Surgical separation of pygopagus twins: A case report

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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 conjoinits 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...
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.
Both the twins were separated after 13 h of the surgery and as per planning, Twin 2 was shifted to adjacent operation theater by assigned anesthesia team.

Final reconstruction was done in prone jack-knife position. Reconstruction of the urethra and vaginal opening was done with valvuloplasty (Fig. 6). Levator muscle present on one side was mobilized and reconstructed posterior to the rectum. Anoplasty was performed followed by covering right transverse loop colostomy. The total duration of surgery was 18 h with blood loss of around 750 ml.

During postoperative period, twins were shifted to pediatric intensive care unit and were kept on elective ventilation for 48 h. Twin 1 had a transient mild weakness in the right lower limb and Twin 2 had CSF leakage which gradually stopped after 5 days.

Presently after three months of colostomy closure, both the twins are without any neurological deficit and are continent for stools and urine. They have started walking with support.

2. Discussion

Conjoined twins are identical twins (monozygotic and monochorionic), developing with a single placenta from a single fertilized ovum.

As any other conjoined twin, pygopagus is more common in girls. Survival has been better in females as compared to males, and males that survive have less severe anatomic malformations [4].

With advancements in surgical skills, and technology, surgical separation should be the aim of management of conjoined twins. A success rate of as high as 70% has been reported whenever an elective separation has been planned. Though the separation of pygopagus may not carry high survival risk which is involved in craniopagus or thoraco-omphalopagus separation, any inadequate planning or decision can lead to lifelong debilitating morbidity. Hoyle, though has reported a lower mortality rates of around 23% in pygopagus as compare to craniopagus, thoracopagus and omphalopagus, the extent of attachment as well as the presence of other anomalies are of paramount importance [3].

Pygopagus twins share the lower part of the spine in 100%, lower gastrointestinal (GI) tract in 25% and genitourinary (with a single bladder and urethra) in 15%, but the spinal cords and cauda equina usually remain separate [5]. Besides skin coverage of large defect, a main challenge is separation of all three involved systems without compromising their anatomy and physiology.

An imaging strategy to accurately define the sharing of organs and vascular anomalies is important for surgical planning and prognostic information. As done in this case, three dimensional reconstruction of CT angiography images delineated sharing of gastrointestinal system, genitourinary system and sacral bone. It helped in deciding the shape, site and size of the skin flaps. A large sharing vein was identified which was crucial from anesthesia and surgical point of view. Rest of the information can be gathered from cystourethrogram study. Similarly MRI helped in planning for cord separation and dural repair.

A staged repair is the key in the successful outcome of the separation. We adopted a staged surgical approach for a good outcome by deciding the placement of tissue expanders and

Fig. 5. Separated fused anorectum.

Fig. 6. Vaginal and labial reconstruction.
conjoined twins [6]. Placement of tissue expanders provides adequate tension-free tissue cover for reconstruction which is important to prevent the morbidity of wound disruption, flap necrosis, CSF leakage (37.5%) etc [7]. In our case, the expansion was well tolerated and good skin coverage was achieved inspite of a large area of fusion. Also the CSF leakage was managed easily without any intervention because of good tension free wound cover.

Although the tissue expanders are not free from complications especially in children [8], proper expansion technique and gradual inflation can prevent these complications. Also placement of tissue expanders under general anesthesia provides a good opportunity for anesthetist to rehearse and hence cutdown the overall operative time during actual separation [6].

Similarly staging procedure by putting a temporary colostomy can prevent complications like wound disruption, enterocutaneous fistula, meningitis etc.

Hirokazu et al. reviewed literature of 33 cases of pygopagus and reported that of these, surgery was performed in 26 cases. He reported, that dural sharing was present in 17 cases while spinal cord sharing was present in 10 cases [1]. Our case adds to one more rare case with fused spinal cord and sharing of conus. Successful separation of the spinal cord was big challenge in our patient, as the long term outcome of the surgery was truly dependent on the preservation of neural structures. The decision of compromising neural structures becomes more difficult especially when both the twins had a normal development. Although intra-operative physiological monitoring has been recommended by some authors [7], we found it to be a useful adjunct in identifying the neural structures. We also recommend operating on neural structures as suggested by some authors [1,7], before proceeding for perineal reconstruction to avoid postoperative risk of meningitis.

Perineal reconstruction is another major undertaking in pygopagus. Accurate delineation of the anatomy with the help of imaging and careful planning can give good functional and cosmetic results. In these cases, perineal area has the maximum degree of fusion. The genitalia and anus are usually displaced posteriorly and in the midline. This case also had the similar anatomy as was described by Henneberg in Blazek sisters [9]. He described the theoretical mechanism of rotation-fusion of anus, urethra and vagina. The anus formed the apex of two shared isosceles triangle, the bases of which are formed by a line connecting the midpoint of the two pubic arches. The urethral and vaginal openings are normally oriented with respect to each other but rotated 90° from each pubic symphysis.

Pygopagus can have spectrum of anorectal anomalies depending on the extent of anorectal fusion. Only 41% of the female pygopagus twins have unfused rectum [4]. This case had a low fusion of anorectum with Y configuration about 2 cm above the pectinate line. They were sharing a common puborectalis sling. Principles of posterior sagittal anorectoplasty were followed to identify the sphincter muscles and reconstruct the sphincter muscles around the anorectum. Anorectal functions with two unfused anorectum and low fused anorectum have been reported to be good [4]. Both the twin sisters are continent for urine and stool after a follow up of 3 months.

A follow up examination and cystourethroscopy have revealed normal genitourinary anatomy with excellent cosmetic results. These twin sisters after completion of their first year have started walking with support. They still had mild positional scoliosis for which they are on regular physiotherapy.

A good invasive and non invasive monitoring required for cardio-respiratory assessment improves the survival rate, and the usage of neuro-monitoring system and muscle stimulator prevents the long term morbidity by preventing the neurological damage.

Besides team approach, meticulous planning and multiple sessions of rehearsal, we recommend:

1) Detailed preoperative imaging to know the degree of sharing
2) The surgery should be staged with the use of tissue expanders and diversion colostomy
3) Use of all monitoring systems like neuromonitoring and muscle stimulators
4) Neural dissection and dural repair should be done before the perineal dissection to avoid any contamination.

References