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

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-omphalogus 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 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]. Conjoined twins are described according to the site of fusion.

The following are the types of conjoined twins along with their frequency [7]: Thoraco-omphalopagus (joined at thorax and abdomen) 28%, thoracopagus (joined at the thorax) 18.5%, omphalopagus (joined at the abdomen) 10%, Heteropagus (parasitic twins) 10%, craniopagus (joined at the level of the cranium) 6%. Less commonly observed types of conjoined twins include: [8] Pyopagus (joined at sacrum and perineum), Rachipagus (joined at vertebral column), Ischiopagus (joined at lower abdomen and pelvis), cephalopagus (joined from head to umbilicus).

The site of fusion and organs involved are a primary consideration for separation during surgery. Typically, 25% of live births live long enough to be candidates for surgery [7]. 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 [9]. The conjoined twins that survive until birth can be broadly categorized into two categories, those who can be surgically separated and those who cannot [4]. The conjoined twins that extensively share vital organs can generally not be separated, as separation might lead to the death of one or both twins. Identification of the anatomical structures is accomplished with postnatal imaging, preferably MRI, keeping prenatal imaging in mind [9]. 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 [10]. Ethical considerations should be kept in mind as they relate to prognosis [11]. Nutritional support is very important in the management of conjoined twins during the neonatal period [12]. Medications should be administrated with caution and there should be increased monitoring because of the variation of pharmacokinetic variable [13]. Prenatal counselling of the family plays an important role because of the complicated nature of management and prognosis [14].The management of conjoined twins requires the intimate cooperation of an inter-professional team [15]. 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. The mother was referred to our tertiary centre (UDUTH) for expert care.

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. The twins were received from the theatre and resuscitated, APGAR scores were good. A combined birth weight of 3kg was recorded. They 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 6^{th} 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.

Figure 1: The thoraco-omphalogus conjoined twins at birth

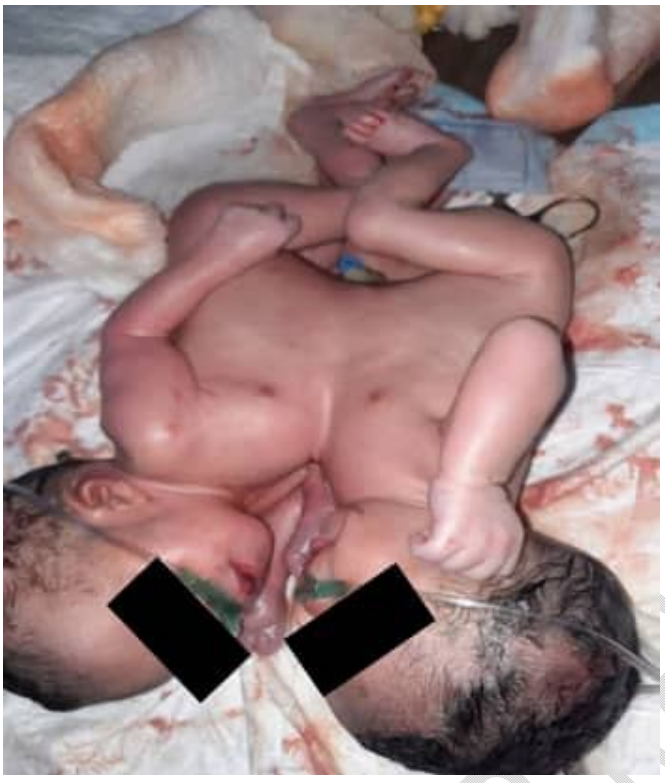

Figure 2: Babygram of the conjoint twins showing fusion of the xiphosterna

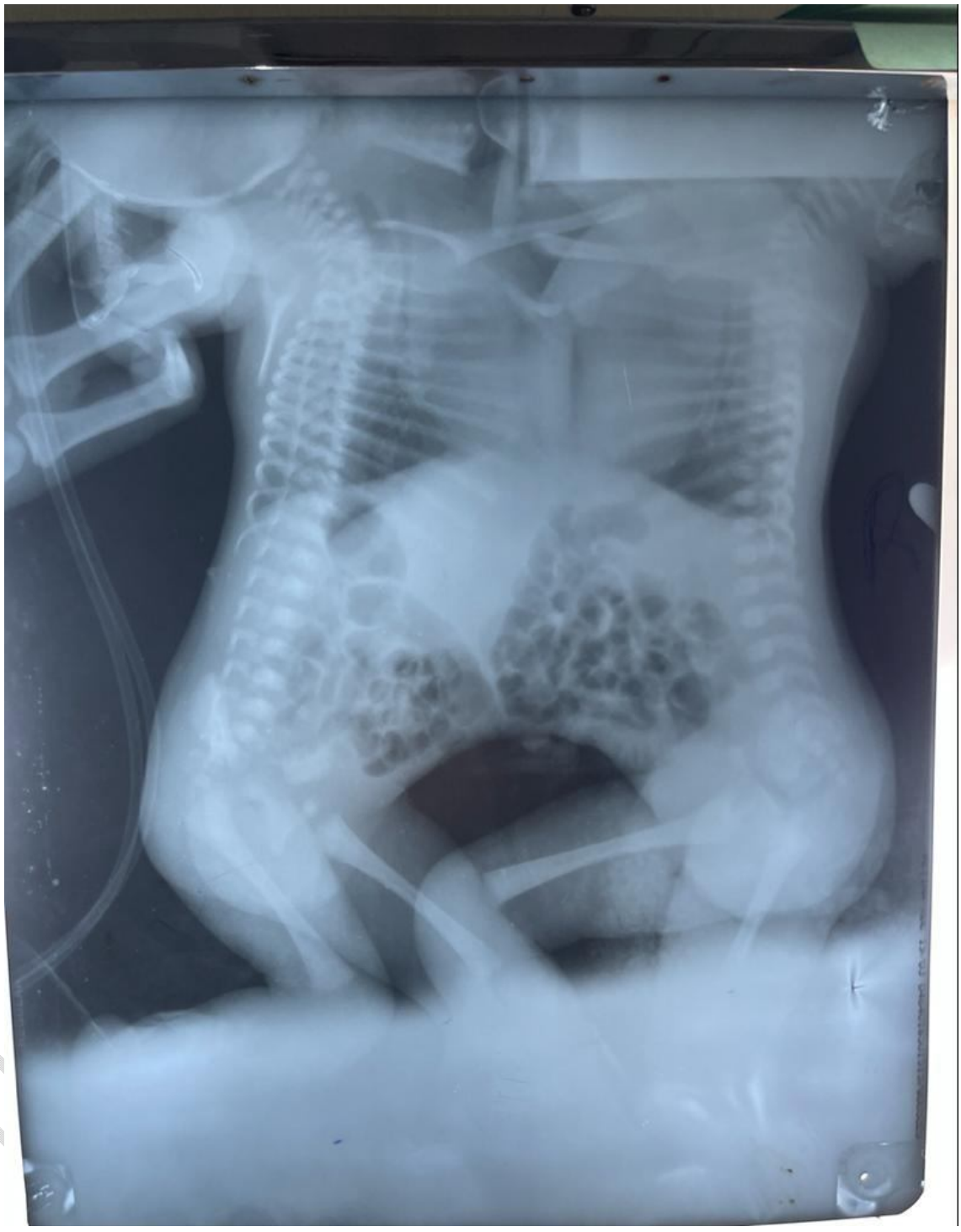

Figure 3: Showing malformed triplet

Figure 4: The conjoint twins at 8 months

Figure 5: The conjoint twins at 12 months

Discussion

This was the first surviving case of conjoined twins in our center, UDUTH Sokoto, even though the first reported case of conjoined twins in Nigeria was in a General Hospital in Sokoto. The index twins were females, this was unsurprising as a female preponderance has been observed [4]. Physical examination and MRI revealed a thoraco-omphalopagus conjoined twins, which is the commonest form accounting for 28% of all conjoined twins [7]. Prenatal diagnosis was made from the first trimester ultrasonography, as first-trimester ultrasonography remains the best modality of diagnosis early in the pregnancy. Our patients 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. 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.

Declaration of patient consent

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

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