TRIPLET: THORACO-OMPHALOPHAGUS CONJOINT TWINS AND MALFORMED FETUS OF SOKOTO, NORTH-WESTERN NIGERIA

Abstract

Conjoined twinning is one of the most fascinating human malformations. The incidence of conjoined is 1:50,000 to 1:100,000 live births, with 40–60% being stillborn and about 35% of the live births not surviving beyond 24 hours. The exact incidence of conjoint twins in Nigeria is not known as there are instances of under-reporting, largely due to the relatively poor prognosis and stigmatisation associated with conjoined twinning.

We are reporting a case of thoraco-omphalophagus conjoint twins and a malformed third fetus diagnosed by ultrasound in the first trimester of pregnancy.

A set of female conjoined twins and a malformed triplet, delivered by 25-year old primigravida via emergency Caesarean Section on the 22nd May 2021 on account of PROM and fetal distress at 34/52 GA, that were admitted in our level one neonatal unit. The twins were joined at the thoraco-abdominal region. Echocardiography showed a moderate- sized Atrial Septal Defect in twin 1 and a large Ventricular Septal Defect in twin 2. MRI showed a thoraco-omphalophagus set of conjoined twins with fused xiphisternum, myocardium, pericardium, liver and small bowel.

Keywords: Triplet, thoraco-omphalophagus, conjoint twins, malformed fetus, Sokoto, North-western, Nigeria

Introduction

Conjoined twins refer to twins that are physically fused in utero and consequently at birth [1]. This type of pregnancy is a complicated phenomenon that requires an inter professional team approach to manage it effectively [1]. Conjoined twinning is one of the most fascinating human malformations and has also been reported in other animals—mammals, fishes, birds, reptiles, and amphibians [2, 3]. The condition is proposed to result from either fission or fusion [4]. It happens when a monozygotic twin pregnancy cleaves more than 13 days after fertilization. Conjoined twins are monochorionic and monoamniotic [4]. The incidence of conjoined twins' falls in the range of 1:50,000 to 1:100,000 live births, with 40–60% being stillborn and about 35% of the live births not surviving beyond 24 hours [4]. The exact incidence of conjoint twins in Nigeria is not known as there are instances of under-reporting, largely due to the relatively poor prognosis and stigmatisation associated with conjoined twinning [5]. In Nigeria, the earliest reported case of conjoined twins were born in Sokoto on 20th December 1935, they shared only abdominal wall but no shared internal organs. They were subsequently separated successfully at a general hospital in Sokoto by a British missionary doctor [6]. In an article published by Omokhodion *et al*[7] *(REF)* in 2001, over the preceding 60 years there were 12 published cases of conjoined twins, they also reported another case of an omphalopagus set of female conjoined twins, undiagnosed prenatally, who presented as obstructed labour needing operative delivery. Their anatomic characteristics and clinical features, including overwhelming sepsis in twin 2 forced

an early separation, which led to their demise. Several other cases of conjoined twins have been reported in Nigeria following that publication. In 2014, Amuabunos *et al* in Benin city, Edo State Nigeria reported a case of the first set of conjoined twins survivor managed in University of Benin Teaching Hospital, 3 other sets of non- surviving conjoined twins had been managed in that facility [5]. Amuabunos *et al* also observed that 18 cases of conjoined twins had been reported in Nigeria over 76 years; from 1935 to 2012, across various health facilities in Nigeria [5]. Conjoined twins are associated with a higher predominance of the female sex [4]. We report our experience in the management of a set of conjoined twins in Usmanu Danfodiyo University Teaching Hospital, Sokoto.

Case report

A 30-minute old set of female conjoined twins were brought to the level one neonatal unit of Usmanu Danfodiyo University Teaching Hospital (UDUTH) Sokoto, delivered via emergency Caesarean Section (CS) on 22nd May 2021. CS was done on account of PROM and fetal distress at 34 weeks of GA, the 25-year old primigravida mother was delivered of a set of conjoined twins and a malformed triplet in our facility

Pregnancy was spontaneously conceived, booked at Sir Yahaya Memorial Hospital, Birnin-Kebbi at 6/12 gestation, booking parameters were essentially normal. She was regular on antenatal visits and she was also regular on oral hematinics. No maternal history of cigarette smoking, consumption of alcohol, any known teratogenic drug or traditional medicine during pregnancy, neither was she exposed to radiation during the first trimester of pregnancy. The mother was not diabetic, no history of chronic drug use during pregnancy, no history of maternal febrile illness or consanguinity in the parents. There was no family history of twin delivery. The mother had 4 obstetric scans from different radiological centers with similar findings of conjoined twins and a malformed 3rd fetus. The first two scans were first trimester scans while the last two were done in the second trimester. Antenatal counselling sessions regarding the diagnosed anomaly was done but however, the parents did not opt for termination of pregnancy when the diagnosis was made in the first trimester of pregnancy due to their religious belief. The mother was referred to our tertiary centre (UDUTH) for expert care.

The twins were received from the theatre and resuscitated, APGAR scores were good. A combined birth weight of 3kg was recorded. The conjoined twins were females each with one head and neck, a chest and abdomen to each of the babies, a pair of upper and lower limbs and were joined at the thoraco-abdominal region with a single umbilicus. Also delivered was a third abnormally formed triplet with no head, rudimentary thorax that is joint to the abdomen, with no upper limbs, and abnormally formed lower limbs. The conjoint twins were managed for respiratory distress syndrome and presumed sepsis. While on admission they had various radiological and blood investigations. Echocardiography showed a moderate- sized Atrial Septal Defect in twin 1 and a large Ventricular Septal Defect in twin 2. CT scan showed two hearts, one for each twin which are contiguously oriented side by side anteriorly giving a mirror image. A hypodense septum is noted separating the two cardiac outline, however there appears to be communication in the region of the myocardium anteriorly. The cardiac contour and aorta of twin 2 appears relatively larger than that of twin 1. They have a single liver with two separate portal veins for each twin. The portal vein from the side of twin 2 is well demonstrated and contrast opacified compared to that of twin 1. Following oral contrast medium was administered to twin 1. The bowel loops show normal distribution, caliber as well as contrast medium opacification on the side of twin 1. Contrast medium was noted in the midline and

subsequently across the midline and extending into the small bowel loops of twin 2 suggesting communication. Furthermore, about 50ml of gastrograffin was introduced via the anus into the rectum using a nasogastric tube, it outlined the rectum up to the caecum of twin 1 with no communication demonstrated.
The pancreas of either twin was not convincingly demonstrated. The gall bladders, kidneys, spleen and lungs are demonstrated separate for each twin. They show normal morphology, density and contrast medium enhancement. There is fusion of the sternum at the level of the xiphisternum. The lungs show normal morphology, density and aeration. The spine and demonstrated bones appear within normal limits. The skull vault, brain parenchyma, ventricular system of the brain, brainstem and demonstrated intracranial vessels also show normal density, morphology and contrast medium enhancement.

MRI showed a thoraco-omphalogus set of conjoined twins with fused xiphisternum, myocardium, pericardium, liver and small bowel. No other gross abnormalities were seen on examination. Blood transfusion, antibiotics, oxygen and other supportive management were given. Milk feeds were given till the 6th month of life when complementary feed of cereal was commenced. Family diet was commenced at 9 months and was well tolerated. The parents had several sessions of counselling. The conjoint twins were been worked up for surgery in our facility and remained in our new born unit until the age of 14 months with a combined weight of 10kg, when the father opted for referral to National Hospital Abuja for separation surgery and continuation of care and the conjoint twins were subsequently referred.

Figure 1: The thoraco-omphalogus conjoined twins at birth

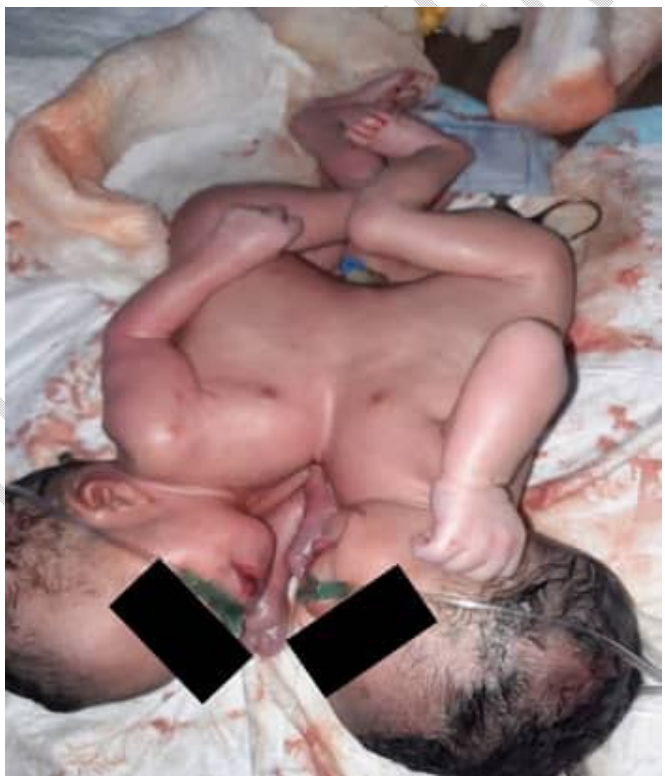

Figure 2: Babygram of the conjoint twins showing fusion of the xiphosterna

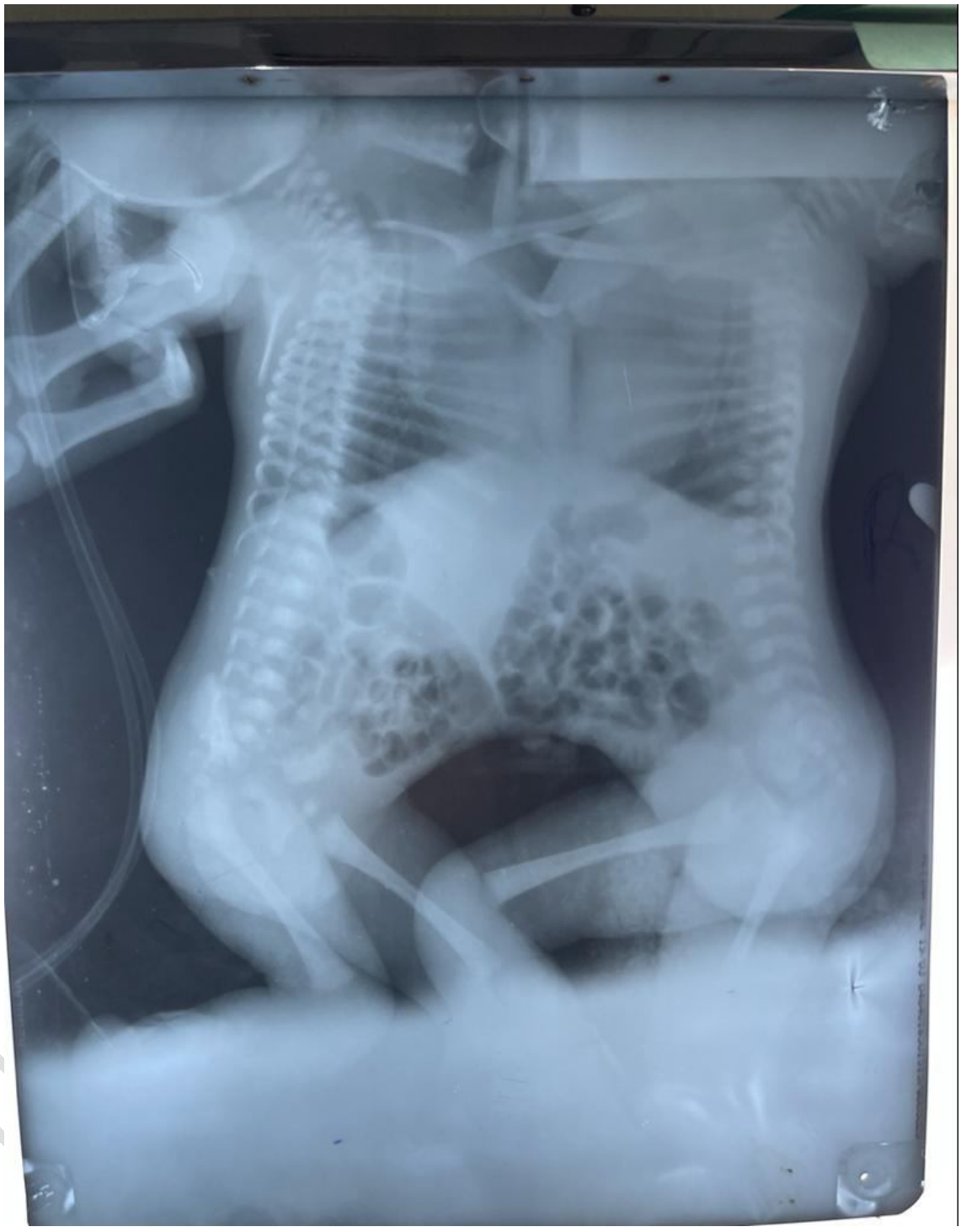

Figure 3: Showing malformed triplet

Figure 4: The conjoint twins at 8 months

Figure 5: The conjoint twins at 12 months

Discussion

This was the first case of conjoined twins in our centre, UDUTH, Sokoto, even though the first reported case of conjoined twins in Nigeria was in a General Hospital in Sokoto.[6] The index twins were females, this was unsurprising as a female preponderance has been observed [4]. Physical examination and MRI revealed thoraco-omphalopagus conjoined twins, which the commonest form is accounting for 28% of all conjoined twins [8]. Similarly, Adeku *et al*[9] in Nigeria reported thoracopagus conjoint twins with malformed triplet delivered by a 32 years old mother at 21 weeks of gestation via C/S, the live conjoined twins died 3 minutes post-delivery, this is in contrast to the conjoint twins reported in our centre that survived and remained in our new born unit until the age of 14 months and were been worked up for surgery in our facility but subsequently referred to National Hospital Abuja for separation surgery and continuation of care based on the parental wish, the report of Adeku et *al*[9] also noticed a non-viable mummified triplet similar to our report, the possible reason survival of the conjoint twins in our report may explained by the difference in the gestational age, the conjoint twins from our centre were late preterm, whereas the conjoint twins reported by Adeku *et al*[9] were extreme preterms.

Prenatal diagnosis was made from the first trimester ultrasonography in this report [9], this is quite different from other reports from Nigeria that were diagnosed at birth.[10,11]. First-trimester ultrasonography remains the best modality of diagnosis early in the pregnancy. Prenatal magnetic resonance imaging can also help with identifying the type of conjunction, embryological malformations, and tissue characterisation [12]. Also, in another report by Rachael *et al*[13] of Triplets with Conjoined Twins in Nigeria that were diagnosed at birth, the conjoint twins were joined at the buttocks, which is in contrast to our report that the twins

were joint at the thoracic and abdominal region. Ibinaiye *et al* from Zaria, Nigeria reported conjoint twins with two heads, two pairs of upper limbs, a shared pelvis and a single pair of lower limbs (dicephalus, tetrabrachius, and dipus twins), a shared liver and spleen and a single pair of hydronephrotic kidneys, the condition was not diagnose antenataly, the mother present to the hospital 2 days after delivery at home [14].

Our patients also shared vital organs including the myocardium, pericardium, liver and small bowel, this was a source of concern considering the guarded prognostic outlook of separation surgery in this index case. The sharing of organs, in particular, the heart is an important consideration because it determines whether surgical treatment is possible and its prognostic outlook [4]. This presents some ethical dilemmas in the surgical management of conjoined twins [15]. Ethical considerations should be kept in mind as they relate to prognosis [16]. Metmet *et al* [17] in Turkey reported a case of conjoined thoracopagus twins diagnosed by ultrasonography at 11 weeks, with only one fetal heart oabserved, the parents were informed about the malformation and the twins' poor chance for survival, after an informed consent the parents decided to terminate the pregnancy, but this is contrary to our report whereby the parents did not opt for termination of pregnancy when the diagnosis was made in the first trimester of pregnancy due to their religious belief.

Nutritional support is very important in the management of conjoined twins during the neonatal period [18]. Medications should be administrated with caution and there should be increased monitoring because of the variation of pharmacokinetic variable [19]. Prenatal counselling of the family plays an important role because of the complicated nature of management and prognosis [20].The management of conjoined twins requires the intimate cooperation of an inter-professional team [21]. Furthermore, our patients had background congenital heart disease. The aforementioned, posed major ethical dilemma in the surgical management of the conjoined twins. A successful management of conjoined twins is a major undertaking requiring careful planning and cooperation of a multi-disciplinary team, in this case the paediatricians, cardiothoracic surgeons, paediatric surgeons, radiologists, anaesthetists and plastic surgeons

Conclusion

Conjoined twins are rare and complex anomalies of the new-born. To optimize postnatal management, conjoined twins should be cared for by a multidisciplinary team. They require a highly experienced team and a centre equipped to deal with such challenging anatomy. Conjoined twins often present a complex psychosocial challenge. Surgical separation is associated with ethical dilemmas, especially in cases of shared vital organs.

consent

Consent was obtained from the parents to publish the images and other clinical information of the patient.

Ethical Approval:

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

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