

# Multisystemic Anomalies in Conjoined Twins: The Role of Imaging in Management

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## ABSTRACT

Conjoined twins (CT) represent one of the rarest and most challenging congenital malformations arising as an unfortunate complication of monozygous twinning, with an estimated incidence ranging from 1 in 50,000 to 100,000 pregnancies, with an overall true incidence of about 1 in 200,000 live births with around 60% of them being stillborn. The types of the twins may differ depending on the site of fusion or nonseparation. The most commonly encountered types are thoraco-omphalopagus (28%), thoracopagus (18%), omphalopagus (10%), parasitic twins (10%), pygopagus (6–19%), and craniopagus (6%), with the pygopagus twins having the highest survival rate. Because of the highly variable and complex anatomy and associated malformations, skilled clinical assessments aided by detailed radiological studies, appropriate planning, and teamwork are required for the successful separation of the CT. The role of imaging in the diagnosis and management of missed diagnosed intrauterine case of pygopagus type CT was presented. At delivery, they were found to have sacral fusion on radiography with distal, rectal fusion on limited barium enema. Magnetic resonance imaging demonstrated distal spinal cord fusion with vascular anomaly found on computed tomography angiography. The surgical separation of the patients was successful with satisfactory postoperative lives.

**Key words:** Conjoined twins; imaging; multisystem anomalies

## Introduction

Conjoined twins (CT) represent one of the rarest and most challenging congenital malformations arising as an unfortunate complication of monozygous twinning, with an estimated incidence ranging from 1 in 50,000 to 100,000 pregnancies; however, around 60% of them are stillborn, giving an overall true incidence of about 1 in 200,000 live births.<sup>[1,2]</sup> Females predominate over males in the order of 2:1–3:1.<sup>[2]</sup> Embryologically, their formation results either from failure of separation of the embryonic plate between 15 and 17 days' gestation or from the secondary union of two separate embryonic discs at the dorsal neural

tube or ventral yolk sac areas at 3–4 weeks' gestation.<sup>[2]</sup> Depending on the site of fusion or nonseparation, the types of the twins may differ.

Information from the largest study to date indicates that the most commonly encountered is thoraco-omphalopagus (28%), thoracopagus (18%), omphalopagus (10%), parasitic twins (10%), pygopagus (6–19%), and craniopagus (6%).<sup>[3]</sup> Other less common types of CT include cephalopagus, xiphopagus, ischiopagus, and parapagus. A previous study on large cohort found that pygopagus twins have the highest survival rate.<sup>[3]</sup>

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Imaging strategy to accurately define the sharing of organs and vascular anomalies is important for surgical planning and prognostic information.<sup>[4]</sup>

Because of the highly variable and complex anatomy and associated malformations, skilled clinical assessments aided by detailed radiological studies, appropriate planning, and teamwork are required for the successful separation of the CT.<sup>[2]</sup>

## Case Report

Babies B.H. and B.H. are female pygopagus twins delivered via cesarean section at term in a peripheral hospital to a 28-year-old para 3+0 (3 alive) woman. The babies were referred to the neonatal unit of Aminu Kano Teaching Hospital (AKTH) immediately after birth. They were joined back to back at the gluteal region and perineal area. They had a common anal canal and single shared anal orifice. Physical examination revealed low birth weight newborns, with combined weight of 4.5 kg. Assessment of their central nervous systems, hearts, and lungs was normal. Examination of the perineum showed two separate urogenital tracts with a thin membrane which constituted the posterior wall of the vaginal canals. Their mother had booked the pregnancy late (second trimester) and had irregular antenatal visits. There was no history of twinning in the same family and congenital abnormality in siblings. She had no history of use of unprescribed drugs during pregnancy.

She had prenatal ultrasound scans on two occasions at the referral hospital which were reported as normal twin pregnancy. On arrival at AKTH, they were admitted for optimization and surgical workup.

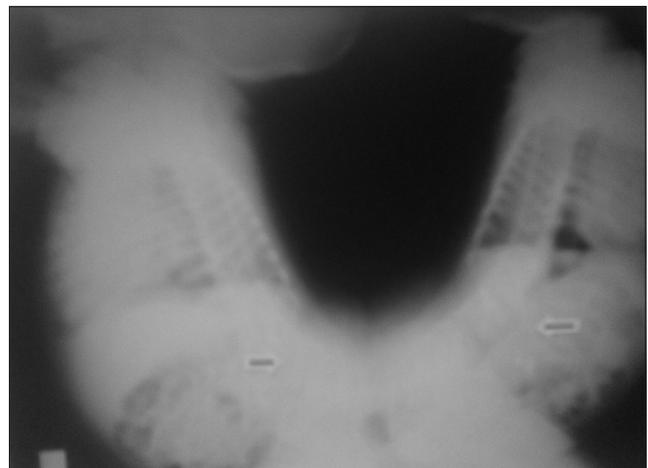
Due to their low birth weight, it was decided to delay separation until their combined weight is >5 kg. They tolerated oral feeds during this period and started to gain weight. After 2 months, the babies weighed 7 kg at which period decision for surgery was taken. While awaiting surgery, preoperative investigations including plain radiograph of the spine, intravenous urography, and barium enema were done. The radiograph of the spine showed five lumbar vertebrae in both twins with a suggestion of fusion at the sacral region. The pelvic bones were separate and complete, and there was no evidence of any other vertebral anomaly. Barium enema showed two separate rectums with a distal fusion in Y configuration [Figure 1]. Intravenous urography showed normal collecting system, ureters, and urinary bladder in both twins [Figure 2].

The parents opted for treatment abroad, where magnetic resonance imaging (MRI) and computed tomographic angiography (CTA) were done as preoperative workup.

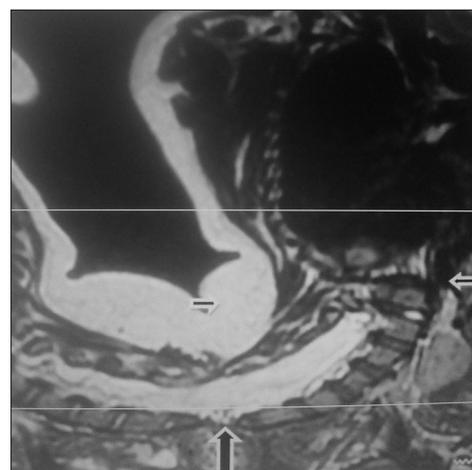
MRI of the spine showed the presence of single dysplastic sacrum with fusion of the spinal cord and thecal sac [Figure 3]



**Figure 1:** Limited barium enema demonstrating the rectums with distal fusion in a "Y" configuration in the conjoined twins



**Figure 2:** Intravenous urography (excretory phase) showing the collecting systems (arrows). Note the common rectal gas shadow



**Figure 3:** T1-weighted magnetic resonance image of the spine (sagittal section) demonstrating terminal fusion of the spinal cords (arrow). Courtesy BLK Super Specialty Hospital, New Delhi

while MRI of the pelvis revealed a single puborectalis sling encircling the common anal canal.

The patients, however, had successful surgery which was carried out by multidisciplinary medical specialists including pediatric, neuro, and plastic surgeons. They had multiple reconstructive procedures and were discharged home in healthy conditions after about 3 months postoperative. On return to Nigeria, they have been seen at the pediatric surgical outpatient clinic of our hospital and are doing well without any complication at present.

## Discussion

Conjoined twinning is one of the most fascinating human malformations. Till now, its cause is not exactly known, but it is generally accepted that CT arise from a single zygote that fails to undergo complete splitting of two inner cell mass during the blastocyst period (5–6 days after fertilization).<sup>[5]</sup>

In pygopagus twinning, the neonates are joined back to back facing away from each other.

Commonly, the twins share the lower part of the spine in 100%, lower gastrointestinal (GI) tract in 25%, and genitourinary (with a single bladder and urethra) in 15%, but the spinal cords and cauda equina usually remain separate.<sup>[6]</sup> In the case presented, the twins were seen sharing the same gluteal region, lower part of the spine (sacrum), rectum [Figure 2], and spinal cord [Figure 3], with separate lower urological and reproductive tracts.

Prenatal diagnosis of CT has been made possible in recent times by prenatal sonography.<sup>[7]</sup> The development of high-resolution transvaginal transducers has made diagnosis possible as early as the first trimester.<sup>[7]</sup> Prenatal sonographic diagnosis is usually based on the findings of an absent dividing membrane with conjoined body parts, inseparable bodies despite changes in position of the fetus, or a bifid appearance of the embryo in the first trimester.<sup>[7]</sup> In addition, sonographic findings suggestive of CT include single umbilical cord with more than three vessels, complex fetal structural abnormalities, extreme flexion of the fetal spine, and proximity of the extremities or persistence of the position of the twins relative to each other throughout scanning. Polyhydramnios is an associated finding seen in more than 50% of these pregnancies.<sup>[7]</sup>

Several pitfalls in sonographic examination of this condition account for the occasional misdiagnosis. However, inseparable skin contours must be a persistent finding at the same anatomic level to avoid a false positive diagnosis of CT.<sup>[7]</sup>

The choice of imaging modality for the evaluation of these cases is dictated by the region where fusion occurs.<sup>[2]</sup> Skeletal surveys, echocardiography, ultrasonography, computed tomography, and MRI provide excellent anatomical details, demonstrating organ position, shared viscera, and vascular anatomy while contrast imaging evaluates the GI and urinary

systems.<sup>[2]</sup> These are all important for surgical planning. In this case, CTA demonstrated vascular anomaly which was crucial from the anesthetics surgical point of view while contrast studies (barium enema) showed sharing of the lower part of the GI system. These helped deciding the best surgical approach in managing these patients. Similarly, MRI helped planning for spinal cord separation and dural repair.

Management of CT poses a major management challenge worldwide<sup>[6]</sup> and more so in developing countries.<sup>[5]</sup> These challenges include ethical issues, surgical management, as well as postoperative care.

Prenatal identification is particularly important in planning obstetrical management of such fetuses, and this is likely in countries where pregnant mothers are scanned routinely by ultrasonography.<sup>[8]</sup>

Surgical methods to separate CT may range from relatively simple to extremely complex, depending on the point and complexity of attachment and the internal organs which are shared.<sup>[8]</sup> In the index case, the management was rather complex and involved staged surgical procedures which included duraplasty, vulvoplasty, and anorectoplasty with transverse colostomy.

Frequently encountered complications include wound disruption, enterocutaneous fistulas, and meningitis.<sup>[9]</sup> These were adequately guarded against in the index case as careful planning with the help of imaging was in place before surgery.

The importance of preoperative diagnosis needs to be emphasized because CT should be referred *in utero* to a specialized center where future management is feasible.<sup>[10]</sup>

The overall survival rate of pygopagus twins was estimated around 87% (being 100% for males and 85% for females).<sup>[8]</sup>

It is strongly recommended that delivery takes place as close to the surgical unit in which separation will be carried out as possible.<sup>[1]</sup> Where this is not possible, early referral to the surgical unit is advisable.

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## Conflicts of interest

There are no conflicts of interest.

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