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ORIGINAL ARTICLE

Multimodality Management of Two Pairs of Pyopagus Twins

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Abstract:

Background and Aim: Conjoined twins, due to their rarity and complex anatomy, pose not only a technical, but also a physiological challenge for their separation, with each case being uniquely distinct. The aim of the present article is to describe the surgical approach and management strategy for two cases of pyopagus conjoined twins operated at our center.

Case Report:

Case 1: Antenatally detected conjoined twin girls presented postnatally to our centre. They were found to have a common vestibule with single anal opening facing partially away from each other. On evaluation they were found to have a single sacrum and fused conus and filum terminale. They were taken up for separation at 2years of age & the 24hour long surgery, culminated in successful separation. The children had good post-operative outcome at 10months post separation.

Case 2: Conjoined pyopagus twin girls presented postnatally, and were found to have fused cords, having a terminal syrinx and partially separate sacrum. They were separated at 2.5 years of age, with a multi-departmental effort and coordination. They are doing well 2 months post-operatively.

Conclusion: A multidisciplinary team support with thorough preoperative planning significantly aids in improving the outcome of surgical separation. This has been possible by using modern technology. Each reported case contributes significantly to literature.

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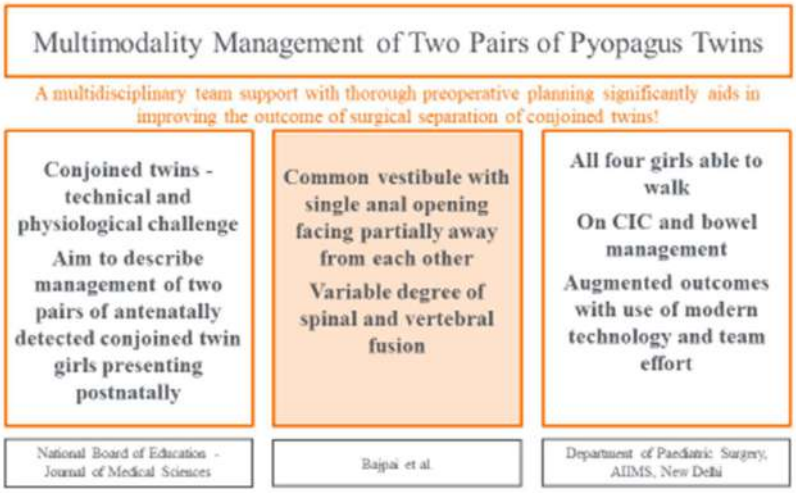
Graphical Abstract



Introduction

Twinning is an embryopathy and the spectrum varies, just as the constellation of anomalies, based upon the insults at different stages of embryogenesis [1]. Conjoined twins, due to their rarity and complex anatomy, have always been the source of academic and social fascination. [2,3].

The incidence is 1:50,000–100,000 pregnancies but as 60% are still-born or die soon after birth being incompatible with life [5], the true incidence is 1:200,000 live births with female predominance [4]. These same sex twins develop from a single fertilized ovum with a single placenta, however no well-defined etiological factors are known. Of the different types of conjoint twins, approximately 15–20% are pyopagus, their fusion is thought to occur at the region of the caudal neuropore, and the structures derived from the cloacal membrane in them are said to be normally developed [5].

We are describing two cases of pyopagus twins with complex sacral and spinal cord anatomy, involving detailed work-up of individual organ systems, along with the multidisciplinary approach to management and the final outcome.

Case Reports:

Case 1:

Conjoined twin girls, joined at the sacrum, with fused spinal cords, were diagnosed antenatally through a standard screening ultrasound to a 28 years old mother.

The twins were delivered via Caesarean section at 36 weeks of gestation with a combined birth weight of 5000 g. The twin on the right in anatomical position was designated as Twin A and the one on the left as Twin B.

Initial examination revealed the twins to be fused at the lower spinal region, sharing a perineum with a single anus, so that they faced away from one another in a partially oblique fashion [Fig. 1]. Although both were moving their lower limbs, they had associated neuro-orthopaedic deformities. They eventually developed a degree of deformational plagiocephaly.

A series of investigations were performed to clearly delineate the anatomy, while assessing their growth and neurological milestones. Imaging revealed spina bifida of L3 to L5 vertebra, fusion of the sacral vertebrae (S2 - S5) with common coccygeal vertebrae. [Fig. 2a] The rectum from both the twins was seen to open into a single anal opening. Both had crossed fused ectopic kidneys to their shared side. Twin B had a prior right MCA territory infarct with cystic encephalomalacic changes with dilatation of the ipsilateral lateral ventricle and atrophy seen in right half of brainstem. Magnetic resonance imaging of lower spine revealed two separate conus medullaris with a common fused filum terminale [Fig.2b]. Arterial supply and venous drainage of the right leg of twin B was found to be from the left internal ilac vessels of twin A [Fig.2c].

One of the most useful imaging adjuncts, was the development of a three-dimensional acrylic model of the ilia, sacrum and lumbar spines using a 3D printer, which allowed a thorough appreciation of the skeletal

anatomy, site of fusion and orientation, aiding the operative planning [Fig.3].

A management team was organized that consisted of paediatric surgery, plastic surgery, cardiothoracic and vascular surgery, anaesthesiology, radiology and neurophysiology.

Prior to the separation surgery, neuronal mapping was done of the lower limbs and anal sphincter which revealed some weakness in myotomes of twin B. The anal sphincter was predominantly controlled by twin A. There were some cross over fibres between twin A and B, MEP to twin B stimulated lumbar myotomes of twin A [Fig.4].

Six months prior to separation, the plastic surgery team inserted tissue expanders however they had to be removed due to resulting infection, leading to slight delay in the final separation.

The operation was performed at our facility at a date accommodating all surgical specialties, which corresponded to the twins' age of 24 months when the preoperative combined weight was 15kg.

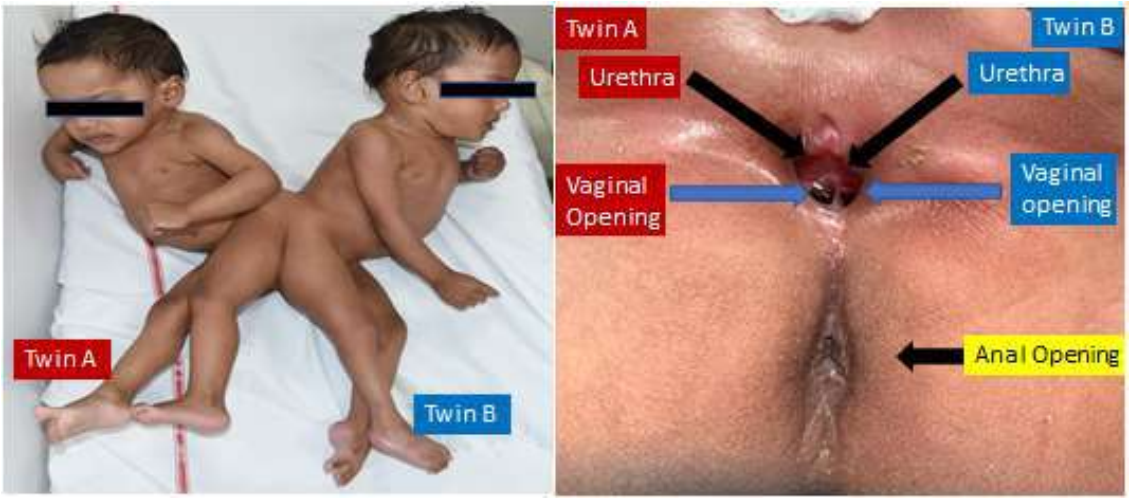


Fig. 1. a. Presentation of the pyopagus conjoined twins, joined at the hip, facing partly away from each other, b. Common perineum with two urethral and vaginal openings and single anal opening.

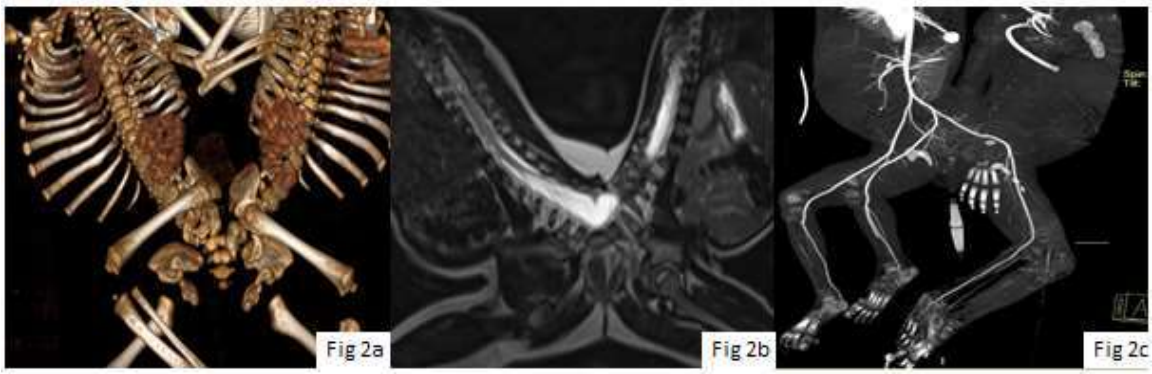


Fig. 2a. CT reconstruction demonstrating spina bifida of L3 to L5 vertebra, fusion of the sacral vertebrae (S2 - S5) with common coccygeal vertebrae;

Fig. 2b. Magnetic resonance imaging of lower spine showing two separate conus medullaris with a common fused filum terminale;

Fig. 2c. CT Angiography demonstrating the arterial supply and venous drainage of the right leg of twin B to be from the left internal iliac vessels of twin A.

Fig. 3. A three-dimensional reconstruction model of the shared bony anatomy.

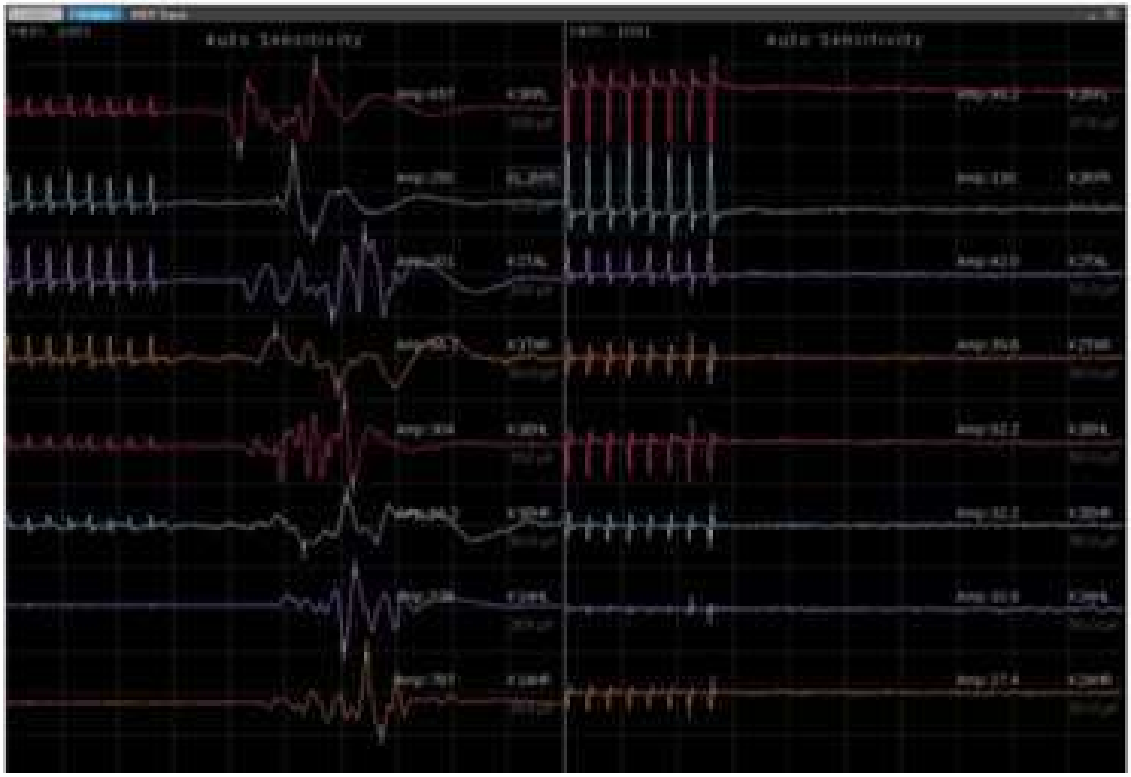


Fig. 4. Neuromonitoring done under anaesthesia showing MEP to twin B stimulated lumbar myotomes of twin A indicating crossover of neural fibers from B to A.

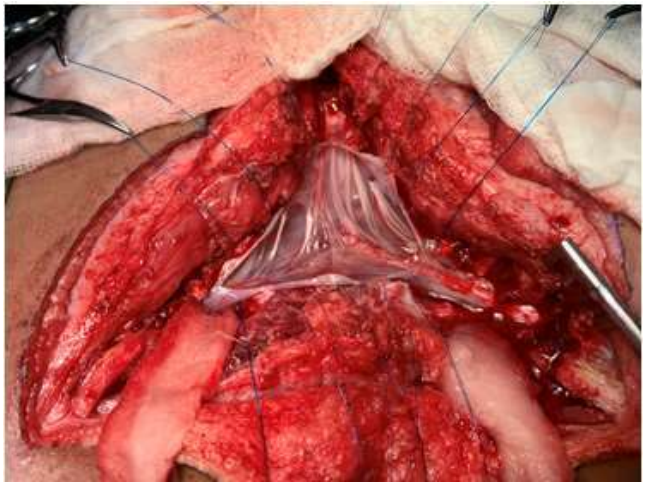


Fig. 5. Laminectomy in first set of conjoined twins revealing a U- shaped spinal cord and fused filum terminale.

An important anaesthetic consideration was restrictive small perimembranous VSD with left to right shunt in twin B. Anaesthetic workflow for the separation surgery had been well rehearsed by the anaesthesia team with models and a mock drill. The lines and drugs for the twins had been colour coded. The patients were then each intubated, with central, arterial and peripheral lines placed. Initially, the patients were positioned in supine position, bladders were catheterized, marking of flaps was done followed by electrode placement by neurophysiology team. The twins were then placed prone, following which surgery was commenced by the paediatric surgery team with exposure of the lower lumbar spine and sacrum by raising subcutaneous flaps as pre-decided for closure in consultation with plastic surgery team.

Spine of twin I identified after diving the paraspinal muscles, ligamenta flava and longitudinal ligaments. Laminectomy was performed in both twins, dura was found to be complete and durotomy was done under the effect of mannitol, the incision was carried down to expose a U-shaped spinal cord with asymmetric and complex cauda equina and fused filum terminale [Fig.5]. Once detethered, neurophysiological monitoring was used to identify nerve roots of individual twin. Bipolar lead used to identify response in twins, if absent confirmed with monopolar lead, keeping in mind the high rate of false positive responses. The main concern were the nerve rootlets arising at the distal and lateral aspects of the fused cord, some of which passed distally and contralaterally. The fibres passing contralaterally were found to be cross over fibres from twin B to A. The nerve roots were separated to the side of individual twins as per the neurophysiological response. Cordotomy done as per the plane of demarcation supplemented by neurophysiological response. Primary dural closure was possible in both twins using 6’0 prolene continuous sutures after inserting a thecoperitoneal shunt (Chabra Lumboperitoneal shunt).

After clearing the tissue caudally in the midline, the complex anatomy of the sacrum identified was suggestive of fusion of 2 sacrum with a preserved lateral sacral foramina to each twin which were juxtaposed to each other and crowding of the cauda equina in a paramedian fashion leaving a very thin strip of bone in the median position. This fused sacrum was divided in midline, preserving neural outflow with the help of responses to neuronal stimulation.

On dissection of the presacral space, peritoneum opened to further visualize pelvic structures. Using Hegar's dilator, the rectum of both twins were identified, which were fused in a Y configuration, leading into a common anal canal [Fig. 6]. This common anal canal and sphincter with adjacent levator ani fibres were kept to the side of Twin A. The rectum of twin 2 was disconnected from the common rectum with the help of a linear stapler and pull through was done by hanging bowel technique [6]. The perineal skin incision completed in as per the previously designed flaps to separate the vagina and urethra.

Continuing the rest of the surgery in supine position, the aberrant supply to left

lower limb of twin B was restored primarily, perineum and soft tissue separated and skin closure done by flaps and cadaveric grafts. The total anaesthesia duration was 24 hours.

The post-operative course was prolonged and constituted intensive critical care management with the help of the pediatrics team. The cadaveric grafts were eventually rejected and replaced by autograft harvested from Twin A. Twin B suffered from perineal wound dehiscence for which she was re-operated and an enterostomy was created. Once the perineal flaps had healed, bowel trimming was performed. Children were then started on clean intermittent catheterisation and bowel management programme for Twin A. Physiotherapy and mobilisation started using splits and walkers. At 1 year of follow up, the twins are doing well [Fig.9a].

Case 2:

Antenatally detected conjoined twin girls, were delivered at term with a combined weight of 5500gms and presented to us postnatally. As in the first case, they were also pyopagus twins, joined at the sacral region, facing partially away from each other. They had a common perineum with separate urethral and vaginal openings but a single anal opening [Fig.7]. The twins also had associated neuro-orthopedic deformities and deformational plagiocephaly.

Complete investigational panel was done showed multiple lumbar vertebral defects in both twins with fused lower sacral vertebra, S2 of twin A and S3 on twin B onwards, with open posterior elements. Magnetic resonance imaging revealed that the conus was low lying and fused at L5 level. There was a syrinx in the cord, which over 1 year increased in size to reach the conus, as a result there was a 1.3 mm of neural tissue separating the fused conus with terminal syrinx and dysplastic neural tissue. Hyperintensity seen in the bilateral deep white matter in the occipital lobe of Twin B due to suspected metabolic insult. There was a small branch arising from left external iliac artery of twin A, seen to cross midline anteriorly to supply the antero-lateral part of right upper thigh of twin B, which had its own vascular supply. Bilateral kidneys were normally located, echocardiography was essentially normal except for dextroposition of Twin A. On contrast enema, there was a common channel length of 2.1cm which bifurcated into the respective rectum of the twins.

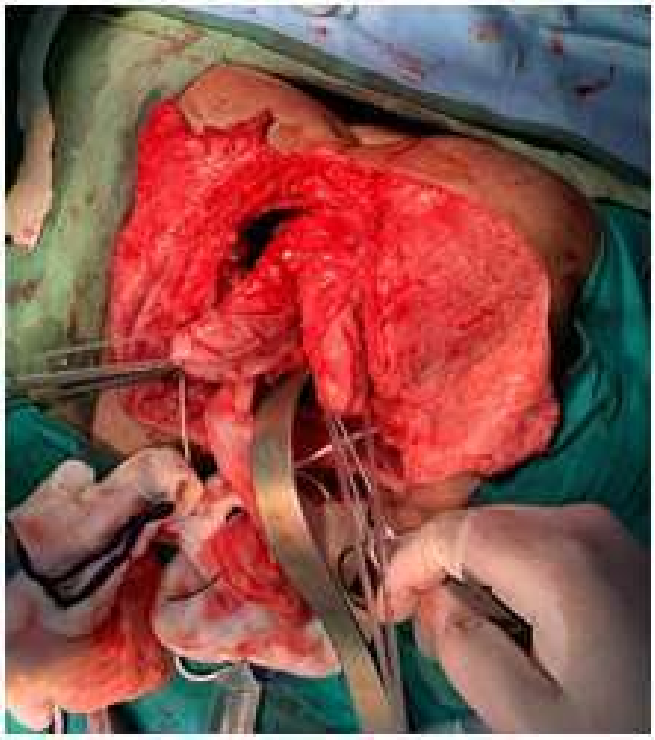


Fig. 6. A Y-shaped configuration of the rectum of both twins were identified leading into a common anal canal.

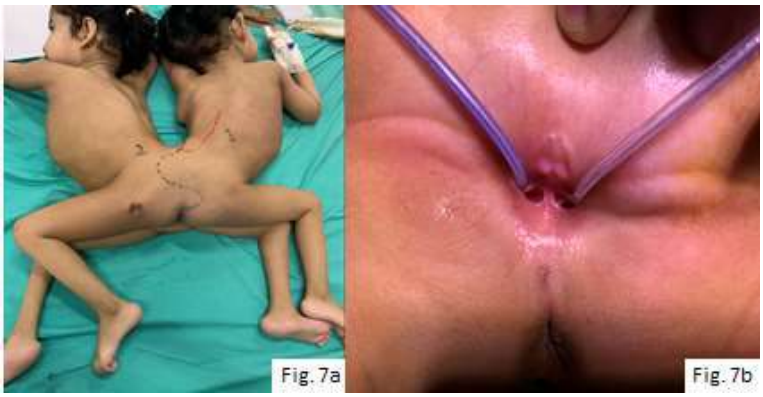


Fig. 7. Second set of pyopagus twins showing nearly identical anatomy, with a single perineum, separate urethral and vaginal opening and single anal opening.

Fig. 8. Lower shared part of spinal cord in second set of twins showing a terminal syrinx.

Fig. 9. Separated twins prior to discharge with intact motor functions.

The preoperative planning and management progressed in a similar manner as previous set of twins; however, tissue expanders were not used and neurophysiological monitoring was conducted after induction during the separation surgery.

The separation surgery progressed in a similar manner as in the first case. The dura was common in the lower aspect enclosing a U-shaped spinal cord, terminal part of which had a syrinx [Fig. 8]. Intra-operative neurophysiological monitoring showed no cross over and equal control of the anal sphincter, which was given to the neurologically better twin A. Enterostomy was not required in either twin. The crossing vessel

could not be separately identified and was ligated in the cleavage plane during the separation. The total anesthesia duration was 13 hours.

Subsequently in the post-operative period, twin B underwent trimming of the hung bowel and both twins underwent autologous skin grafting harvested from Twin A, after rejection of their cadaveric grafts. There was no evidence of lower limb weakness.

Currently both twins are doing well at 4 months follow up and have been started on bowel training program and clean intermittent catheterization. They are being mobilized with the help of splints and walkers [Fig.9b].

Discussion

Conjoined twins, joined at homologous sites are clinically classified based on the most prominent site of union [7]. Pyopagus conjoint twins represent a group in which separation of the embryonic axis in the caudal region was incomplete during 3rd-4th week of gestation, with resultant fusion of the sacral and neural elements to varying degrees. The first successful separation of conjoined twins was performed in 1689 by Johannes Fatio [8]. To our knowledge 37 cases of pyopagus conjoined twins have been described in detail in literature, and ours is the first report of two cases managed successively with all 4 surviving twins.

A thorough planning and preoperative evaluation is needed prior to separation in view of a very complex anatomy and high incidence of associated anomalies [9], surgery should be planned on an elective basis at a time when the twins gain weight and are stable to handle the surgical stress of major reconstruction. The survival rate correlates with age at separation being less than 50% if attempted in the neonatal period but increased to 90% if separation was delayed until 6 months of age or later [10].

Majority of these cases have bony fusion and 68% have sharing of dural sac [11]. Conjoinment of the distal cord and cauda equina forming a U-shaped spinal cord is not uncommon. A spectrum of anomalous, intraspinal variations exist involving the spinal cord & this group of anomalies possesses unique challenges in separation [12,13].

Functional and anatomical midline may not coincide. In our case, use of preoperative neurophysiological monitoring along with its intraoperative use helped to divide the spinal cord in the functional rather than anatomical midline, preserving the outflow to each twin, mitigating post-operative neural deficits, including neurogenic bladder [14].

It also helped to allocate the anal sphincter to the twin in better control of it. Role of preoperative neuromonitoring cannot be over emphasized, as it helped in preemptive planning and saved prolonged general anesthesia time during the final separation [12]. The anatomy of the common sacrum was further complicated by the apparent fusion of 1st sacral vertebra which led to the medial sacral outflows of both twins being crowded and exiting through a fused common foramen, where surgical separation was immensely benefited by neuronal monitoring. The benefit was self-evident during the immediate post-operative period when the children were able to move their lower limbs.

Placement of a lumbo-peritoneal shunt helps in preventing CSF leak which enables the flaps and grafts placed to heal adequately. A thorough mechanical bowel preparation and performing neurosurgical separation and dura closure first helps in preventing ascending CNS infection in these children, also requiring concomitant gastrointestinal surgery.

Some of the pygopagus conjoined twins have significant cross-circulation, and in up to 25 %, large anastomotic vessels including transverse veins may be observed intraoperatively leading to unexpected blood loss by damaging unidentified vascular structures [11].

The outcome of separation is highly dependent on the use of modern technology by an expert multidisciplinary team at a tertiary care center. Exhaustive parental counselling is mandated prior to surgery, explaining the possible complications, and expected outcomes resulting from an extensive and prolonged surgery.

Conclusion

Outcomes of surgical separation of conjoint twins have drastically improved over the years. Surgical separation, however, continues to be very challenging involving multiple organ systems, needing support from a multidisciplinary and dedicated team. Due to the rarity of cases and complexity of anatomy, every successfully separated case adds immensely to the literature to help guide and improve subsequent outcomes.

Statements and Declarations

Conflicts of interest

The authors have no competing interests to declare that are relevant to the content of this article

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