J Ped Surg Case Reports 2 (2014) 119–122

Contents lists available at ScienceDirect

Journal of Pediatric Surgery CASE REPORTS

journal homepage: www.jpscasereports.com

# Surgical separation of pygopagus twins: A case report

Prashant Jain*, Anjani Kumar Kundal, Rachna Sharma, Praveen Khilnani, Prem Kumar, Praneet Kumar

*12/25, West Patel Nagar, New Delhi, India*

ARTICLE INFO

*Article history:*
Received 5 January 2014
Accepted 4 February 2014

*Key words:*
Pygopagus
Conjoined
Twins

ABSTRACT

We report a rare case of Nigerian symmetrical pygopagus conjoined twins. They had sharing of anal canal, genitourinary system along with the fused spinal cords with a single dysplastic sacrum. Both the twin sisters were healthy and without any major health issues. The main challenge was to separate them without any neurological deficit, with continent bladder and bowel habits.



The separation of conjoined twin is a unique challenge due to its complex anatomy and physiology. Although advancement in imaging and monitoring has improved the survival rates, separation can be successfully achieved only with meticulous planning and team work.

Pygopagus conjoined twins represent 6–19% of all the conjoined twins [1]. They are joined at the sacral area with sharing of terminus of spine, gastrointestinal system, genitourinary system and spinal cord to a variable extent. They represent a group of conjoints in which the separation of the embryonic axis in the caudal region was incomplete [2].

The reported incidence worldwide is estimated at 1:50,000 to 1:100,000, live births, with higher incidence of 1: 14,000 to 1: 25000, experienced in Asia and Africa [3].

We share a rare case of symmetrical pygopagus twins who had sharing of all the three systems. We present our challenges and the treatment strategy for separation.

## 1. Case summary

8 months old, symmetrical pygopagus female twins were referred from Kano, Nigeria to our hospital. Parents were aware of the twin pregnancy but their conjoined status came to them as a surprise. It was a term normal vaginal delivery with birth weight (combined) of 4.5 Kg. Before being referred to us, they were kept in a hospital of Nigeria for about 8 months and did not have any major health related issues.

On clinical examination, their combined weight was 13.8 kg with normal development and milestones. They had a large area of fusion at sacral and perineal region with circumference measuring 45 cm (Fig. 1). The conjoined sisters were aligned in opposite directions. Both the buttocks were well developed on which twins could sit and lie on them. They were passing stools from a common anal opening, which was slightly stenotic. A single vestibular area with two pairs of well developed labia majora was present. In this single vestibular opening, four separate openings could be identified (two vaginal and two urethral). The two vaginas were sharing a common wall (Fig. 2). Their lower limb movements were normal without any sensory or motor deficits. Assessment of the central nervous system, heart and lungs was normal.

To avoid any error or confusion during the perioperative period, the twins were labeled as twin one and twin two and were given two different color codes (pink and blue). Twins were investigated in detail in the form of ultrasound abdomen, computed tomogram angiography, magnetic resonance imaging and cystourethrogram study. Imaging was suggestive of single dysplastic sacrum with fusion of spinal cord and thecal sac. They both had separate urinary bladder, urethra, uterus, rectum with single anal canal. There was an anomalous communication of internal iliac vein of both the twins, with shunting of blood from one to the other and vice versa. The entire spinal cords of both the twins appear normal, except on its lower most aspect where there was fusion of lower conus

* Corresponding author. Tel.: +91 9582413828 (mobile).
*E-mail address:* docpedsurg@gmail.com (P. Jain).

2213-5766 
http://dx.doi.org/10.1016/j.epsc.2014.02.008

**Fig. 1.** Pyopagus twins.

medullaris at S1 vertebral level and below this level the cord was continuing as a single filum terminale (Fig. 3).

MRI pelvis revealed a single puborectalis sling encircling common anal canal. Cystourethrogram revealed two separate normal urethra and urinary bladder.

A multispecialty team led by pediatric surgeon was constituted and a detailed strategy was framed. As both the children were healthy, so any neurological deficit after separation was unacceptable. Two separate teams from each specialty were constituted. To achieve perfection, multiple rehearsals were carried using dummies. Detailed counseling of the parents was done regarding the risk of paraplegia and incontinence.

In order to have adequate skin cover, two tissue expanders were placed over lumbar area in each twin and were gradually inflated over a period of two months.

For the separation surgery, flaps from thigh and lumbar area were initially raised in prone position and then in supine position. After raising flaps, neurosurgery team separated the spine and the spinal cord with the help of integrated neuromonitor (Fig. 4). Duraplasty was done giving extra cover with glue and pseudocapsular tissue.

This was followed by perineal separation by pediatric surgery team. Anal canal was mobilized using principles of posterosagittal anorectoplasty. Both the rectums were dissected and adequately mobilized (Fig. 5). Anal canal was divided in the midline. Vaginas were mobilized and separated along with the urethral opening. A large sharing vein between the two twins was safely ligated and cut.

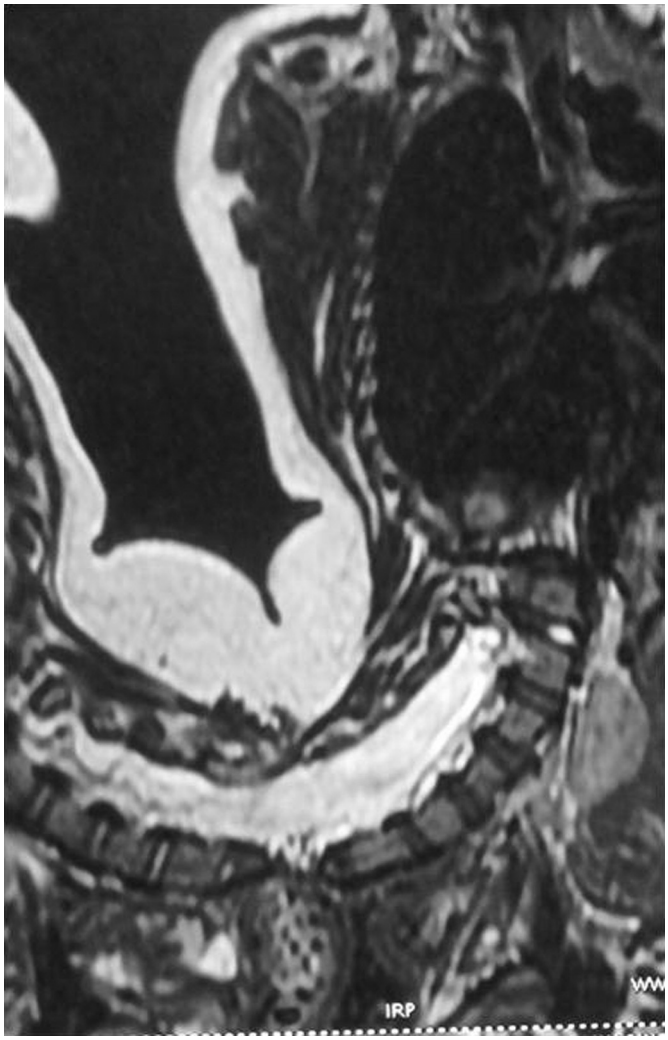


**Fig. 3.** MRI showing fused lower spinal cord.

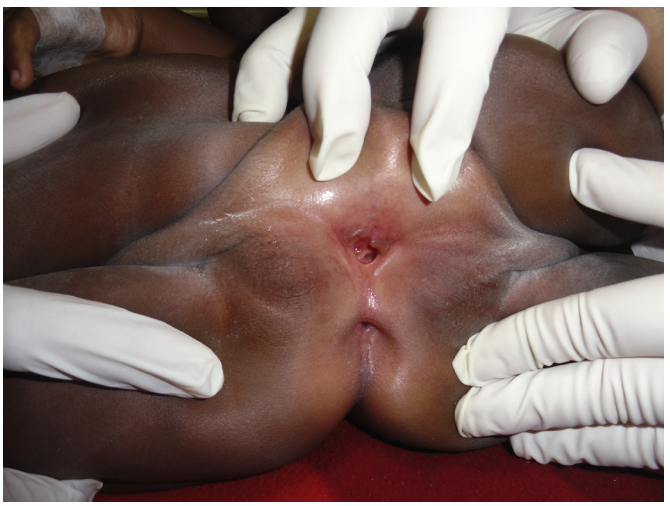

**Fig. 2.** Perineal view: sharing of genitalia and anal canal.

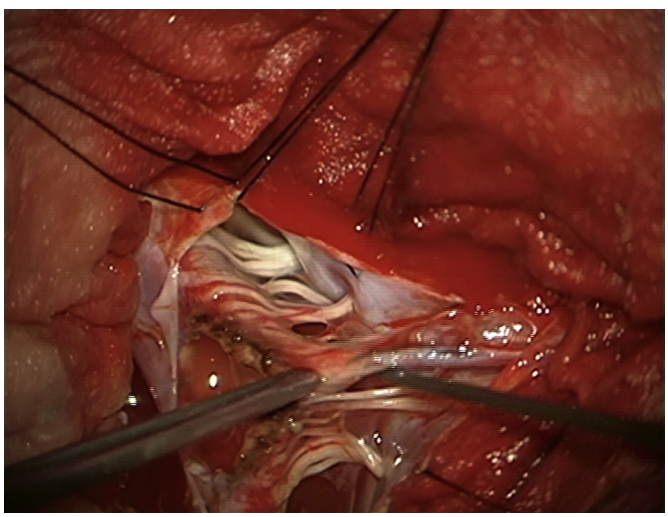

**Fig. 4.** Microscopic view of fused terminal ends of spinal cords.