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

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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 risky 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 derivated 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 en 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 retablie après trois jours postopératoires. La fermeture d’iléostomie a eu lieu trois semaines après et les deux jumelles ont sorti 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 placenta. 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,
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, omphalopagus 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.
**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.

**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, omphalopagus 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 on the case but the overall survival rate of 64% was quoted by Hoyle in 1990 [8]. 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 [8].

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 [18]. 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 [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 pygopagus twins necessitates detailed radiographic examination of all urinary, reproductive, and gastrointestinal systems. Magnetic resonance imaging should be part of the preoperative investigations of all pygopagus 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 pygopagus 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 pygopagus 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 [3, 5].

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.

REFERENCES