

Surgical Separation of Pygopagus Conjoined twins at Holy Family Hospital Rawalpindi; a Neurological Surgery Perspective and Outcomes: a Case Report

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ABSTRACT

Objective: Objective: Conjoined twins are considered to be the result of aberrant embryogenesis with incomplete embryonic separation. Estimated incidence of Conjoined twins is 1:50,000 pregnancies, and 1:200,000 live births, in new-borns with a female predominance of 3:1.

Conjoined twins are classified into different subtypes according to the site of fusion: Thoracopagus, Omphalopagus, Pygopagus, Ischiopagus, and craniopagus. Even in conjoined twins, Pygopagus has an incidence of 6-19%; subsequently, not many cases have been reported worldwide.

Case: This case report is about 6 months old female Pygopagus conjoined twins who fused at the lumbosacral level with a shared spinal canal. They were successfully separated in Pakistan with great success and had no neurological deficits on consequent follow-ups. An adequate build-up of physiological parameters until twins are old enough to undergo separation with multi-disciplinary management is the cornerstone of the successful management and separation of Pygopagus twins which is only possible in planned elective settings. Whereas emergency separation in the background of concomitant sepsis and sharing of vital organs carries high mortality and raises valid ethical questions regarding which life to prioritize saving and what organs to give to either viable child in case of shared CVS, GIT, and genitourinary systems.

Keywords: Pygopagus Conjoined twins, paediatric, surgery

INTRODUCTION

Conjoined twins, also known as Siamese twins, popularly thought to be a product of erroneous embryogenesis is a rare condition occurring in 1:50000 births but accounting for approximately 60% of cases ending in stillbirth; the true incidence lies between 1:200000 with female predominance [1]. History of conjoined twins goes as far back to caveman drawings, from elevation to divine status by ancient Romans i.e. Janus, to have been feared into culling and killing or put on display for public interest in circuses, at a point they were even revered by churches as saints; most famously is the Biddenden Maids.

Records of attempted separation of twins go back to a millennium in Byzantine; the first successful separation of conjoined twins was reported in the literature by Johannes Fatio a Swiss surgeon who successfully separated Omphalopagus twins, Elizabeth and Catherine Mayerin, in 1689 (After tracking the umbilical vessels to the navel, he applied a ligature to the connecting ensiform cartilage then tightened it over several days until it cut completely through) [1].

With the accepted understanding that conjoined twins are a result of errors in embryogenesis resulting in an abnormal form of monozygotic twinning, two theories are accepted i.e. fusion and fission (Kaufman, 2004; Spencer, 2000a; Spitz, 2005), which state that aberrations occur due to incomplete splitting or cleavage at primitive streak (fission theory) or joining of an embryo at vulnerable sites after complete splitting (fusion) but both have their limitations due to non-conclusive testing and rare phenomenon [2].

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Boer et al proposed that the underlying initial duplication of axial morphogenetic potent primordia could be the starting element in the genesis of ventrally, laterally, and caudally conjoined twins which offers a better explanation and understanding of the formation, etiopathogenesis, and embryogenesis of conjoined twins (3).

Siamese twins are classified based on the site of their attachment site i.e. abdomen (omphalopagus), thorax (thoracopagus), head (craniopagus), sacrum (pygopagus), vertebral column (Rachiopagus), based on site of lateral fusion (parapagus and its subvarieties encompassing varying degrees of thorax and cranium involvement) (4). Pygopagus comprising 6-19% of the conjoined twins poses a unique challenge to neurosurgeons with complex anatomy of the fused spinal cord with variable extent having a common terminal portion of the spine, fused genitourinary system, fused gastrointestinal system (1).

Due to multi-organ system involvement and unique physiology of conjoined twins a multi-disciplinary approach which including Neurosurgery, Paediatrics Surgery, Paediatric medicine, and Obstetrics is required from initial diagnosis on prenatal scans to delivery of baby, survival, and gradual build-up till surgery requires coordinated multi-disciplinary effort and resources, especially in a setting like ours where our health system is already burdened and choked with the inaccessibility of health to all, the lack of trained staff and specialized instruments are a challenge to deal with such delicate tactful cases.

CASE

This case report is about conjoined twins diagnosed at 20 weeks on prenatal scan in a private setup and presented to our setup at 22 weeks. The mother was 30-years-old G5P4A0 who had antenatal care at a local hospital. At the 28th week, the mother developed anaemia and was admitted and treated, an early caesarean section was performed at 37 weeks, the

APGAR score at the birth of both twins was 10, and the combined weight was 3500 grams on examination they faced opposite to each other in obvious oblique pattern, and there were no gross musculoskeletal abnormalities noted. However, both were fused at the lumbosacral region with a sacral circumference of 14 cm at the conjoined area. Detailed neurological examination was unremarkable, with normal power in both lower limbs, normal reflexes, and no neurological deficit of any sort.

They had a single common anal opening and a separate vagina. Four separate openings could be identified (two vaginal and two urethral). Assessment of CVS and the respiratory system was unremarkable. They were shifted to a high dependency unit for postnatal care as they were deeply jaundiced, oxygen-dependent, and in sepsis which was being managed accordingly.

They developed abdominal distention with respiratory distress, with scanty stool being passed from a single anal opening. Pelvic divided colostomy was made on the 7th day for both infants with per-operative findings of separate small and large gut until the rectum. They were shifted to CPAP for respiratory distress, breathing settled over time, and free of oxygen dependence.

In subsequent workup, echocardiography showed no abnormality and both CVS functions were in the normal range.

CT angiography of the abdominal aorta showed separate blood supplies with no dependence on each other, and CT Urogram was unremarkable.

- MRI of the Lumbo-Sacral spine showed almost complete fusion of Coccyx with sharing of the common spinal canal, common dural sac, fibers of cauda equina which were fused with spinal canal at the Coccyx

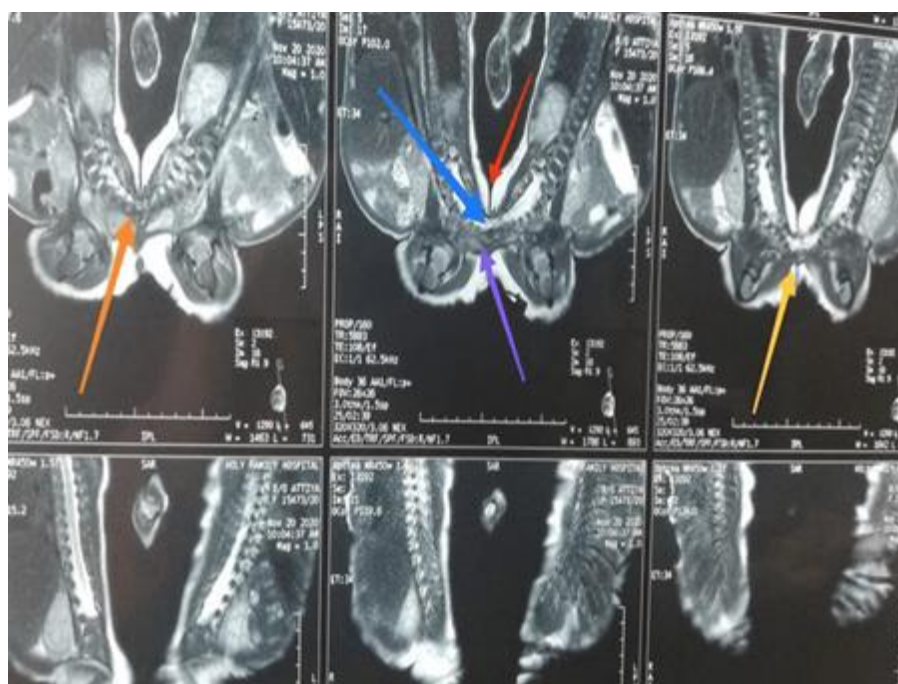


Figure 1: MRI lumbo-sacral Spine showing complete fusion of Coccyx with common shared spinal canal

CT Angiography was performed in the second month of life with coronal, sagittal reformats and 3D reconstruction, which showed normal vascular anatomy but vertebral bodies till SV4 with complete non-visualization of SV5 and Coccyx. Multidisciplinary management board comprised paediatric surgery, paediatric medicine, neurosurgery, plastics surgery, anaesthesia department, and it was decided to separate the twins at the 6th month of life. In this study, we will focus on neurosurgical aspects of the separation of conjoined twins.

When they reached the 6th month of age, surgery was planned, and the management team was organized comprising Neurosurgery, Paediatric surgery, and anaesthesia. Twins were operated in Holy Family Hospital. Magnifying loupes were used by neurosurgeons. Pre-operative preparations were done as per standard bowel preparation, and following standard protocol consisting of 4-hour nil per oral pre-op, 2 pints of Blood were arranged and kept in hand. Regarding anaesthesia consideration, two anaesthesia teams were ready and involved, and rehearsal by anaesthetic was done pre-operatively. Peripheral lines were placed. Both were labelled as Baby 1 and Baby 2 for convenience, to avoid confusion among management teams, and to have separate identities after separation.



Figure 2: complete non-visualization of 5th sacral vertebrae and Coccyx.



Figure 3: Incision marking and planning

They were anesthetized separately on separate anaesthetic medicine and machines, The twins were repositioned on the OT table as planned by the neurosurgical team, the skin was prepared and draped, an incision was made, and subcutaneous dissection was done up to the conjoined sacral area where the bone was skeletonized using monopolar cautery under magnification of loupes posterior part of the conjoined dysplastic sacrum was removed to expose the conjoined dural sac.

Dural Sac was carefully exposed to the extent that dural repair would be easily possible. After exposing the dural sac, a linear incision along the longitudinal axis of the sac was given, and CSF flow was observed. Cauda Equina rootlets were observed to be found to be conjoined.

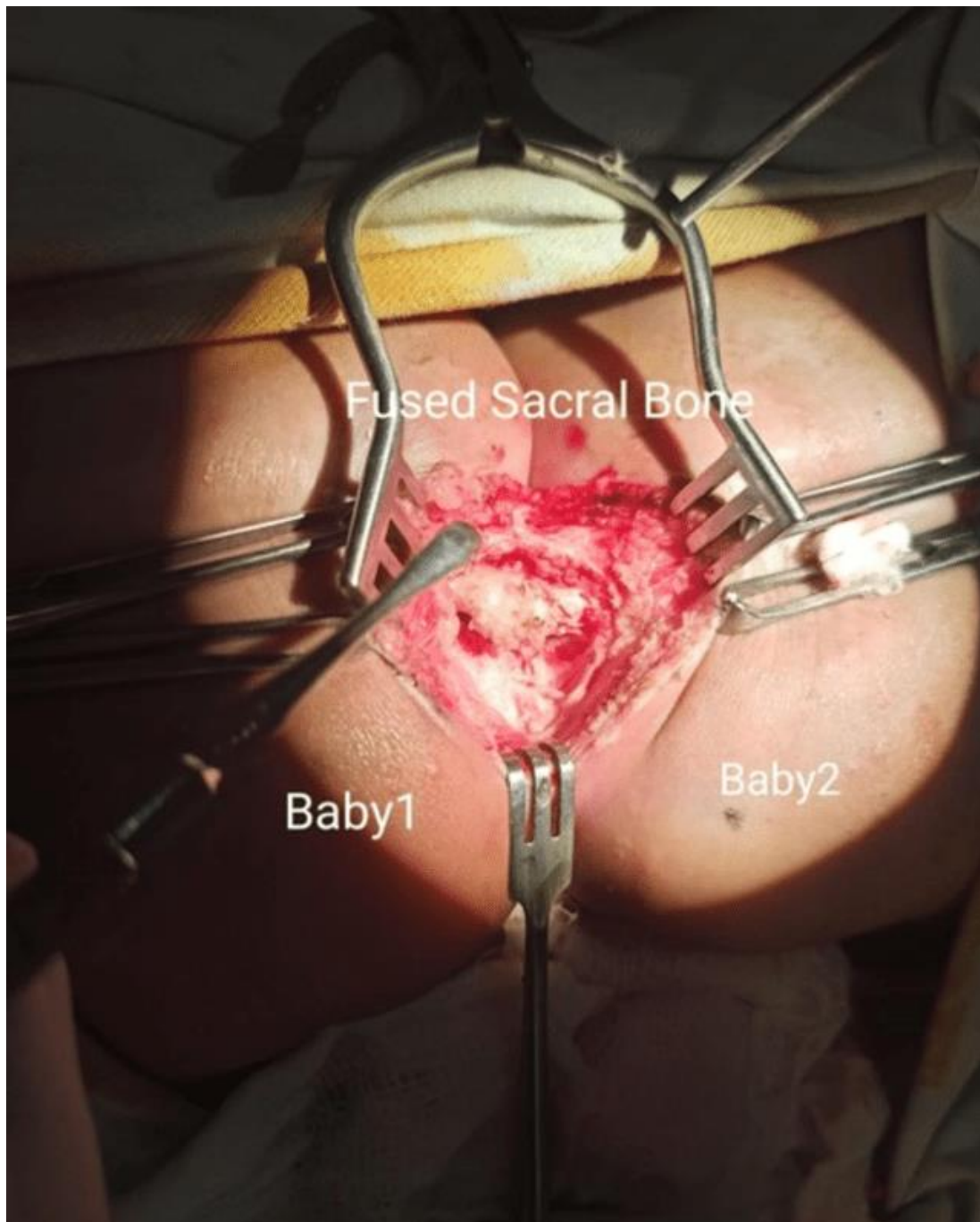


Figure 4: Fused Dysplastic Sacral bone.

We did not have a nerve stimulator or intra-operative neuromonitoring, so we carefully resected the Filum Terminale and Cauda Equina rootlets. After separation and arachnoid dissection, rootlets retracted back into the spinal cord of both babies.

After making sure that rootlets were not attached to the dura, both dural sleeves were closed in a watertight fashion. After that, soft tissue dissection was done, and reaching near the anal canal, babies were handed over to a team of paediatric surgeons who found a common anal opening.

Anoplasty was done at the spot, and the anal canal was reconstructed separately both babies were shifted on separate tables, and by two teams of neurosurgeons and paediatric surgeons, skin flaps were closed in a routine fashion. The wound was closed in reverse order after confirming that there was no CSF leak.

No neurological deficit was observed and normal anal and urethral sphincter tone was examined at follow-up after surgery. Mild CSF leakage in baby 2 was observed which after applying stitches to the skin was stopped within 12 hrs. No further complications were observed on subsequent follow-ups.

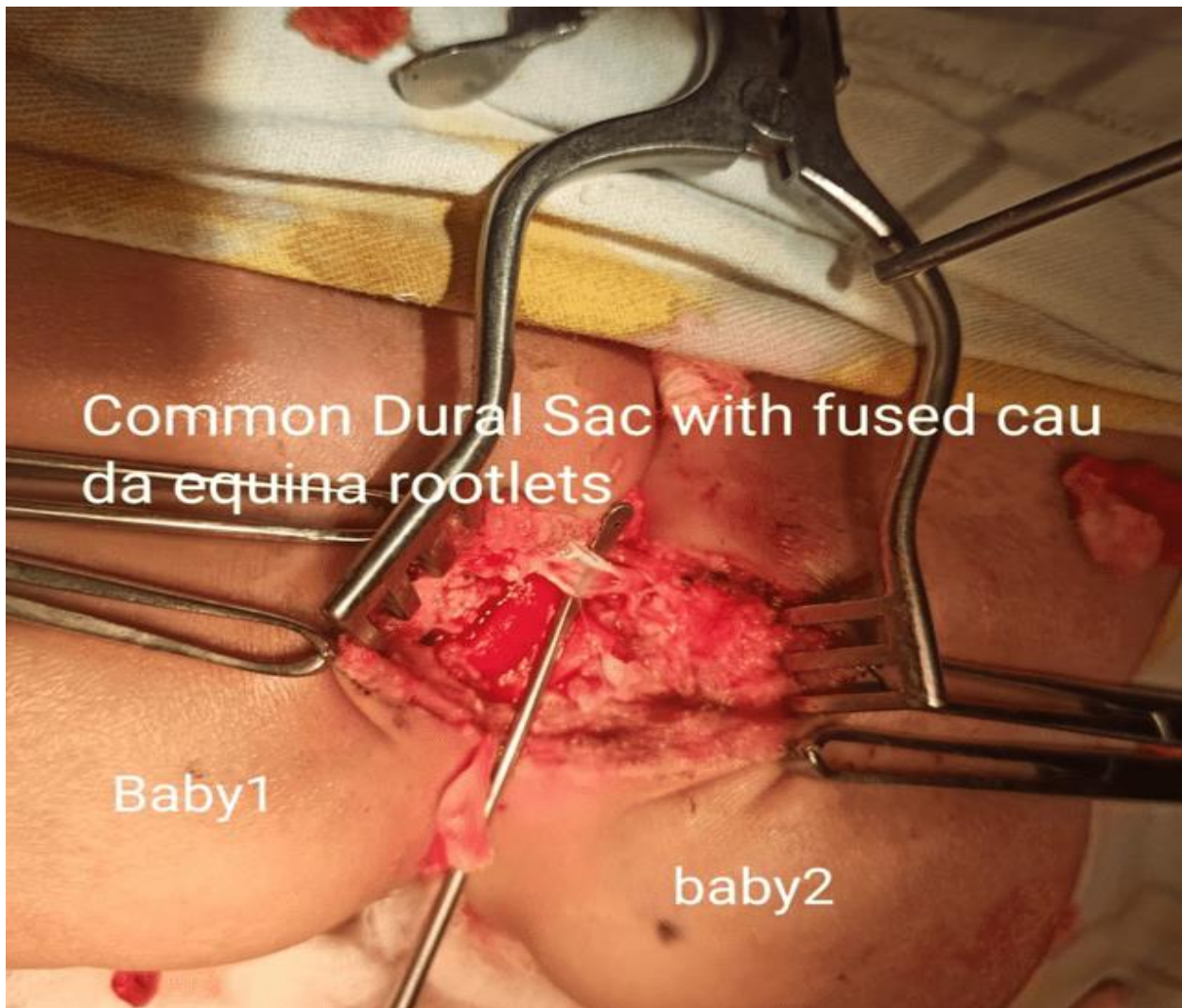


Figure 5: Common Shared Spinal Canal.

DISCUSSION

With the dawn of the scientific age era, Siamese twins are no longer looked at as high mortality or poor prognosis condition, even in the congenital diversity of Siamese twins Pygopagus has a unique presentation as in they are fused at the sacrum and perineum with the union of spine, rectum, urinary tract, and reproductive organs which make up unique physiology and challenging complex anatomy for surgeons and consequently difficult anaesthesia induction, and at the same time they pose a moral dilemma in prioritizing about which of the twins to be saved, and deciding what organs to give particularly in case of the fused anus, urethra, vagina which pose a unique ethical issue for all involved parties hospital authorities, doctors and parents (5). Due to the rarity of the cases and even fewer reported operated cases for Siamese twins, which are 25 cases reported up to date (6), many aspects of Pygopagus from aberrant embryogenesis to surgical management, differ as there is no mutual consensus about definite guidelines, in the modern era our neurosurgical goal is to redefine the anatomy of the spinal canal and vertebral column in a way that prevents neurosurgical deficit, dural CSF leak, prevent infection, pseudomeningocele formation, however in our case, one of the twins developed CSF leak 3rd day postoperatively which is reported in 37 percent of the operated cases of Pygopagus (7,8).

However, it was managed conservatively by applying stitches and giving antibiotics for reduced risk of infection. Neural separation and reconstruction were performed before perineal reconstruction to reduce the risk of post-operative meningitis and avoidance of any possible fecal spillage and contamination; similarly, Prashant Jain et al reported that separation of neural structures before perineal reconstruction carried a better prognosis considering the reduced risk of post-operative meningitis (7).

This was the first successful separation operation of Pygopagus in Pakistan despite having a lack of resources in LMIC countries like ours.

Government and existing health models tend to focus on preventive and curative aspects of communicable diseases, thus focusing less on 'costly' and 'complex' provision of surgical care; neurosurgery has thus been deprioritized (9).

Pygopagus is an extremely delicate and complicated procedure, and many authors have reported the use of novel devices like three-dimensional solid models (6), intraoperative neuromonitoring, SSEP (somatosensory evoked potential) (10), teleconsultations (11), CAD (computer-aided designs) techniques (12) for 3D

reconstruction of the whole anatomy and allowing rehearsals and planning of the operation.

But in our resource-limited setup, we had no available option of the utilities mentioned above, yet an attempt was taken, and Pakistani Neurosurgeons performed separation, and there was no neurological deficit or complication observed in consequent follow-ups. 33344dsasdf6yu7olöo.şigGü

This report emphasizes the need for a multidisciplinary approach as well as delaying the surgery till when the physiological reserves of the twins are well developed i.e. 6 months (1) compared to emergency separation which carries high mortalities 28% mortality vs 85% in elective settings (8), according to a single-center study conducted from 1974 to 2006 in the Philippines on a set of 22 conjoined twins (11 thoracopagus, 5 omphalopagus, 3 ischiopagus, 2 craniopagus, and 1 pygopagus twin) 6 twins were separated on an emergency basis and only 1 twin survived (13) which reached almost 80 percent mortality as reported by Spitz et al. (14), the successful separation and favourable outcomes in elective surgery owes to well-planned multidisciplinary approach, well-planned strategies and better physiologic parameter and optimization of twins for surgery keeping in mind the abysmal factors like intractable acidosis, sepsis, haemorrhage which are the leading cause of mortalities in conjoined twins (13,14).

CONCLUSION

Pygopagus separation may be a technically difficult case demanding excellence and sophistication with a minimum margin of error.

Especially, if the patient has no pre-op neurological deficit, but this is the first of its kind successful reported case in Pakistan owing to its success of well-coordinated multidisciplinary effort at the same time highlighting the lack of resources and basic infrastructure to deal with such complicated cases,

Paediatric neurosurgery should be given priority basis, and units should be established with a specialized workforce and equipped state of the art machinery.

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Ethical approval: All procedures performed in studies involving human participants were in accordance with the institutional and/or national research committee's ethical standards and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

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