

Dicephalic - dipus: A case report

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Summary

Bipagous conjoined twins are rare with an incidence of 1 in 50,000 to 1 in 100,000 births¹, but rarer still are heteropagous dicephalic dipus with an incidence of 0.1 - 0.2 per 10,000 births². No more than 4 sets of such surviving twins-sharing an undivided torso and two legs have been recorded in history³ consequently, the usual recommendation is for termination of pregnancy following prenatal diagnosis since historically, postnatal survival is unlikely⁴.

We present a case of dicephalic-dipus seen in our institution.

Key-words: *Twins, Conjoined, Duplicata incompleta dicephalus.*

Résumé

Des jumeaux bigageux unis sont rare avec une incidence de 1 en 50,000 à 1 en 100,000 naissance. Pas plus de 4 séries de tels jumeaux-partage survivant un torse non divisé et deux jambes ont été notés dans l'histoire. Par conséquent, la recommandation habituelle est l'interruption de grossesse à la suite du diagnostique prénatal parceque la survie historique et postnatale sont improbables. Nous présentons un cas d'un deciphalic dipus vue dans notre institution.

Introduction

There are 2 types of conjoined twins⁵: - Symmetrically joined conjoined twins (duplicata completa) described as bipagous (figure 1) and asymmetrically joined conjoined twins (duplicata incompleta) described as heteropagous conjoined twins. This later group could be:

- An inferior conjunction (i.e. lower body is single) also called duplicata incompleta dicephalic dipus implying two separate heads but only two lower limbs
- A middle conjunction
- Superior conjunction (i.e. upper body is single) e.g. Ischiopagous tetrapus.^{6,7,8}

We present a case of inferior conjunction featuring as a dicephalic dipus. Both bipagous and heteropagous types of conjoined twins (figures 1 and 2) have presented in our center within an interval of six weeks (September 2004 and October 2004 respectively)

Case report

A 26-year-old mother N.F. (G⁴P³⁺⁰A²) booked at Ebonyi State University Teaching Hospital (EBSUTH) at 34⁺³ weeks gestation.

Sonographer's comment on the ultrasound assessment was:- "Gestational age -31 weeks, 4day(+7-14days)

Two viable fetuses, cephalic presentation but in oblique lie

Large polyhydramniotic uterus

Single anterior fundal placenta

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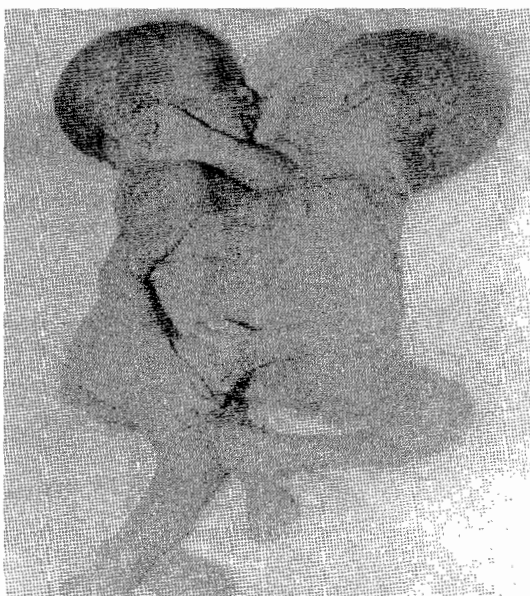


Fig. 1 Bipajour conjoint twins (EBSUTH)

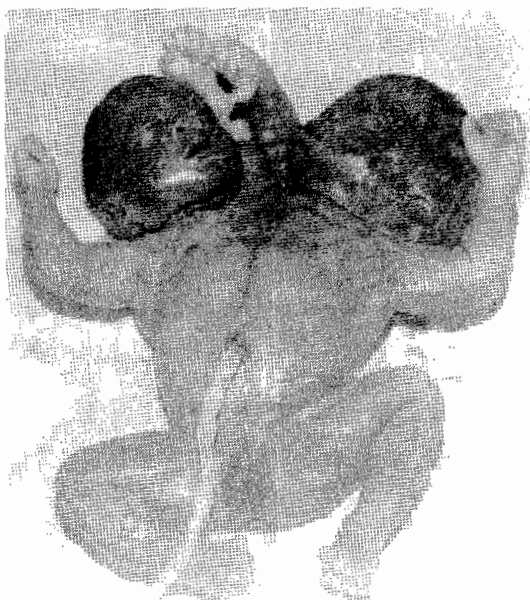


Fig. 2a Dicephalic dipus a form of heteropagous conjoint twins (EBSUTH)

Good bio-physiological profile score

No abnormalities"

(Actual ultrasound pictures not available)

She is a farmer and there is no known exposure to teratogens or infections. Her first pregnancy however, ended in a still birth; she has had no twin pregnancy but there is a



Fig. 2b *Dicephalic dipus a form of heteropagus conjoint twins (EBSUTH)*

positive family history of surviving twins on the maternal side.

At 42 weeks gestation, she fell into spontaneous labour and had symptoms suggestive of abruptio placenta.

Comment on an emergency ultrasound assessment then was:-

“Gestational age -35 weeks +1 (+7-14 days)

Both fetuses were breech; and viable

Cardiac contractility was 136 per minute very irregular

Movement of limb and body were reduced

Placenta was fundal

No praevia but pockets of gas noted anteriorly.

Ultrasound-impression was “possible concealed abruptio placenta; poor biophysiological profile score”. (Actual ultrasound pictures not also available)

An emergency lower segment caesarean section was carried out. A set of heteropagus conjoint twins with inferior conjunction was extracted; combined weight 3.5kg; one minute combined apgar score was 5 and 5 minute Apgar score was 1. Resuscitation with ambu-bag and intranasal oxygen were unsuccessful and they died one after the other within the next one-hour.

Post mortem examination revealed:

External morphology (figure 2a)

Dicephalic conjoined twins made up of two separate head and neck regions joined to single thorax, with separate right and left nipples, but a third shared nipple at the midline. Each had her own pair of well formed upper limbs (making a total of 4 upper limbs) there was a single abdomen and a single umbilical cord. The pelvis was single. However, there was only a pair of well-formed lower limbs and the perineum was single. This constituted a brachio-thoraco-omphalo-ischiopagus-dipus. There were two separate anal dimples that represented imperforate anus. The external genitalia

looked grossly ambiguous⁶. The cervical vertebrae were separate but they led to a joint upper thoracic vertebra that resulted in a single vertebral and ended in a ruptured spinal bifida cystica.

Internal morphology (figure 2b)

The clavicles were separate but they articulated medially, with a single manubrium sternum that remained joined up to the xiphisternum (figure 2b arrow 1). The hearts and pericardial cavities were separate. Each had separate lungs. The left lung of one of them (B) was hypoplastic. They shared one single diaphragm that was deficient posteriorly. This had an associated diaphragmatic hernia containing the two stomachs, duodenum and upper jejunum. The whole hypoplastic liver of B, the proximal aorta and inferior vena-cava were separate but joined at the lumbosacral region. The stomachs, duodenum, jejunum and upper ileum were separate but they joined at the terminal ileum to lead into a single large colon that ended in an imperforate anus. (Figure 2b arrow 2)

Both shared two kidneys although one was ectopic (in the pelvis).

The internal genitals were feminine with a single uterus and a pair of fallopian tubes and ovaries.

Discussion

Monovular (identical) twins results from complete division of the blastocyst provided this division occurs within 14 days.^{9,10,11} After this time, the conceptus becomes an embryo and any attempts at division can result in varying shades of incomplete division and, the later the attempted division, the more severe the defect.^{12,13,14}

Conjoint twins are therefore monovular twins that were either grossly separate (or separable) except at some points of joining (the *duplicata completa*) figure 1 or grossly inseparable (*duplicata incompleta*) figure 2.

Duplicata incompleta thus becomes the most severe form of conjoint twinning and of all these types, the *dicephalic dipus* (2heads, undivided torso, but two legs) though very frequent in amphibians and reptiles, is very rare in man with only 70 cases recorded world wide¹⁵. Fraternal (non-identical) twins are more common in blacks; but monovular and, therefore, conjoint twins are not usually influenced by heredity, race, maternal age or parity.

Since the embryopathy is traceable to the first 2-3 weeks of gestation, *dicephalus dipus* can be diagnosed in-utero within the first trimester using a high-resolution real time ultrasound.^{16, 17, 18} or better still, a magnetic resonance imaging.¹⁹ Pit falls in ultrasound diagnosis are not unusual as depicted in our case-and it is usually due to non visualization of the inseparable fetal anatomic parts and mirror-image super-imposition of structures. However, the polyhydramniotic detected here, is usually present in about 50% of conjoint twins unlike 2% in normal single pregnancy and 10% in normal twin pregnancy.^{20, 21, 22} As depicted in this case, about 70% of conjoint twins are females²³, although this was a female hermaphrodite (or inter sex anomaly).

Since postnatal survival is almost always nil, for those diagnosed before 24 weeks, vaginal delivery is recommended and not necessarily in a tertiary center^{24, 25}.

For those diagnosed near term, as in this case, delivery

is by elective caesarean section to avoid maternal trauma and associated morbidity. In between these dates, option depends on maternal and fetal factors.²⁶ Postpartum separation is uniformly fatal with only one recorded successful separation in 1987 that survived for 3yrs. Non-separation is usually the rule because, most are usually still born while only a few, like this case, surviving just for a short while.^{27,28}

Not more than 4 sets of surviving dicephalic dipus have been recorded in world history: The Perosian twins survived for one year and twenty two days while the Hensel twins lived up to 6yrs²⁹

Conclusion

Dicephalic dipus is the most severe form of conjoint twins in the duplicata incompleta group. It is also the rarest. Postnatal survival is almost impossible.

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