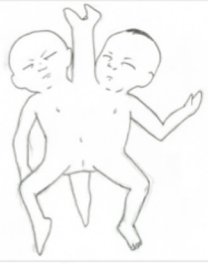
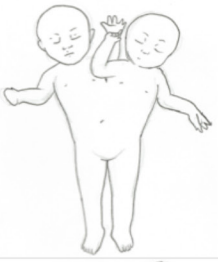
Type of Conjoined Twins (Incidence)[2-4,6]	Illustration[3] (inspired by the original artwork of Dr. Rowena Spencer)	Site of Embryologic Disc Involvement[3]	Degree of Union[3]
Craniopagus (2.7%-5.0%)		Dorsal	- union of the calvarium
Rachipagus (1.4%-2.0%)		Dorsal	- union of the vertebral column (typically thoracic or lumbar; rarely with cervical involvement)
Pygopagus (1.4%-6.0%)		Dorsal	- union of the sacrum and coccyx
Parapagus Dicephalus (13.5%-24.2%)		Ventral (Lateral)	- two distinct heads and faces - variable union of thoraces - shared lower abdomen and pelvis - 4-7 extremities
Parapagus Diprosopus (0.4%-3.8%)		Ventral (Lateral)	- two faces on a single head - shared thorax - shared abdomen and pelvis - 4 extremities
Omphalopagus (7.3-37.8%)		Ventral (Rostral)	- union at umbilicus extending superiority to include upper abdomen - may include part of the thorax (limited or no sternal involvement)
Thoracopagus (19.0%-56.3%)		Ventral (Rostral)	- union of the thoraces and upper abdomen
Cephalopagus (7.3-11.0%)		Ventral (Rostral)	- union of the entire head with a single face, extending caudal to the umbilicus
Ischiopagus (2.4%-11.0%)		Ventral (Caudal)	- union of the lower abdomen and pelvis (including urogenital structures)

Table 2. Summary of the various subtypes of parapagus dicephalus conjoined twins with illustrations

Parapagus Dicephalus Subtype[3]	Illustration[3] (inspired by the original artwork of Dr. Rowena Spencer)	Shared and Duplicated Anatomy[3]
Parapagus dicephalus dibrachius dipus		- typically share a single heart (often with congenital defects) - two esophagi emptying into a single stomach - partial to complete spinal duplication
Parapagus dicephalus dibrachius tripus		- typically share a single heart (often with congenital defects) - greater likelihood of duplication of the perineal structures - partial to complete spinal duplication
Parapagus dicephalus tribrachius dipus		- partial to complete cardiac duplication - two esophagi - (+/-) duplication of the stomach and intestinal tract - (+/-) duplication of trachea and lungs - partial to complete spinal duplication
Parapagus dicephalus tribrachius tripus		- partial to complete cardiac duplication - two esophagi - (+/-) duplication of the stomach and intestinal tract - (+/-) duplication of trachea and lungs - greater likelihood of duplication of the perineal structures - partial to complete spinal duplication
Parapagus dicephalus tetrabrachius dipus		- partial to complete cardiac duplication - two esophagi - (+/-) duplication of the stomach and intestinal tract - (+/-) duplication of trachea and lungs - partial to complete spinal duplication
Parapagus dicephalus tetrabrachius tripus		- partial to complete cardiac duplication - two esophagi - (+/-) duplication of the stomach and intestinal tract - (+/-) duplication of trachea and lungs - greater likelihood of duplication of the perineal structures - partial to complete spinal duplication

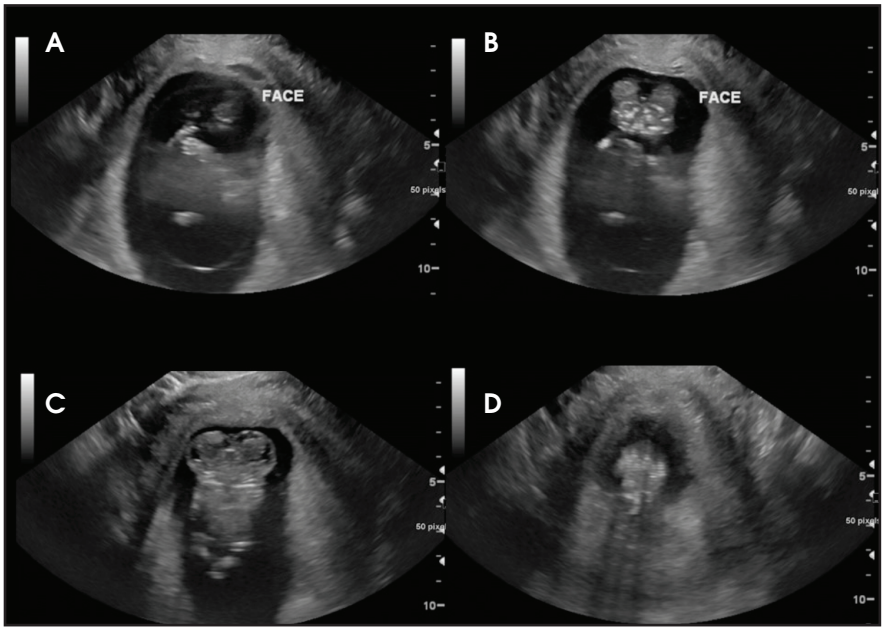


FIGURE 1. *Coronal gray scale transvaginal ultrasound scanning anterior to posterior demonstrates a fetus with (A) two upper extremities, (B) two maxillofacial structures fused at midline, (C) two heads fused at midline, a single thorax/abdomen with two lower extremities and (D) partial duplication of the spine.*

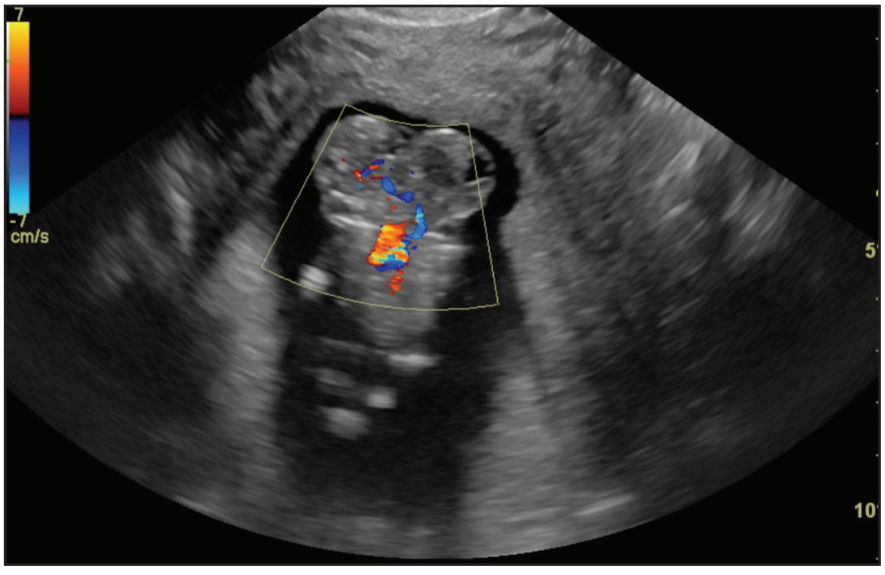


FIGURE 2*. Coronal duplex Doppler transvaginal fetal ultrasound demonstrates a single heart with shared intracranial vasculature.*

or a bifid appearance of the fetal pole in the first trimester."[5]

Other findings suggestive of conjoined twins include: An umbilical cord with greater than three vessels, persistence of fetal position and anatomy relative to one another on repeat scans. Identification of shared organs is critical in guiding management as the main clinical dilemma is determining if the twins can be surgically separated.

Multiple case reports encourage the use of three-dimensional sonography to diagnose conjoined twins.[1,6] Three-dimensional sonography improves specificity regarding the level of fusion and degree of shared anatomy. Three-dimensional sonography also aids in parent counseling as it helps demonstrate disease severity and illustrate reason for prognosis.

CONCLUSION

The early diagnosis of a conjoined twin pregnancy is important in guiding clinical management. Classification of conjoined twins is a relatively simple system based on sites of fetal union. Establishing a correct diagnosis and identifying shared organs will help determine the possibility for fetal separation and minimize morbidity.

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