

SUCCESSFUL SEPARATION OF CONJOINED PYGOPAGUS TWINS IN AFRICAN ENVIRONMENT WITH LIMITED RESOURCES

M.Nyundo^{1,2,*}, A.Jahn², K.Kayondo², F.Ntirenganya², K.Muzungu³, G.Ntakiyiruta², W.Kiviri⁴, JN. Karangwa⁴, JB. Kayinamura⁴, A.Bahati⁴, JC. Byiringiro^{1, 2}, L.Mutesa^{5,6}, J.Gashegu^{1, 2}

¹Clinical Anatomy Laboratory; Faculty of Medicine, National University of Rwanda, Butare, Rwanda

²Department of Surgery, University Teaching Hospitals, Kigali- Butare, Rwanda

³Department of Pediatrics, University Teaching Hospital Kigali, Rwanda

⁴Department of Anesthesiology, University Teaching Hospital Kigali, Rwanda

⁵Center for Medical Genetics, Faculty of Medicine, National University of Rwanda, Butare, Rwanda

⁶Medical Research Center, Rwanda Biomedical Center, Ministry of Health, Kigali, Rwanda

ABSTRACT

Conjoined twins are identical or monozygotic twins whose bodies are joined in utero. Pygopagus or Iliopagus twins are a type of conjoined twins in which two bodies joined back to back at the buttocks. Surgical separation of conjoined twins is extremely risk of death and life threatening. We report on cases of female pygopagus twins of three months operated and separated at Kigali University Teaching Hospital. For both babies a posterior sagittal anorectoplasty was successfully performed with derived ileostomy.

Despite limited resources to carry out such operation in our settings, no complications occurred during the operation and oral feeding was done at third postoperative day. Ileostomy closure was performed three weeks after and babies were discharged from neonatology unit at 35th postoperative day.

In conclusion, adequate preoperative investigation with a well organized and trained team contribute a lot to the success of such conjoined twins separation.

Key words: Conjoined Pygopagus twins – Successful separation – Limited resources – Kigali University Teaching Hospital

RESUME

Les jumeaux siamois sont des jumeaux identiques ou monozygotiques dont les corps sont joints ensemble in utero. Les jumeaux siamois appelés Pygopagus ou Iliopagus sont des jumeaux habituellement joints au niveau du dos. La séparation chirurgicale de ce type de jumeaux siamois est extrêmement délicate et comporte souvent un risque de décès ou de complications pouvant mettre en jeu le pronostic vital.

Ici nous rapportons deux cas de jumelles siamoises Pygopagus âgées de 3 mois qui ont été séparées chirurgicalement à l'Hôpital Universitaire de Kigali. Chez les deux siamoises, une opération consistant en anorectoplastie sagittale postérieure avec iléostomie dérivée a été pratiquée avec succès. Malgré les moyens techniques limités pour mener une telle opération dans nos facilités, aucune complication n'a été observée durant l'opération et l'alimentation par voie orale a été rétablie après trois jours portopératoires. La fermeture d'iléostomie a eu lieu trois semaines après et les deux jumelles sont sorties de l'Hôpital après 35 jours postopératoires.

En conclusion, les investigations préopératoires adéquates avec une équipe bien formée et organisée contribuent énormément à la réussite d'une telle opération.

Mots-clés: Jumelles siamoises Pygopagus – Séparation réussie – Moyens techniques limités – Centre Hospitalier Universitaire de Kigali

INTRODUCTION

Conjoined twins (CT) are 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, but around 60% of them are stillborns, giving an overall true incidence of about 1 in 200,000 live births with a male-female ratio of 1:3 [1-3]. Conjoined twinning arises when the twinning event occurs at about the primitive streak stage of development, at about 13-14 days after fertilization and is exclusively associated with the monoamniotic monochorionic type of placentation. Two contradicting theories exist to explain the origins of CT. The older and the most generally accepted is fission,

in which there is an incomplete splitting of the embryonic axis and, with the exception of parasitic conjoined twins, all are symmetrical and the same parts are always united to the same parts. The second theory is fusion, in which a zygote completely separates, but stem cells find like-stem cells on the other twin and fuse the twins together [4].

The incidence of the various types of CT is discussed and they are classified according to the most prominent site of attachment. Information from the largest study to date indicates that the most common encountered were thoraco-omphalopagus (28%), thoracopagus (18%), omphalopagus (10%), parasitic twins (10%) and craniopagus (6%) [4]. Other less common types of CT include pygopagus, cephalopagus, xiphopagus, ischiopagus and parapagus.

Pygopagus twins are joined back to back facing away from each other, commonly share the gluteal region,

* Correspondence to: Martin NYUNDO, MD, MMed
Department of Surgery
University Teaching Hospital
Kigali / Rwanda
GSM: +250 788418727
Email: nyundomartin@gmail.com

terminal spine, and lower gastrointestinal, urological, and reproductive tracts. A previous study on large cohort found that pygopus twins represented 17% of all conjoined twins [5]. The first recorded pygopus conjoined twins date back to 1100s [6-8]. They lived 34 years joined together at the hips and buttocks and shared a vagina and an anus, but when one of them died, the other refused to be separated and died 6 hours later. The prognosis of pygopus twins is generally good with an overall survival rate of 87%, but they are also known to have associated vertebral, spinal cord, as well as anorectal anomalies that form a spectrum that differs between males and females [5, 9, 10]. In the present paper, we report on a first case of successful surgical separation of conjoined twins "pygopus" type in African country with limited resources.

CASE PRESENTATION

Female pygopus twins (Figure 1(a)) were born by cesarean section at 37 weeks gestation at a rural district hospital in Rwanda. They were transferred to our neonatal unit immediately after birth. They had a combined weight of 4 kg. They were joined back to back at the buttocks, and perineal areas. They shared one anus and two fused genitalia (Figure (b)). Physical examination revealed hypotrophic newborns but the assessment of the central nervous system, heart, and lungs was normal. Examination of the perineum showed two separated urogenital tracts with a thin membrane which constituted the posterior wall of the vaginal canals.

Plain radiograph of the spine showed the sacra to be separate (Figure 2(a)). There was no evidence of any other vertebral anomalies. Lower gastrointestinal tract studies by barium enema showed two separate rectums with a distal fusion in Y configuration (Figure 2(b)).

They tolerated oral feeds and started to gain weight, so it was decided to delay separation until their combined weight is greater than 5 kg. After two months the babies weighted 7.5 kg and the separation was decided. Before surgery, two teams of Rwandan surgeons with anesthesiologists and operating room nursing staff met to discuss the operation procedure, the management of the team and the theatre room.

The operation started by separating the soft tissues on the posterior gluteal region and we found that the vertebral column was fused distally at the level of the coccyx by a cartilaginous and fibrous tissues which were easily divided. The two slightly fused rectums were separated and vaginas were opened and repaired individually. For both babies, a posterior sagittal anorectoplasty was performed with derivated ileostomy.

After perineal reconstruction, the skin was closed primarily using the already raised V-shaped skin flaps. No skin grafting was needed. The operation lasted 3 hours and postoperatively, they were briefly ventilated in the

recovery room. Twenty four hours later, they were transferred to the neonatal unit and started oral feeding on the third postoperative day. No postoperative complications were observed and the perineal wound healed without any problem and the ileostomy closure was performed after three weeks. The babies were discharged to home on the 35th postoperative day with a weight of 4.250 kg for baby A (Figure 3(a)) and 3.700 kg for baby B (Figure 3(b)). One year later, they were found to be healthy and thriving normally without any residual problem.

DISCUSSION

Conjoined twinning is one of the most fascinating human malformations and has also been reported in other animals especially mammals, fishes, birds, reptiles, and amphibians [11].

Up to now, the cause of conjoined twins is not exactly known, but it is generally accepted that conjoined twins arise from a single zygote that fails to undergo complete splitting of two inner mass cells during the blastocyst period (5-6 days after fertilization). The conjoined twins also arise from the incomplete splitting of the inner mass cell or embryoblast while the embryo is undergoing the hatching from the zona pellucida at day 6 after fertilization. In rare cases, conjoined twins may result from an incomplete separation of the embryonic disc after 12 days of embryogenesis before the gastrulation begins [2-4]. This, as well as the classification, management, and prognosis of conjoined twins, have been extensively reviewed by Spencer [12, 13].

Treating conjoined twins can be a daunting challenge for the surgeon. Furthermore, these cases often raise religious, moral, ethical and legal issues [13, 14].

The diagnosis of conjoined twins can be made prenatally in centers where pregnant mothers are subjected to routine ultrasonography as early as 12 weeks' gestation [15]. However, The diagnosis can be missed on occasions and that happened in our patients. The importance of preoperative diagnosis needs to be emphasized because conjoined twins should be transferred in utero to a specialized center where future management is feasible.

Surgical methods to separate conjoined twins may range from relatively simple to extremely complex, depending on the point and complexity of attachment and the internal organs which are shared. Most cases of separation are extremely risky and life-threatening. In many cases, the surgery results in the death of one or both twins, particularly if they are joined at the head. Of all types of conjoined twins, omphalopus twins are the most favorable candidates for elective surgery due to good survival rates [16]. The surgery results in the death of one or both twins, particularly if they are joined at the head.

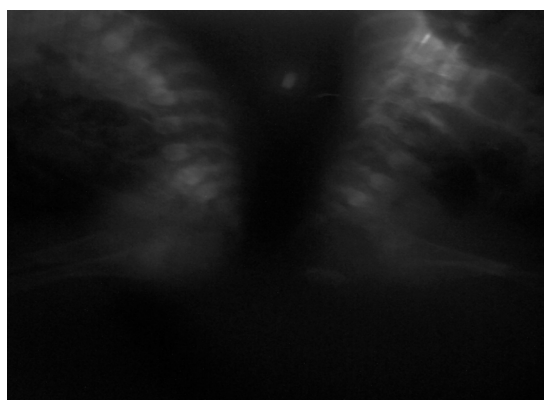


(a)

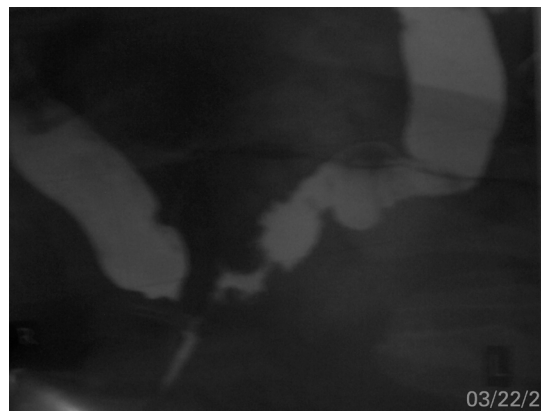


(b)

Figure 1: Clinical photograph showing female pygopagus twins joined back to back facing away from each other with attachments at the buttocks and perineum (a). Photograph showing two fused genitalia with one anus.



(a)



(b)

FIGURE 2: Plain radiograph showing two separate spinal columns (a); and lower gastrointestinal tract studies by barium enema showing two separate rectums with a distal fusion in Y configuration (b)



FIGURE 3: Photographs showing the two separated conjoined pygopagus twins, baby A (a) and baby (b)

Of all types of conjoined twins, omphalopus twins are the most favorable candidates for elective surgery due to good survival rates [16]. Emergent conditions may arise at any time and include intestinal obstruction, rupture of an omphalocele, congestive cardiac failure, severe degree of respiratory compromise, and terminal illness in one of the conjoined twins.

The first successful separation of conjoined twins was performed in 1689 by Johannes Fatio. The survival rate of conjoined twins, however, correlates with age at separation. It was less than 50% if surgical separation was attempted in the neonatal period, but increased to 90% if separation was delayed until 6 months of age or later [17].

In the present case, separation was performed when twins were 3-month-old because we consider that the babies were gaining weight and able to undergo the operation. The success of the separation depends of the case but the overall survival rate of 64% was quoted by Hoyle in 1990 [8]. Thoracopagus, craniopagus, and omphalopus were associated with the highest mortality rate (51%, 48%, and 32%, respectively), whereas lower mortality rates occurred with ischiopagus (19%) and pygopus (23%) twins [8].

Pygopus twins are very rare, accounting for about 10% to 18% of all conjoined twins, which gives an incidence of about 1 in 1,000,000 live births of this particular type [18]. They commonly share the gluteal region, terminal spine, and gastrointestinal, urological, and reproductive systems to variable degrees. About 50% of pygopus twins have anomalies unrelated to the classic fused organs including a high incidence of vertebral anomalies [5, 9]. Our patients had two separate rectums but fused distally in a "Y" configuration with one anal orifice, two separate urological systems, two sacra fused at the coccyx by a small band of cartilage and no spinal cord

fusion or other vertebral anomalies.

The surgical management of pygopus twins necessitates detailed radiographic examination of all urinary, reproductive, and gastrointestinal systems. Magnetic resonance imaging should be part of the preoperative investigations of all pygopus twins with particular attention to the anatomy of the spinal cord. Due to the limited investigation facilities in our settings, we performed on our patients just radiography to visualize the aspect of the rectums and the distal column and ultrasound to look for other associated malformations; MRI was not on our reach to see the anatomy of spinal cord but fortunately no spinal fusion was found during operation.

The overall survival rate of pygopus twins was estimated around 87%, and for males it was 100%, whereas for females it was 85% [5]. In this previous study, half of the twins had nonfused rectums, and half had fused rectums. The nonfused had 2 rectums (80%) or one rectum and one rectovaginal fistula (20%). The fused had high (46%) or low (54%) anorectal "Y" junction. All reported living male pygopus twins have had nonfused rectums. All these cases can be managed applying the principles of posterior sagittal anorectoplasty [3, 5].

In our case, the twins were females and they had two rectums with a lower junction with one anal orifice and the anorectal malformation was managed by posterior sagittal anorectoplasty protected by an ileostomy. The separation of our patients was successful despite limited facilities and the babies were discharged 1 month later in good condition without any sphincter troubles or motor deficits.

In conclusion, adequate preoperative investigations, a team approach, accurate operative techniques and good postoperative care contribute to the success of conjoined twins separation in general and pygopus twins separation in particular.

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