Dicephalus dipus dibrachius: conjoined twins through the ages

Abstract

Conjoined twins are the most rare form of monozygotic twinning occurring when there is incomplete division of the embryonic disc after day 13 post conception. This is associated with a very high risk of perinatal morbidity and mortality. Prognosis is dependent on the site and extent of fusion and the degree of sharing of vital organs. Most conjoined twins die in utero or in the early neonatal period. However less severe cases can be successfully separated.

This is a review of the types of conjoined twinning, an historical perspective and a case of a rare form known as dicephalus dipus dibrachius (two heads and a single body with two arms and two legs).

Keywords: conjoined twins, dicephalus dipus dibrachius, prenatal ultrasound diagnosis.

Figure 1: Transverse view of the two heads very close to each other with no membrane between them.

Case study

Our patient is a 30-year-old gravida 4, para 1, with one previous full term lower segment caesarian section (LSCS), one miscarriage and one termination of pregnancy (TOP). She had a BMI of 33.8, was a non-smoker, on no current medication and had no significant medical history. This pregnancy was a spontaneous conception. She presented to our department for her nuchal translucency screening (NTS). She had not had any previous scans in this pregnancy.

Ultrasound was initially performed transabdominally and showed conjoined twins. Transvaginal scanning confirmed this finding. They were joined from the neck/upper chest

with two heads and one body. The heads were very close together with no membrane between them (Figure 1). There was one heart, two arms and two legs. There was a moderate amount of oedema around both twins extending from above the heads and down most of the common body (Figure 2, 3).

By crown rump length (CRL) this pregnancy was dated to be 11 weeks and 5 days which was in agreement with her last menstrual period. The scan was performed using 2D, 3D and 4D ultrasound (Figure 4, 5). Review of the 3D sweeps showed a possible four chamber heart view (Figure 6). However the amount of oedema was suggestive of early cardiac failure and it is most likely that this was an abnormal heart. Posteriorly

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Figure 2: Transverse view at the level of the joined necks with oedema seen posteriorly.

Figure 3: Sagittal view of the increased nuchal thickness/oedema posteriorly.

Figure 4: 3D image showing two heads, a single body, two arms and two legs.

Figure 5: 3D image of the twins facing each other.

there appeared to be two spines which joined in the mid thoracic region (Figure 7). The diagnosis of dicephalus dipus dibrachius conjoined twins was made based on this ultrasound.

The parents were counselled regarding these findings and opted for TOP. This was performed by dilatation and curettage (D&C) the following day.

Discussion

Monozygotic twins account for approximately 30% of all twins and dizygotic twins account for the remaining 70%. While race, maternal age, heredity and fertility enhancing medication affect dizygotic twinning, the rate of monozygotic twinning is

independent of these factors. It should be noted however that the rate of monozygotic twinning is increased in association with in vitro fertilisation (IVF) treatments particularly when the age of the egg donor, whether one's own or donated, is less than 35 years.¹

Monozygotic twinning occurs in approximately 1 in 200 births. However the incidence at conception is considerably higher. In fact the ratio of monozygotic twins compared to dizygotic twins in spontaneous miscarriage is 17:1.²

There are four types of monozygotic twins which are primarily dependent on the timing of the division of the early zygote or embryo.¹⁻⁵

Figure 6: Review of a 3D sweep showing an apparent four-chamber view of the single heart.

Dichorionic/diamniotic

Division occurs within the first four days after conception. This accounts for 18–36% of all monozygotic twins. It is not possible to distinguish this type from dizygotic twins by ultrasound and in fact 10–15% of all dichorionic/diamniotic twins are monozygotic.

Monochorionic/diamniotic

Division occurs from days 5–8 after conception. This is the most common type of monozygotic twins accounting for approximately 70% of cases.

Monochorionic/monoamniotic

Division occurs early in the second week after conception. This accounts for 2–3% of monozygotic twins.

Conjoined twins

There are two main theories regarding the embryology of conjoined twins.

The most commonly accepted theory is that division occurs or is attempted at the end of the second week on or after day 13 post conception. There is incomplete division of the monozygotic embryonic disc resulting in conjoined twins. This is known as the fission theory.

The second theory is secondary fusion between two embryonic discs that have previously separated. This is known as the fusion theory.^{6,7}

While the most famous case of conjoined twins is that of Chang and Eng born in Siam in 1811 historically there have been cases known through the centuries. The earliest case is that of a 17 cm marble statuette known as the double goddess or the Sisters of Catalhoyuk from the sixth millennium BC in Turkey (Figure 8). This appears to be the first documented case of dicephalus dipus dibrachius twins. Another early case is a stone carving dating to 80BC in Italy. These early examples were felt to be an omen and met

Figure 7: 3D image of the posterior aspect showing the joining of the two spines at the mid thoracic level.

with fear of impending disaster. Over time this feeling became one of fascination and exhibitionism as in the case of Chang and Eng.⁷

Conjoined twins occur in 1:50,000 to 1:100,000 gestations. However it is estimated that up to 60% of cases die in utero and when termination rates are factored in the resulting incidence is 1:250,000 live births. Approximately 35% are still born or die within the first 24 hours after birth.⁶ There is a female to male ratio of 3:1. It is sporadic with no known associations.

There is fusion and sharing of body parts, the site and extent of which is variable. The prognosis is highly dependent on the degree of sharing of vital organs. Additional abnormalities remote from the site of union are also common. There are several types of conjoined twins, which are named for the site of union followed by the suffix *pagus* meaning 'fixed'. There can also be overlap with more than one type involved.^{2,3,6–8}

Types of conjoined twins

Ventral union

Thoracopagus

These are joined at the thorax and upper abdomen. The organs of these two cavities, particularly the heart, are shared to varying degrees making separation impossible.

Omphalopagus

Joined at the abdomen, sharing abdominal organs, anterior abdominal wall and umbilicus.

There is often an associated omphalocele and an abnormal cord with up to six vessels. These twins have the highest chance of survival of separation at 82%.

Figure 8: A 17 cm marble statuette known as the double goddess or the Sisters of Catalhoyuk from the sixth millennium BC. This is the earliest known image of dicephalus dipus dibrachius conjoined twins. Grateful acknowledgement to Dick Osseman (www.pbbase.com/dosseman) for the use of this photo.

Thoraco-omphalopagus

There is overlap of the first two types and together are the most common type of conjoined twins accounting for 70% of cases.

Cephalopagus

Fusion from the top of the head to the level of the umbilicus. There is a common cranium with one or two faces. Each twin has two arms and two legs. The lower abdomen and pelvis are separate.

Ischiopagus

Fusion of the pelvis, lower abdomen and genitourinary (GU) systems. There may be sharing of the intestine from the ileum to the rectum and the external genitalia may be shared, fused, rudimentary or absent. This type has a 10% association with congenital heart disease (CHD).

There are two sub types of this classification:

- Ischiopagus Tripus (three legs)
- Ischiopagus Tetrapus (four legs).

Figure 9: Two-headed turtle affectionately known as Thelma and Louise at the San Antonio Zoo. Grateful acknowledgment to Craig Pelke, Curator of Amphibians and Aquatics, San Antonio Zoo, www.sazoo.org for the use of this photo.

Dorsal union

Pygopagus

Fusion of the sacrum and coccyx with sharing of a common rectum and anus. Most have fused lower spinal cords.

Craniopagus

Fusion of the cranium with sharing of the cranial vault, meninges, and dural venous sinuses.

The site of union can be sub classified as frontal, temporal, parietal or occipital and the twins will be lying at right angles to each other. There are commonly associated anomalies of the heart and GU tract.

Rachipagus

Fusion of the vertebral columns involving variable degrees of spinal sharing. The twins are facing opposite to each other each with two arms and two legs.

Lateral Union

Parapagus

Lateral fusion of the pelvis and lower abdomen. Varying degrees of the trunk and thorax are affected. There may be two, three or four arms and two or three legs.

Diprosopus

Lateral fusion of the head and body with two faces. There is a single body with two arms and two legs. This is commonly associated with neural tube defects, particularly anencephaly, and cardiac anomalies.

Cephalothoracopagus

There is a single head and face with two bodies and as many as eight extremities.

Dicephalus dipus dibrachius

This is the type of conjoined twins documented in our case. There are two heads and a single body with two arms and two legs. The vertebral columns are separate. There is a single heart with a high association with CHD. There are varying degrees of duplication of the respiratory tract, gastrointestinal tract and genitourinary tract.^{9,10} Survival of dicephalus twins generally ranges from stillbirth to a few hours. There are only a few cases found in the literature.^{9–11}

While dicephalus dipus dibrachius twinning occurs frequently in amphibians and reptiles it is exceedingly rare in humans.⁸ Of note is the recent two headed turtle born in the USA (Figure 9).

Conclusion

The prognosis for conjoined twins is highly dependent on the site and extent of fusion, the degree of sharing of vital organs and any associated anomalies. Prenatal ultrasound, particularly 3D and 4D, plays an important role in this determination. MRI may also be useful prenatally. The timing, mode and site of delivery must be carefully planned particularly if separation is being considered. Postnatally multiple imaging modalities are used before a decision can be made regarding clinical management and potential separation.

If the fusion is minimal with no sharing of vital organs separation may be possible. However this requires a multi-disciplinary approach with varying degrees of success.

Severe types of conjoined twinning with the sharing of vital organs have an extremely poor prognosis and are often considered lethal. Our case of dicephalus dipus dibrachius with a single shared heart was obviously incompatible with life.

Acknowledgement

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