Annals of Clinical Obstetrics and Gynecology

ISSN: 2694-491X

Case Report

A Case Report of Dicephalic Tribrachius Dipus Parapagus Co-Joined Twin

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Abstract

This is a case of undiagnosed dicephalic (two heads) tribrachius (three upper extremities) dipus (two lower extremities) parapagus co-joined twin which is a very rare type of co-joint twin. A 28-year-old gravida two, para one presented to the emergency gynecology outpatient department of Zewditu Memorial Hospital (ZMH) in May 2014 with the chief complaint of leakage of liquor and pushing down of three hours duration. Emergency cesarean section was done for an indication of twin pregnancy first non-vertex presentation (and intra-operatively co-joint twin diagnosed) to affect the delivery of alive co-joint twin (dicephalic, tribrachius, dipus parapagus) weighing 3900 grams with APGAR score of 3,4,5 (for both) in 1st, 5th &10th minutes but expired after 30 minutes of life. Mother was discharged on the 4th postoperative day in good condition. After a year she got pregnant and delivered by C-section twin alive and healthy babies.

Keywords: Co-joined; Dicephalic; Dipus; Ethiopia; Parapagus; Tribrachius; Zewditu memorial hospital (ZMH)

Introduction

Conjoined twins are the most extreme form of twinning, occurring in about 1% of monozygotic twins with incidence of about 1 in 50,000 to 1 in 100,000 births and the condition is more common in Africa and India. Over the last decade, the widespread use of assisted reproductive techniques has decreased the rate of monochorionic twins, and consequently may have changed the prevalence of conjoined twins. 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. Approximately 75% of conjoined twins are females; and 75% die within the first 24 hours. The first and the second commonest conjoined twins reported were thoracophagus and omphalophagus, respectively. Here we are reporting a dicephalic tribrachius dipus Parapagus conjoined twin diagnosed intrapartum.

Conjoined twins have an anomaly of duplication in single-zygote conceptions which is thought to occur because of splitting and incomplete separation of the inner cell mass at 13-15 days post-fertilization. The prenatal diagnosis of conjoined twins began in 1976. Without doubt, accurate and early diagnosis of fetal malformations will affect management and perinatal outcomes.

Case Presentation

A 27 years old gravida 2, para 1; gestational age from LNMP 41 weeks and 3 days; presented to emergency gyn OPD with the chief complaint of pushing down pain and leakage of liquor of 3 hours duration.

She started antenatal care from six months of gestations at health center; referred to Zewditu Memorial Hospital after ultrasound showed polyhydramnios at 32 weeks plus 6 days and had four visits.

The first delivery was seven years back by spontaneous vertex delivery at a health center at nine months of gestation. The outcome was a 3.5 kg alive female neonate. No ante-/intra-/post-partum complications. The child is a grade one student.

Otherwise, no history of taking drugs for fertility, no family history of twinning, no personal or family history of chronic medical illness.

She had three obstetric ultrasounds at different places; none of them diagnosed co-joined twins.

Physical examination on arrival to the OPD blood pressure systolic 130 MMHg-140 MMHg , diastolic 80 MMHg-90 MMHg. Multiple fetal parts appreciated fetal heart beat 140'/120'. Cervix was seven centimeters dilated and footling breech.

For an indication of twin pregnancy 1st non-vertex cesarean section done under general anesthesia; through Pfannenstiel incision, lower uterine transverse incision made but it was difficult to delivery; while examining two heads, three upper extremities and one thorax found then inverted T-uterine incision made for suspect of co-joint twin to affect the delivery of alive male co joined twin neonate weight 3.9 kg with APGAR score of 3,4,5 (for both). Had two separated heads, three upper extremities (one in between with six fingers), attached starting from thorax, one umbilical cord, one external male genitalia, one anus and two lower extremities (Figures 1 and 2). The neonates resuscitated & transferred to NICU, after 30 minutes of life expired. Placenta was single monochorionic and monoamniotic weight 800 grams.

After the parents signed not to take for burry, autopsy was taken to for post mortem examination. The finding was Malformations of the heart (Left heart: abnormal venous return (persistent left

Citation: Endalkachew Mekonnen Assefa. A Case Report of Dicephalic Tribrachius Dipus Parapagus Co-Joined Twin. Ann Clin Obstet Gynecol. 2021;2(1):1007.

Publisher Name: Medtext Publications LLC

Manuscript compiled: Aug 12th, 2021

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upper cardinal vein) [1]. Eccentric hypertrophy of the Rt. Ventricle. Right heart: abnormal venous return (persistent Lt. Super. Vena cava in addition to both vena cava). Fused atria, common AV-valve (tricuspid), single (common) ventricle. Hyperplastic pulmonary artery, hpoplastic aorta (retrograde perfusion of the arch, atretic aortic valve). Fusion of the descending thoracic aorta at the level of diaphragm [2] Alimentary tract; Atresia of the left proximal esophagus (without Communication with trachea (Figure 3) [3] and Fusion of the intestinal tract 50 cm distal to the pylorus (Figure 4), Meckel diverticulum [4] Fused liver with 2 gall bladders and an accessory mid lobe protruding into thoracic cavity (Figure 5) [5] both thyroids normal position and size [6]. Thymus: both normal size and position. Single left sided spleen [7]. Separated respiratory systems [8]. Urogenital tract separated (normal) both testitcles Intraperitoneal [9]. Fusion of the vertebral column at level of sacrum (Figure 6).

Post-operative hematocrit was 34%. The mother discharged with smooth post-operative course and had two post-natal visits [10].

Figure 1: Dicephalic tribrachius dipus parapagus co-joined twin (Photo 1) immediately after delivery.

Figure 2: Dicephalic tribrachius dipus parapagus co-joined twin (Photo 1) immediately after delivery.

Figure 3: Autopsy of atresia of the left proximal esophagus (without communication with trachea).

Figure 4: Autopsy of fused intestinal tract 50 cm distal to the pylorus (arrow).

Discussion

Conjoined twin is a rare phenomenon, occurring in 1 in 50,000 to 100,000. However, since 60% are stillborn or die shortly after, the true incidence is around 1 in 200,000 live births. In a recent epidemiological study, it was found that the total prevalence was 1.47 per 100,000 births. A significant female predominance particularly of the thoracopagus type and a significant male predominance in parapagus and parasitic types have been detected. Whereas the incidence of conjoined twinning in our country is unknown; there have been a few previous reports of parapagus conjoined twins from Ethiopia.

The embryonic disk starts to differentiate on the 13th day. If the split occurs after day 13, then the twins will share body parts in addition to sharing of their chorion and amnion (mono chorionic, mono amniotic) [11,12].

There are two theories of how conjoined twins are formed. The more widely accepted theory is the "fission theory" which states that conjoined twins occur when a fertilized ovum begins to split into identical twins, but is somehow interrupted during the process and develops into two partially formed individuals who are stuck together [4]. It is argued that conjoined twinning cannot possibly

Figure 5: Autopsy of fused liver with 2 gall bladders and an accessory mid-lobe protruding into thoracic cavity.

Figure 6: Autopsy of fused vertebral column at level of sacrum.

result from a "fission event", and can result from the fusion of mono amniotic twins [5]. He proposed that two mono-ovular embryonic discs may lie adjacent to one another at various angles, and may be become secondarily united dorsally, caudally, laterally or dorsally and symmetrically or asymmetrically but always homologous [6]. It might seem logical to assume that dicephalus twins arise from two separate, nearly parallel notochords on one embryonic disc, very close together caudally, but with varying degrees of separation rostrally. No significant genetic-, environmental-or demographic-associated factors have been identified.

Parapagus twins represent less than 0.5% of all reported cases of conjoined twins. The classification of conjoined twins is based on the site of union. The suffix-pagus is used meaning fastened. Thoracopagus is the commonest variety, accounting for 40% of reported cases. It is followed by omphalopagus (32%), pygopagus (19%), ischiopagus (6%), and craniopagus (2%). Parapagus twins joined antero-lateral result from two nearly parallel notochords in close proximity. This anomaly represents less than 0.5% of all reported cases of conjoined twins. Regardless of the site of union, variations occur with regard to the internal organs. In the literature, there are case reports presenting dicephalic (two heads with one body) conjoined twins, either tetrabrachius or dibrachius. Some of them are reported to be stillborn while others died shortly after birth.

Dicephalic conjoined twins may have a long life. Their main predictor of survival is the degree of conjunction and abnormality of the hearts. The majority of stillborn have cardiopulmonary malformations that are incompatible with extrauterine life. It is desirable to separate less extensively conjoined cases. In dicephalic and in certain other types of extensively conjoined twins, the anatomic structure is such that it is unlikely that both twins will survive an attempt at separation. In a review by Bondeson, dicephalic conjoined twins in the past and present were investigated. The Tocci brothers (1877-1940) were also parapagus conjoined twins (dicephalic, tetrabrachius, dipus). Each boy controlled the leg on his respective side, but they were never able to coordinate their movements and could never walk without assistance throughout their life. Poor muscular development, caused by prolonged bed rest and inactivity, is presumed to be the reason for their immobility. Their immobility was also advantageous for their parents because this made their exploitation much easier.

The Hensel twins (dicephalic, dibrachius (tribrachius during delivery), dipus) are parapagus conjoined twins currently living as unseparated which is almost the same to our case. Although each twin controlled the arm and leg on her side, they were remarkably agile, coordinated their movements perfectly, and could not only walk, but ran, swam, and rode a bicycle. They are now 27 years old, alive and well.

Based on our literature search, there are few dicephalic (either tetrabrachius or dibrachius) parapagus conjoined twins who remained alive as unseparated. Cases of conjoined twins occur so rarely; it is important to learn as much as possible from each case. Furthermore, they can perform their activities of daily living, can walk independently and are continuing their education.

In conclusion, un-separated conjoined twins should not be left to their fate. Rehabilitative approaches can help them to become functionally active, and if needed, with the help of assistive devices, they can live independently.

Acknowledgement

We acknowledge Prof. Jacob Schneider for his enormous work helping us to know the detail post mortem autopsy results.

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