Challenges In The Management Of Conjoined Twins In A Poor Resource Setting: A Report Of 3 Cases

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Abstract

Background: Despites the advances in transport, intensives care unit and new treatments modalities, conjoined twins continue to pose a significant challenge for paediatric surgeons. This study was undertaken to highlight the peculiarities of the management of conjoined twins in a poor resource setting.

Methods: All confirmed cases of conjoined twins were prospectively documented from 2001 to date at the Charles De Gaulle Paediatric Teaching Hospital in Ouagadougou (Burkina Faso).

Results: Three cases were recorded: one case of thoraco-omphalopagus conjoined twins, born prematurely by elective caesarean surgery and transferred at the second hour of life, one case of pygomelus conjoined twins with two phalluses, and one last case of epigastric heteropagus conjoined twins naturally delivered at home at term and transferred at 15 days and three months of age, respectively. All our patients were male. Antenatal diagnosis was made in one case. Associated malformations were found in two patients. Thoraco-omphalopagus twins who shared the breastbone, liver, heart, and transverse colon died at day 38 of life from heart failure before any treatment. The two others were successfully operated by surgical resection separation of the parasitic twin at 30 days of life for the epigastric heteropagus conjoined twins and four and a half months for pygomelus conjoined twins.

Conclusion: The management of conjoined twins is difficult in the context of Sub-Saharan Africa developing countries. Their survival depends on an early detection by prenatal diagnosis, the creation of intensive care units, the inclusion of major congenital malformations in national neonatal programs for the integrated care of mother and child diseases and the development of partnership between hospitals in the South and those in the North.

INTRODUCTION

Conjoined twins (CT) also known as Siamese twins or double watches are a rare and specific congenital malformation of mono chorionic and mono amnion twin pregnancies. They occur in 1 in 50,000 to 200,000 births [1]. They constitute a real diagnostic and therapeutic challenge. If in developed countries the multidisciplinary approach together with the creation of reference centres and the proven experience of surgical teams have significantly improved prognosis [2,3], this is not the case in developing countries where their prognosis is poor due to late diagnosis and the under-equipment of surgery and neonatal resuscitation units [4,5,6].

Located in the Sub-Sahara Africa, Burkina Faso is a developing country where the prenatal diagnosis of congenital malformations and paediatric surgery are poorly developed. The purpose of this study is to highlight the peculiarities of the management of conjoined twins in such context of poor resource setting.

CASES

The newborns were received after the second hour of birth at the Charles De Gaulle Paediatric Teaching Hospital in Ouagadougou for thoraco-abdominal joining. They were male. The history of the family revealed a case of an early abortion of the mother while the father had twins in his

family. The mother, 24 years old, was a nullipara. The father was 27 years old. There was no consanguinity. Prenatal diagnosis was made at week 27 of the pregnancy. They were delivery by surgery after 36 weeks of gestation. Apgar score was normal at the first minute. The combined weight of the conjoined twins was 4,200 grams.

The examination of newborns showed any signs of respiratory distress. They were joined to one another by the chest and abdomen with a single umbilicus. The diagnosis of the thoraco-omphalopagus twin was made (Figure 1).

Figure 1

Thoraco-omphalopagus twin

Ultrasound and computed tomography revealed an individual, normal and complete skeleton except the sternum, the heart, the liver, the gallbladder and the common transverse colon

(Figure 2-3). Their death occurred at day 38 of their reception in the hospital with a sudden cardio circulatory failure prior to their transfer to a more equipped centre in Europe.

Figure 2

Normal and complete skeleton except the sternum

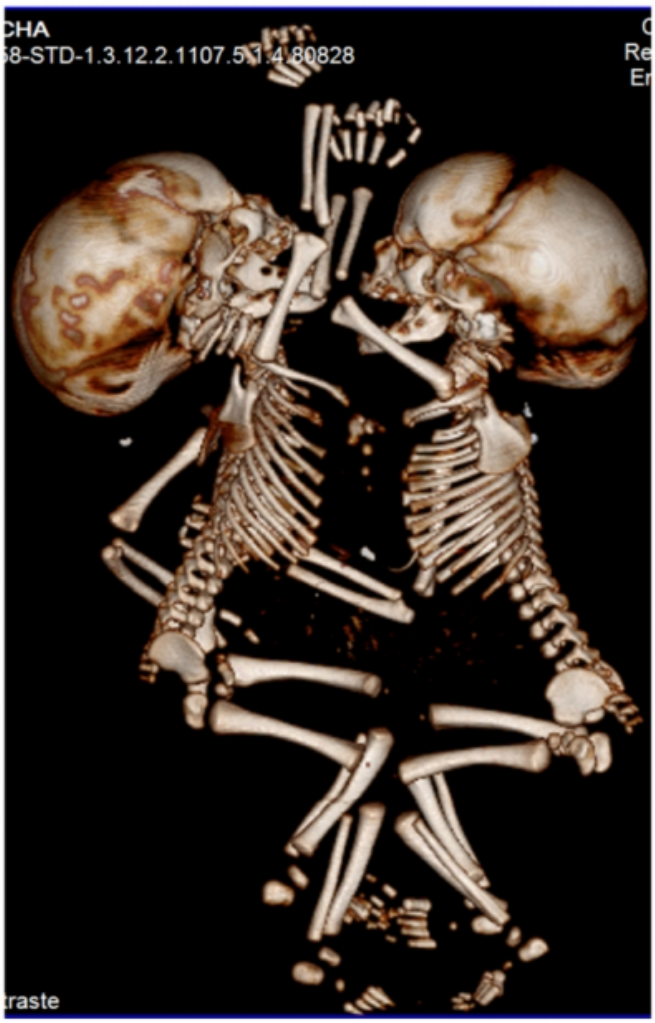
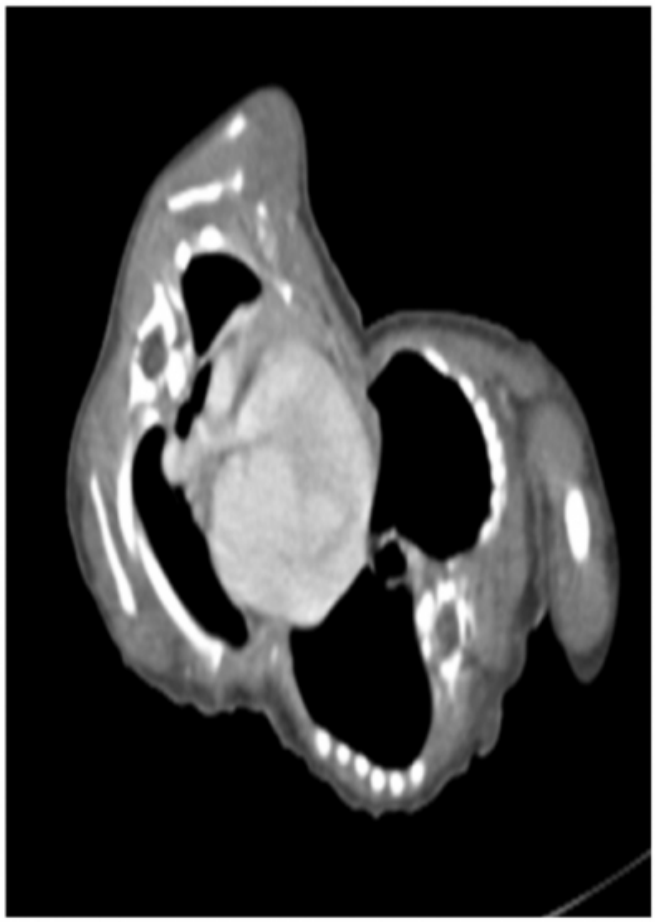


Figure 3

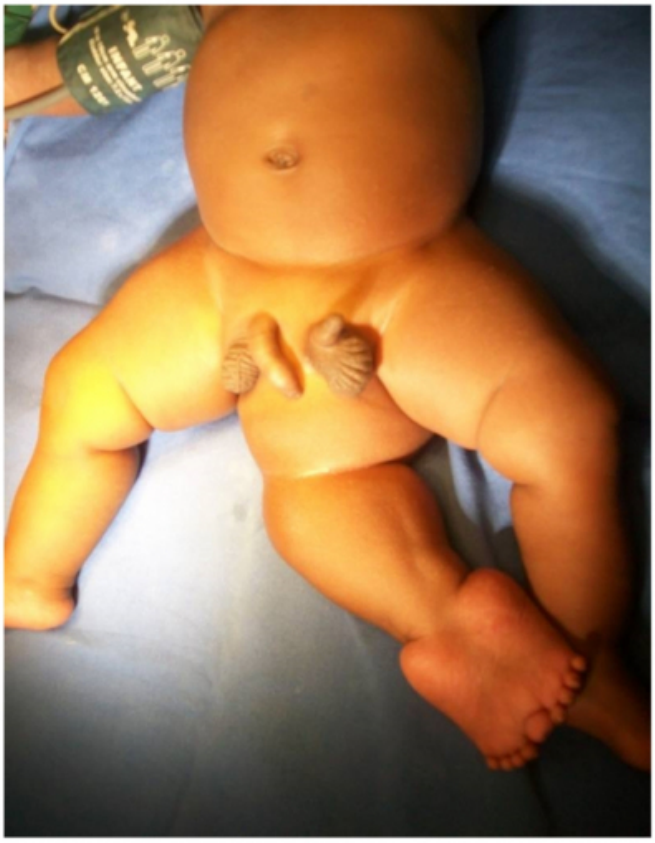
Common heart



The patient was a male newborn. He was received at the Charles De Gaulle Teaching Paediatric Hospital Centre at Ouagadougou at three months for supernumerary limb. In the family history, he was the fifth child of the uterine siblings of 5 children. The mother was 42 years old and the father 52 years old. There was no consanguinity between parents. No prenatal consultation was made during the pregnancy. The mother naturally delivered at term at home. The clinical examination showed an infant with an overall good condition, weighing 6 kg, a lower supernumerary limb comprising a thigh, a leg, two feet joined together by their medial edge, two half scrotum testicle each containing a testicle and two phalluses including a blind one without urethral meatus. The second phallus had an apical urethral meatus allowing normal urination. The diagnostic of pygomelus with sex disorder was diagnosed (Figure 4).

Figure 4

Pigomelus with sex disorder

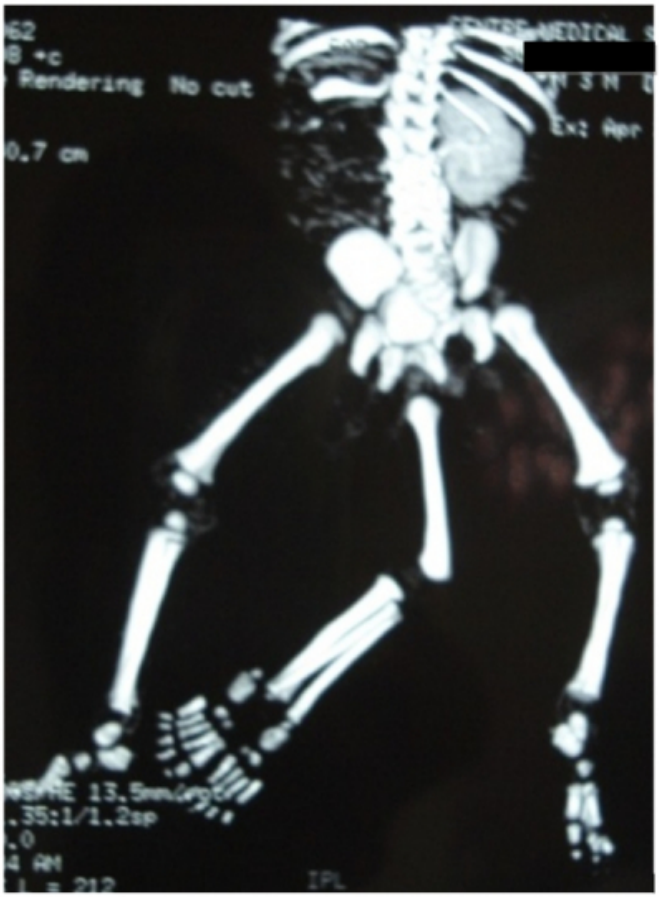


The ultrasound and tomodensitometry examination showed:

- In the supernumerary limb: a femur articulating by a neo-joint in the pubic symphysis, two tibias, tarsus and metatarsus in the joined feet (Figure 5).

Figure 5

Skeleton X-ray



-13 thoracic vertebras including one hemi-vertebra in the 10th thoracic vertebra.

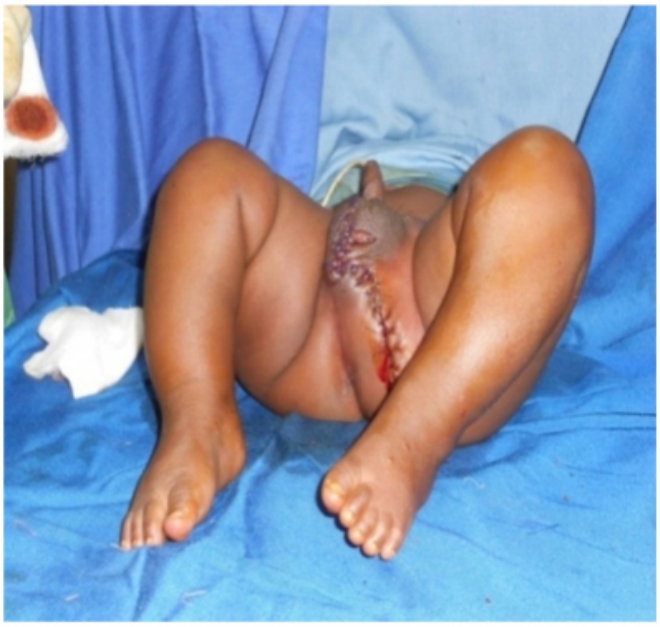
-12 ribs in the right and 11 in the left.

-an hypertrophic single kidney in the left and a mega ureter

Surgery performed one month after their admission consisted in a resection of the supernumerary limb, the resection of the non-functional supernumerary phallus and a plasty of the scrotum (Figure6).

Figure 6

Post operative aspect



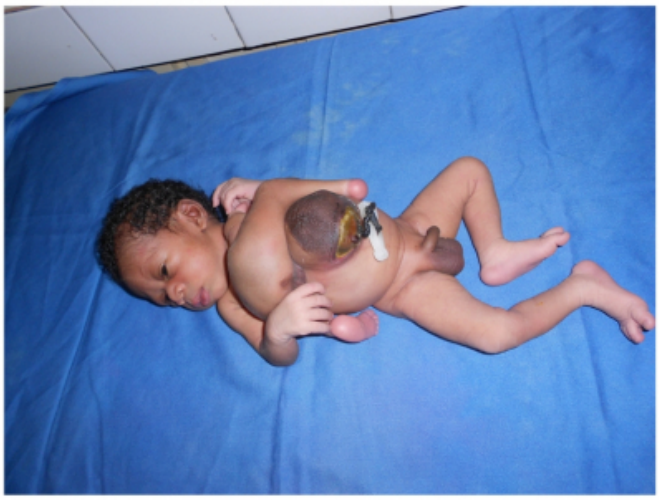
No complication was observed in the postoperative period. The patient is reviewed each every three months for the monitoring of its malformation uropathy and its vertebral malformation.

A male newborn was received at day 15 of life at the Charles De Gaulle Teaching Paediatric Hospital Centre of Ouagadougou for malformation of the anterior abdominal wall. In the family history, he was the fifth child of the uterine siblings of 6 children. The mother was 48 years old and the father 55. There was no consanguinity between the parents.

Two prenatal visits were performed but no any prenatal diagnosis was made. She naturally delivered in a health centre. Over the clinical examination at admission at the Charles De Gaulle Teaching Paediatric Hospital Centre of Ouagadougou, the parasite twin was dew-like, not greyish or necrotic. He was attached to the upper abdomen of the host twin by a short pedicle of nearly 3 centimetres long and 10 centimetres wide and showed two lower limbs and one upper right limb clearly distinct from the feet, an anal perforation, a less developed penis and empty purses. The host twin has a normal aspect except an omphalocele. The diagnosis of heteropagus epigastric conjoined twin was diagnosed (Figure7).

Figure 7

Heteropagus epigastric conjoined twin



Ultrasound and tomodensitometry assessment showed that both twins had no common organ. There was no associated congenital heart disease.

Surgery was performed 30 days after delivery to separate the parasite from its host twin (Figure 8). The dissection of the short pedicle wide base connecting the two twins showed that it was set in the abdominal wall of the host twin (Figure 9). The omphalocele was managed in two stages (use of aqueous eosin at 2% until obtaining a complete skin constitution then the closing of the muscular defect by closing suture of large muscles of the abdomen). No complication was observed during the postoperative period.

Figure 8

Parasit twin

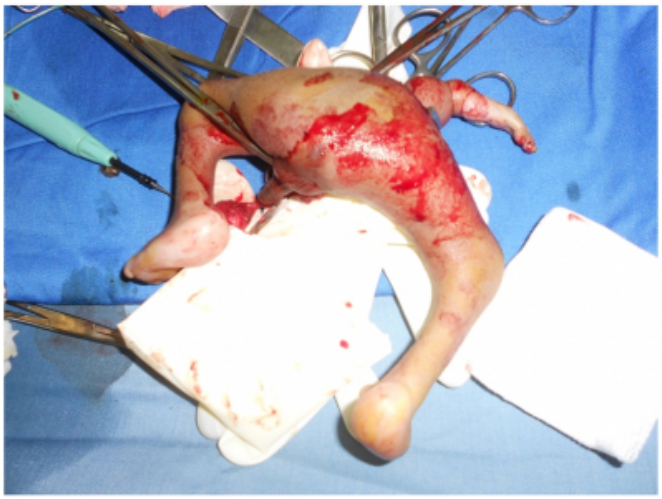
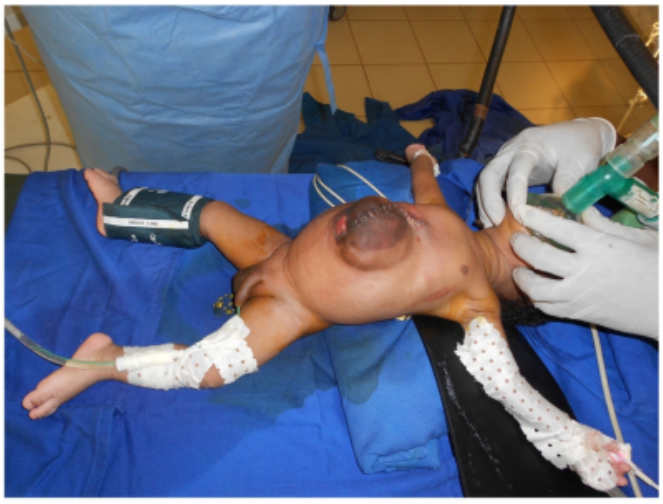


Figure 9

Post operative aspect



DISCUSSION

Conjoined twins are a rare anomaly which incidence varies between 1 out of 50,000 and 1 out of 200,000 births [1]. The low hospital prevalence in our series certainly is due to the lack of prenatal diagnosis, the study of spontaneous abortion products, the notification of stillborn and strong socio-cultural factors with a trend to kill newborns with severe malformations or to hide them [7]. There are several classifications of conjoined twins. The most commonly used gives two types: symmetric (typical) and asymmetric (atypical) and eight subtypes: cephalopagus, thoracopagus, omphalopagus, ischiopagus, parapagus, craniopagus, pygopagus and rachipagus [8]. The cause of this anomaly is discussed and seemed to be due to a history of twin pregnancies which constitutes a risk factor [5, 9] as found in two patients in our short series. The lack of prenatal diagnosis in two patients did not enable to accurately describe the relationships between both twins, find an associated anomaly, monitor this pregnancy monthly which may get complicated, inform parents of the anomaly found and organise the delivery in a health facility equipped with a surgery unit. This weakness has led to late diagnosis (except in the first case), the lack of psychological preparation of parents, and delivery in inappropriate conditions. If in literature [10, 11] thoraco-omphalopagus conjoined twins are the most commonly found and the most difficult to manage because of some of common organs (liver, heart, lungs, small bowel, colon) requiring surgery in specialised centres, this is not the same for pygopagus and epigastric heteropagus for which few cases were reported [8]. Indeed, these are not usually associated with a high rate of prematurity and major malformation in autosite twins.

Despite the lack of obstetric complications in mothers in cases 2 and 3 who delivered naturally, the resort to caesarean should be systematic in all cases to prevent a possible obstructed labour. After delivery, the medical paediatric and surgery care should be organised as soon as possible. Most of the cases reported in literature [8] mention a steady success of the surgery with survival autosite twins without any after-effects like in cases 2 and 3.

CONCLUSION

The lack of antenatal diagnosis coupled with neonatal, social and psychological conditions do not enable to improve conjoined twins care a poor resource setting. Their survival goes through an early detection, the construction and equipment of neonatal intensive care units, the integration of major congenital malformations in national programs for the integrated care of mother and child diseases and the development of partnerships between hospitals in the South and those in the North.

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