3D Assisted Prenatal Sonographic Diagnosis of Dicephalic Conjoined Twins and Subsequent Planned Vaginal Delivery☆,☆☆

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ARTICLE INFO

Article history:
Received 4 March 2014
Accepted 11 March 2014
Available online 26 March 2014

Keywords:
Conjoined twins
Prenatal diagnosis
Three-dimensional ultrasound

1. Introduction

Conjoined twinning is a rare occurrence with an incidence of about 1 in 50,000 pregnancies, 60% of which result in stillbirth [1]. There is an approximate 2–3:1 female to male predominance [1]. The classification of conjoined twins is complex, but is usually based on degree and anatomic location of the fusion [2]. Parapagus twins always share a conjoined pelvis with one or two sacrums and a single symphysis [2]. Dicephalic parapagus twins share a common thorax and account for approximately 3.7% of all conjoined twins [1].

2. Case Report

A 37-year-old Caucasian female, para 1–0–2–1 was referred to our department at 27 weeks gestation for evaluation of conjoined twins. The patient was a late registrant for care at 22 weeks gestation and her initial ultrasound was performed at 26 weeks gestation showing polyhydraminos and a dicephalic fetus. The patient denied any pertinent past medical or surgical history and any history of drug or toxin exposure.

Permission for autopsy, excluding head, was obtained from the parents on the day of delivery. External examination was notable for a disproportionally large head. Both the hands and the feet appeared disproportionately large for the fetus; the fetus weighed 690 g (26 weeks), with the right lung weighing 2.5 g and the left lung weighing 5.3 g (normal 24 week fetus would have a 17 gram combined lung weight). Furthermore, the right lung demonstrated a rudimentary fourth lobe. Cardiac imaging was difficult secondary to fetal positioning and was incomplete. There was no apparent duplication of the abdominal organs and a single 2 vessel umbilical cord was present. The largest diameter of the dicephalic presenting part was 8.8 cm, equivalent to a 35 week singleton biparietal diameter (Fig. 4).

Given the findings of an assured non-viable fetal condition, the option of pregnancy termination was offered. The patient was admitted later that day and underwent an induction of labor after cardioplegia with laminaria and pitocin augmentation. She had a spontaneous vaginal delivery of a stillborn, dicephalic female fetus in cephalic presentation. The family declined chromosomal analysis, but desired a limited autopsy. Her postpartum course was uncomplicated.

3. Pathologic Findings

Both 2D and 3D ultrasound were performed on a Voluson 730 scanner (General Electric Health Care, Milwaukee, WI) with a 4–7-MHz transducer at our institution with findings consistent with dicephalic conjoined twins with acrania (Figs. 1 and 2). Two spines were identified and appeared parallel (Fig. 3) with fusion in the thoraco-lumbar region with associated rachischisis. Cardiac imaging was difficult secondary to fetal positioning and was incomplete. There was no apparent duplication of the abdominal organs and a single 2 vessel umbilical cord was present. The largest diameter of the dicephalic presenting part was 8.8 cm, equivalent to a 35 week singleton biparietal diameter (Fig. 4).

Examination of the internal organs revealed abnormalities predominantly within the thoracic cavity. Hypoplasia of the lungs was evident, with the right lung weighing 2.5 g and the left lung weighing 5.3 g (normal 24 week fetus would have a 17 gram combined lung weight). Furthermore, the right lung demonstrated a rudimentary fourth lobe. An adherent 0.4 cm diameter focus of ectopic pancreas was noted along the adventitia of the distal esophagus.
The only abdominal duplication involved the formation of a bifid gallbladder. All other abdominal organs appeared appropriate in size and orientation. Of note, an additional focus of ectopic pancreas formation was evident as an adherent 0.2 cm diameter nodule along the greater curvature. Microscopic analysis revealed extramedullary hematopoiesis in the liver, and congestion of the spleen. A single kidney was present on the right and left side and demonstrated vascular congestion.

Mild abnormalities of the pelvic organs were noted, including a uterus with constriction along the superior aspect of the fundus. The remainder of the thoracic, abdominal, and pelvic organs appeared normal in orientation, although in size corresponded to a variable gestational age of 22–28 weeks.

4. Discussion

To our knowledge there are no published reports of the use of three-dimensional ultrasonography in clarifying this nonviable form of conjoined twins, although first trimester diagnosis [3] and the use of MRI [4] to assist has been described. Recent reports have shown the value in both 2D and 3D ultrasound in the first trimester to classify conjoined twins and allow earlier reproductive choices [5–8].

Classification of conjoined twins is paramount for guiding obstetrical management. Prenatal diagnosis can help guide decisions so that both fetal and maternal morbidity and mortality can be minimized. When considered as a whole, 75% of conjoined twins do not survive the first 24 h of life [9]. The fetal chance for survival has to be weighed against the potential surgical morbidity to the mother and feasibility of vaginal delivery [9]. In this case of non-viable conjoined twins, the use of 2D and 3D ultrasound correlated very closely with the postmortem autopsy report and measurement of the combined cephalic diameter allowed for a successful trial of vaginal delivery.

References

Fig. 5. Dicephalus dipus dibrachius female stillborn.