

Case Report

Dicephalus tribrachius conjoined twins: Case report and review of literature

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ABSTRACT

Conjoined twins is a phenomenon believed to be caused by either delayed splitting of the fertilized ovum or fusion of the embryonic stem cells. Disorganization of mutant genes is believed to be a cause in some cases. We report a case of stillborn dicephalus tribrachius female conjoined twins with features that are not reported previously in the literature with a single umbilical artery and two urinary bladders. The cause of intrauterine death in this case was probably multifactorial i.e. severe anemia in the mother, congestive heart failure, complications of obstructed labor and multiple skull fractures.

Keywords: Conjoint twins; Dicephalus tribrachius; Sudan.

INTRODUCTION

Conjoined twins is a rare condition characterized by fusion of separable or an inseparable part or parts of the body of genetically identical, monozygotic, monoamniotic and monochorionic twins [1]. It is

still debatable whether it is due to delayed splitting of the fertilized ovum (after 13 days of ovulation) or due to fusion of embryonic stem cells that search for similar cells and attach to it as seen in case of neural tube closure [2-4]. However, gene mutation is contemplated to be the cause in some conjoined twins [5]. The incidence of conjoined twins is estimated to be one in 100,000 deliveries and the condition is more common in Africa and India. Seventy percent of conjoined twins are females and 75% die within the first 24 hours. The famous Simian parapagus twins Eng and Shang lived up to age 63 (1811-1874) and have had 22 children, and some of their grand daughters produced normal twins.

CASE REPORT

An 18-year-old primigravida presented to Ombada Teaching Hospital in labor pains that continued for more than 24 hours prior to admission. She failed to attend antenatal care clinic. Examination revealed severe anemia, multiple gestation and irregular slow heart rate. No ultrasound scan could be done on arrival

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to hospital. Emergency lower segment Caesarean section was carried out, with blood transfusion and an antibiotic coverage. The product was a fresh stillborn female conjoined twins with one trunk, two heads, two well formed arms, a rudimentary arm in between the two heads (Figure 1), female external genitalia and two well formed legs (Figure 2).

Birth weight was 3.7 kg and length was 48.5 cm. Right head circumference was 30 cm and there was excessive molding, multiple fractures and no apparent dysmorphic features. Left head circumference was 34.5 cm. with evidence of excessive molding and no apparent fractures or dysmorphic features. Chest was symmetrical with two equidistant nipples. Abdomen was normal in contour and the umbilical

cord contained one vein and a single artery. Liver was palpable in the right hypochondrium. Examination of the back showed two spines with no kyphosis or scoliosis. There were a well formed female external genitalia and an anus. Documentary ultrasound scan showed a normal liver in the right hypochondrium, two kidneys with dilatation of the upper part of the right side ureter and two urinary bladders. Two hearts could be identified with dilatation of the one located on the right side. X-ray (Figure 3) showed multiple fractures of the skull bones of the right head and two separate vertebral columns that join at the pelvic brim. Upper and lower limbs were complete and normal. There was a long bone and two clavicles in-between the two heads indicating a rudimentary third arm.

Figure 1: Dicephalus tribrachius conjoined twins: Rudimentary third arm.



Figure 2: Dicephalus tribrachius conjoined twins: Female external genitalia and two well formed legs.



Figure 3: X-ray showed multiple fractures of the skull bones of the right head and two separate vertebral columns that join at the pelvic brim. Note the long bone and two clavicles in-between the two heads indicating a rudimentary third arm.



DISCUSSION

The earliest documented report of conjoined twins dates back to the year 945 AC from Armenia. In the Islamic history, Ibn Kathir mentioned that Hashim and Abd Shams, the sons of Abd Minaf Ibn Qusai were partially conjoined twins who were separated by their father [6]. Dicephalus twins were reported from Sardinia in 1829 and Scotland in 1940 [7]. The oldest known living Dicephalus twins are Abigail and Brittany Hensel who were born in Minnesota in 1990 with two heads, two arms and two legs. Brittany got engaged in 2011 and she is planning to get married. Dicephalus twins with 4 arms were reported from Turkey, and with 4 arms and 3 legs from Russia [8]. Conjoined twins are usually classified according to the body part at which their bodies are joined [9]. Diplopagus (pagus: fixed to) means symmetrical and well formed twins. Heteropagus indicates asymmetry i.e. one complete twin and a parasitic incomplete twin. Acardiac twin denotes the presence of a complete twin and another incomplete one formed of body and legs with no heart. Well recognized patterns of conjoined twins include the following:

- Craniopagus: (2%) Joined by portion of the skull only (occipital, vertical, frontal or parietal bones). Brain may be shared.
- Cephalopagus: Joined by head and neck. Brain is usually malformed and therefore they are either stillborn or die soon after birth.
- Cephalothoracopagus: Joined at heads, neck and chest with separate arms, legs and one or two faces. Brain and heart are shared and it is almost always non-viable.
- Syncephalus: One head with a single face, four ears and two bodies.
- Diprosopus: Two faces in a single head. Very rare in humans but common in animals.
- Dicephalus: Two heads on a single body with one genital system; two, three or four arms, two hearts

and two legs

- Tricephalus: Three heads on a single body
- Thoracopagus: (35%) Joined at the upper part of the chest with separate heads, arms and legs. There may be one or two hearts.
- Xiphopagus: Joined at the xiphoid process only. They have separate organs except for the liver that might be shared.
- Omphalopagus: (6%) Joined at the abdomen and share parts of the gastro-intestinal tract but not the heart.
- Ischiopagus: (6%) Joined at the lower spine end-to-end at a 180° angle. There are four arms, two, three or four legs and one external set of genitalia and anus.
- Omphalo-ischiopagus: Same as Ischiopagus but facing each other with a joined abdomen
- Rachipagus: Joined back-to-back above the lumbar spine. Parasitic twins are commonly seen as rachipagus.
- Pyopagus: (19%) Joined back-to-back at the lower back and pelvis
- Parapagus: (5%) Joined side-by-side at the lower half of the body. Heart is sometimes involved
- Triplets: Three joined twins.
- Unclassified twins are those with no definite anatomical structures that indicate two fetuses.

The presence of a single umbilical artery denotes the presence of other congenital malformations that could have been revealed by post-mortem examination in the present case, which was denied by the parents. Intra-uterine death in this case was probably multi-factorial i.e. complications of severe anemia in the mother, obstructed labor, multiple skull fractures and right side heart failure. Death of one of the conjoint twins is usually followed by the death of the other. Most of the dicephalus twins have one uro-genital system. In our case there were two urinary bladders and, to our knowledge, this is the first documented case

with such anomaly.

Surgical separation of conjoined twins was attempted in 1950. The first successful pyopagus twin separation was performed in Louisiana in 1953.

With the development of microsurgery and cardiovascular surgery, many cases are successfully

separated in different parts of the world [10].

However, for ethical reasons, surgery should not be attempted if it deemed to be hazardous or may lead to scarify or disable one twin for the sake of the other [11]. Successful surgical separation of conjoined twins needs multi-disciplinary approach of an experienced team [12].

References:

1. Kamal K. Conjoined twins. Available at <http://emedicine.medscape.com/article/934680-overview> , Accessed 05 Sep, 2011.
2. Kaufman M H. Embryology of Conjoined Twins. *Child Nerv Syst* 2004; 20 (8-9):509-25
3. Gilbert-Barness E, Debich-Spicer D, Opitz J N. Conjoined Twins: morphogenesis of the heart and a review. *Am J Med Genet A* 2003; 120 A(4):568-82
4. Crauciuc E, Pricop F, St.Antone D. Dicephalic Conjoined Twins: a case report. *Rev Med Chir Soc Med Nat Lasi* 2002; 106(1): 169-73
5. Petzel M A, Erickson R P. Disorganization: a possible cause of apparent conjoined twinning. *J Med Genet* 1991; 28(10):712-4
6. The Life of Prophet Muhammad: Al-Sira Al-Nabawiyya by Ibn Kathir, Trevor Le Gassick, Muneer Fareed, pg. 132
7. Harma M, Mill Z, Oksuzler C. Vaginal Delivery of dicephalic parapagus conjoined twins: a case report and literature review. *J Exp Med* 2005; 205 (2): 179-85
8. Available at: http://en.wikipedia.org/wiki/Conjoined_twins. Accessed July 10, 2011.
9. Children Hospital of Philadelphia. Web site for conjoined twins. Available at: <http://www.chop.edu/consumer/jsp/division/generic.jsp?id=83379>. Accessed July 10, 2011.
10. Kamak I, Cifteci A O, Buyukpcukcu N. Epigastric Heteropagus: a case report with review of the literature. *Eur J Pediatr Surg* 1999; 9(5): 347-50
11. Groner J I, Teske D W, Teich S. Dicephalus dipus dibrachius: an unusual case of conjoined twins. *J Pediatr Surg* 1996; 31(12):698-700.
12. Al Rabeeah A. Conjoined Twins – past, present and future. *J Pediatr Surg* 2006; 41(5): 1000-4.