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ORIGINAL ARTICLE

Role of Neuromonitoring in the Spinal Separation of Pyopagus Twins

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Abstract
Introduction and Aim: Separating the spinal cord while separating spine sharing conjoined twins may have long-lasting implications for the survivors. We aim to study the role of neurophysiological monitoring in delineating the spinal anatomy as an important component to pre-operative planning and twin separation, thereby improving neurological outcome.
Case Report: Study involves two pairs of pyopagus conjoined twins with a shared spine. The twins were subjected to neurophysiological monitoring under general anesthesia to identify the neuronal supply and cross-innervation of various organs including lower limbs and anal sphincters with respective brain. Pre-operatively, the spinal cord was seen to be joined terminally with varying degree in both set of twins. Neurophysiological monitoring done preoperatively on the first set of twins revealed cross innervation from twin B to twin A, although twin A had its independent complete innervation. The sphincter was predominantly controlled by twin A. In the second set of twins, there was no cross innervation, both twins had equal control of sphincter hence the sphincter allocation was done using other general and anatomical factors. Neurophysiological monitoring was also utilized intraoperatively to divide the cord in its functional midline instead of anatomical midline thereby preserving neural outflow and function. Post-operatively, both set of twins retained motor and sensory supply to lower limbs and mobilize both limbs well, aided with physiotherapy.
Conclusion: Better functional outcomes are possible with the use of neurophysiological monitoring during the workup and management of spine-sharing conjoined twins.

Keywords: Neurophysiological monitoring, pyopagus, conjoined, twins

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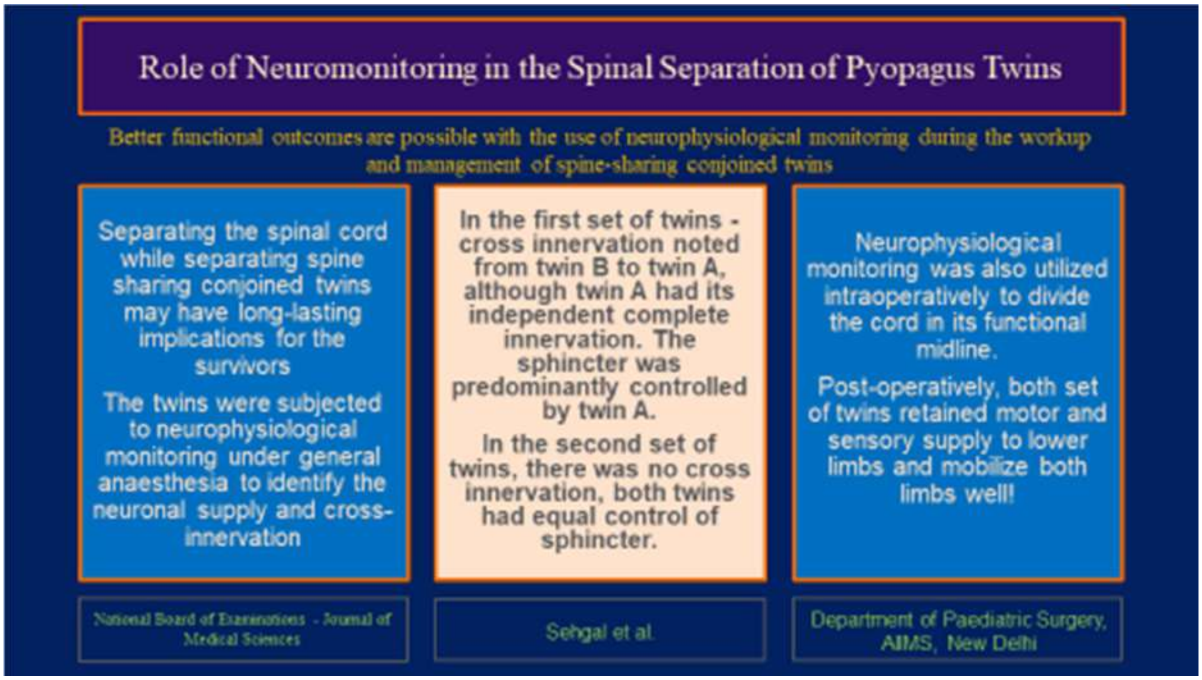
Graphical Abstract



Introduction

Insults at different stages of embryogenesis may lead to varying types of twinning. Caudal duplication syndrome: more evidence for theory of caudal twinning [1,2,3]. Pyopagus twins, a rare variant of conjoined twins, are notable for their union at the sacrum and perineum. They may have spinal and spinal cord abnormalities with variable fusion of their spinal cord [4]. The shared organs can generally be separated and reconstructed, which leads to a good outcome in these cases. The challenge in separation of these twins lies in the separation of the shared spines, which have to be divided in a way to preserve the neural innervation to each twin.

Intraoperative neurophysiological monitoring (IONM) assesses the functions of the neural tissue during the procedure, to limit neurological insult while handling tissues. Although routinely used in neurosurgical cases, only limited case reports have utilized this technique in the separation of spinal separation in conjoined twins. Motor evoked potentials are generated on transcranial stimulation at different levels in the brain and are transported by the pyramidal tract. They monitor the motor pathway and can be recorded at the spinal cord level or the muscle [5]. Electromyography monitors the integrity of the nerves during surgery. Electrical potential is produced on depolarization of a motor nerve which is then monitored using subdermal or intramuscular electrodes placed in the affected muscle [6].

Intraoperative neurophysiological monitoring has occasionally been used in the separation of these twins, to improve functional outcomes and prevent neurological deficits [7]. We present our experience of utilization of neurophysiological monitoring for the separation of two pairs of pyopagus conjoined twins.

Case History

Two pairs of antenatally detected conjoined twin girls were presented to our hospital. They were joined from the lower back, facing partially away from each other, sharing their lower back and had a common perineum. They were noted to have one common anal opening and 2 separate urethra and vagina openings. A detailed evaluation was necessary to understand the anomalous intraspinal anatomy Spina bifida occulta: radiographic and operative correlation [8] were carried out for both the pairs of the twins. Both pairs underwent computed tomography (CT), magnetic resonance imaging (MRI) and gastrointestinal studies for better understanding of their anatomy. To avoid confusion, the first set of twins were referred to as Twin A and B and second set as Twins 1 and 2, respectively.

In both the pairs of twins the spinal cords were said to be fused as per the images. In the first pair, CT scan revealed spina bifida of L3 to L5 vertebra and fusion of the sacral vertebrae (S2 - S5) with common coccygeal vertebrae. Magnetic resonance imaging of lower spine revealed two separate conus medullaris with a common fused filum terminale. The Twin B was also found to have a right old MCA infarct. Both twins had moved their lower limbs, with a power of 4/5 in both limbs of Twin A and power of 3/5 in limbs of twin B as per the Medical Research Council, UK, Manual Muscle Testing scale. However, both had limb wasting, along with neuroorthopaedic deformities, noted to be more in twin B.

In the second pair of twins, multiple lumbar vertebral defects in both twins were noted on the CT imaging with fused lower sacral (S3-S5) vertebra, 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 increased in size over a year to reach the conus. As a result there was a a neural tissue of length 1.3 mm separating the fused conus with terminal syrinx and dysplastic neural tissue. Hyperintensity was seen in the bilateral deep white matter in the occipital lobe of Twin B due to suspected metabolic insult. Both twins had moved their lower limbs, with a power of 4/5 in all limbs, as per the Medical Research Council, UK, Manual Muscle Testing scale. However, these twins also had limb wasting, along with neuroorthopaedic deformities, noted to be more in twin 1.

There was also a dilemma of the control of the common anal sphincter, which had to be given to either one of the twins during separation or divided in a way that each twin retained control of the sphincter to prevent future fecal incontinence. Hence, a multidisciplinary team was formed with the inclusion of neurophysiologists to aid in the separation.

Technique of Neurophysiological Monitoring

The first pair of twins then underwent pre separation neurophysiological monitoring using the NIM-ECLIPSE® NS System (Medtronics, USA) under general anaesthesia, avoiding muscle relaxants during the stimulation Time. Sterile, paired subdermal stainless steel needle electrodes were placed using sterile technique in the limb muscles and around the EAS after the patients were prepped. Muscle motor evoked potentials (m-MEP) recordings were attempted using fast charge transcranial stimulation at a frequency of 275Hz and voltage of 100-1000V.

On transcranial stimulation of twin A followed by twin B motor cortex and recording of m-MEPs from lower body muscles (Bilateral Rectus Femoris, Tibialis Anterior, Extensor Halluces Longus, Abductor Halluces) of both twins, we found that there was no cross over of nerve fibers from twin A to twin B in lumbar and upper sacral region (S1) and twin A had her independent neural supply, but there was a cross over of nerve fibres (L4, L5, S1) from twin B to twin A (Fig. 1). Muscles in the right lower limb of twin B did not elicit m-MEPs even at high voltages and seem to have poorly developed motor supply. On transcranial stimulation of twin A followed by twin B motor cortex and recording of m-MEPs from, bilateral abductor halluces (AH) of both the twins and anal sphincters (AS) (S1,S2,S3,S4), we found cross-over of sacral fibres from twin B to twin A. Twin B's motor cortex seems to have adequate motor control of right side of only the anal sphincter and its right AH has poor motor supply and no m-MEPs were recorded from it despite the high degree of stimulation. Twin A however, had a good control of the entire anal sphincter (Fig. 2). In view of these findings, we decided that the anal sphincter would be given to twin A to improve her fecal continence. Intra-operative nerve root mapping was also done to identify and separate the nerve fibres during the spinal separation. Triggered electromyography (EMG) was utilised for nerve root mapping, using a single pulse of duration 500 µs and frequency of 1 Hz. Biphasic stimulation was performed with bipolar stimulator probe and cathodal stimulation with monopolar stimulator probe.

Following principles of laminectomy & intra-spinal dissection, we opened the dura, & identified a closely apposed U-shaped spinal cord configuration [9], with nerves fanning out inferiorly. Nerve roots from L4-S3 were identified on the basis of responses obtained from Abductor hallucis, Extensor Hallucis Longus, Tibialis anterior, Rectus femoris and Anal sphincter (Fig. 3). Few nerve roots were identified which evoked triggered EMGs from both twins (twin B showed greater amplitude than twin A). In such cases, neural fibres going to twin A were divided, as twin A has its own intact nerve innervation for all muscles apart from this accessory cross innervation from twin B. All the sacral nerve roots that triggered EMG response from anal sphincter were spared to ensure intact anal sphincteric function. This led us to carefully divide the cord in its functional midline than the instinctive anatomical midline.

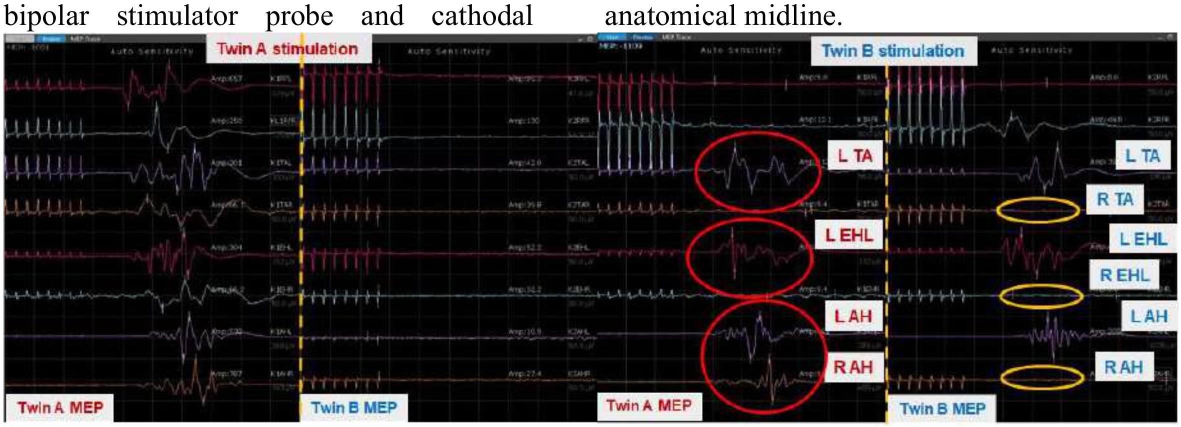


Figure 1. Findings of transcranial stimulation of twin A followed by twin B motor cortex showing no cross-over of nerve fibers from twin A to twin B and a cross-over of nerve fibres (L4, L5, S1) from twin B to twin A

Figure 2. Findings of transcranial stimulation of twin A followed by twin B motor cortex showing complete control of anal sphincter by twin A and partial control by twin B.2.

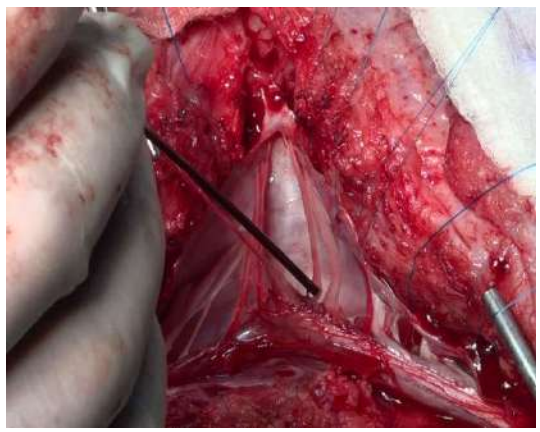


Figure 3. Direct stimulation of nerve roots using co-axial electrode.

We also evaluated the m-MEP, before and after the separation surgery. Transcranial stimulation was performed on twin A followed by twin B's motor cortex and recording of MEPs from muscles (Bilateral Brachioradialis, Adductor Longus, Rectus Femoris, Tibialis Anterior, Extensor Hallucis Longus, Abductor Hallucis, Anal Sphincter) of both twins. On first stimulating twin A, m-MEPs were obtained at 500 V, and no responses were recorded from twin B. These responses remained intact after sacral bone separation. The m-MEPs were obtained at 400 V from twin B on stimulation, with reduced response from right side lower limb muscles, not showing any response even at 1000 V, except for left Abductor Hallucis. Cross responses were recorded from twin A in lower limb muscles. After sacral bone separation, responses from twin B remained intact but the cross responses from twin A disappeared. The result from this study also helped us to prognosticate the parents with regard to the future outcome and also has a potential medicolegal implication.

In the second pair of twins, we proceeded directly to the separation surgery. Prior to the procedure, after induction, the children were subjected to neurophysiological monitoring to identify inter twin neural communication and control of the anal sphincter. We found both twins to have an equal control of the sphincter and no neuronal cross over. Hence we proceeded to divide the spinal cord in the functional midline as identified with monopolar and bipolar triggered EMG electrodes. The anal sphincter was given to Twin 1 as she was neurologically better. Post-separation, the m-MEPs were noted to be intact in both twins.

Outcomes

Both pair of twins were moving their limbs in the immediate post-operative period. Once the wound related issues subsided, both pairs were mobilised. In the first set of twins, both stand with support and Twin A walks with support as well. Among the second set of twins, Twin 1 is able to walk without support for short distances and Twin 2 walks with support. All received extensive physiotherapy in the post-operative period and with the help of special shoes and walkers, they were discharged home in a mobile condition.

Among the first set of twins, twin B required a stoma as she did not receive the sphincter. Twin A has adequate bowel control with requirement of bowel management for constipation, but no soiling. Among the second set of twins, both are on bowel management. Twin 2 has had severe constipation and occasional soiling at follow up.

Discussion

The goal of spinal separation in pyopagus twins is to have two neurological intact children. With the complex anatomy and anatomical anomalies often identified in these twins, it is imperative to use technology which helps us attain this goal. The advantage of neuromonitoring during the intra-operative period of spinal separation allows the separation with minimal neurological damage. Since we are conclusively able to identify the source of the neural tissue encountered, we can thereby divide it in the correct cleavage plane.

The role of intraoperative neuromonitoring has been debated in case reports of pyopagus twin separation. According to some authors, if there is no neurological difference between the twins, IONM would have little role in changing their management as there is no option other than sectioning the cord as equally as possible [5,6,10,11]. Others also omit IONM in cases where either twin is not expected to survive or is not stable enough to withstand a prolonged general anesthesia time [4].

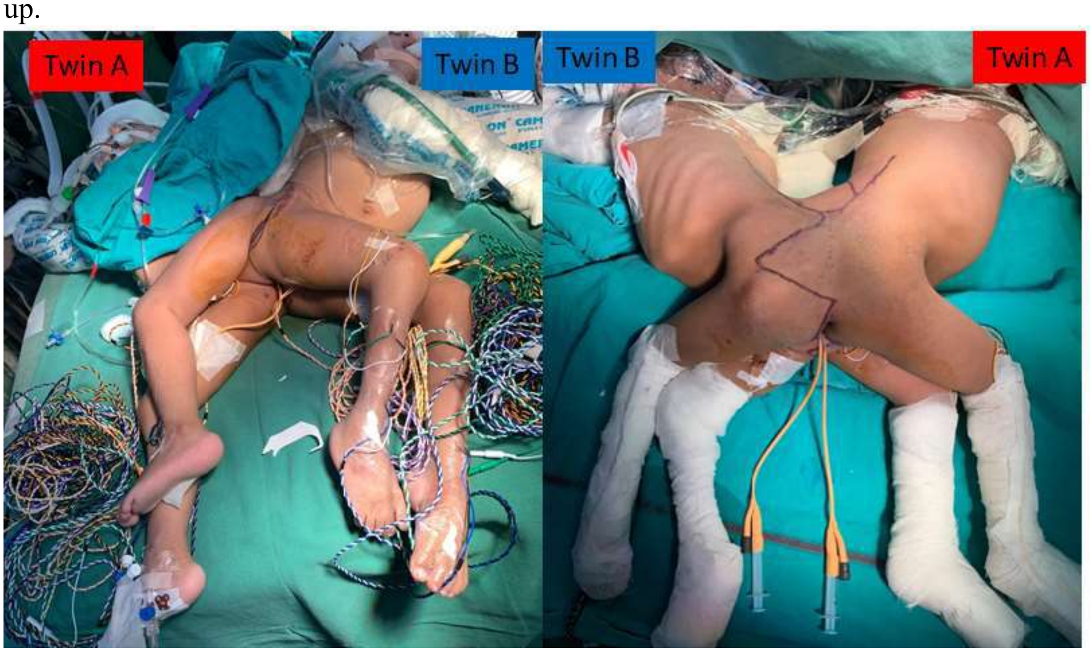


Figure 4. Placement and arrangement of needle electrodes in the lower limb muscles of twins 1 and 2

However, with this approach, some minor neurological deficits have also been reported by Fieggen et al. [7]. Utilization of this technique however, has led to improved outcomes as reported by some authors, especially in cases with a fused U shaped spinal cord, where sectioning the cord at the apex is challenging as the cleavage plane may not be in the anatomical midline. Fieggen et al. have also reported patient specific evidence in separating the anal sphincter complex utilizing IONM [7].

In our study, although we could have done without IONM for spine separation in the second set of twins, in the first set, given the anatomy and challenges in separating a fused spinal cord in way that precludes neural deficits in either twin, led us to utilize this technique. However, IONM is not replaceable in situations where the sphincter control has to be decided among two neurological intact twins. The additional information obtained in the first set of twins that the entire sphincter was controlled by Twin A helped us plan the final anal sphincter allocation to her and prepare Twin B for a stoma.

Limitations of this technique are that it can be used only in places where the equipment is available and there is availability of a neurophysiologist to interpret the results. The placement of electrodes and adequately preparing the twins in OT is however a time-consuming task. It took us an additional 30 mins to prepare the set-up and place the subcutaneous electrodes in the children, fix them with sterile adhesive dressings and wrap them in a way it does not entangle during the process of the separation (Fig. 4). Close communication between teams is also essential as this procedure must be performed without a muscle relaxant, hence if the technique is planned to be used, it must be communicated to the anesthetist to provide only a short acting muscle relaxant during induction [11].

Conclusion

IONM is a valuable tool which should be utilized while separating spine sharing conjoined twins. Apart from aiding in diving the spinal cord in the functional midline, it also helps to allocate the anal sphincter to the more appropriate twin. With the help of a multidisciplinary approach, successful separation and good neurological outcomes are attainable, utilizing technology to our advantage.

Conflicts of interest

The authors declares that they do not have conflict of interest.

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