

# First successfully separated set of thoraco-omphalopagus twins in Ilorin, Nigeria: A case report

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## ABSTRACT

The birth of babies sharing body part(s) and surviving independently is indeed a scientific marvel. Conjoined twinning is a rare phenomenon with various presentations, having an estimated incidence of 1 per 200,000 live births. The successes recorded by experts in developed countries in the separation of conjoined twins in recent times, especially among those with complex unions are indeed fascinating. The management of conjoined twins presents a great challenge to medical and surgical teams in tropical African countries such as ours where we are faced with challenges of limited resources and facilities, even amidst the presence of well-trained experts. The incidence of conjoined twins is difficult to report in our practice because of failures that accompanied previous cases that were managed and not reported. This is the first successful separation after two previous failed attempts at our center. We report a case of successful surgical separation of thoraco-omphalopagus conjoined twins who were undiagnosed prenatally and delivered by emergency cesarean section following prolonged labor by a primiparous woman. Babies were joined from the lower chest and the upper abdomen and they shared a single umbilicus and omphalocele sac. No internal viscus was shared apart from the torso wall. They were cared for from 12 h of live until 127 days of live when they were successfully separated by a team of medical and surgical experts. This is a report of a successful separation of conjoined twins at the University of Ilorin Teaching Hospital in North Central Nigeria. Careful interdisciplinary conduct of expert activities will ensure the survival of rare congenital malformations such as conjoined twinning even in underdeveloped climes.

**Key words:** Female, Ilorin Nigeria, outcomes, surgical separation, thoraco-omphalopagus conjoined twins

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## INTRODUCTION

A conjoined twin is a subset of monozygotic twin gestation. It is a rare phenomenon with an incidence of 1 per 200,000 live births.<sup>[1]</sup> The term “conjoined twins” refers to babies who are physically joined at some point. It results from the incomplete splitting of monozygotic (identical) twins shortly during the period of embryogenesis (fission theory), while some authors

postulate that it may actually result from the fusion of stem cells of already separated embryo (fusion theory).<sup>[1]</sup> Conjoined twinning is no doubt one of the most fascinating malformations in humans, but there have also been reported cases in other species; mammals, reptiles, birds, and fishes.<sup>[1]</sup>

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The term “pagus” originates from the Greek word meaning “that which is fixed,” and as proposed by Spencer and colleagues, the site of union forms the basis of classification.<sup>[2,3]</sup>

Thoracopagus (joined at the chest) - 40%, omphalopagus (joined at the abdomen) - 34%, Pygopagus (joined at the buttocks) – 18%, Ischiopagus (joined at the ischium) – 6%, Craniopagus (joined at the head) – 2%.

A rare type of conjoined twins, known as heteropagus twinning, occurs when one incompletely formed (parasitic) twin is dependent on a well-formed one.

Conjoined twins were called Siamese twins because the celebrated conjoined twins were born in Siam (now Thailand) in 1811. They were males and lived for about 62 years.<sup>[4]</sup>

The actual cause of this malformation is unknown, with some sporadic occurrences, unrelated to hereditary or maternal age factors.<sup>[4]</sup> It has been found to be more in live-born females with a female-to-male ratio of 3:1; however, this condition occurs more in male fetuses as evidenced by its higher rates in male stillborns. Male conjoined twins are also more likely to die shortly after birth, implying that female conjoined fetuses have a better chance at survival than their male counterparts.<sup>[2]</sup>

There have been several reports of conjoined twins in different parts of the globe. In Nigeria, the earliest reported case of conjoined twins was born in Sokoto on December 20, 1935, to a 25-year-old woman.<sup>[5]</sup> The incidence of conjoined twins in Nigeria is unknown; an article published in 2001 reported that over the preceding 60 years, there were 12 published cases nationwide.<sup>[6]</sup> This probably does not reflect the true incidence in Nigeria as many cases are not reported on account of the associated stigma and poor outcome,<sup>[6]</sup> showing the overall reporting of conjoined twins to be low in Nigeria. Our review of available literature shows that 18 cases have been reported across the country over 76 years from 1935 to 2012<sup>[6-9]</sup> but there are still cases of conjoined twins that were found in the Nigerian news reports that never made the medical literature.<sup>[4]</sup>

We aim to report a successful separation of a set of conjoined twins at the University of Ilorin Teaching Hospital, Ilorin, Nigeria, after 2 failed previous attempts in 2005 and 2015.

## CASE REPORT

A set of female conjoined twins referred to our center; University of Ilorin Teaching Hospital from the general hospital (secondary level) New Bussa, Niger state, Nigeria, on August 31, 2020, at 12 hours of life [Figure 1] on account of the fusion of the twins in the lower chest and abdomen.



**Figure 1:** Shortly after caesarean delivery

They were delivered by a 25-year-old primiparous woman through emergency cesarean section following prolonged labor in a peripheral clinic (primary level) complicated by maternal hemorrhage. The combined birth weight was 5.57 kg and both cried immediately after birth.

The pregnancy was booked at a private hospital (primary level) and the mother was regular on routine medications and antenatal visits. There was no history of maternal febrile illness, abuse of drugs, or the presence of comorbidities. None of the obstetric ultrasound scans done at antenatal visits diagnosed conjoined twins. Mother was in labor for 17 hours and sustained injuries to her perineum before referral following no progression of footling breech of the first twin.

At presentation, both twins were conscious, not in distress, pink in room air. They were joined at the level of xiphisternum down to a point just below the umbilicus. There was an omphalocele about 8 cm wide with a single umbilical cord stump of 4 cm. They were admitted into the neonatal intensive care unit (ICU) and were designated as “twin R” and “twin L” (right and left). They were placed on intravenous cefuroxime, metronidazole, and Vitamin K, and on the daily dressing of the omphalocele with povidone-iodine and sufratulle™.

They, however, developed early neonatal sepsis, blood culture yielded mixed growth of *klebsiella pneumoniae* and *pseudomonas eruginosa* sensitive to amoxyl-clavulanic acid (augmentin), and gentimicin following which antibiotics were changed appropriately. Serum electrolytes, urea and creatinine, full blood count, and clotting profile were normal, viral screening was negative. Sepsis resolved by 3<sup>rd</sup> week [Figure 2]. Feeding was by expressed breast milk with cup and spoon initially and later by direct sucking.

Transfontanelle ultrasound was normal but the initial echocardiogram showed a patent ductus arteriosus in the

right twin which later closed before the time of surgery. A serial abdominopelvic ultrasound scan suggested normal separate viscera. Doppler ultrasound showed normal hepatic arteries and veins, portal vein with normal Doppler, however, the liver abuts in the midline with a well-demonstrated line of cleavage, and both gallbladders were within normal limits [Figure 3]. Abdominal computed tomography (CT) scan also shows separate viscera with no vascular exchange and clear hepatic capsular demarcation [Figure 4]. The bowel loops were crossing over but no communication [Figure 5]. Cardiac and major vessel configurations are normal. There was xiphisternal bone bridge but no diaphragmatic defect [Figure 6]. They remained in the neonatal ICU, until the age of 100 days at which their combined weight was 10 kg. This was to ensure they had close monitoring, prevention of sepsis, protection from public pressure on the management team, and stigmatization of parents before the eventual separation surgery was performed at 127 days of life.

Findings at surgery [Figure 6] were that of a xipho-omphalopagus connection of the conjoined twins involving the xiphisternum, anterior abdominal wall skin and muscle, healed omphalocele scar, separate liver, gallbladder and biliary tract, and separate small and large bowel.

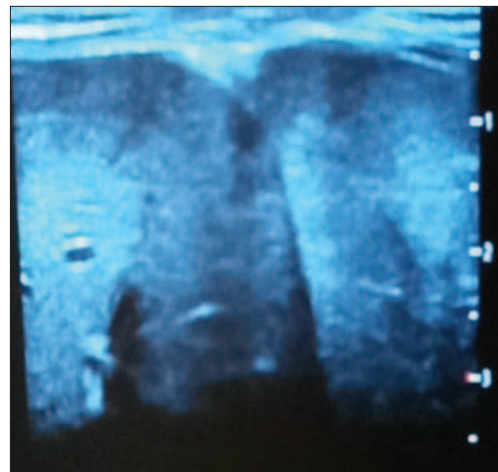
The cecum of the left twin was on the left upper quadrant with wide mesenteric base and duodenojejunal junction on the left of the midline. For the right twin, small and large bowels are in normal position. They were transferred to the ICU postoperatively for elective monitoring [Figure 7]. The immediate postoperative period was turbulent characterized by fever, and abdominal distention from fecal impaction until the 3<sup>rd</sup> day. They were placed on antibiotics, analgesics, and suppositories. They were transferred to the pediatric surgical ward on the 5<sup>th</sup> day postoperative after resolution of fever and commencement of oral feeds on 4<sup>th</sup> day. Both developed intermittent pyrexia on the ward and were positive for malaria parasite which was treated appropriately. Stitches were removed on day 10 and they were discharged from the ward 14<sup>th</sup> day postoperative. Individual weights at discharge were 5.68 kg and 5.23 kg for the right and left twins, respectively. They were followed up at the outpatient clinic and they are now over 1 year old [Figure 7].

## DISCUSSION

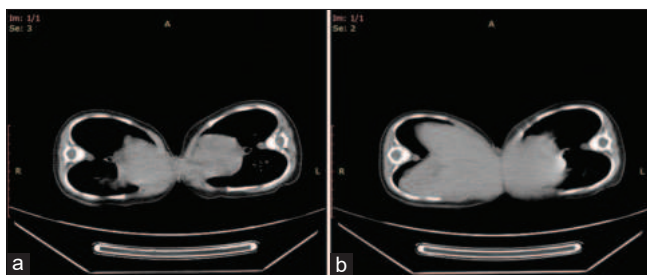
Thoracophagus and omphalopagus together represent the most common presentation of conjoined twins accounting for about 74% of cases.<sup>[2]</sup>



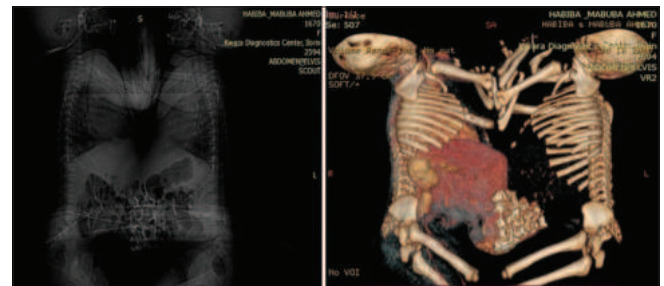
**Figure 2:** The twin at 25 days



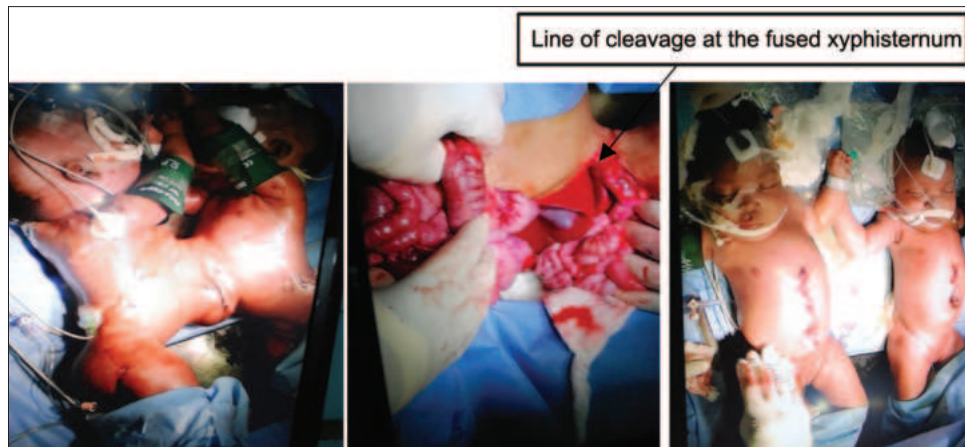
**Figure 3:** Ultrasound showing well-demarcated liver capsules



**Figure 4:** Computerized tomographic scan showing the (a) fused cartilaginous xiphoid process and (b) well-delineated livers of the twin



**Figure 5:** Computerized tomographic scan and reconstructed images of the conjoined twins



**Figure 6:** Intraoperative and immediate post separation pictures



**Figure 7:** Separated twin at (a) 3<sup>rd</sup> day in intensive care unit, (b) At 8 months, (c) at 1 year

The management of conjoined twins poses a challenging task, with each surgery of conjoined twins having its own unique circumstances, and its success depending on factors, that make surgery range from possible to impossible. It is a joint decision between the medical team and the family to go ahead with the separation or not, as surgery also has its associated complications. It requires a multidisciplinary approach, comprising extensive medical work-up on patients, multiple meetings and discussions with all the involved specialties and supporting staff, rehearsal of the planned surgical procedure, involvement of parents, and psychosocial counseling of parents, taking into consideration the financial implications on the parents in a setting where health insurance does not cover all citizens.

Surgical separation depends on the shared site and the organs. It can range from a relatively simple to a very complicated operation.

Antenatal diagnosis is very important in the management of conjoined twins, but unfortunately, in the case under study; the fusion was not detected during the antenatal period; thus, the mother was subjected to a very risky labor for delivery of twin pregnancy in an incompetent setting.

Even the emergency cesarean section was not prepared for a conjoined twin. The referral to our center was prompt though the distance was far (more 200 km) on bad roads, a journey which lasted for more than 6 hours and was conducted by untrained parents and relations.

Early ultrasonic prenatal diagnosis of conjoined twins is possible; cases of conjoined twins are reported as early as the 10<sup>th</sup> week of gestation.<sup>[10]</sup> With the use of high-resolution vaginal sonograph, diagnosis is possible as early as the 8<sup>th</sup> week of gestation.<sup>[2]</sup>

The success of surgical separation is usually determined by the complexity of the union, availability of expertise and modern imaging and intensive care facilities, and timing would also appear to be a key determinant of success.<sup>[11,12]</sup> When the diagnosis of the conjoined twins is made, the type and severity of the abnormality are assessed with two- and three-dimensional ultrasound, CT scan, or magnetic resonance imaging. The ultrasound and Doppler scan in this case were repeated at least 3 times to clear doubts about shared organs and the computerized tomographic scan with contrast could not be done early until babies were 110 days old because the parents could

not afford the cost and the machine in our facility was under repairs.

The surgical management of conjoined twins recognizes three categories.<sup>[10]</sup>

Category I: no surgical intervention should be considered when the cardiac fusion is such that it is not possible to construct one single-functioning heart.

Category II: emergency separation in case of death of one of the twins or one of the twins dying and threatening the life of the other. The presence of an anomaly that is incompatible with life if left untreated.

Category III: planned separation when the infants' condition is deemed stable to perform the necessary imaging investigations that allow precise mapping of the separation.

The case being reported is classified as category III. The major organs shared in this case included the xiphisternum and the anterior abdominal wall, with no viscera organ attachment, however, one of the twins had intestinal malpositioning with the cecum of the left twin found on the left upper quadrant with wide mesenteric base and the duodenojejunal junction was found on the left of midline.

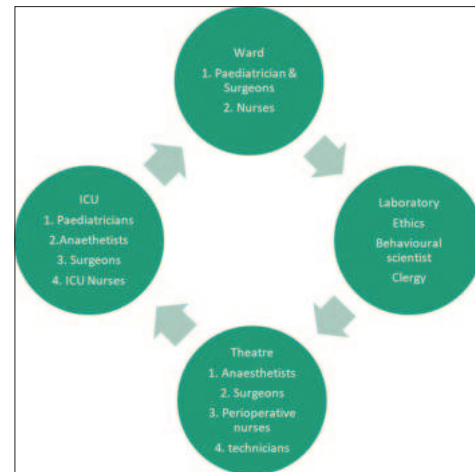
Two previous failed separations at our facility in 2005 and 2015 were under Category II because one of the twins was compromised in each case.

The ideal timing for separation would be around 3 months of age when the physiological condition and pliability of tissues are optimal.<sup>[11]</sup> A tertiary referral center with an experienced medical team of surgeons, anesthetists, and ICU is required in case of surgical intervention.<sup>[11,12]</sup>

Formation of a formidable multidisciplinary team with interdisciplinary cohesion in the management of our conjoined twins assisted in the success of the procedure. The multidisciplinary team comprised neonatologists, pediatric cardiologists and gastroenterologists, pediatric surgeons, plastic surgeons, anesthesiologists, radiologists, hematologists, chemical pathologists, perioperative, neonatal and pediatric wards, and ICU nurses, technicians, behavioral scientists, pharmacists, ethical review committee members, and clergy. There were multiple specialist independent reviews and interdisciplinary consultations with frequent rehearsals and dedication from all team members, including the hospital management and other stakeholders who provided pro bono service as well as financial support to the parents [Figure 8].

## CONCLUSION

This is the first successfully separated conjoined twins at the University of Ilorin Teaching Hospital, North Centre



**Figure 8:** Simulation and drill sketch for the multidisciplinary team members

after failed two previous attempts. Separation of conjoined twins is a complicated procedure; the constitution of complementary multidisciplinary team with rehearsal of all aspects of the operative procedure cannot be overemphasized. Although it is rare, it should be suspected in all monochorionic, monoamniotic twin pregnancies, and careful sonographic assessment should be performed to identify the possibility of shared fetal part(s) during antenatal visits.

The paucity of data fails to represent the true incidence of conjoined twins in Nigeria, hence adequate and proper reporting of cases is encouraged.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the parents have given their consent for the images of their babies and other clinical information to be reported in the journal. The parents understand that name and initials will not be published, and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

## Conflicts of interest

There are no conflicts of interest.

## REFERENCES

1. Spencer R. Conjoined twins: Theoretical embryologic basis. *Teratology* 1992;45:591-602.
2. Amuabunos AE, Eregie CO, Omoigberale AI, Effiong V. Conjoined twins in Edo state of Nigeria; a report of the first surviving set. *Niger J Paediatr* 2014;41:239-43.
3. Spencer R. Anatomic description of conjoined twins: A plea for standardized terminology. *J Pediatr Surg* 1996;31:941-4.
4. Sultan OM, Tawfeek AS. Conjoined twins—thoraco-omphalopagus (type A). *BJR Case Rep* 2015;2:20150016. doi: 10.1259/bjrcr.20150016.
5. McLaren DW. Separation of conjoined twins. *Br Med J* 1936;2:971-986.4.
6. Omokhodion SI, Ladipo JK, Odebode TO, Ajao OG, Famewo CE, Lagundoye SB, *et al.* The Ibadan conjoined twins: A report of omphalopagus twins and a review of cases reported in Nigeria over 60 years. *Ann Trop Paediatr* 2001;21:263-70.
7. Mabogunje OA, Lawrie JH. Conjoined twins in West Africa. *Arch Dis Child* 1980;55:626-30.
8. Adejuyigbe O, Sowande OA, Olabanji JK, Komolafe EO, Faponle F, Adetiloye VA, *et al.* Successful separation of two pairs of conjoined twins in Ile Ife, Nigeria: Case reports. *East Afr Med J* 2005;82:50-4.
9. Ekenze SO, Ibeziako SN, Adimora GN, Anyanwu PA, Ekwunife H, Ajuzieogu VO. Ruptured omphalocele in thoracoomphalopagus conjoined twins. *Int Surg* 2009;94:221-3.
10. Chiabi A, Nkemayim D, Tchokoteu PF, Guegang EG, Ndeki NN, Minkande JZ, *et al.* Conjoined twins in Cameroon: Issues inherent in diagnosis and management in the African context. *Afr J Reprod Health* 2009;13:127-35.
11. Bindlish A, Sawal A. A detailed description and discussion on conjoined twins. *Cureus* 2022;14:e29526.
12. Fallon SC, Olutoye OO. The surgical principles of conjoined twin separation. *Semin Perinatol* 2018;42:386-92.