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Case Report

Management of Conjoined Twins: Challenges And Limitations in A Resource-Limited Context, Experience of Albert Royer Children's Hospital in Dakar about Two Cases

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INTRODUCTION

Conjoined twins, commonly called Siamese twins, are a fusion of fetuses by one or more parts of the body. It is a rare congenital malformation with an incidence of 1/50,000 to 1/250,000 births [1-4]. This malformation causes fetal death in 40 to 60% of cases, and 35% of conjoined twins die in the first 24 hours of life [5,6]. This anomaly mainly affects girls, with a sex ratio of 1/3 [5]. Conjoined twins occur in monozygotic monoamniotic and monochorionic twins' pregnancies, they are therefore always of the same sex [7]. The causes of conjoined twins are not yet known. The most common form of conjoined twins is the fusion of the anterior thorax or abdomen, associated or not, of the two fetuses, determining a thoracopagus, an omphalopagus, or a thoracic-omphalopagus [6,8]. Their antenatal diagnosis is possible thanks to ultrasound [7], and fetal MRI details the lesions. However, in an under-equipped setting, the management of this rare malformation remains a challenge with cases of discovery of the anomaly in the delivery room [9]. All this leads to improvisation in the entire chain of diagnostic and therapeutic management.

In postnatal management, the treatment is multidisciplinary and the imaging assessment is key to guiding therapeutic attitudes. If the separation of twins is possible, it constitutes a challenge on the anesthetic and surgical level and requires a lot of prior consultation between the different teams, including the most experienced people.

The authors report the challenges and results of the management of two pairs of xypho-omphalopagus and omphalopagus conjoined twins, in an under-equipped setting.

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CASE PRESENTATION

Case 1

These are female conjoined newborns, received three hours after their birth.

They were born to a 25-year-old mother, first pregnancy, first parturition without any particular pathological history. The pregnancy was poorly monitored, with five prenatal consultations and an obstetric ultrasound performed in the first trimester finding a monochorionic monoamniotic twin pregnancy. The birth occurred by c-section at 37 weeks and 6 days of gestation, indicating severe preeclampsia. On admission, the newborns were in good general condition. Both weighed 5300 grams. They were joined ventrally from the xiphoid appendix to the umbilicus, with a single umbilical cord. They had two heads, and four limbs each, the upper thorax and the pelvis were separated. Furthermore, they had two permeable anuses with meconium issues. The twins were identified as Twin 1 and Twin 2 (Figure 1). Clinical and radiological findings specific to each twin are shown in Table I.

Table 1: Detailed findings in each twin

Exploration	Twin 1	Twin 2
Clinical	Normal	Scoliosis
Radiographic	Normal	Free L2 hemivertebra
Cardiac ultrasound	Situs solitus and dextrocardia	Levocardia, ventricular septal defect, bidirectional shunt

A whole body CT scan revealed two cardiac masses, with normal morphology and position of great vessels, with four lungs of normal density, diaphragmatic ascension right-sided in Twin 1 and left-sided in Twin 2, a bulky hepatic mass transversely oriented, with a fatty part in its center, distinct from the blood vessels (Figure 2). The other intra-abdominal organs were unremarkable. Furthermore, the upper gastrointestinal series depicted two distinct digestive systems.

Figure 1: Images of the twins on admission, with the fusion area showed by red arrow.

After clinical examination and imaging, the diagnosis was xipho-omphalopagus twins with two hearts, two separate digestive systems, and fusion of the left livers' lobes, with a congenital heart defect and a hemi-vertebra in Twin 2. After multidisciplinary meetings with surgery pediatric, neonatology, anesthesia-resuscitation, and imaging teams, we concluded that the conjoined twins were separable. Clinical monitoring with regular weight gain was recommended in addition to oral feeding. Venous accesses were prohibited to reduce infection risk. A one month and 12 days of life, both twins weighed 7500 grams. A separation was scheduled and carried out at 1 month and 17 days of life. The operative room was adapted for such an operation.

After anesthetic preparation, central catheter placement, orotracheal intubation, and cleaning, the surgical approach was made by a longitudinal skin incision from the xiphoid process to the umbilicus. Exploration of the abdominal cavities noticed an adjoining of the two sternums on approximately 5 cm, an adjoining of the two pericardia on a-2.4 cm length, an adjoining of the two livers' left lobes with a thin demarcation line (Figure 2), two gallbladders, two liver's pedicles, well separated gastrointestinal tract (GIT), with the Twin 2's covered of intact peritoneum.

The separation of the twins was gradually at the sternums, the pericardia, and the liver by section-hemostasis with electrocautery covered by Surgicel at the surfaces of the section (Figure 3). Finally, complete skin separation was achieved to separate the twins.

Skin closure alone was done in Twin 1, realizing an evisceration (Figure 4), and abdominal closure layer by layer was done in Twin 2, with umbilicoplasty and subcutaneous skin closure. After 2 hours 40 minutes of anesthetic preparation, and 2 hours 30 minutes of surgery, the twins were separated. The two stable patients were transferred to the intensive care unit for monitoring.

In the postoperative period, in Twin 2 we noticed the appearance of signs of respiratory distress and facial puffiness, cardiac ultrasound revealed a large ventricular septal defect, a left-to-right shunt, and aorta dextroposition of more than 50% and moderate circumferential pericardial effusion. She died on postoperative day 3.

Twin 1 was hemodynamically and respiratory stable in the postoperative period. On day 2, she presented a superficial skin necrosis of the wound, which has evolved well. She left the hospital 41 days after separation surgery. She showed good psychomotor development and her postoperative eventration repair was done 2 years later.

Currently aged 6 years, Twin 1 has no complaints, she is in school and has good weight gain and good psychomotor development. Hepatic biologic investigations and ultrasound are unremarkable (Figure 5).

Case 2

They were female conjoined twins, identified as Twin 1 and Twin 2, female, received at the age of four months at the pediatric surgery department of the Albert Royer National Children's Hospital (CHNEAR) for treatment.

The diagnosis of conjoined twins was made antenatally during the morphological ultrasound performed at 16 weeks. The pregnancy was monitored with other obstetrical ultrasounds. Delivery was done at 39 weeks + 2 days by C-section. The newborns were hospitalized for a neonatal infection for 45 days. After several unsuccessful attempts for referral abroad, the children were received for care.

In their history, there is a notion of first-degree parental consanguinity. The mother was a 24-year housewife, G3P4. The children had good psychomotor development and an up-to-date vaccination status according to the Senegalese program.

The examination on admission at the age of 4 months and 25 days revealed twins in good general condition, both weighing 13 kg, with

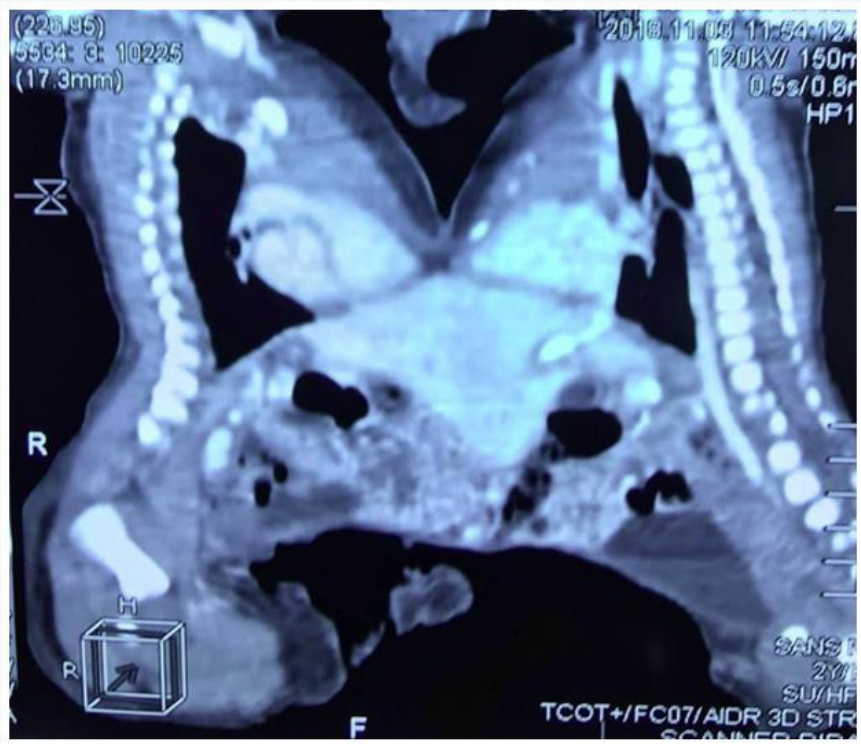

Figure 2: CT-scan with frontal view showing abdominal fusion with difficulty to precisely identify zones of hepatic fusion.

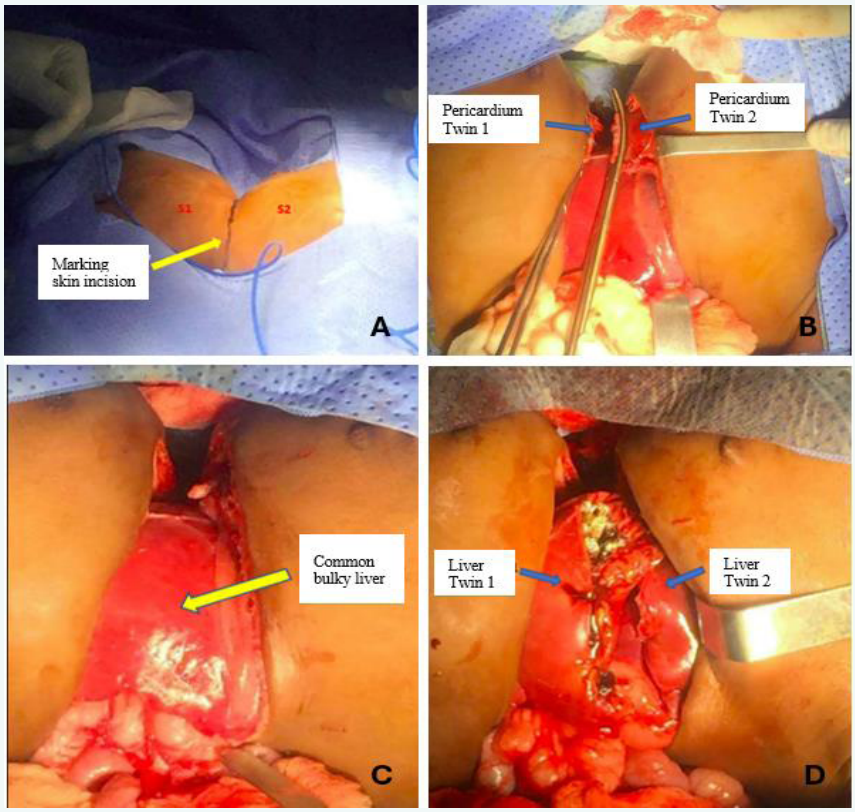

Figure 3: Intraoperative findings. In (A), the incision line. Progressive separation of fused organs in (B), separation of pericardia, in (C) remark the fused livers without any demarcation line and in (D), separated livers.

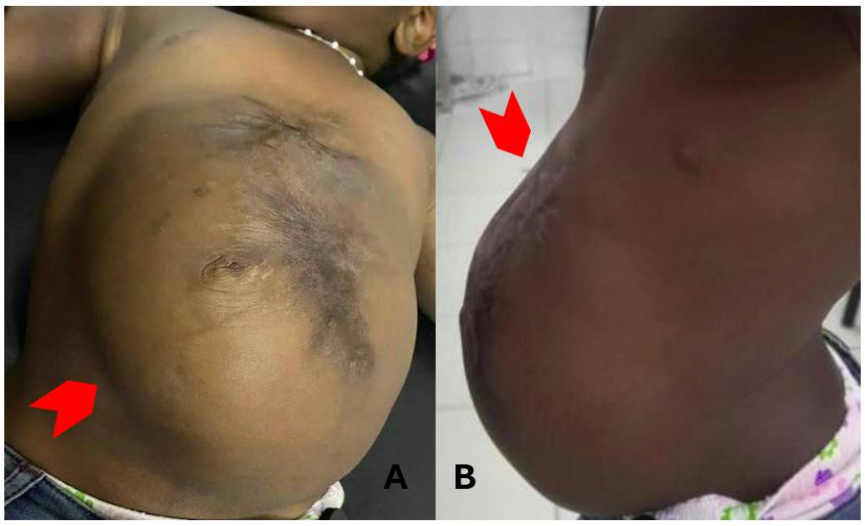

Figure 4: Images of eventration (red arrow) in front view (A) and side view (B)

Figure 5: Images of Twin 1 aged of 6 years.

normal hemodynamic vitals. An omphalopagus-type fusion (Figure 6), with a union zone measuring 10.5 cm in diameter, the presence of four distinct limbs, separate thoraxes without visible deformation, separate external genitalia of the female type, two perinea of normal appearance with the presence of a urethral, vaginal, and anal orifice in each of the twins. No other visible malformation was noted.

Abdominal ultrasound noticed a fusion of the two liver's left lobes. In addition, the gallbladders were flat, the intra and extrahepatic bile ducts were not dilated with patent portal trunks of normal caliber, the hepatic veins were patent, and exploration of the kidneys, spleens, uteri and the rest of the abdominal cavity was normal. A whole-body CT scan revealed distinct full digestive organs, livers of normal volume without dilatation of the bile ducts, and confirmed the absence of any other abnormality and the fusion of the left lobes of the livers.

The UGI series showed opacification of the GIT of Twin 1 without passage towards Twin 2 (Figure 7). Doppler cardiac ultrasound showed left ventricle hypertrophy with good biventricular systolic function in Twin 1 and probable aortic bicuspid with good systolic function. Preoperative cardiac ultrasound was normal in both twin sisters. Thus, the diagnosis of omphalopagus twins was retained and the indication for separation surgery was made. As part of the organization of the surgical intervention, consent was signed by the legal parent and two multidisciplinary staff were organized including pediatric surgeons, anesthesiologists, pediatricians, administrative staff, social service, operating room, and hospitalization nurses. After a pre-anesthetic consultation, separation surgery was performed at the age of 11 months. Installation was done on a regular table in lateral decubitus, under general anesthesia and orotracheal intubation, with epidural block and urinary catheterization. The twins were operated by a longitudinal incision through the union zone. After dissection of the different planes of the abdominal wall, exploration of the cavity revealed a fusion of the xiphoid appendages over approximately 2 cm, a fusion of the two left lobes of the livers, two intact and separate pericardia, and the absence of other visualized anomalies.

A separation of the two sternums was performed by electrocautery and a separation of the two left lobes of the liver by ligature section. The control of hemostasis was completed by the use of Surgicel. Then each twin was installed on an operating table, with two separate surgical

Figure 6: Global view of the twins on admission

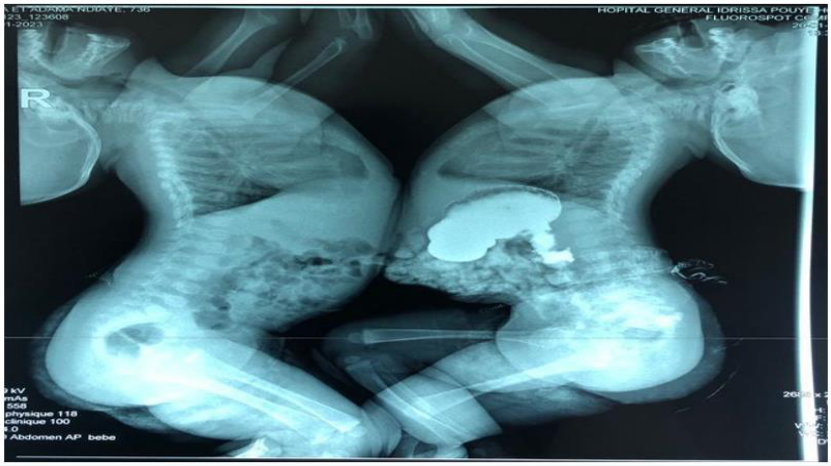

Figure 7: Radiography showing GIT opacification in one twin, without communication with the other twin's GIT

Figure 8: Picture of the 22 month-aged twins.

and anesthesia teams. A parietal closure was made after aponeurotic dissection on either side of the defect, followed by an umbilicoplasty with subcutaneous skin closure in both twins.

Extubation was done at 7 hours postoperatively and the immediate postoperative course was simple. Discharge was done on day 8 after the separation surgery after a liver function test returned normal, with a weight of 6500 g for Twin 1 and 7840 g for Twin 2. Both twins were followed up by the nutritionist.

At the age of 22 months, the two sisters have a weight delay compared to the age of -1.5 standard deviation for Twin 1 and -1 standard deviation for Twin 2. We noted in Twin 1 a weight of 8120 g, and a height of 75 cm, she vocalizes and has a slight deformation of the lumbosacral spine without difficulty in walking. Twin 2 also presents a good general condition, a weight of 9200 grams, a height of 77.5 cm, and a spontaneous deviation of the neck to the left without limitation of movements. We noted in both sisters, a satisfactory aspect of the abdominoplasty. In addition, they have good psychomotor development and are full of life-like children of their age (Figure 8). Currently aged 25 months, they do not present any particular pathology.

DISCUSSION

Conjoined twins are one of the rarest congenital anomalies and one of the greatest challenges in pediatric surgery [9]. Conjoined triplets have been described but are even rarer [10,11]. Although conjoined twinning is usually known to be related to monochorionic monoamniotic twin pregnancies, its etiopathogenesis remains controversial [12-14]. The theories of fission and fusion seem incomplete and other theories on the origin of conjoined twins are resurfacing [12]. Indeed, there is no implication of chromosomal abnormality. Furthermore, race, heredity, parity, and consanguinity seem to not be causal [9]. Among our patients, two out of four parents had a first-degree consanguinity. However, it is difficult to incriminate the notion of consanguinity in the etiological factors for conjoined twins. More than 70% of conjoined twins have a fusion of the thorax, abdomen, or both [6]. In this thoracic-omphalopagus entity, there is first the xiphoid-omphalopagus which is linked from the xiphoid process to the umbilicus with sometimes a partial fusion of the pericardium with two separate hearts as in the case of our first patients. Secondly, the omphalopagus is linked by the umbilical region as described in our second observation. Whatever the form, the antenatal diagnosis of conjoined twins can be made very early antenatally even for the rare forms thanks to the ultrasound and fetal MRI [15-17]. However, this diagnosis is not always made in our practice. Our first case had a single antenatal ultrasound. Poorly monitored pregnancies without morphological ultrasound still exist in certain settings. Thus, the malformation is discovered at birth sometimes during difficulties in labor. Thirty percent of conjoined twins die in utero, 40 to 60% are stillborn and 35% survive only one day [12]. These deaths are all the more frequent when the malformation is complex [13,18]. Postoperative mortality can reach 75%. In case of severe associated anomalies such as congenital heart disease, or incomplete imaging, single plan abdominal wall closure can be performed to reduce the risk of increased intrathoracic pressure. Then, the eventration is repaired later after monitoring vital functions. This technique was used successfully in one of our first twins. In our two pairs of twins, one of the children with a heart anomaly died in the days following their separation.

The diagnostic and therapeutic approach to conjoined twins is complex. This complexity means that some conjoined twins are not separable [19,20]. Thus, some teams recommend termination of pregnancy even for some advanced gestations [21]. Postnatal surgery, if possible, remains a challenge. It requires careful planning, a multidisciplinary approach, repetition, and experience are important factors in the management of conjoined twins [22,23]. While some centers have experience in separating conjoined twins [22,24], the success of such surgical interventions is rare in Africa [19], even if exploits are increasingly reported. Despite this, fewer and fewer patients emigrate to seek treatment in developed countries. In Senegal, three successful surgical interventions on conjoined twins have been noted, the last two of which are reported here. Both were performed by the same anesthetic and surgical teams. The experience gained during the first surgery to separate conjoined twins allowed us to better plan the second, surgically, and in terms of anesthesia and logistics. However, limitations persist, in particular the accessibility of fetal and postnatal MRI with reconstruction. Imaging examinations such as ultrasound, CT scan, and digestive opacification constitute our main imaging investigations. Our first twins were operated on at the age of 1 and a half months. This decision to intervene earlier can be justified on the one hand by the particular environment with its realities, requiring hospitalization of infants until their separation, and on the other hand by the phobia of being confronted with an emergency separation. Indeed, emergency separation is all the more complex because, in addition to the difficult procedures, socio-cultural and ethical realities must be added to manage while making urgent and well-considered decisions [12]. This emergency separation of conjoined twins often results in death even for experienced teams [12-14]. In Senegal, three emergency separations of conjoined twins have taken place, all of which resulted in death. However, at a distance from the separation surgery, conjoined twins live almost normally like all other children of their age [25], like our patients currently aged 6 years and 26 months.

CONCLUSION

Conjoined twins are a rare malformation dominated by anterior fusion of the thorax and abdomen. Their separation requires planning involving all stakeholders and remains a major challenge, particularly in under-equipped settings.

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