Case Report

# Autopsy Findings on a Pair of Dicephalic Parapagus Twins: A Case Report

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## Summary

Conjoined twins are a rare occurrence that presents significant challenges to both parents and medical care givers with many theories being advanced to explain this occurrence.“Parapagus” is a fairly recent term, in which the twins lie side by side with ventro-lateral fusion and are extremely rare representing 0.5% of all reported cases. We present a case report on post mortem findings on a set of parapagus twins delivered through caesarian section at Narok district hospital. We illustrate the various anomalies of the thoracic, abdominal cavity and central nervous system and discuss the embryologic etiologic theories.

**Key Words**: Conjoined, Dicephalic, Malformations, Autopsy

## Introduction

Conjoined twins represent a rare congenital anomaly in 1/50,000 to 1/100,000 pregnancies and are commonly referred to as Siamese twins, after Chang and Eng Bunker of Siam (Thailand) (1,2). Such conjoined twins present both parents and healthcare personnel with severe challenges in care and treatment of twins as it is accompanied by severe morbidity and mortality as well as the attendant religious, moral, ethical and legal issues (3). Conjoined twins of the parapagus (anterolaterally fused) or dicephalus (two-headed) type are extremely rare and due to shared thoraco-abdominal and spinous anatomy tend to have a poor prognosis (4-6). These dicephalic twins share a common body from the neck or upper thorax downwards with two pairs of limbs and shared reproductive organs. We present the findings at autopsy of dicephalic parapagus twins

## Case presentation

A twenty two year old lady, who was Para 3+0 Gravida 4 at 37weeks of gestation by dates, presented to our facility in labour. She had not attended antenatal clinic, had no history of twinning and no family history of children born with congenital defects. On physical exam, she had multiple foetal poles. This raised the suspicion for the possibility of a twin pregnancy. In view of the physical exam findings coupled with the fact that she had never attended antenatal clinic, an ultrasound scan was requested. An ultrasound exam revealed maternal polyhydramnios and a viable fetus with two heads and one body. An emergency caesarian section was performed and the outcome was live conjoined twins who had two heads and necks originating from one trunk. The twins passed away fifteen minutes after delivery.

Pre autopsy radiographs showed dicephalic parapagus twins with two heads; both heads had features of hydrocephalus, notably an eroded sella turcica and a beaten-copper skull appearance. In addition, the radiographs revealed two cervical spines fused into one at the spinal level of T3 (figure 1).On gross examination, the twins had two heads and necks originating from one trunk (figure2).Further, each had an open metopic suture and sutural diastasis suggestive of hydrocephalus, mandibular retrognathia and high arched palates. The common trunk had two nipples, one umbilical cord with one artery and two veins, common male genitalia and spina bifida aperta in the lumbar region.

Insert Figure 1 and 2 here

Upon dissection, we noted a cervical ectopia cordis. The heart had four chambers, tricuspid atresia and a large ventricular septal defect. The lungs were hypoplastic and extrathoracic above the ribcage and anterior to the two cervical roots. The right lung had four lobes while the left lung had two lobes. There were two sets of tracheae (one from each twin) which merged and birfurcated at the level of T3. In addition, there were two oesophagi which merged and entered the stomach at its cardia. An anterior diaphragmatic hernia was observed. There was one large liver, entirely within the thoracic cage, which had three lobes and two gall bladders. The

stomach, small and large intestines were single and normal. There were no abnormalities detected in the genito-urinary system.

**Discussion**

Conjoined twins are monozygotic, monoamniotic, and monochorionic (7). They occur in one in every 2,800 to 250,000 births, with incidence cited in literature as 1/50,000 to 1/100,000 deliveries (6, 8, 9). They have been reported as being more common in parts of Asia and Africa10. Reports from Africa place the incidence at 1:192,000 in Southern Africa (8). To the authors' knowledge there are no reported incidence data for a Kenyan population. Increased prenatal detection of conjoined twins by ultrasound may explain an apparent rise in occurrence (5). The point of union is used to classify conjoined twins; the postfix used is the Greek word pagos, which means "that which is fixed". The most common varieties encountered were thoraco-omphalopagus (28%), thoracopagus (18.5%), omphalopagus (10%), parasitic twins (10%) and craniopagus (6%) (15).They may also be named by the prefix di- (meaning two), followed by the portion of the twins that is unfused. Examples include dicephalus (two heads on one body) and dipygus (single head and torso with separated pelves and four legs) (15).Spencer proposed a parapagus classification for antero-lateral conjoned twins; dicephalus (two-head) and diprosopus (two-face) (13). Dicephalus and diprosopus twins comprised 11.2% of Machin's reciew and 13% in the Latin American Collaborative Study (4, 14). Our report is of dicephalus parapagus twins who shared common male genitalia. This is contrary to most authors, who report that 70-75% of conjoined twins are female (9, 11, 12). Nevertheless one study cited a nearly equal male: female ratio (4).

Autopsy reports have described fused hearts with complex anatomy; including right aortic arch and reversal of or transposition of great vessels (6, 7, 12, 13). Our case demonstrated a single cervical ectopia cordis, tricuspid atresia and a large ventricular septal defect. Most reports have shown two sets of lungs, which may be underdeveloped or anomalous (7, 12, 13), similar to the findings of our case at autopsy. Defects of laterality, with absent spleen or situs inversus of the right twin's abdominal organs have been reported to accompany heart abnormalities (13). We report findings of a shared liver entirely within the thoracic cage, which had three lobes and two gall bladders. This is in accord with reports that the liver, pancreas, gall bladder, genitourinary tracts and rectum may be shared (7, 12, 13). Finally, the twins had a spina bifida aperta in the lumbar region. Neural tube defects, cystic hygroma, clubfoot and imperforate anus have been reported in parapagus twins (5, 9, 14).

Two theories have been proposed to explain conjoined twinning. The first asserts that incomplete fission of a single embryonic disc occurs 13 to 15 days after the ovum is fertilized. Spencer proposed a second theory: that a fertilized ovum divides into two embryonic discs whose unusual proximity results in secondary fusion into conjoined twins as the embryos enlarge. In parapagus twins who are joined anterolaterally, it has been posited that they result from two parallel notochords in close proximity caudally, but with various points of separation towards the

rostrum. In addition, most conjoined twins face each other. In contrast, dicephalus twins' axes are side-by-side and nearly parallel. By the end of the fourth week, except for the area of the umbilical region, the entire embryo becomes covered by intact ectoderm and all the possible sites of conjoined twins may have either closed or moved to locations which are inaccessible for possible secondary fusion. This sequence of events makes "fission" the most likely explanation for the formation of conjoined twins (6).

In general, few conjoined twins survive due to heart, lung, abdominal and neurological malformations often present even in unshared structures (5, 8). Roughly 40% of conjoined twins are still born and 35% die in the first 24 hours of life (8). Moreover, only 60% of surgically treated conjoined twins survive (8). Still birth and mortality rate are extremely high in dicephalus twins (5, 9). Groner et al's dibrachius (two-arm) dicephalus twins had a remarkable 11-day survival. Rare three- and four-arm dicephalus twins live to adulthood (5).

## Conclusion

Conjoined twins are a challenge to manage and have a high mortality and morbidity. Due to extensive nature of shared organs, survival and quality of life for parapagus twins is a contentious issue. Majority do not survive to an age where surgical separation is possible.

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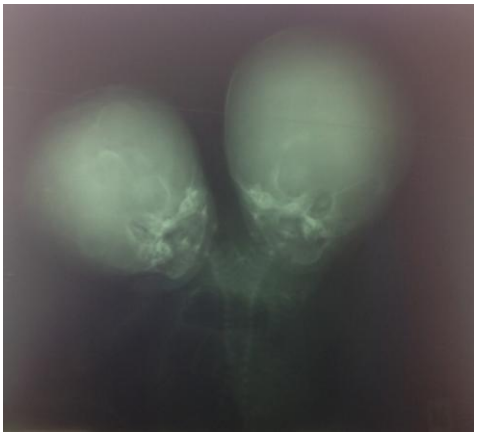

Figure 1: A radiograph showing dicephalic parapagus twins with two heads and two cervical spines which fuse into one at the spinal level of T3

Figure 2: A photograph of the twins demonstrating the two heads and necks originating from a common torso, note the normal shared external male genitalia.