The separation of pygopagus conjoined twins with fused spinal cords and imperforate anus

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Each set of conjoined twins has specific anatomic features dictating unique challenges to separation. Overcoming these challenges requires creative solutions that necessitate interdisciplinary collaboration. We present a unique case of pygopagus conjoined twins with fused spinal cords, imperforate anus without fistula, and a single anal sphincter complex. Separation included the use of novel applications of 3D printing and neurophysiologic monitoring. The 3D print helped to clarify the complex anatomy and facilitate communication during planning sessions. The neurophysiologic monitoring helped to distinguish a plane of separation for the spinal cords as well as the shared anal sphincter. Implementing these technologies and thus successfully separating these twins safely required a multidisciplinary team that extended beyond clinical specialties.

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Conjoined twinning is a rare occurrence that is typically associated with highly specific and complex connections. Separation of these patients is beneficial in order to provide the opportunity for normal and long term function. However, due to the complexity of the connections, separation is not always straight forward and requires novel solutions. We present a previously unreported case of pygopagus twins with fused spinal cords, imperforate anus without fistula, and single anal sphincter complex. We discuss novel uses of both new and well established technologies as well as the importance of collaboration.

1. Case report

On September 23, 2014 pygopagus conjoined twins were delivered by Caesarean section in Seroti Regional Referral Hospital, Uganda. With the exception of attachment at the buttocks, perineum, and lower back, they were observed to be healthy and were allowed to breastfeed. On day five of life, they were transferred to Mulago Regional Referral Hospital in Kampala, Uganda in septic shock. It was recognized that each infant had severe abdominal distention due to imperforate anus, and both underwent emergency colostomy placement, resulting in clinical stabilization. Our institution was contacted shortly afterward by an American advocate with ties to Uganda to consider separation. Evaluation in Uganda, including echocardiogram and abdominal ultrasound, revealed no other abnormalities. The twins arrived at our institution on December 28, 2014 and underwent MRI evaluation and examination under anesthesia (EUA) to determine the extent of connection. MRI revealed a shared thecal sac with fusion of the distal spinal cords and lipomyelomeningocele at the level of S2. A distinct plane of separation could not be defined radiographically. EUA revealed separate urethras, vaginas, and an imperforate anus with a single anal sphincter complex, an anatomic configuration not previously reported in the literature (Fig. 1). There was no evidence of neurologic deficit with each twin moving both of their legs well.
The extent to which each twin contributed innervation to the single anal sphincter complex was unknown.

In addition to the anatomic challenges of separation, there were logistical challenges related to accepting unfunded patients with complex medical conditions from abroad. Overcoming these challenges required a concerted multidisciplinary team effort organized by a team leader (GB). The team consisted of over 30 members from 13 disciplines, including General Pediatric Surgery, Plastic Surgery, Neurosurgery, Colorectal Surgery, Pediatric Anesthesiology, Neurology/Neurophysiologic Monitoring, Infectious Disease, Radiology, Pediatric Intensive Care, Physical Medicine & Rehabilitation, Nursing, and Mathematical Medicine. All participants agreed to donate their time to care for these children.

Soon after admission, the twins underwent placement of three tissue expanders by the plastic surgeons in preparation for later separation. One-on-one sessions were held between a radiologist and the clinical team members to explore details of the complex anatomy. CT scans were then used to create a full scale 3D printed model representing the spinal, urinary, and colorectal anatomy (Fig. 2). The model was used in pre-operative planning sessions to clearly define the anatomy. Two interdisciplinary planning sessions were held leading up to the separation. The planning sessions included all physicians and staff that were to be involved in the separation in order to ensure complete understanding of each individual’s role, and to define the sequence of events for separation. Because the spinal cords were fused, significant attention was placed on plans for intraoperative neurophysiologic monitoring of the lower extremities and of the shared anal sphincter. These sessions also included rehearsal in the operating room in order to optimize anesthesia set up, surgical instrument set up, and planning for prepping, draping, and transfers. Working with the mathematical medicine team, digital modeling was utilized to determine if there had been sufficient skin expansion for coverage after separation.

Separation surgery took place on September 3, 2015. After induction of general endotracheal anesthesia and placement of appropriate vascular access, the neurophysiology monitoring team placed electromyography (EMG) needle electrodes in the lower extremities of both twins and around the circumference of the anal sphincter complex. During the separation of the fused spinal cords and lipomyelomeningocele resection, the electrodes were used to monitor spinal nerve roots, guiding the neurosurgeons in identification of a cleavage plane. Neurophysiologic mapping of the anal sphincter complex revealed approximately one half belonged to each twin. The thecal sac of one infant could be closed primarily, whereas the other infant required duraplasty using acellular dermal matrix. After dural closure, the anal sphincter was divided along the cleavage plane mapped by EMG, followed by final division of the remaining soft tissues and the twins were separated.

One infant was transferred to a separate OR table in the same operating room, maintaining sterility. Wound closure included muscle flap coverage of the repaired thecal sacs and fasciocutaneous flap coverage. Anorectoplasty was performed by positioning the rectums appropriately and fashioning the levator muscles that were present into slings. The anuses were left closed deep to the anal sphincter complex to allow for optimal wound healing. The entire procedure required 16 h. Postoperatively, there was apparent preserved neurologic function in all lower extremities. The twin with the duraplasty had a CSF leak postoperatively, necessitating operative intervention. Anoplasties at the skin level were performed several weeks after surgery, followed by colostomy closure.

2. Discussion

Conjoined twins are rare, occurring in 1 in 50,000 to 1 in 100,000 live births [1]. Although there are common configurations of joining, each pair is unique, requiring a thorough evaluation to determine the extent of connection, which in turn determines the feasibility of separation. These pygopagus twins presented with a previously unreported configuration of spinal cord fusion, imperforate anus without fistula, and a single anal sphincter complex. To overcome the limitations associated with the use of two-dimensional MRI and CT data in representing complex...
three-dimensional anatomic relationships, we utilized 3D printing technology. The involved surgeons felt that the 3D print allowed them to quickly and clearly establish global anatomic understanding of the conjoined areas by demonstrating 3D visuospatial relationships and offering haptic feedback, which would not have been as easily performed with conventional imaging. Although the impact of this technology cannot be quantified in this single case, we have compared the 3D print produced here to a conventional CT scan and to a CT scan with digital reconstruction, in order to determine how 3D printed models facilitate surgeon understanding of complex anatomy. Preliminary findings indicate that the printed model helped surgeons to more accurately identify anatomy and to increase their understanding of scale and shape.

The use of neurophysiologic monitoring for the separation of spinal cord fusion in pygopagus twins has been minimally reported, and its utility has not been evident in the literature [2,3]. We found the neurophysiologic monitoring to be an integral component of the separation in that it allowed us to define the plane of separation between the spinal cords in order to avoid neurologic injury. The use of neurophysiologic monitoring to define the plane of separation for innervated structures such as the anal sphincter has not been reported and may have significant implications for functional outcome. Continence in these twins will not be certain until at least three years of age. However, urodynamic studies at this time show no evidence of abnormal innervation, suggesting normal function to these pelvic structures.

3. Conclusion

The management of conjoined twins presents substantial logistical, medical, and surgical challenges unique to each pair, which require creative and novel solutions. Although the success of such solutions is anecdotal due to the rarity of their application, it is clear that overcoming these challenges requires a level of planning, communication, and collaboration that cannot be achieved by any one physician or specialty [1,4,5]. It must also be remembered that collaboration for these surgical patients extends beyond clinical specialties, as in this case. By working with our mathematical medicine colleagues, we were able to apply 3D printing technology to clarify the anatomy, and to ensure that adequate wound coverage was achieved with the tissue expanders. Only by working collaboratively can optimal care and outcomes be delivered to complex patients such as these.

Disclosures

All persons gave their informed consent prior to their inclusion in this project. Additional informed consent was obtained from all individual participants for whom identifying information is included in this article. This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors. The authors declare that they have no conflict of interest.

References