



Management of Xipho-Omphalopagus Conjoint Twins in North Central Nigeria: A Case Report and Discussion of Our Experience

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Authors' contributions

This work was carried out in collaboration among all authors. Authors SO, PMM, OOO and YJC designed the study, performed the statistical analysis, wrote the protocol and wrote the first draft of the manuscript. Authors KEO, JA and SO managed the analyses of the study. Authors BO, AMM, AY and NM managed the literature searches. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

This case report describes the management of Xipho-omphalopagus conjoint twins in North central Nigeria. Conjoint twins have been reported in literature and they present very enigmatic and grotesque appearances. The aim of management of most cases is to separate them into two independent persons, but in case of parasitic twins, is to remove the parasite from the autosite. On the 11th of June 2018, a set of conjoint twins were referred to the Paediatric Surgery Division of our Hospital. The patients were delivered by a caesarian section after a prolonged labour at St Mary's Hospital in Gwagwalada. Two operation teams were constituted which undertook six sessions of

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conference discussions and several drills on how to mobilize, position, transport, intubate and monitor these patients through the course of operation and in the post-operative care. The management of conjoint twins is multidisciplinary and requires that the managing team, the parents and community have a common understanding with a view of the successful separation of the babies. The society is very limited in the knowledge of the requirements for care, yet put so much pressure on the doctors and the health care institutions to deliver on the separation of conjoint twins, it would be appropriate as was done in this case to constitute a media team of the hospital to respond to concerns from media houses and interested parties so as to allow the managing team concentrate on their professional duties.

Keywords: Conjoint twins; xipho-omphalopagus; separation; Nigeria.

1. INTRODUCTION

A xipho-omphalopagus conjoint twins is one in which the babies are fused at the lower thoracic cage involving the xiphisternum and the anterior abdominal wall with the presence of an omphalocoele at the level of the umbilicus. The conjoining is an embryonic event and may result in fusion or sharing of organ(s). Conjoint twins present very enigmatic and grotesque appearances with complex clinical scenarios and challenges [1]. Thus, the care of such cases requires a multi-disciplinary approach in order to achieve a successful outcome. In monozygotic twins pregnancy, the fertilized ovum divides and develops into two identical human being, however failure of separation of contiguous embryonic cells at any site results in fusion or sharing of symmetric parts of the bodies that develop from those cells. In dizygotic non identical twins, two different ova are fertilized by independent spermatozoa and hence no conjoining occurs in them. Globally, incidence of 1 /50,000-200,000 live birth is reported for conjoint twins, but the successes in management of most of the cases have been in the developed world with well-equipped hospitals [2,3].

The aim of management of most cases is to separate them into two independent persons, but in case of parasitic twins, is to remove the parasite from the autosite. The first reported separation of omphalopagus twins was in 1957 by Bertam Kartz and his team. Since then, several cases of successful separation of other conjoint twins such as the craniopagus, cephalopagus , parapagus, parasitic twins, et cetera with varying challenges in their respective management have been reported in the literature [4,5,6].

In Nigeria, few cases of separation have been documented but with very poor outcome [5,7]. We hereby document our experience with the

successful separation of a set of xipho-omphalopagus twins. From literature available to us, this represents the first successfully managed case of conjoint twins in the north central Nigeria which is located between latitude 8.0 and 11.0 N and longitude 3.0 and 10. 0E with a population of about 30 million people.

2. PRESENTATION OF CASE

On the 11th of June 2018, a set of conjoint twins were referred to the Paediatric Surgery Division of our Hospital. The patients were delivered by a caesarian section after a prolonged labour at St Mary's Hospital in Gwagwalada. They were delivered to 31 year old Para 2+1, married woman whose first child died at birth however, the cause of death was not known as autopsy was not done. The pregnancy was booked and uneventful, and there was no family history of twinning, congenital anomalies or consanguinity. Their mother took only routine ante natal medications and there was no history of exposure to radiation or known teratogen. A late third trimester antenatal ultrasound scan diagnosed the twin pregnancy but did not pick the fusion of the babies which was discovered at the Caesarian section following difficulty with the delivery of the first twin. Both twins had Apgar scores 7 at the first minute of life and were stable before being transferred to our hospital.

2.1 Management

They were referred to our centre within 12 hrs postnatally, they were pink and afebrile. Their combined weight was 5.1 kg, they were in opisthotonus position with fusion of both male neonates from the xiphisternum down to the umbilicus with moderate size omphalocoele. They had normal penis and the testes were palpable within the scrotum. They had separate cardiac activities and were not in respiratory distress. Their head circumferences were normal

for age and their anterior and posterior fontanelles were patent and normotensive. They had normal upper and lower limbs and primitive reflexes. A diagnosis of an Xipho-omphalopagus tetrapus conjoint twins in stable state was made (see Fig. 1). They were commenced on intravenous 10% dextrose in water and prophylactic antibiotics. Both twins passed meconium within the first 24 hours of life and later started on expressed breast milk feeding. By the second week of life they had two - dimensional echocardiography (2D Echo) which was essentially normal for both of them. Both abdominal ultrasound and CT scans revealed separate kidneys and urinary bladders, however, there was fusion of the hepato-biliary system (see Fig. 3) but they had independent gall bladders. An oral contrast enhanced CT scan of the gastrointestinal system ruled out its fusion (see Figs. 2 and 4). A thoracic CT sections ruled out fusion of the hearts (see Fig. 5). With progressive weight gain and the increasing activities of the babies and the worsening opisthotonus posture and absence of other life threatening conditions, an early separation operation was contemplated as against the traditional 12 months or more of age.

2.2 Pre-Operative Planning and Operation

Two operation teams (named red and blue) were constituted which undertook six sessions of conference discussions and several drills and rehearsals using manikins on how to mobilize, transport to and from the theatre, position, intubate, monitor these patients through the course of operation and in the post-operative care. They were made up of the Paediatric surgeons, plastic surgeons, the neonatal care nurses, the peri-operative nurses, the anesthesiologists. A haematologist and a blood bank scientist were part of the team to ensure availability of blood for transfusion. The red team was to perform the separation operation and handed over the second baby to the blue team. Each team would then continue with further reconstruction as was necessary for each baby. Eventually, at 18 weeks of age with combined weight of 12.5 kg, they had successful separation operation with both twins alive. Intraoperative findings were fusion of the whole breadth of the liver at the level of the falciform ligament. There was also complete fusion of the xiphi-sternal bones but the bowels were not. The sternal bone was bisected and with the aid of harmonic

scalpels and diathermy, the shared liver was divided into near equal halves. Haemostasis was secured and the abdominal walls were reconstructed. Intra operative blood transfusion 100 ml of fresh whole blood was done for the twins.



Fig. 1. The new born male conjoint twins



Fig. 2. A contrast enhanced CT showing non spillage of contrast into the intestine of the second

2.3 Post-Operative Care

Both patients did not require mechanical ventilation post-operatively and commenced oral intake within 48 hours. They remained stable, and their operative wounds healed without surgical site infection. Following good post – operative course, they were discharged home at 3 weeks post operatively with their weight at 6.5 kg and 6.7 kg respectively.

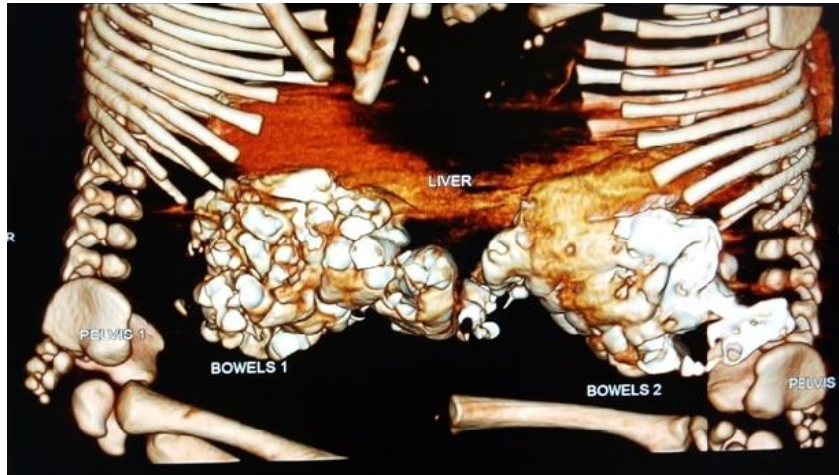


Fig. 3. CT reconstructed image of the twins showing fusion of the liver, but independent intestine

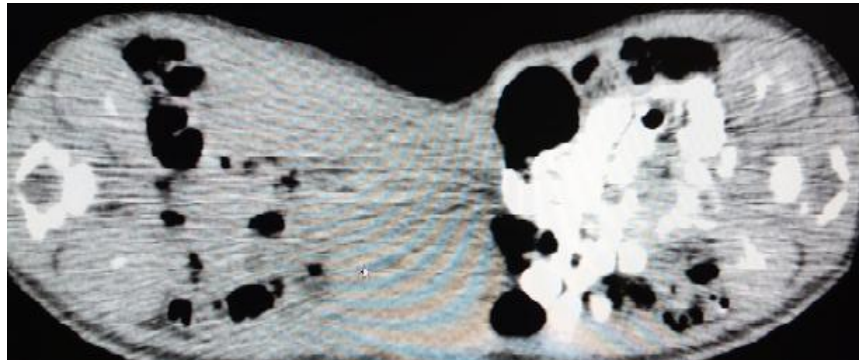


Fig. 4. CT transverse section showing contrast in the intestine of one of the twins but absent in the other



Fig. 5. CT scan of the thorax showing the separate hearts

3. DISCUSSION

Globally, the management of conjoint twins always excites the interest of the clinicians and non- clinicians alike as it poses an enigma. There

are different types of conjoint twins each with its unique characteristics and peculiarity of management and hence morbidities and mortalities associated with them. The xipho-omphalopagus and omphalopagus conjoint twins

have been reported to have better prognosis following separation than other types. The more complex the conjoining, the more likely there would be morbidities and the need for multiple surgeries to correct the both anatomic and motor functions deficits [2,5,8]. The ethical issues in the management of the conjoint twins are challenging from the point of diagnosis to the separation operation hence the need for the parents to be carried along in decision making [9,10]. The key to resolving many of the enigma that surround the conjoint twins is the application of multiple investigative and diagnostic tool such as the ultrasonography(USS), MRI, CT scan, and Echocardiography to detect organs that are fused and to help plan the separation and further procedures that may be required as was done for this case. Prenatal Ultrasound (or MRI) can detect conjoint pregnancy early especially at the 20-21 weeks of gestation but our patient did only one USS at third trimester when detection of anomalies is difficult with ultrasound scan hence the inability to detect the fusion of the babies. Appropriate counseling of parents and planning for the caesarian delivery of those babies in a centre where there is expertise to handle them, avoids the dangers that vaginal delivery poses to unsuspecting mothers, however, that was not the case here as vaginal delivery was advised because the prenatal diagnosis of conjoint twins was not made prior to onset of labour [11,12,13, 14].

A multi-disciplinary collaboration which was deployed in this case enhances professionalism especially in complex surgical condition. The initial management involved the paediatric surgeons, neonatologist, neonatal nurses and the paediatric cardiologist whose role was to keep the babies healthy and stable for safe but rigorous radiological investigations. The haematologist and the laboratory scientist were resourceful in ensuring that laboratory test results were available as well as blood for transfusion. The intraoperative team headed by the Paediatric surgeons ensured the successful separation while the plastic and reconstructive surgeons ensured appropriate siting of the skin incision and adequate skin cover for the abdominal wound closure.

One of the main challenges of management of conjoint twins is skin closure after the separation, which is addressed in some cases with use of tissue expanders to lengthen the available skin but were not used in these patients as they both had enough skin for the abdominal wound closure [5]. Anaesthesia was considered one of

the most important parts of the surgical process hence a lot of planning with manikins and drills on how to safely transfer the second twin to the other operating suite were undertaken. The critical processes were positioning, intubation, monitoring and intraoperative massive blood loss and transfusion which were adequately handled by the anaesthetic team and the haematologist [15,16].

The financial implication for the care of these babies was enormous though both parents had tertiary level education, they were both economically bankrupt as they had no job to lean upon for living hence a charity organization and well-meaning individuals rose up to the occasion as well as the good gesture of the hospital management which waived the entire bill as part of her social cooperate responsibility, ensured the success of the case. Our experience was similar to report by Ademuyiwa et al. [5].

The case being its first in our Centre, offered all the professional disciplines the benefit of working together as a team and provided ample opportunity to communicate ideas and decisions under a team leader.

The society is very limited in the knowledge of the requirements for care, yet put so much pressure on the doctors and the health care institutions to deliver on the separation of conjoint twins, it would be appropriate as was done in this case to constitute a media team of the hospital to respond to concerns from media houses and interested parties so as to allow the managing team concentrate on their profession. Our experience about the media was similar to that of the Kano and Keighley Siamese twins of 1953 and 1955 [17].

4. CONCLUSION

The management of conjoint twins is multidisciplinary and requires that the managing team, the parents and community have a common understanding with a view of the successful separation of the babies. Designating and equipping hospitals as centres of excellence for correction of severe congenital anomalies such as conjoint twins would lead to proficiency and expertise in the treatment of such conditions with better outcomes in the country.

CONSENT

Consent was obtained from the parents for this publication.

ETHICAL APPROVAL

It is not applicable.

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COMPETING INTERESTS

Authors have declared that no competing interests exist.

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