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Pygopagus separation surgery: Role of 3D printing

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A B S T R A C T

Pygopagus twin separation surgeries can prove to be quite challenging especially when deciding the best surgical approach. Advancement in medical imaging allows proper planning and anticipation of difficult encounters during these surgeries. With the advent of 3D printing, surgeons are able to visually and physically appreciate the extent of fusion and soft tissue involvement. We present the case of the first successful pygopagus surgery in Kenya assisted by a 3D printed model which was invaluable during the preoperative and intraoperative period. Using the 3D print, we were able to anticipate a sacral bony spur and maneuver the fused sacrum without causing neurologic injuries.

1. Introduction

Conjoined twins are a product of incomplete embryonic separation of monozygotic monochorionic twins [1]. The phenomena is more commonly encountered in female patients who also have been reported to have better survival rates [2]. While rare in incidence, conjoined twins present a unique and challenging surgical experience even. This is especially because of the complex anatomical structures and connections in play. Scrupulous planning and preoperative preparation is key in not only successful separation of the twins but also in avoiding any long term neurologic deficits. With the advent of 3D printing in medicine, surgeons all over the world are turning to 3D printed models to appreciate visuospatial relationships that cannot be realized on regular imaging modalities. These models have been used successfully in multiple spinal and craniofacial surgeries and more recently in pygopagus twin separation surgeries [4,5]. These models are able to simulate and allow clinicians to anticipate complex anatomical movements and connections. We discuss the value of 3D prints in the first successful pygopagus twin separation surgery in our region. (see Figs. 1–7)

2. Case summary

On 5th September 2014, three month old, symmetrical pygopagus female twins presented to us at Kenyatta National Hospital, Kenya. They had been born via spontaneous vertex delivery at term, both had good Apgar scores. They were referred to our facility for further care after being noted to be conjoined at the sacrum. They had no previous antenatal imaging that predicted their conjoined nature.

Initial examination revealed healthy females twins conjoined at the sacrum with one anal orifice. They had separate external genitalia and were passing urine normally via 2 separate urethral orifices. Baseline laboratory tests were all within normal range. Initial MRI evaluation revealed a fusion at the conus medullaris with no significant nerve impingent. A multidisciplinary team was formed and a management plan formulated. A decision to postpone the surgery to allow maturity was made.

At the age of 1 year and 4 months, a second MRI was done which showed fusion at S3 with a common dural sac and the spinal cord joined at the filum terminale. A date for surgery was set and tissue expanders placed by the plastic surgery team to allow for adequate soft tissue cover after the separation procedure.

During the months prior to the surgery, CT scan was done and the DICOM images used to generate a stereolithography file using open source software. A final print of the anatomical structures involved was used in planning for the surgery and its' stages. We were able to preempt a bony spur later encountered in the surgery and appreciate multiple connections not clearly visible on the CT and MRI scans. This model was used in a dry run of the surgery a day before the procedure

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Fig. 1. Pygopagus twins after birth.

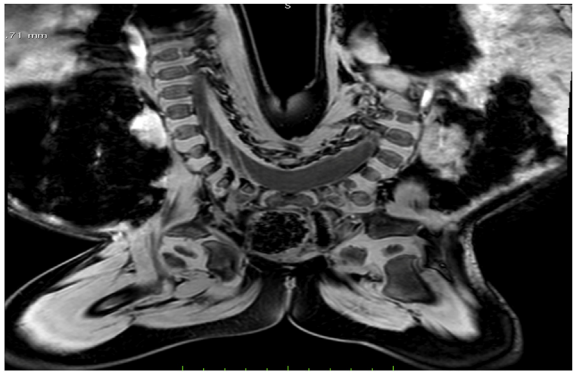

Fig. 2. Initial MRI showing fusion at conus.

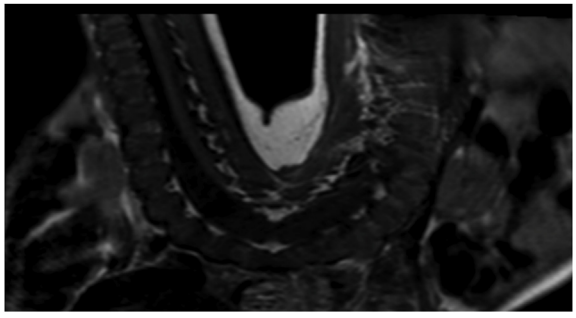

Fig. 3. Initial MRI with focus at conus.

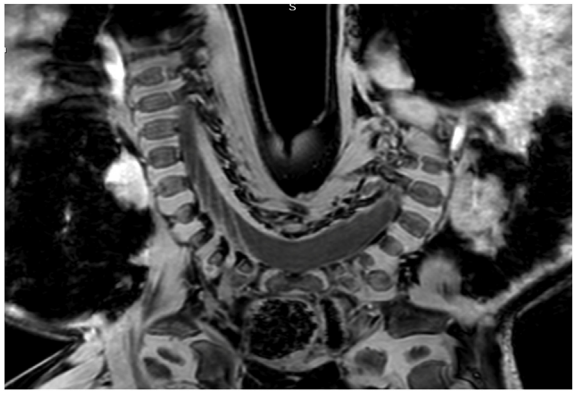

Fig. 4. Second MRI showing fusion at filum terminale.

Fig. 5. "STL 3D renders" generated prior to printing.

Fig. 6. Final 3D print.

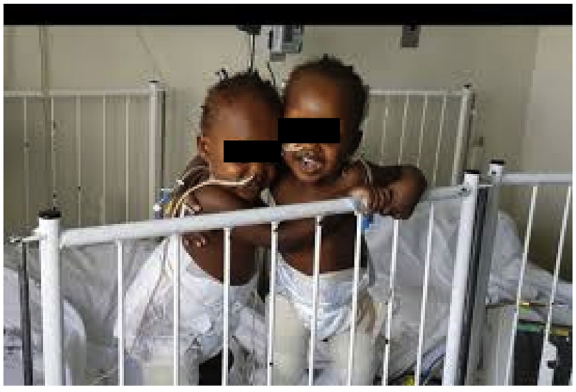

Fig. 7. One month post operative.

and further deployed to educate the mother and obtain informed consent prior to separation.

On November 1, 2016, a 50 member multidisciplinary team conducted the surgery on the then 2 year old girls. The procedure was 3 phased and took a total of 23 h. Induction of anesthesia took 3 h after which the surgery began. Aside from the anesthetic and pediatric team who monitored the twins throughout the procedure, a neurophysiology monitoring team used electromyography (EMG) needle electrodes placed in the lower limbs and anal sphincter complex to guide the

neurosurgeons and monitor nerve activity. The final 3D print was referred to multiple times during the procedure and in the check images after the procedure.

After separation and closure of the dural sacs, one twin was aseptically transferred to an adjoining theatre for wound closure. After successful reversal of anesthesia, both twins were wheeled to the intensive care unit where they remained under care for 2 weeks. Both twins developed post-operative cerebrospinal fluid leaks which were managed conservatively. The second twin developed some weakness on her left ankle which was managed with physiotherapy and she was able to regain normal function within 3 months.

The children have reported satisfactory developmental milestones while still under pediatric follow up and have since not reported any neurologic deficits.

3. Discussion

Conjoined twins are classified into 8 groups based on the conjoined parts; cephalopagus, omphalopagus, thoracopagus, ischiopagus, parapagus, pygopagus, craniopagus and rachipahgus [3]. About 18% of all conjoined twins (1 in 200,000 live births) are joined at the sacrum. Pygopagus twins have been shown to have good prognosis after separation surgeries and proper clinical management [1]. The incidence of conjoined twins in our set-up is unknown due to a paucity of data on the same. In keeping with this, few cases of separation surgeries are known. The separation of conjoined twins poses a financial, clinical and physical challenge to patients in low income countries. Poor referral systems and health insurance policies prevent most of these patients from accessing quality care. With the advent of 3D printing in medicine and its' successful use in pre-surgical planning, success in complex surgeries as the one presented above are expected.

The team involved in this surgery was able to adequately prepare for the complex surgery by visuospatial appreciation of the unique anatomical connections in the twins. Such minute connections which would have otherwise not been realized on the CT scans and MRI scans generated. They would have likely required more focused images during the planning period which would have financially strained the family of the patients. The patients' parents were able to fully comprehend the complexity of the procedure and any expected risks.

4. Conclusion

3D printing proves a viable avenue in planning and execution of conjoined twins' separation surgeries currently and in the future [4,5]. Resource limited set-ups are advised to adopt newer technologies in preparation or complex surgeries such as these. A multidisciplinary team is key in the success of such surgeries and adequate preparation time should be allowed to discuss all possibilities and possible techniques to be employed if there is no indication for emergency surgery.

Disclosures

Informed consent was obtained from the guardians of the twins and all other involved parties prior to their inclusion in the write-up. This case study did not receive any funding or grant support towards its publication. All authors attest that they meet the current ICMJE criteria for Authorship. The authors declare that they have no conflict of interest.

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