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Surgical planning and separation of ischiopagus conjoined twins using 3D printed models and intraoperative neurophysiological monitoring

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ABSTRACT

The birth of conjoined twins is one of the rarest neonatal conditions worldwide, with an incidence of 1 in 100,000 live births. Twins joined by the pelvis, known as ischiopagus, represent 6–11% of all conjoined twins. This clinical condition increases the degree of complexity in separation surgeries. Each case poses unique anatomical challenges, and clinical outcomes may vary according to the health facility where the infants are born or can access. This case report describes a successful separation surgery of ischiopagus conjoined twins in Peru. 3D printing and neurophysiological monitoring were used for surgical planning and intervention, allowing a full-scale anatomical model and preserving the mobility of the lower limbs of conjoined twins after surgery. This article demonstrates that established procedures and novel technologies can be valuable platforms for preoperative planning and intraoperative guidance to safeguard the central and peripheral nervous systems while increasing survival rates of ischiopagus conjoined twins in middle-income countries such as Peru.

1. Introduction

The incidence of conjoined twins worldwide is 1 in 100,000 live births, making this a rare condition, and the probability of survival of newborns with this condition is only 65% [1]. This incidence varies greatly depending on the geographical region. This condition is less common in Europe than in India [2], while in Latin America, the prevalence of this condition is 1 per 74,506 births [3]. Conjoined twins commonly present numerous anomalies in tissues and organs, such as ischiopagus conjoined twins, infants that share

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a pelvic region along with the gastrointestinal tract and genital region. Ischiopagus represents approximately 6–11% of all conjoined twins [4,5].

Separation surgery is the only treatment to improve patients' quality of life, but these clinical interventions bring significant challenges and may have potential complications [6]. The prognosis will depend on how the twins are united, which tissues or organs share, and the knowledge and experience of the surgery team. When conjoined twins are expected, the family and physicians must carefully analyze, discuss, and prepare infants for surgical intervention. Due to the challenges of separation, meticulous preoperative planning is usually required. In the literature, different planning and surgery techniques and technologies have been shared that can help to increase the survival rate of ischiopagus twins, such as 3D printing (3DP). This digital manufacturing technology allows physicians to create anatomical models of patients that will be treated. The models are physical representations where physicians can visualize the different complexities of conjoined twins bodies [7].

Neurophysiology is a relatively young science, and its application in the operating room is even more recent. One of these applications is known as intraoperative neurophysiological monitoring (IONM). This technique is used to monitoring the function of neurological structures that may be at risk during surgery time, avoiding possible lesions. Due to its capabilities for recording data in real time, IONM allow physicians not only to monitor but to document their procedures for future discussion to improve surgeries techniques and make them safer and better [8]. This approach aims to provide the surgeon with proper orientation and minimize the risk of injury, delivering optimal patient care [9].

2. Case report

2.1. Clinical information

Father and mother aged less than 25 years, pregnant for the first time with more than six prenatal visits, was diagnosed with a pregnancy of ischiopagus conjoined twins. The patient had a urinary infection in the second semester of pregnancy, which was treated with ceftriaxone; she also received two doses of betamethasone for fetal lung maturation. The father did not have a relevant medical history. Both parents are based in Lima, Peru, have incomplete basic education, and live in extreme poverty.

In 2018, ischiopagus conjoined twins with opposite cephalocaudal axes were born by emergency cesarean section, dystocic delivery due to placental insufficiency and ischiopagus conjoined fetuses of the tetrapus type, with a gestational age of 34 weeks, 1540 g of weight, 41.5 cm of length between cephalic extremities, 29 cm of the perimeter at the junction, APGAR 7 at the minute and 9 at 5 minutes, it was necessary to use positive pressure ventilation. On physical examination, there is difficulty in the passage of the orogastric tube, limited to the upper third of the esophagus. There is evidence of an umbilical cord and shared lower abdomen, single ambiguous genitalia, an outline of a single opening that eliminates urine and feces, phallic rudiment with a meatal opening in an impermeable blind bag. There is evidence of a single shared anorectal canal. Neurological examination without relevant findings, upper limbs with preserved mobility, decreased mobility of lower limbs due to union between twins.

On tomography, it is observed that the liver and kidney of density and morphology are preserved, separated pelvic arches, not fused, with very marked pubic diastasis by the communication between conjoined twins. A double bladder also evidences it with a regular urinary ureter outlet, cryptorchidism with pelvic level testicles, and a single shared anorectal canal. Twin A is diagnosed with type III esophageal atresia, complex anorectal malformation, and complex genitourinary malformation. Twin B is also diagnosed with complex anorectal malformation and complex genitourinary malformation. It is important to note that a week after birth, a thoracotomy was performed to close the tracheoesophageal fistula in Twin A.

2.2. Surgical procedure

The surgical intervention for separating conjoined twins was carried out on Sept 25, 2019 (at nine months of age) and was directed by a multidisciplinary team. The operation for the separation of ischiopagus conjoined twins was a highly complex surgery that required the participation of a multidisciplinary team. The objective of the surgery was to achieve structural or anatomical separation of the different apparatuses that kept the patients united. To achieve this objective, specialists in plastic surgery, cardiovascular and pediatric surgery, traumatology, urology, orthopedics, neurophysiology, and neurosurgery were required.

The pediatric surgeons identified and repaired the common anal canal and individualized it for each patient, opened the abdominal cavity to access the internal organs, identified and separated the thin intestinal loops of each individual, identified, and separated the common large intestine, and performed a Y-shaped intestinal suture with a cutting linear mechanical suture, opened an ostomy by counter opening. The urologists identified and separated the urethras of each patient and performed a ureteroneocystostomy; also, a vesicostomy or bladder sizing was performed. As for plastic surgery, a fasciocutaneous flap was designed to cover the exposed areas after separation. The plastic surgeons participated in the reconstruction of the pelvis after separation, and the fasciocutaneous flap was used to cover the exposed areas. The traumatologists performed osteotomy and placed external fixators to stabilize the bones and soft tissues.

The cardiovascular surgeons focused on identifying the common arterial or venous vascular elements to ensure adequate blood flow to the members once separated. The pediatric neurosurgeons identified the common neurological elements between the patients and repaired the nerves if necessary to ensure that each separated member maintained adequate function and sensitivity. As for neurophysiology, sensors for neuromonitoring were placed in sterile conditions in the patients to monitor the electrical activity of the nerves and muscles during surgery. The electrical activity of the nerves and muscles was monitored continuously during surgery to ensure that the nerves and muscles remained functional during separation.

2.3. 3D anatomical model

The conjoined twins underwent computerized axial tomography (CT) evaluation for anatomical clarification in infants. For this case, CT scans were used to create a 3D representation and a physical model of pediatric patients. The surgical team decided to include solid objects for preoperative planning, allowing the team members to understand anatomical relationships better and have adequate tactile interaction with infants' anatomies. Experts from the Digital Fabrication facility of Universidad Peruana Cayetano Heredia were contacted to fabricate the anatomical model. 3D printing, an additive manufacturing technique, was selected for this task.

Creating a 3DP anatomical model requires following a technical path that starts with image acquisition, segmentation, pre-printing preparation, and printing. The medical requirement was to access the bony pelvis of the infants and visualize the relationships of these complex structures. After data acquisition, CT scans in the DICOM format of conjoined twins were imported into 3D Slicer (v4.10), an open-source platform for analyzing and visualizing biomedical images [10]. This software was selected for post-processing segmentation. Segmentation was performed using the *Threshold* tool in the *Segment Editor* section. An appropriate range in Hounsfield units (HU) must be chosen for bone segmentation. For this case, a range between 281 and 1623 HU was defined (Fig. 1). Then, a range between −86 and 350 HU was selected to segment the complement tissues. Both segmentations were exported in STL format for processing and merging 3D models. Blender (v2.9) is open-source software for 3D modeling and animations [11]. This software allowed the research team to visualize 3D model details. A demo of the visualization is in Supplementary Material 1 (video). The STL files were imported into Blender to modify errors or other artifacts unrelated to this procedure. 3D models were aligned and smoothed to achieve the final model (Fig. 2a). To fabricate the bony pelvis model, these structures of interest should be selected (Fig. 2b). Then, the 3D models have exported again in STL format for 3D printing (Fig. 2c).

Supplementary video related to this article can be found at https://doi.org/10.1016/j.epsc.2023.102604

An STL file contains data about geometry, texture, and other attributes of the 3D object. A slicing program must process this file to create horizontal slices of the model and transform them into instructions (G-code) as coordinates, orientation, and speed for the 3D printer. Repetier Host was the slicing program selected, another open-source software used by the scientific community. Current additive manufacturing techniques use multiple pieces of equipment according to medical requirements and spatial dimensions of the model. Most used consumer devices are low-throughput 3D printers. They are based on the Fused Filament Fabrication (FFF) process, have a working space of 200x200 × 200 mm, and use thermoplastics. FFF printers use a thermoplastic filament which is melted and deposited layer by layer to compose the desired physical object. For this fabrication, a 3D printer MD-6C was used, a FFF machine manufactured by Mingda that was sold for less than $2000. This 3D printer has a printing size of 300x200 × 500 mm and uses up to 8 thermoplastics.

The larger workspace allowed fabricating the whole conjoined twin's anatomical model using a white polylactic acid (PLA) filament (Fig. 3a). The figure shows that the model's orientation helped to visualize the region of interest. Usually, the external support material is trimmed manually. The supports of this model were not removed. Instead, they were used to hold bone structures in a fixed position to represent the conjoined twins' morphology. The printer parameters were a layer height of 0.2 mm, a fill of 25%, and

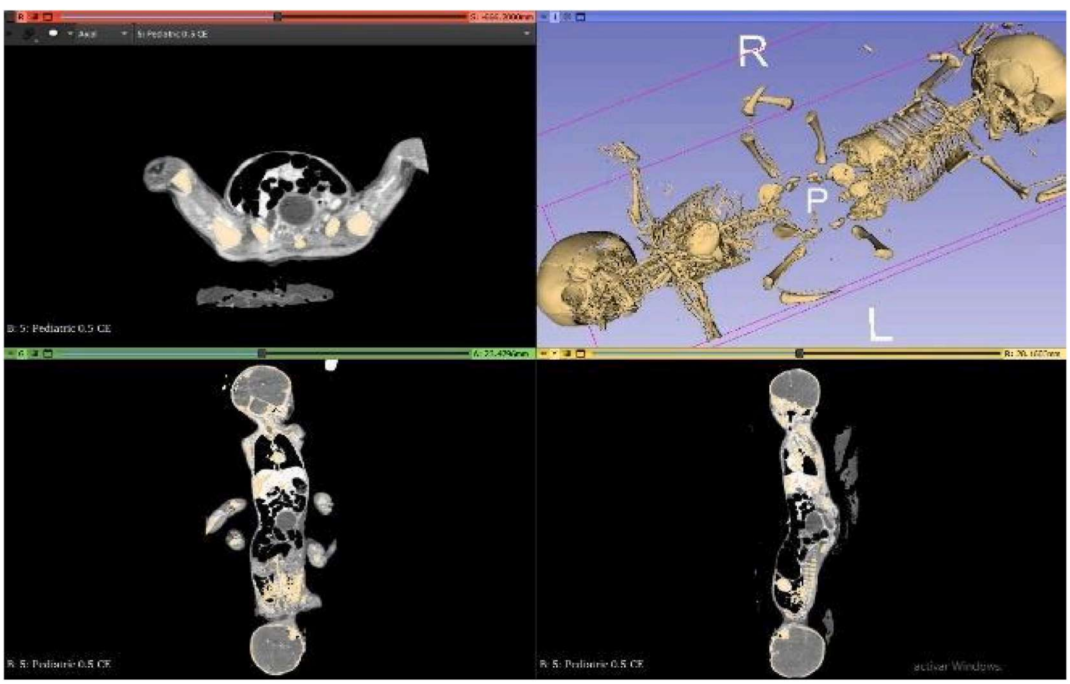

Fig. 1. Bone segmentation using 3D Slicer.

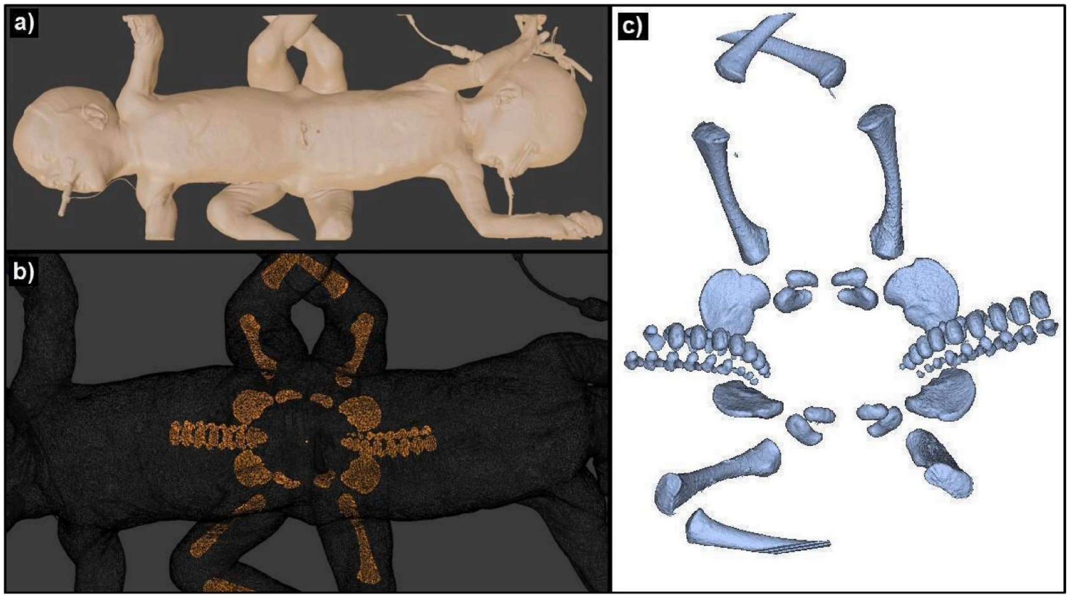

Fig. 2. Pelvis segmentation using CT scans for 3D printing. a) 3D reconstruction of patients' bodies. b) Identification of the bony pelvis of the conjoined twins. c) Segmentation of the tissue of interest and generation of the 3D model to be printed.

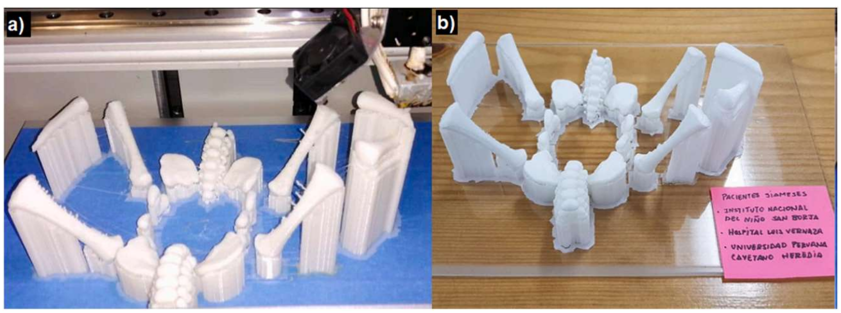

Fig. 3. 3D printed anatomical model. a) 3D printing process of the model b) 3D printed model for medical use.

a hexagonal pattern. Almost 4 hours and 10 mt of filament were needed for 3D printing. Then, the finished 3D model was glued to an acrylic piece for use by the surgical team (Fig. 3b).

2.4. Role of neurophysiological monitoring and 3D printing

The 3D-printed real-scale model was used in preoperative planning sessions, and these practices were carried out in the operating room. However, despite its usefulness in the planning phase, the model was ultimately not employed during the actual surgery. The urogenital and digestive systems (urinary bladder, ureters, urethra, small intestine, and large intestine) were separated into different surgical turns. Surgery started at 8:00 a.m. After 17 hours, the infants were separated and transferred to an intensive care unit for recovery. The first part involved making an incision in the pelvic areas to separate the infants. The specialist in physical medicine and rehabilitation, who was involved in the segmentation and pre-print preparation of the model, assisted surgeons in specific procedures, such identification of testicles for dissection. In the second part, the surgeons focused on individual reconstruction. The pelvic floor was reconstructed by external fixation to correct the pelvic bone and the conjoined twins' anatomical position of the lower limbs. Finally, the skin closure was performed by plastic surgeons in pelvic areas.

During separation, one of the main concerns was that the incisions would affect the mobility of the lower limbs, which is why the physician specialized in physical medicine and rehabilitation uses IONM to guide surgeons in preventing neurological sequelae. The IONM protocol consisted first of a preliminary evaluation. Subdermal needle electrodes were placed in specific muscles of the lower limbs (psoas muscle, anterior tibial muscle, and abductor hallucis muscle) of both twins to record and monitor motor evoked potentials (MEPs), somatosensory evoked potentials (SSEPs) and free-running electromyography (f-EMG) bilaterally. The recorded impedances were below 5kΩ and four trains of four (TOF) without neuromuscular blocking effect. The baseline recording showed that the SSEPs were asymmetrical, had low replicability, and had low amplitude bilaterally (Fig. 4). The MEPs were not appropriately recorded at the start of the monitoring. They were replicable in amplitude and latency until the end of the intervention (Fig. 5). f-EMG did not show spontaneous activity, suggesting transient radicular irritation (Fig. 6). No fluctuations in the MEPs nor neurogenic shock

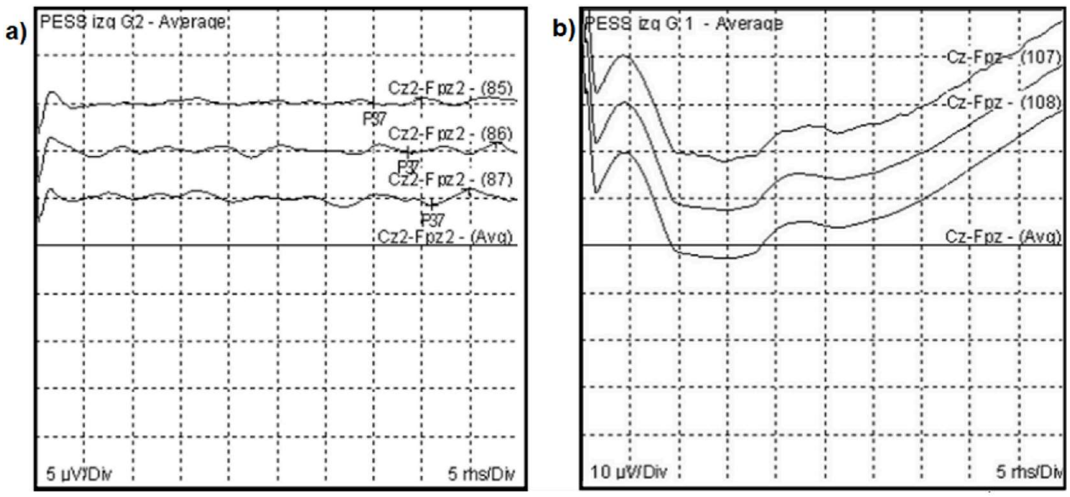

Fig. 4. Somatosensory evoked potentials a) Twin A b) Twin B.

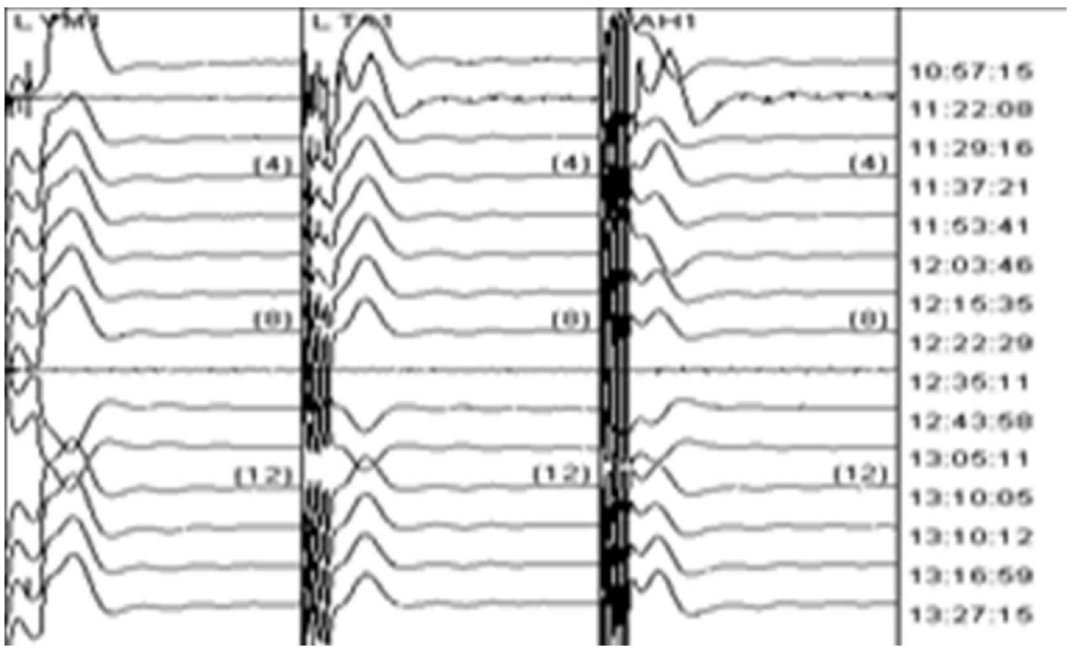

Fig. 5. Motor evoked potentials.

were identified during the surgical shifts. The SSEPs were also monitored by stimulating the posterior tibial nerve with a control on the right upper limb. No anal sphincter complex mapping was required. IONM showed a preserved neurological function in the lower limbs of conjoined twins.

2.5. Surgical interventions after the separation of the conjoined twins

One week after the operation (October 2019), Twin B presented dehiscence of the surgical wound at the perineal level to the abdominal wall, fibrin in the muscular plane, devitalized muscle wall edges, as well as protrusion of the intestinal loop through the surgical wound, in addition to dehiscence of the urethral plane cutaneous. Therefore, the correction of the dehiscence of the surgical wound was made. First, the intestinal loops were reduced to the abdominal cavity, then the edges were cleaned and revived, and the bladder anchor points were placed on the abdominal wall. Second, the modeling of the urethral edges and anastomosis to the cutaneous plane was carried out. A Penrose drain was placed through the perineal wound and below the urethra. Polypropylene mesh was placed, fixing the muscular plane and mesh vertices on the bone surface of the ischium and pubic branches. After seven months of mesh placement, it was removed together with the external fixators.

Two weeks after the operation (October 2019), Twin A presented necrotic tissue periurethral, dehiscence of the perineal flap with serous secretion, and devitalized tissue. Therefore, the removal of staples and suture points from the previous surgery, surgical clean-

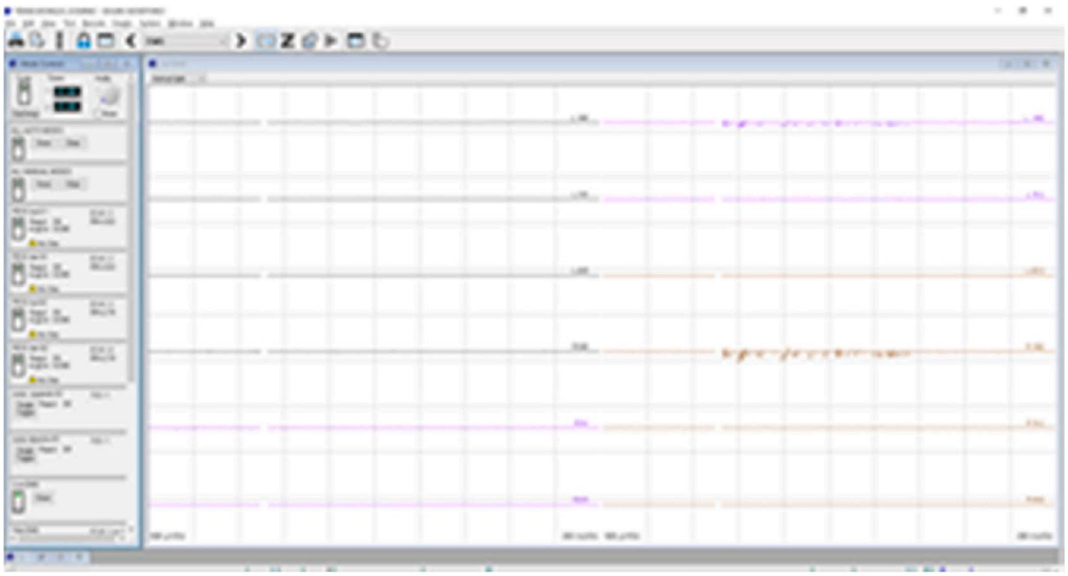

Fig. 6. Free-functioning electromyography of infants.

ing of the devitalized tissue of the perineal flap containing the hypospadic urethra, facing of flaps by planes, and fixation of periurethral flaps to adjacent fasciocutaneous flaps, placement of a laminar drain in the perineal region were made. Also, cleaning and healing of the external fixator entry points were done; at the end, they were covered with paraffin gauze and dry gauze.

Five weeks after the separation operation (October 2019), Twin An underwent esophagoplasty by thoracic approach to repair the tracheoesophageal fistula by opening the upper esophageal stump and dehiscence of the tracheoesophageal fistula and non-tense anastomosis of stumps with a ratio of 1.2/1.

3. Discussion

Surgical management of conjoined twins is still complex and requires extensive preoperative planning and organization. Each case poses unique anatomical challenges, and clinical outcomes may vary according to the health facility where the infants are born or can access. Case reports describe a survival rate of 50% when the operation is performed in the neonatal period and 90% when it is done after four months of age in all types of conjoined twins [12]. The survival rate is increasing, being ischiopagus variants with the highest rate after the separation [13]. The intervention reported in this article was performed at nine months of birth, which may contribute to survival after the separation of conjoined twins.

Medical imaging has played a relevant role in the early diagnosis and separation of conjoined twins. It can help to optimize the results for patients in invasive surgeries, such as separation surgery, providing a better understanding of the scenario for decision-making and medical procedures. The modalities will depend on whether the results are sufficient to answer the anatomical questions of the surgical team. CT exams were helpful for this case report because pelvic bone anatomy was required for preoperative planning. 3D reconstruction and simulations using CT images allowed physicians to get a reference for twins anatomies but could not offer relative scales, distances, and relationships of anatomical structures. In contrast, physical models like 3D printed models can be used by specialists to understand better spatial relationships through visualization and interaction. Different organs and tissues can be represented and included for pre-surgical planning, as well as improving communication with colleagues and parents for educational purposes. Children's hospitals are increasingly implementing new techniques and digital manufacturing technologies to operate safely, even in developing countries such as Peru.

After three years of the separation surgery, both twins are still alive. None of the infants has been undergoing rehabilitation. Twin A has been diagnosed with a severe psychomotor developmental delay. Twin B has achieved better motor skills, including walking independently without support, climbing stairs, and running, and more advanced language development with a more extensive vocabulary than his brother. On the other hand, Twin A manages to be bipedal. He can walk with a limp but is not able to run or climb stairs. Both patients still present urinary infections. Twin A is a bladder-size carrier and colostomy, and Twin B is a colostomy carrier. Beyond the challenge of surgical separation, complex and persistent urological problems in surviving ischiopagus twins are usually reported [14,15], such as the case report presented in this article.

4. Conclusions

Complex surgical interventions require a multidisciplinary team and mixed techniques to achieve successful outcomes in conjoined twins. This case report demonstrates that neurophysiological monitoring and 3D printing can be valuable platforms for preoperative planning and intraoperative guidance to safeguard the central and peripheral nervous systems. The surgical team was able to preserve the mobility of the lower limbs of the infants while separating the urogenital and digestive systems. Different perspectives of the anatomical structures and their surroundings were available with the 3D printed model. Ischiopagus conjoined twins can be separated in middle-income countries such as Peru using established procedures and novel technologies.

Patient consent

The parents of the patients gave their informed consent for this study. This report was reviewed by the institutional research ethics committee of the INSN San Borja. This case study did not receive any funding or grants for its publication. The authors declare they have no competing financial interests or personal relationships that can influence the study.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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