Impact of Rigid Fixation of the Pubic Symphysis for Spinopelvic Fixation in Two Cases of Lumbosacral Agenesis

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Abstract:

Introduction: In patients with lumbosacral agenesis (SA), Renshaw type III or IV, lumbosacral instability is the primary cause of major clinical complications. Although they are usually treated with spinopelvic fusion, nonunion at the spinopelvic junction is a major complication due to the congenital sacropelvic abnormalities. The purpose of this study was to evaluate whether a combination of lumbosacral fixation and rigid fixation at the pubic symphysis could lead to postoperative bone union in patients with SA (Renshaw type III).

Methods: Retrospective case series study. We present the cases of two patients with SA, Renshaw type III, who were surgically treated by lumbosacral fusion using a posterior approach, and they exhibited nonunion at the lumbosacral junction.

Results: Case 1. A 10-year-old male underwent T8-S posterior fixation followed by multiple augmentations using allografts at the lumbosacral junction for delayed union. All additional procedures with bone graft using a posterior approach failed to achieve bone union; however, additional rigid fixation at the pubic symphysis resulted in a successful lumbosacral bone union.

Case 2. A 6-year-old male underwent vertical expandable prosthetic titanium rib (VEPTR) surgery with multiple rod extension procedures. Subsequently, at the age of 10 years, a combined two-stage anterior (L1-3) and posterior (T8-iliac) fixation with T9 hemivertebrectomy was performed. As a result of subsequent nonunion with screw loosening, additional rigid fixation at the pubic symphysis was performed 1 month after posterior fixation. Bone union was finally achieved 1 year after all the surgical interventions.

Conclusions: Rigid fixation at the pubic symphysis may play a significant role in achieving rigid bone union for unstable lumbopelvic connection, such as SA, Renshaw type III or IV.

Keywords:

lumbosacral agenesis, Renshaw type III, telescoping sign, pubic symphysis, lumbosacral junction

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Introduction

Lumbosacral agenesis (SA) is a rare condition characterized by the absence of one or more lumbar vertebrae and the total or partial absence of the sacrum. It is commonly associated with myelomeningocele (spina bifida) and paraplegia below the site of the spinal malformation, congenital musculoskeletal deformities, and varying degrees of sensory and motor deficits[1)]. SA occurs in approximately 0.1-0.25 per 10,000 pregnancies[2)] and has been associated with maternal diabetes[3)], vascular hypoperfusion[4)], and Currarino triad and homeobox gene abnormalities[5)]. In 1978, SA was classified into four types, largely based on the osteological defects between the spine and the sacrum[6)]. In patients with SA, Renshaw type III or IV, lumbosacral instability with a short trunk is the primary cause of the major clinical issues,

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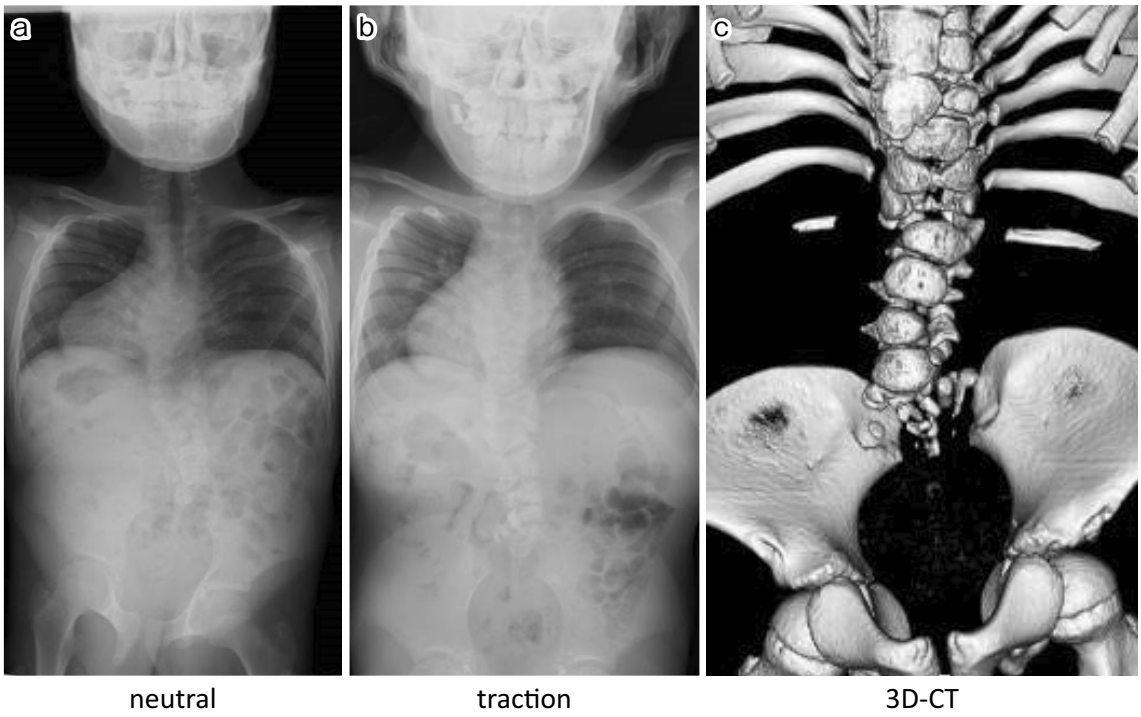

Figure 1. X-ray and 3D-CT images showing the characteristics of the articulation between the spine and the pelvis based on Renshaw type III. a, neutral position; b, manual traction showing a telescoping sign; c: congenitally deformed lower lumbar vertebrae descending into the gap between the bilateral ilia.

such as thoracic insufficiency syndrome, visceral anomalies in the urinary tract and lower alimentary system, compression of abdominal organs, and neurological damage. Further, the orthopedic management of children with SA has always been controversial. A previous report has advocated subtrochanteric amputation or knee disarticulation and subsequent prosthetic fitting for the treatment of more severely affected lower extremities[6]. Another report revealed that the surgical outcome of fixation of the lumbar spine to the pelvis was effective[7]. Although patients classified as having Renshaw type III or IV are often treated by spinopelvic fusion, nonunion at the spinopelvic junction is a major complication due to the congenital anatomical abnormalities. Nonunion reduces the patient's quality of life postoperatively and results in the progression of the deformity. Here, we report two cases of spinopelvic nonunion after lumbosacral fixation in which additional rigid fixation at the pubic symphysis resulted in bone union.

Materials and Methods

We present the cases of two patients with SA, Renshaw type III, who were surgically treated with lumbosacral fusion using a posterior approach in Meijo Hospital. Radiological data for the patients who underwent spinopelvic fusion were evaluated by X-ray and computed tomography (CT) after surgery.

Result

Two renshaw type III cases of SA

Case 1

A 10-year-old male with Klippel Feil syndrome had a characteristic telescoping back (Fig. 1). Although he was ambulatory, his chief complaint was weakness in the right leg. Radiological images revealed a complete absence of the sacrum and incomplete formation of the lower lumbar vertebrae which descended into the gap between the ilia. Under traction, the lumbar vertebrae moved away from the gap (the telescoping sign), which meant an unstable lumbopelvic connection. At the age of 10 years, he underwent T8-S posterior fixation with a spinal implant that had been designed for occipitocervical fixation, followed by multiple augmentations using allografts at the lumbosacral junction. The operation time was 470 min, and intraoperative blood loss was 228 mL. At the 1-year follow-up, the X-ray images revealed rod breakage and screw loosening due to nonunion at the spinopelvic junction (Fig. 2a). The broken implant was removed, followed by fixation using pedicle and iliac screws. Subsequently, because the posterior approach failed to achieve bone union again, bone graft augmentations were performed twice (Fig. 2b). However, bone union at the spinopelvic junction was still not achieved (Fig. 2c). We, therefore, opted for rigid fixation at the pubic symphysis with a small dynamic compression plate (DCP), creating a circumferential fixation of the pelvis (Fig. 2d). Eighteen

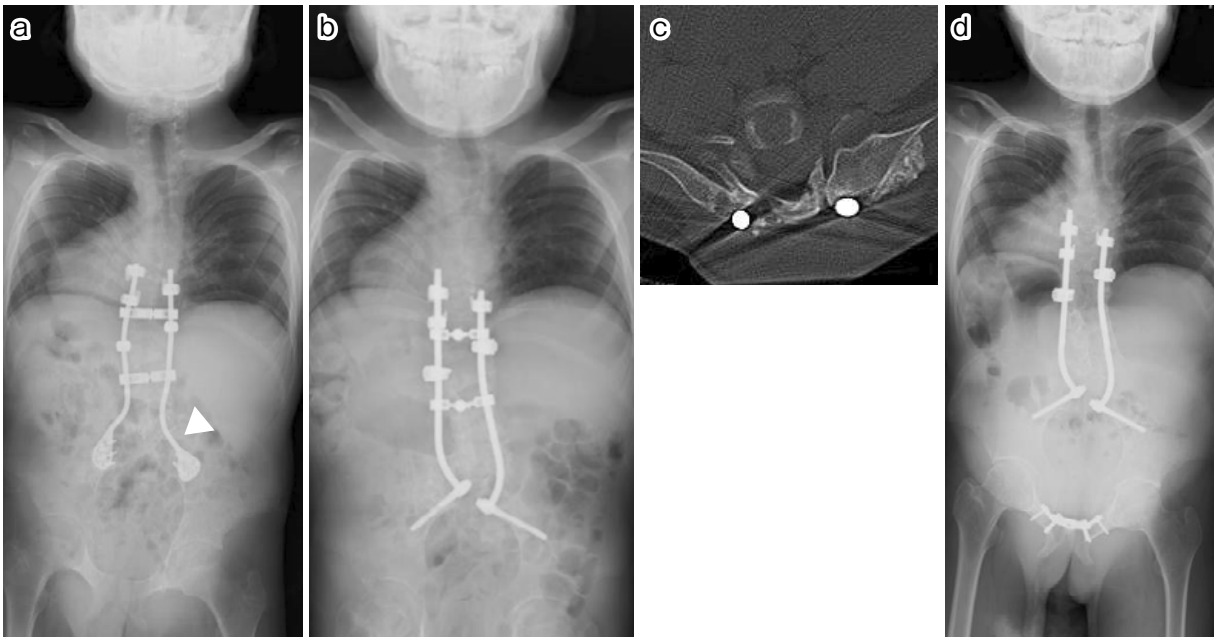

Figure 2. a: 1-year postoperative X-ray image showing a right rod breakage (white arrowhead); b: X-ray showing posterior fixation with pedicle and iliac screws after removing U.H.U spinal implant; c: CT shows delayed union at the spinopelvic junction after two bone graft augmentations. d: X-ray shows rigid fixation at the pubic symphysis.

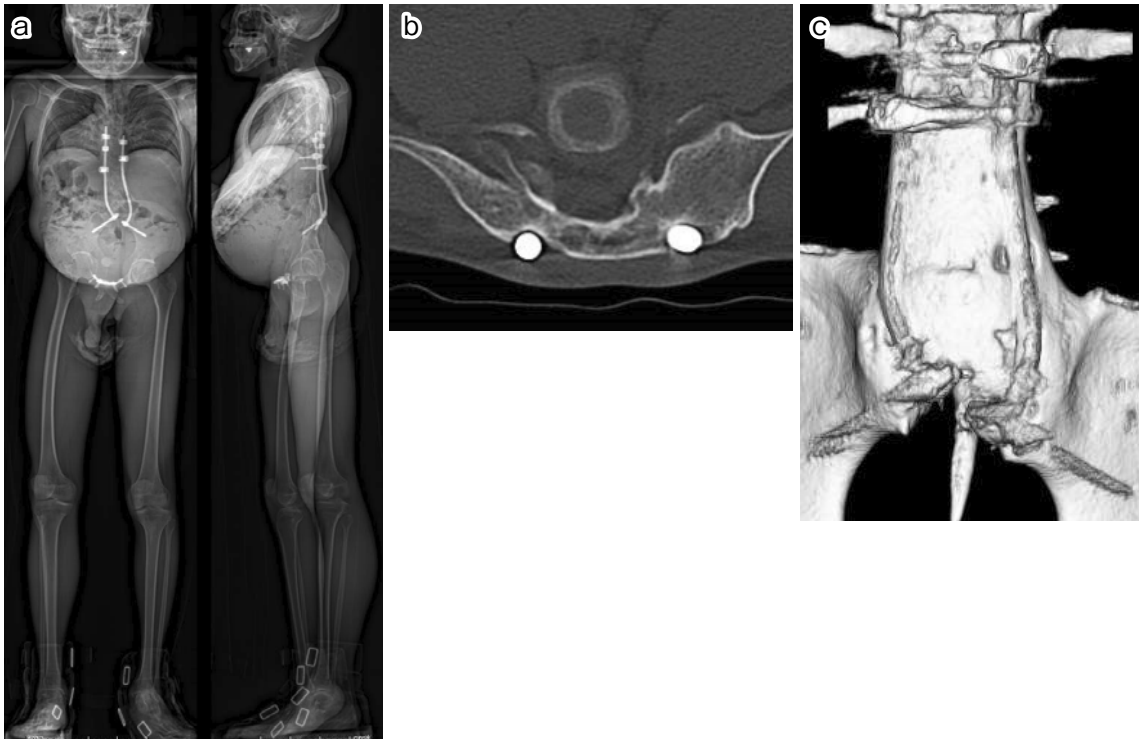

Figure 3. a: Plain X-ray image showing bone union at the spinopelvic junction in an erect position; b, c: CT images showing complete bone union at the spinopelvic junction.

months after the rigid fixation of the pubic symphysis, we detected a fusion mass on the laminae, and bone union at the spinopelvic junction was achieved. He maintained an ambulatory status with the support of a single crutch at his final check-up, 8 years postoperatively (Fig. 3).

Case 2

A 3-year old boy was referred to our institution with progressive spinal deformity (Fig. 4). He had spinal dysraphism in addition to lumbosacral agenesis (Renshaw III) and a history of hydrocephalus and anal atresia. This had been

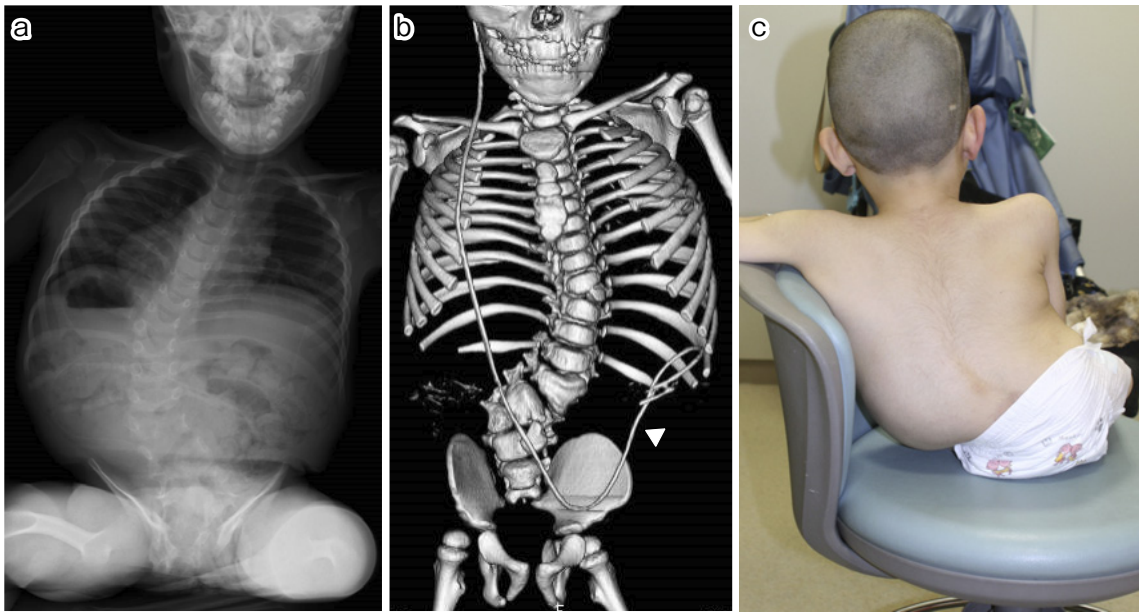

Figure 4. a: X-ray images; b: 3D-CT images showing the relation between the congenitally deformed spine and the pelvis (Renshaw type III). White arrowhead indicates a ventriculoperitoneal shunt tube; c: clinical photography.

treated surgically, and he had colostomy closure for anal atresia at the age of 1 year. He also underwent ventriculoperitoneal shunting for the treatment of hydrocephalus at the age of 2 months. At our initial consultation, he had bilateral incomplete paraplegia of the lower limbs, and he moved mainly using his arms at home and a wheelchair outside. In the rehabilitation training room, he could stand with the support of bilateral long leg braces. He had multiple hemivertebrae in the thoracic and lumbar segments, thoracic scoliosis of 78°, and lumbar scoliosis of 59°, with moderate lumbar kyphosis. On the 3D-CT images, the absence of bilateral sacroiliac joints was observed, with a residual structure of the first sacrum, which sank between the gaps of the ilia. We made a diagnosis of lumbosacral agenesis, Renshaw type III. We performed growth-friendly surgery using bilateral rib-pelvis devices, vertical expandable prosthetic titanium ribs (VEPTR), following halo-gravity traction at the age of 6 years (Fig. 5a). After serial extension procedures (four times, with replacement three times, Fig. 5b), a combined two-stage anterior (L1-3) and posterior (T8-iliac) fixation with T9 wedge hemivertebrectomy was performed with autologous bone graft between the lumbar vertebrae and the ilia bilaterally, using a rib graft, at the age of 10 years. The operation time was 324 min, and intraoperative blood loss was 488 mL. Because of unreliable stability on the distal anchor fixation, we planned and performed additional fixation at the pubic symphysis with a small DCP 1 month after the posterior fixation (Fig. 6a), followed by posterior augmentation with an allograft. These multiple procedures to create a rigid stabilization of the pelvic ring enabled complete bone union at the spinopelvic junction within a year of the operation (Fig. 6b).

Discussion

Patients with SA often exhibit spinopelvic instability; severe cases exhibit spinopelvic dissociation due to congenital partial or complete agenesis of the sacrum combined with one or more of a multitude of genitourinary and gastrointestinal malformations[8,9]. Due to the complexity of the treatments, it is important that patients with SA are treated by a multidisciplinary team involving pediatricians, pediatric and orthopedic surgeons, physiotherapists, and social workers. For spine surgeons involved in the treatment of patients with SA, an important consideration is the potential of these patients to walk[10].

Sacral malformation or SA is a rare type of malformation of the lower spine (caudal regression syndrome). Between 1959 and 1977, Renshaw classified 23 patients with sacral agenesis into four groups according to the pattern of morphologic deficiency in the bones and their articulation[6]. This classification reveals the missing part of the lower spine and shows how the spine connects to the pelvis. In patients with Renshaw classification types III and IV SA, spinopelvic kyphosis or instability frequently develops and may require lumbopelvic fusion after the age of 4 years[6].

A characteristic feature of SA is a telescoping spine, in which the unsupported trunk shortens and then extends when supporting the upper body with the upper limbs in the sitting position; this means the spinopelvic connection is very unstable. A spinopelvic fusion and stabilization may help provide sitting balance in these patients and protect the visceral organs from compression by reducing the kyphotic deformity[11,12]. Complications, such as implant breakage, screw loosening, or nonunion at the lumbosacral spinal fixation site, often occur because of the load on the lumbosacral connection, combined with the difficulties in achieving rigid

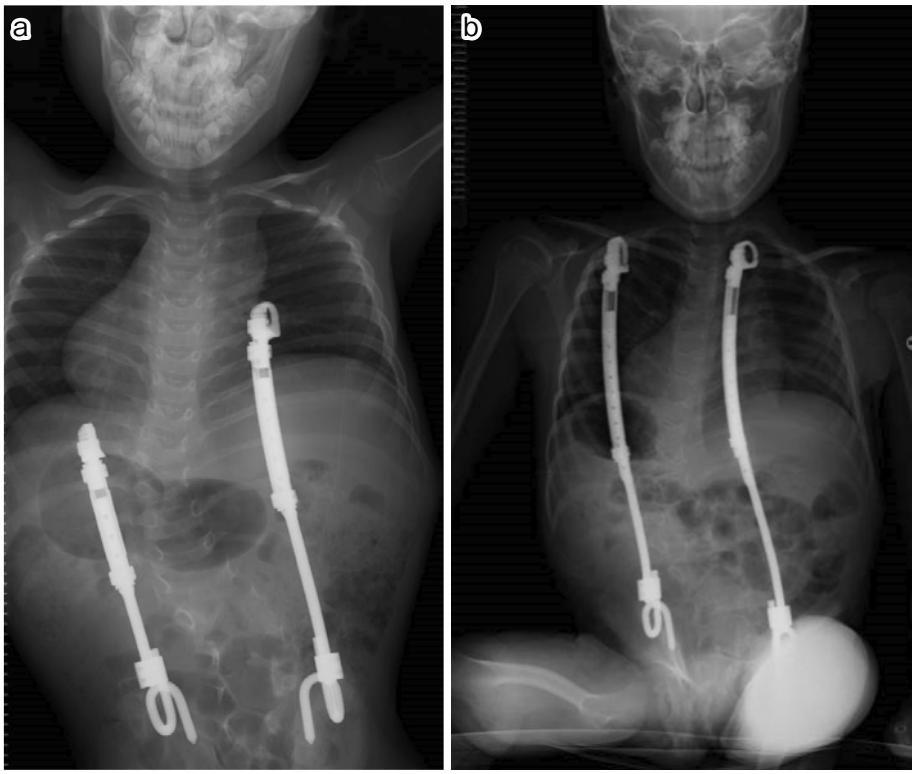

Figure 5. Postoperative plain X-ray images of growth-friendly surgery using VEPTR. a: after index surgery; b: after the fifth extension following replacement.

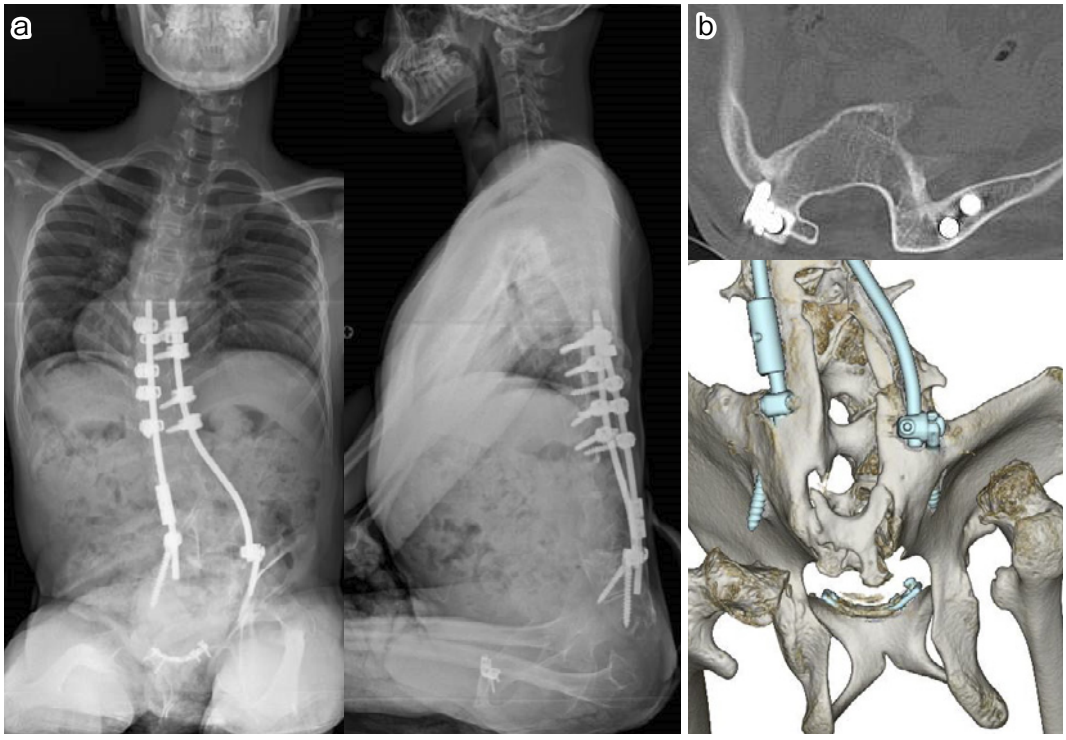

Figure 6. a: Plain X-ray images showing rigid fixation at the pubic symphysis combined with posterior fixation of the spinopelvic junction; b: CT images showing complete bone union at the spinopelvic junction.

fixation.

A previous report revealed that movement at the pubic symphysis involves compression, separation, and vertical glide[13]. Therefore, in cases with an unstable spinopelvic

junction, rigid fixation of the pubic symphysis must be considered to obtain stability of the pelvic ring. In other words, increasing the stability of the inferior part of the pubic symphysis results in increased stability of the superior part of the sacroiliac joint and reduced motion of the sacrum relative to the innominate bone[14]. In these two cases, we used small DCPs for rigid fixation of the pubic symphysis, resulting in bone union at the spinopelvic junctions, which had been unstable despite surgical treatment with posterior spinal instrumentation.

For the management of partially unstable pelvic ring injuries, even suture button fixation of the pubic symphysis has been found to be biomechanically similar to plate fixation in terms of the achieved stability[15]. There is no clear consensus on whether fixation of the pubic symphysis should always be performed to augment spinopelvic fixation in this situation regardless of the method of fixation. Therefore, future studies are needed to determine the best strategy for spinopelvic instability in lumbosacral agenesis.

From this result, we believe that rigid fixation at the pubic symphysis might play a significant role in achieving rigid bone union for unstable lumbopelvic connection, such as SA, Renshaw type III.

Conclusion

In patients with SA, postoperative nonunion at the spinopelvic junction is a major complication. Fixation of the pubic symphysis may play a major role in a successful bone union by achieving rigid fixation of the pelvic ring, thus stabilizing the spinopelvic fusion site.

Conflicts of Interest: The authors declare that there are no relevant conflicts of interest.

Ethical Approval: This study was approved by the ethics committee of Meijo Hospital.

Informed Consent: All the included patients were informed that their data would be used in this study.

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