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Surgical Correction of Ischiopagus Tripus Conjoined Twins with Fused Pelvis: Enhancing Quality of Life through Orthopedic Intervention

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Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Patient: Male, 3-year-old
Final Diagnosis: Ischiopagus tripus conjoined twins after surgical correction
Symptoms: Fused body in pelvis reion
Clinical Procedure: Amputation • osteotomy
Specialty: Orthopedics and Traumatology

Objective: Congenital defects/diseases

Background: The rarity of ischiopagus tripus conjoined twins complicates the surgical separation, owing to the lack of cases and high complexity. We aim to report our experience in performing orthopedic correction for ischiopagus tripus twins.

Case Report: A pair of 3-year-old conjoined boys presented with a fused body at the pelvis region and only 1 umbilicus. There were 2 legs separated by shared genitalia and an anus at the midline, and 1 fused leg, which could be felt and moved by both of the patients. The twins also shared internal organs of the bladder, intestine, and rectum, as visualized through angiography computerized tomography scan. After several team discussions with the institutional review board, the hospital ethics committee, and both parents, it was agreed to perform disarticulation of the fused third limb, followed by correction of the trunk alignment by pelvic closed wedge osteotomy and internal fixation. We successfully reconstructed the pelvis using locking plates and additional 3.5-mm cortical screws and 1.2-mm stainless steel wire.

Conclusions: This report describes the presentation and surgical management of a case of ischiopagus tripus conjoined twins. It highlights the challenges involved in surgery and the importance of investigating these infants for other congenital abnormalities. Although surgical approaches for different sets of twins should be individually tailored, interventions aimed to provide optimal outcomes should consider ethical issues and parental/patient expectations. Even in situations in which the twins are inseparable, there is still room for surgical correction to be performed.

Keywords: Twins, Conjoined • Orthopedic Procedures • Pediatrics

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Introduction

Conjoined twins are typically rare to be encountered within the medical field, with incidence of 1 pair of twins in every 50 000 to 200 000 births [1]. The rarity of such cases can be rooted in the stillbirths or infant deaths that occur in as many as two-thirds of all conjoined twin cases. There seems to be a higher proportion within the female population, with ratio of 3: 1 [2]. Based on the anatomical fusion site, conjoined twins can be categorized into thoracopagus (chest), omphalopagus (abdomen), thoraco-omphalopagus (chest and abdomen), pygopagus (sacrum), ischiopagus (pelvis), craniopagus (cranium), parapagus (one side), cephalopagus (head), and rachipagus (spine) [1,3]. The ischiopagus type is an uncommon variety of conjoined twins, with joint site at the pelvis, and accounts for only 6% to 11% of all conjoined twins [4]. Ischiopagus twins can be further categorized according to the number of legs present: tetrapus (4 legs), tripus (3 legs), or bipus (2 legs) [5]. Expectant mothers are able to find out the possibility of their fetuses being conjoined early during pregnancy with the help of ultrasound and better imaging, such as magnetic resonance imaging [1].

The surgical separation of ischiopagus conjoined twins requires a multidisciplinary approach consisting of pediatric surgeons, urologists, orthopedic surgeons, plastic surgeons, and antesthesiologists. Eades and Thomas reported the first separation of ischiopagus tetrapus conjoined twins back in 1966 [6]. Almost 5 decades later, there have been only a handful of case reports discussing ischiopagus tripus conjoined twins globally, and even more limited reports regarding surgery from orthopedic perspectives [7,8]. Separating conjoined twins surgically is still a very challenging task even for the experienced surgeon. Ethical clearance and parental decision are other obstacles that can hinder the procedure [1]. We aim to report our approach in performing corrective surgery for ischiopagus tripus conjoined twins, without separation, from orthopedic perspectives. Much like past reports of the rare case, our case reports twins that were joined at the pelvic region but, interestingly, with the presence of an extra non-functional lower limb in between the twins. Further details of the twins will be described in the presented illustrations.

Case Report

A pair of 3-year-old conjoined twin boys were presented at Hasan Sadikin Hospital in Bandung, Indonesia. Both patients were born with 2 normal-looking and functioning upper limbs. The bodies of both patients were fused in the pelvis region with only 1 umbilicus, while from their thorax level and above, they were completely separated. The patients shared 1 penis and 1 anus, where both patients defecated through the same orifice. There were 2 legs separated by the shared genitalia and anus, and 1 fused leg, which could be felt and moved by both patients (**Figure 1**). The position of their trunks was horizontal to each other's, making it impossible for them to sit or stand. No other associated congenital deformity or functional

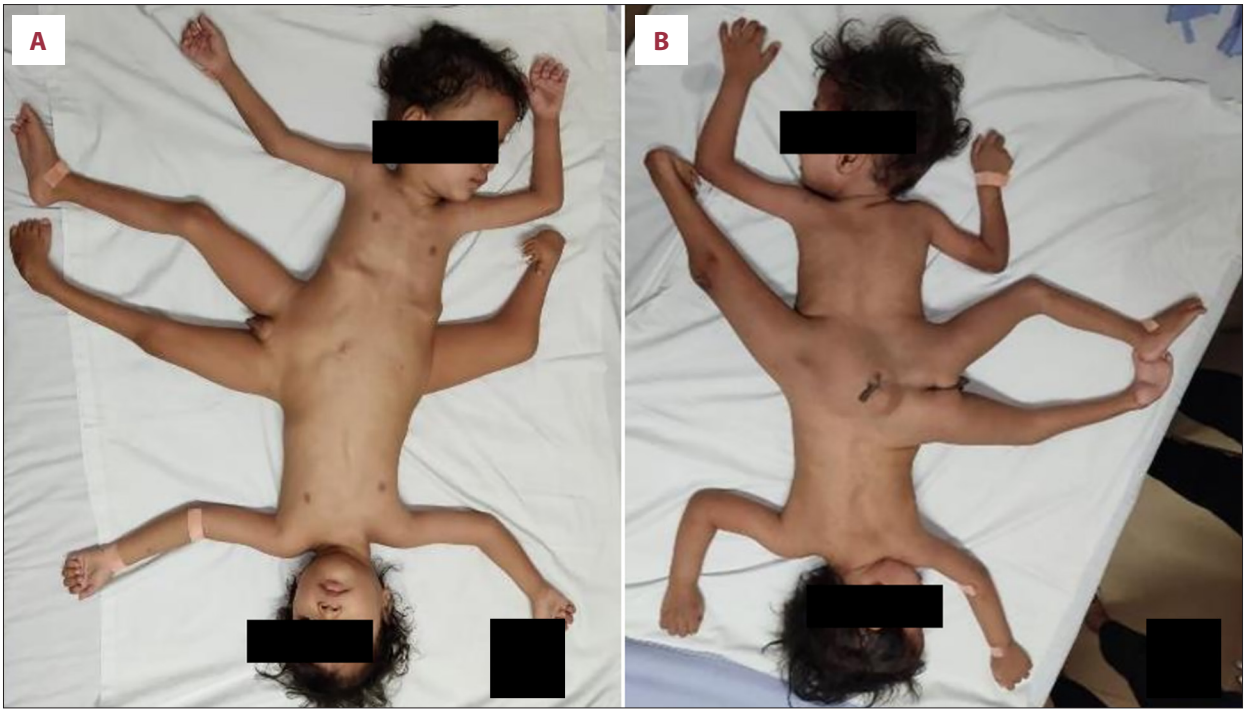

Figure 1. Clinical presentation of 3-year-old ischiopagus tripus conjoined twins. (**A**) anterior view; (**B**) posterior view.

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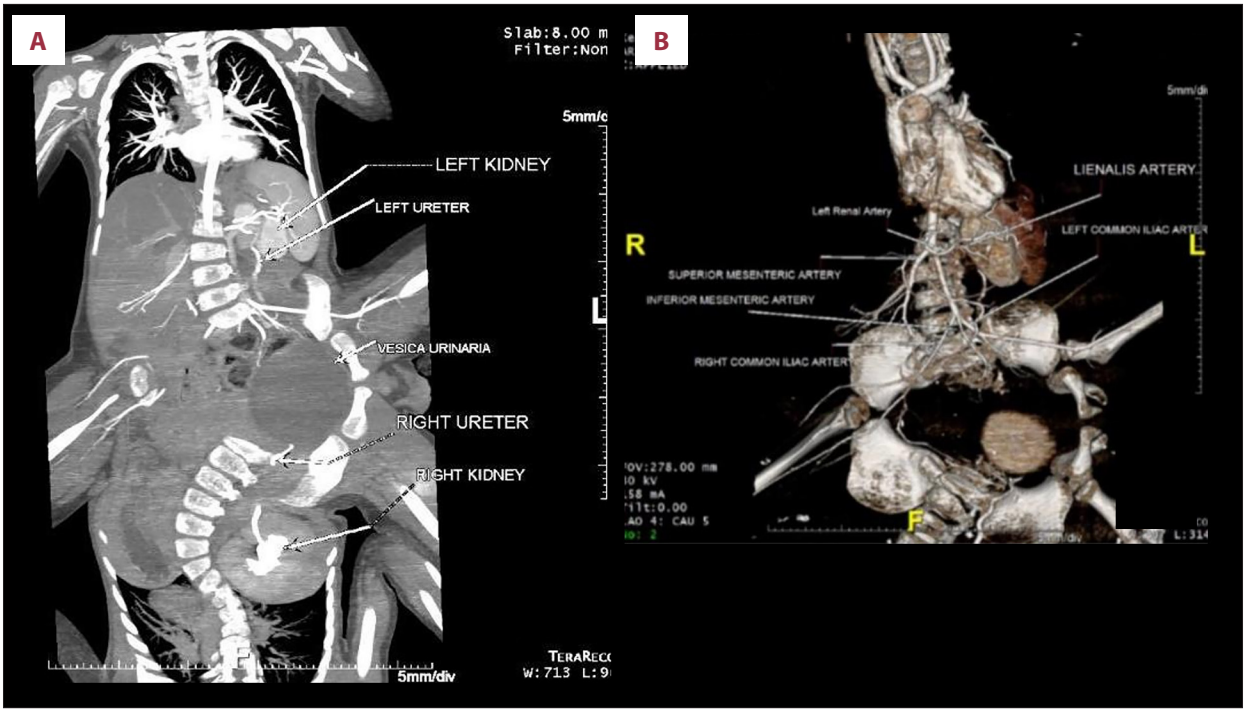

Figure 2. Angiography computed tomography scan of the patient. (**A**) Left kidney hypoplasia in twin I, and left kidney agenesis in conjoined twin II. (**B**) Left renal artery appears smaller than the right renal artery.

defects were found. When supported by their upper limbs, the twins could sit upright.

The patients were the third and fourth children of a couple with no family history of congenital anomalies from the previous children and other family members. No significant events or illnesses occurred during the perinatal period. The mother reported consuming no supplements, medicines, or traditional herbal concoctions and routinely checked her pregnancy with a local midwife. The twins were delivered at term by a midwife, with no postnatal complications. After birth, the parents rarely brought the twins for hospital check-ups since birth, due to their residential area being in a very remote area of West Java. Distance became the primary obstacle for the parents to access advanced health services. The patients never had any surgical procedures performed. As they grew older, mobility became a major issue for their already-restricted activities. The parents then visited our hospital to seek further medical advice.

Angiography computerized tomography (CT) scan revealed ischiopagus conjoined twins, with 3 lower extremities, 1 bladder, intestines sharing 1 rectum, and suggestion of left kidney hypoplasia in conjoined twin I, and left kidney agenesis in conjoined twin II (**Figure 2A, 2B**).

The patients were planned for disarticulation of the fused third limb and pelvic wedge osteotomy, along with open reduction and internal fixation to straighten both trunks vertically. Prior to surgery, we informed the parents thoroughly regarding the possible risks and the planned approach, which the parents then agreed to. This case report adhered to the 2020 Surgical CAre REport (SCARE) guideline, and we provided the filled checklist.

Surgical Procedure

The surgery was done by 3 orthopedic surgeons, who prepared and performed the reported corrective surgery. Osteopower osteotomy, curved locking reconstruction plate, 3.5-mm screw, Kirschner wire (K-wire), stainless steel wire, small chisel, and pointed clamps were prepared prior to incision.

The patients were in the supine position, and an incision mark was planned for a longitudinal and modified racquet incision (**Figure 3**). After dissecting the overlying tissue, the femoral vessel and nerve were set aside and the incision was further extended. After identifying the joint capsule, we performed disarticulation of the fused third limbs (**Figure 4A-4F**). We continued the procedure with pelvic wedge osteotomy at the level of the triradiate cartilage. After performing closed wedge reduction and making the trunk more vertical to the lower limb, we then fixated using a curved locking reconstruction plate and stainless steel wire (**Figure 5**). The intraoperative time and bleeding volume were not beyond expected, and no postoperative complications occurred (**Figure 6**).

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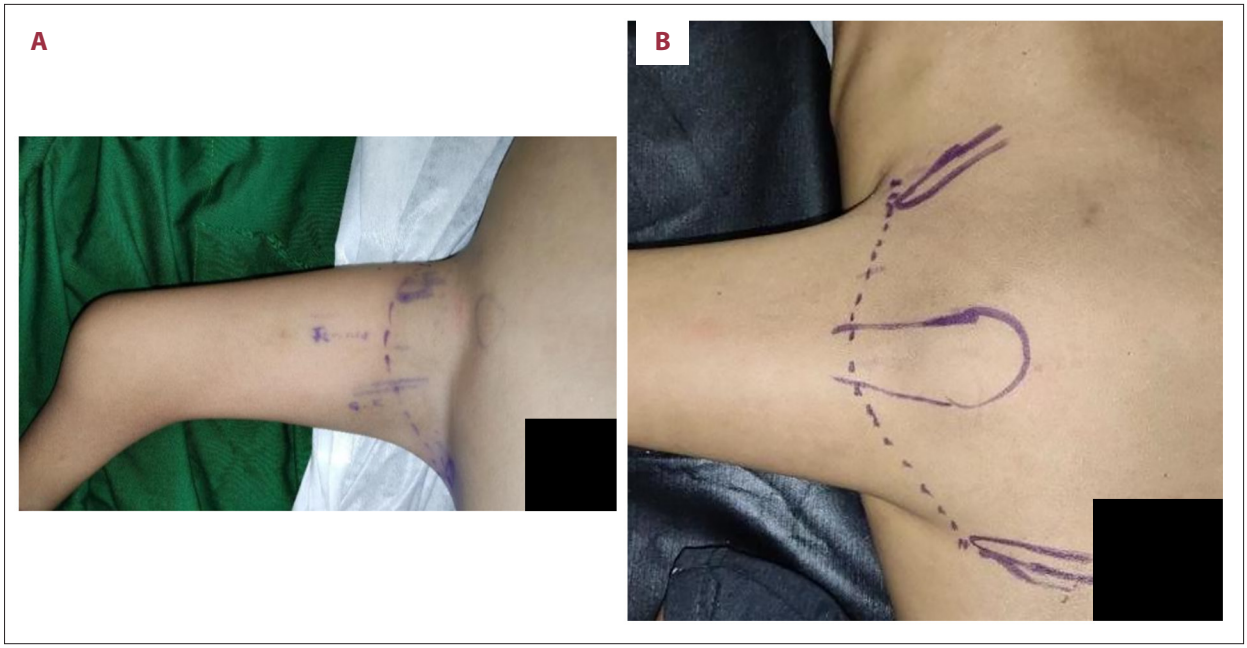

Figure 3. Incision design marking on (**A**) anterior and (**B**) posterior segment.

We documented the patients' plain radiograph and condition before discharge (**Figure 7**). Note that the twins' spinal alignment was not 180° straight to each other. The latest follow-up was 3 months after surgery, and no complications were reported. Improved mobility was observed, as both patients were able to flex their upper trunk (**Figure 8**). The treatment timeline is summarized in **Figure 9**.

Discussion

This case report describes the surgical corrective procedure for inseparable ischiopagus tripus conjoined twins from orthopedic perspectives. A pair of 3-year-old ischiopagus tripus conjoined twins underwent disarticulation of the fused third limbs, pelvic wedge osteotomy, open reduction, and internal fixation with a curved locking reconstruction plate.

Ischiopagus conjoined twins are fused at the pelvis and further divided based on the number of legs owned and shared. Around half of ischiopagus conjoined twins are tetrapus types, followed by the tripus type contributing to 30% of cases. Anatomically, ischiopagus conjoined twins usually have their heads opposite to each other. The common presentation of ischiopagus conjoined twin is that one almost always has a smaller-sized head or has a more severe clinical defect than the other, as described in previous literature [8]. The exact etiology of conjoined twins is still unknown, but it was hypothesized that both genetic and environmental factors equally and, possibly, interactively contribute to the incidence. Several risk factors found to be related with conjoined twins are alcohol consumption, teratogenic drug use, lack of folic acid during pregnancy, exposure to radiation or chemicals, and family history. There are 2 acceptable theories regarding the possible pathophysiology of conjoined twins. The first is fission theory, which is based on an incomplete division during embryo development. The second is fusion theory, which states that there is a secondary fusion of monozygotic twins [9].

The diagnostic examinations that aim for thorough preoperative evaluation involve plain radiography, ultrasonography, contrast imaging of the gastrointestinal and urinary systems, endoscopy, echocardiography, CT angiography, and magnetic resonance imaging [7]. Three-dimensional (3D) imaging can be helpful in preoperative planning since it can provide anatomical details and vascularization of the fused organs [3].

Surgical separation of conjoined twins is a complicated procedure and should be individualized based on the anatomy of the patients. A thorough and careful consideration for the children's development and safety is crucial. Other co-existing birth anomalies, such as kidney hypoplasia or kidney agenesis like in our patients, made them ineligible as candidates for any lengthy procedure under anesthetics. A similar case reported by Chen et al (1994) described a successful separation of ischiopagus tripus twins. The surgery was only complicated postoperatively, costing one of the twins' lower extremities, but apart from this, the twins survived until 3 years later. Further back, Hung et al (1986) reported their experience in separating twins that were joined at the pelvis. We noticed similar problems faced by these authors, namely urinary tract complications. To solve these complications, follow-up surgeries were required, which

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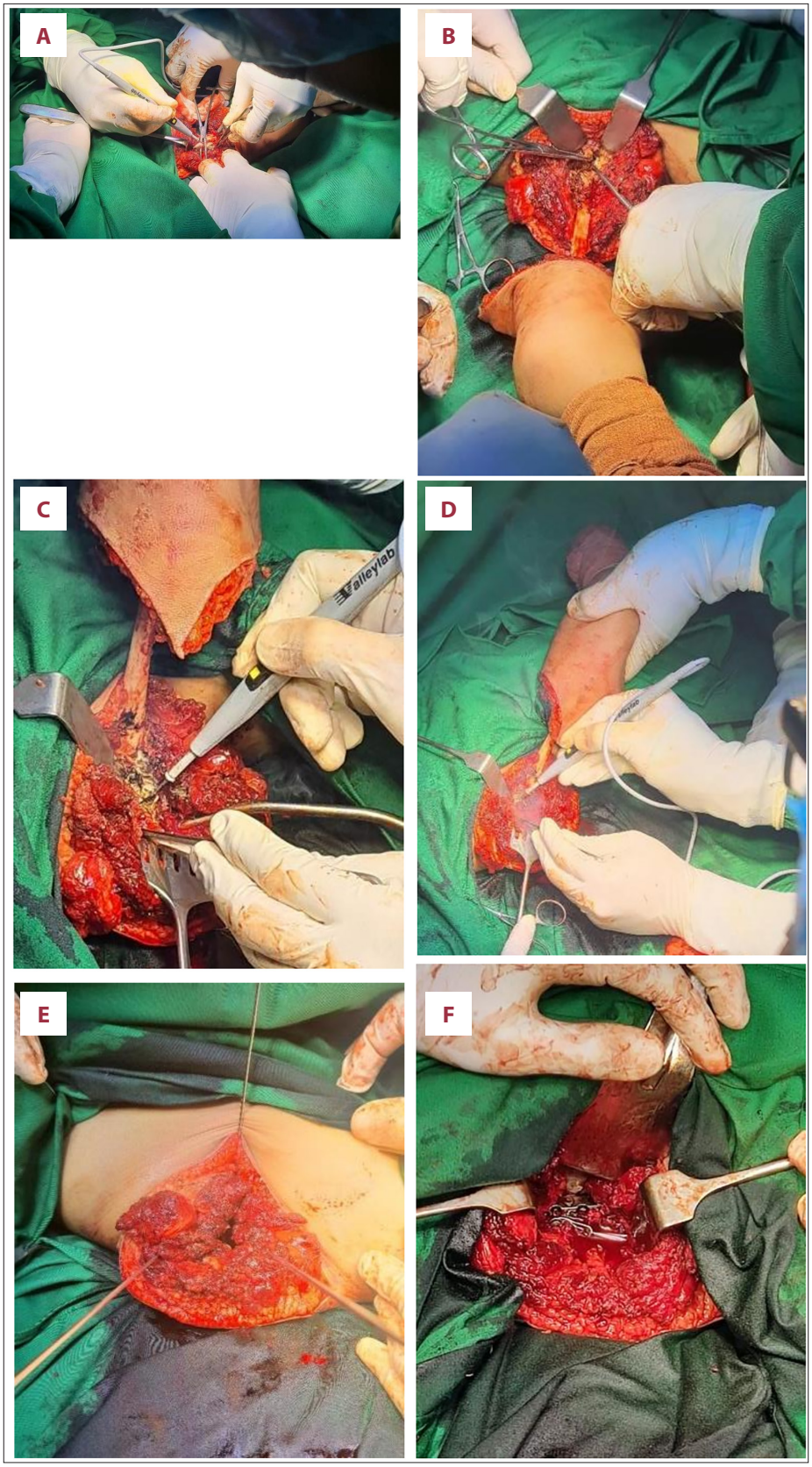

Figure 4. (**A-F**) Intra-operative views of non-functioning third limb disarticulation.

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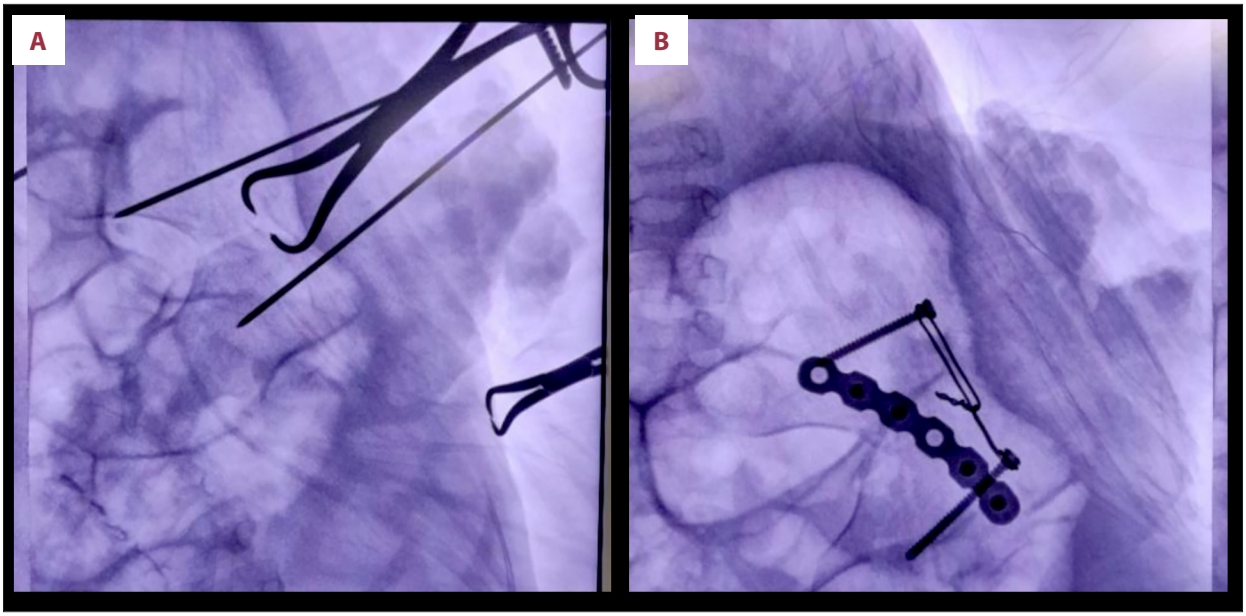

Figure 5. (**A**) Temporary K-wire marking and (**B**) internal fixation using curve locking reconstruction plate.

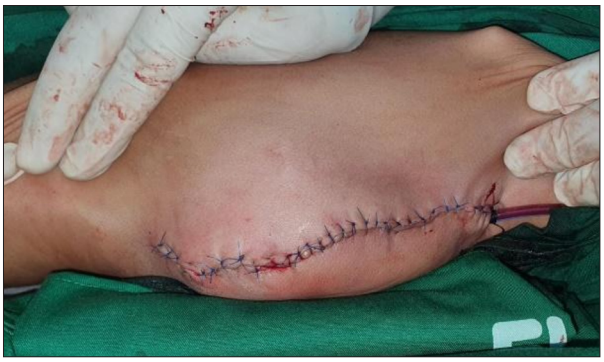

Figure 6. Soft tissue closure.

were successful and allowed the patients to return home. The timing of performing corrective surgery should also be considered, as the neonatal period would be unsuitable [10]. The surgery in our patients was performed at the age of 3 years due to the COVID-19 pandemic, which delayed the surgery.

The surgical team discussed several treatment plans with the Medical and Health Research Ethics Committee and the parents until the final decision for the surgical option [11]. We encountered difficulties in deciding which twin would keep the leg and genitalia that they shared. Another difficulty encountered was the approach to the twins' trunks positions, which were horizontal to each other. The final decision was to not separate the twins but instead to make the trunk more vertical so the twins could stand and even circumambulate with walking aid, as in the other ischiopagus twin case. The main challenge of the surgery is reconstructing the pelvic bones. Accordingly, a preoperative 3D reconstruction model and angiography CT can be very helpful to help visualize and plan for the surgery.

There are several techniques in performing pelvic wedge osteotomy, categorized into 3 types: re-directional osteotomies (Salter triple, periacetabular osteotomy, spherical osteotomy), reshaping osteotomies (Pemberton, Dega), and salvage procedures [12]. The fixation used in this patient was a curved locking reconstruction plate and K-wire, which have been shown to provide successful stabilization of fractured 3D-shaped bones, such as the pelvis.

There are few studies regarding the long-term prognosis of ischiopagus tripus conjoined twins after surgery. Unfortunately, these twins tend to have a shorter lifespan than non-conjoined twins, due to internal complications. Chatterjee et al reported a pair of ischiopagus tetrapus twins in India, who still lived normally until 22 years after separation surgery [13]. In contrast, Hoyle et al reported mortality of one twin at the age of 2 years, while the other survived for 23 years. In Indonesia, Rochadi et al reported a successful ischiopagus tetrapus conjoined twin separation in 2008, and the baby lived normally in the first 2 years of follow-up [14].

Rode et al reported that ethical considerations, which must consider the best option for the twins and their parents, are ever so important in the decision-making process. An ethical guideline for conjoined twin separation developed by Great Ormond Street Hospital recommends custodial care to be offered and allowing the natural course of the anomalies to take their course. Twin separation should be attempted when one twin has a high chance of not surviving due to lethal anomalies or when one has failed to survive.

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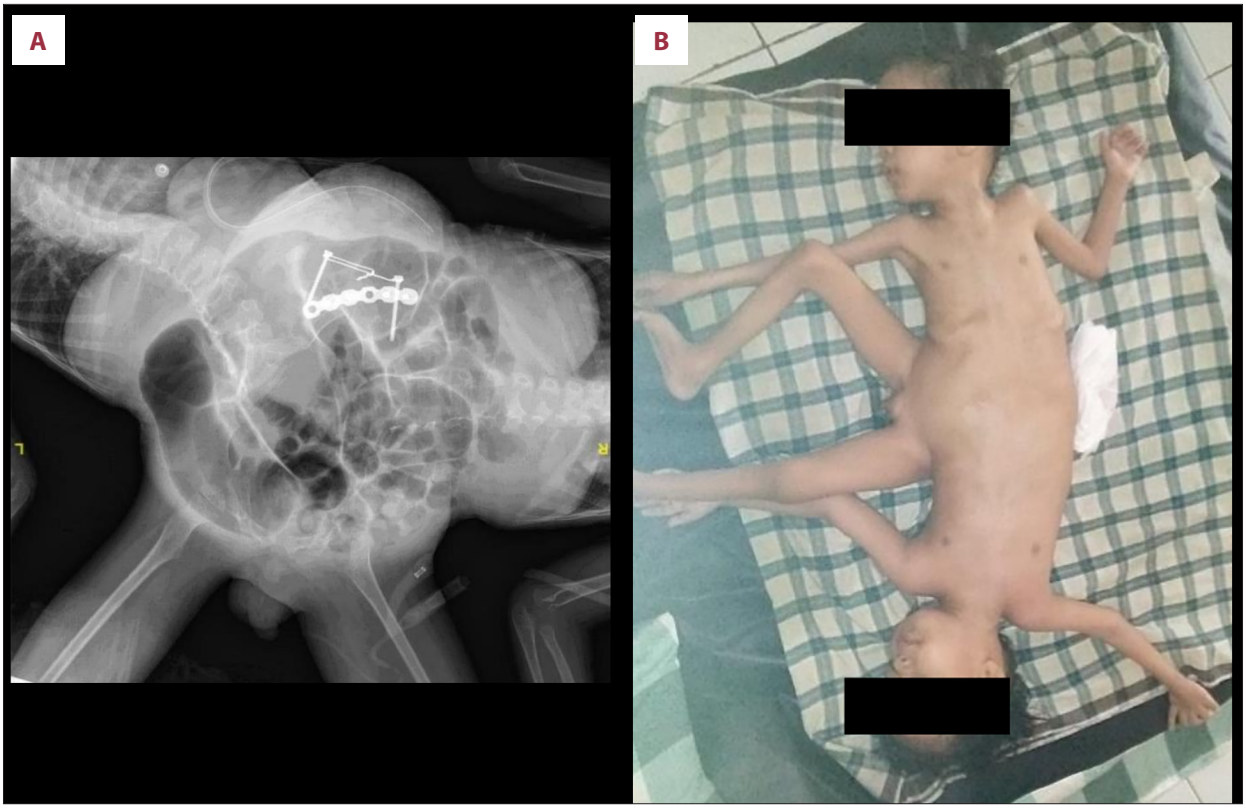

Figure 7. Postoperative (**A**) X-ray and (**B**) clinical condition.

Figure 8. (**A, B**) Follow up at 3 months after surgery.

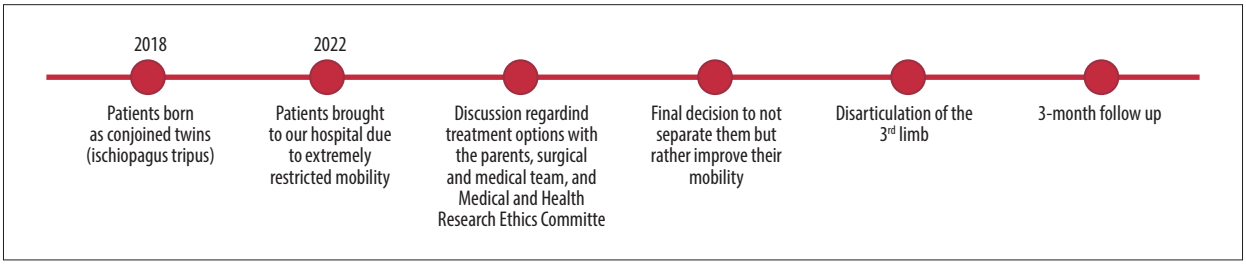

Figure 9. Patients' sequence of events.

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In twins who are not candidates for separation surgery, ambulation will not be an easy task. El Said explained that the twins may learn how to walk in their own way, which can occur later than expected. As a result, the process of improving their skills will be hindered. It is important to note that, although inseparable, these twins are still two different individuals with their own minds and personalities. One twin might dominate the other, pushing the other to follow and submit, which can affect the submissive twins' personality [15].

Physical therapy and rehabilitation are essential to promote strength, endurance, gait, range of motion, gross motor development, positioning, feeding, communication, and developmental skills. Orthotics and specialized devices, such as walking aids or adaptive chairs, can provide additional stability for the twins to perform fine mobility. In addition, extra counseling for caregivers should be considered and they should be well-educated regarding their child's condition [15,16]. In our case, there was a need for special walking aid after the surgical correction, considering that these conjoined twins were 3 years old and were expected to be more mobile. They were also provided with continuous physical therapy and rehabilitation for their physical activity. The parents were very satisfied with the surgical outcome, especially in improving the twins' mobilization.

There are few other reports of successful separation of ischiopagus tripus twins; we hope more will follow sometime soon, when we encounter such cases in the future [7,17,18]. In our case, we learned that surgical management might provide improvement in quality of life, even though the patients were not candidates for separation surgery.

We recognize this manuscript is not without limitations, with only a single case being reported and the case being inseparable. Our intervention, however, was rather successful in improving their quality of life.

Conclusions

In this case of inseparable ischiopagus twins, orthopedic intervention offered great potential benefits for mobility and development. This report has described the presentation and surgical management of a case of ischiopagus tripus conjoined twins. It highlights the challenges involved in surgery and the importance of investigating these infants for other congenital abnormalities. Every case of conjoined twins needs to be individually treated, and interventions should be aimed to provide optimal outcomes while considering ethical issues and parental/patient desires. In some conditions, there is an option to make a surgical correction even if the twins are inseparable due to technical and/or ethical reasons.

Ethical Statement

The surgical team discussed the treatment plan with the Institutional Review Board and the hospital Medical and Health Research Ethics Committee of Dr. Hasan Sadikin Central General Hospital.

Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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