

## Successful surgical separation of conjoined twins: First experience in Rwanda

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*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. Female pygopagus twins of three months were been operated and separated at Kigali Teaching University Hospital. For both babies a posterior sagittal anorectoplasty was performed with derivated ileostomy without problem. No complications occurred during the operation, oral feedings was done at third postoperative day. Ileostomy closure was done three weeks after and babies were discharged from neonatology unit at 35th postoperative day. Adequate preoperative investigation with a well organized and trained team contributed a lot to the success of conjoined twins separation.*

### Introduction

Conjoined twins are one of the rarest and most challenging congenital malformations arise 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<sup>1, 2, 3</sup>. 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<sup>4</sup>.

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%). Other less common types of CT include: Pygopagus, Cephalopagus, Xiphopagus, Ischiopagus and Parapagus<sup>4</sup>.

Pygopagus twins are joined back to back facing away from each other, commonly share the gluteal region, terminal spine, and lower gastrointestinal, urological, and reproductive tracts and a recent reported study found that they represented 17% of all conjoined twins<sup>5</sup>. The first recorded pygopagus conjoined twins dates back to 1100<sup>6, 7, 8</sup>. 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 pygopagus 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<sup>5,9,10</sup>.

We report the first successful surgical separation of conjoined twins “pygopagus” type in Rwanda.

### Case report

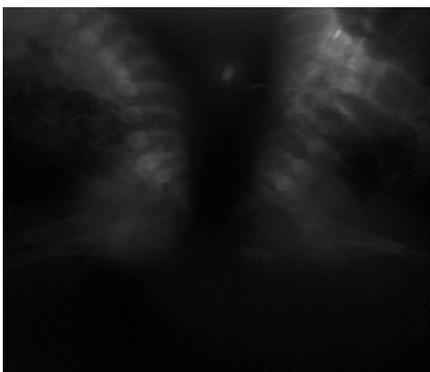
Female pygopagus twins ([Figure 1](#)) were delivered by cesarean section at 37 weeks gestation at a district hospital. They were transferred to our neonatal unit immediately after birth. They had a combined weight of 4 kg. They were joined back to back at the buttocks, and perineal areas. They share one anus and two fused genitalia ([Figure 2](#)). The physical examination showed hypotrophic newborns but the assessment of the central nervous system, heart, and lungs revealed no abnormalities. The examination of the perineum showed two separated urogenital tracts with a thin membrane which constitute the posterior wall of the vaginal canals.



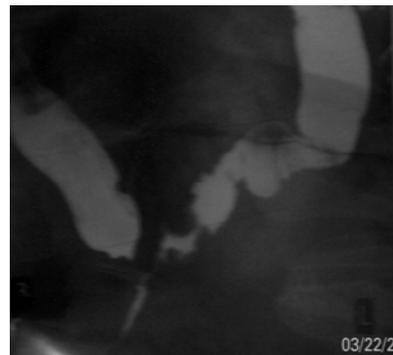
**Figure 1.** Clinical photograph showing female pygopagus twins joined back to back facing away from each other with attachments at the buttocks, and perineum.



**Figure 2.** This radiograph shows two fused genitalia with one anus



**Figure 3.** Plain radiograph showing two separate spinal columns.



**Figure 4.** Lower gastrointestinal tract studies by barium enema showed two separate rectums with a distal fusion in Y configuration

**Figure 5A.****Figure 5B**

Plain radiograph of the spine showed the sacra to be separate (Figure 3). 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 4). 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 surgeons with the team of 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 and the babies were discharged home on the 35th postoperative day with a weight of 4.250kg for baby A (Figure 5A) and 3.700kg for baby B (Figure 5B). The follow-up 1 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<sup>11</sup>.

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 arise also 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<sup>2,3,4</sup>. This, as well as the classification, management, and prognosis of conjoined twins, is extensively reviewed by Spencer<sup>12</sup>.

Treating conjoined twins can be a daunting challenge for the surgeon. Furthermore, these cases often raise religious, moral, ethical and legal issues<sup>13, 14</sup>. 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<sup>15</sup>. The diagnosis, however, can be missed on occasions as what happened in our patient. The importance of preoperative diagnosis needs to be emphasized because conjoined twins should be transferred in utero to a center where future management is feasible.

Surgery 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 of the twins, particularly if they are joined at the head. Of all types of conjoined twins, omphalopagus twins are the most favorable candidates for elective surgery because of good survival rates<sup>16</sup>.

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 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<sup>17</sup>. In this case separation was performed when twins were 3 months 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<sup>8</sup> in 1990. Thoracopagus, craniopagus, and omphalopagus were associated with the highest mortality rate (51%, 48%, and 32%, respectively), whereas lower mortality rates occurred with ischiopagus (19%) and pygopagus (23%) twins<sup>8</sup>.

Pygopagus 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. They commonly share the gluteal region, terminal spine, and gastrointestinal, urological, and reproductive systems to variable degrees. About 50% of pygopagus twins have anomalies unrelated to the classic fused organs including a high incidence of vertebral anomalies<sup>5, 9</sup>. 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 pygopagus twins necessitates detailed radiographic examination of all urinary, reproductive, and gastrointestinal systems. Magnetic resonance imaging should form part of the preoperative investigations of all pygopagus twins with particular attention to the anatomy of the spinal cord. Because of limited investigation facilities in our setting we



performed on our patients just radiography to see the aspect of the rectums and the distal column and ultrasound to look for other associated malformations; IMR 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 pygopagus twins was 87%, and for males it was 100%, whereas for females it was 85%<sup>5</sup>. 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 pygopagus twins have had nonfused rectums. All can be managed applying the principles of posterior sagittal anorectoplasty<sup>5,3</sup>.

In this case the twins were female 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 and the babies were discharged 1 month after in good condition without any sphincter troubles or motor deficits.

### Conclusion

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 pygopagus twins separation in particular.

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