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Case Report

Agenesis of Medial Patellofemoral Ligament (MPFL) with Habitual Patellar Dislocation in Adolescent Woman with Cerebral Palsy – Case Report

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ABSTRACT

Background: The current treatment for habitual dislocation and nonoperative treatment failure is surgical therapy. Therefore, this study describes chronic patella dislocation's comprehensive evaluation and management in an adolescent woman with cerebral palsy (CP).
Case Report: A 15 years old female patient has suffered knee pain for almost three years. Furthermore, the physical examination revealed positive adam's forward bending test and left knee cap dislocation with positive patellar J-sign at 90°. Radiologic examination of the vertebrae and left knee shows scoliosis left thoracolumbar curved with 43 cobb angle and patellar shift. The patient was then diagnosed with habitual patellar dislocation, spastic diplegia type of CP, and neuromuscular scoliosis, consulted to the pediatric department, and then planned for medial patellofemoral ligament (MPFL) reconstruction. During the surgical examination, we discovered the MPFL Agenesis. Finally, lateral release and plication of the medial retinaculum were selected for surgery and planned to receive a Boston brace for scoliosis. After six weeks of follow up, the patient shows a reduction in pain.
Discussion: Lateral release and MPFL reconstruction for patellar stabilization generate better results. However, in this case, the absence of the medial patellar facet and the medial femoral condyle enhances Lateral release and plication of the medial retinaculum more preferable to fixate the left patella and improve functional limitation.
Conclusion: Comprehensive and immediate treatment for a patient with habitual patella dislocation and other predisposition diseases increases the chances of success.

Keywords: Agenesis MPFL; Cerebral palsy; Human and medicine; Habitual patella dislocation; Surgical treatment

INTRODUCTION

Children with disability may occurs suffer in their lives. It is known to be one of the cause neonatal death in the world. The prevalence of congenital abnormalities in Indonesia is about 59.[3] for every 1000 live birth.[1] Furthermore, the congenital abnormalities that arise physical problems include the instability of patella, which is identified by dislocation or subluxation of the patella in the lateral direction.[2–4] Hasler and Studer reported that patients with habitual patella dislocation from combined predisposition factors such as trochlear dysplasia, malalignment, ligamentous laxity, and hypoplastic condyle should be considered for surgical procedures treatment.[4]

The decision-making in surgical procedures is based on orthopedic examination during the presurgical and surgical process.[4] Furthermore, the patients with habitual and recurrent patella dislocation, specifically without trauma, have a poorer stabilization prognosis and develop re-dislocation post-treatment.[5] Therefore, patients with these injuries require immediately advanced imaging and surgical repair because early identification and diagnosis with the conformity of surgical medication in-

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crease the chances of success. Hence, this study describes the holistic examination and management of habitual patella dislocation in adolescent patients with congenital disease.

CASE REPORT

In February 2020, a 15 years old female patient with knee and back pain was present in the orthopedic department of Dr. Soeroto Ngawi Regional Public Hospital. The patient suffered for almost three years and felt sick from walking long distances during sport. The patient also complained about breathing, back deformity, right-hand finger stiffness without complaint of hands function, diplegic gait abnormalities, knee instability, and cognitive impairment. The patient's knee pain and instability often felt better at rest and worse while performing a heavy activity. Also, the patient had not visited the doctor nor taken certain medicine before.

The patient has a record of congenital abnormalities at birth. Additionally, the mother has an advanced maternal age of 40 years old. There is no history of teratogenic exposure and infection during pregnancy, and the mother routinely experiences prenatal checkups. The baby was given a midwife vaginal delivery in the 36 weeks of gestation with a birth weight of 3500 grams and a body length of 50 cm.

Nursing difficulties arose from the start, whereby the baby could not cry spontaneously, having yellow skin color and low muscular tone activity. A few days after birth, the patient had respiratory distress and difficulty swallowing. Also, the patient had a history of complete immunization, six months of breastfeeding, and complimentary food after that.

As the patient grew older, there was still difficulty standing and walking until the age of 2, had monosyllable speech, and could not speak eloquently until the age of 4. Moreover, the patient experienced several difficulties memorizing, counting, and reading during the school period. Meanwhile, there is no history of similar abnormalities among the families, including the grandmother's family.

At the time of presentation, physical examination revealed strabismus in the eye exam, right chest protrusion, asymmetrical shoulder (right shoulder is taller than the left shoulder), and the gap between arm and body trunk (Figure 1) in chest exam, spine deformity, forming "S" Shape with bending to the right, rib hump at the right side of the thorax region, and lower back hump at the left hip are discovered on back examination (Figure 1). Gait assessment revealed diplegic gait abnormality in the upper extremities and difficulties climbing up and down the stairs. At the same time, there is no symptom in other gaits such as neuropathic, myopathic, choreiform, etc. Adam's Forward bending test showed asymmetrical back without change before and after the test (Figure

Figure 1. The back's Inspection shows deformity. Arrow; Red: asymmetric of head and body axis; Green: shoulder asymmetric; black: right rib hump and left hip lower hump positive; Blue: the curve of spine; Orange: the gap between arm and trunk.

Figure 2. Adam's Forward Bending Test. It looks asymmetrical, and there is no change in the back shape before and during the test. Arrow; Red: Right rib hump positive.

2). Upper extremities examination revealed hypertonic muscle on both sides, spastic specifically on the right hand, and positive pathological reflex on both hands. The Marshall test shows hyperextension on both hands and above 10 degrees elbow angle.

Inferior extremities examination revealed hypertonic muscle on both sides, limited Range of Movement (ROM), and limitation in patella mobility which patella shifting of the left knee cap with patellar J-sign positive in 90° extensions and 45° flexions. The Beighton scale measurement was 8 (Table 1), and the Apprehension test was positive.

The plain thoracolumbar radiologic imaging revealed left curve spine thoracolumbar scoliosis with 43 degrees cobb angle (Figure 3). In contrast, left knee radiologic imaging revealed patella shifting with 19 degrees of the Q angle (Figure 4) and no high patella in 30 degrees of flexion in the left knee for Blumensaat's line (Figure 5). Skyline merchant Knee X-Ray views were not evaluated during presurgical examination.

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Figure 3. Plain thoracolumbar radiologic

The final examination, either presurgical or during the surgical approach, confirmed that diagnosis for this patient is agenesis of MPFL with habitual patellar dislocation, spastic diplegia type cerebral palsy, and neuromuscular structural scoliosis.

Table 1. Beighton Scale

Measurement	Left	Right
Passive dorsiflexion and hyperextension of the fifth MCP joint beyond 90°	√	√
Passive apposition of the thumb to the flexor aspect of the forearm	√	√
Passive hyperextension of the elbow beyond 10°	√	√
Passive hyperextension of the knee beyond 10°	√	√
Active forward flexion of the trunk with the knees fully extended so that the palms of the hands rest flat on the floor	-	-
Total Score	8	

Figure 4. A 19 degree of the Q angle

Figure 5. There is no high patella in 30 degrees of flexion in the left knee for Blumensaat's line

Surgical Approach

After experiencing all series of examinations, the patient experienced MPFL reconstruction, preceded by patella examination preoperative. Then, advanced examination under anesthesia is performed to evaluate ligament stability and the patella mobility in extension and flexion patella. The patellar surgical contains medial patellar approach and identification of medial retinaculum and remnant of the MPFL. The MPFL identification showed the absence of medial facet patellar and medial femoral condyle. These findings were referred to as the agenesis of MPFL. Therefore, medial plication from the caps and the retinaculum and lateral retinacular release at the left knee was more preferred than the MPFL reconstruction due to its absence.

A week after the operation, the patient had not complained of pain or other abnormal symptoms. The patient received several therapies, including physical therapy for muscle strengthening gait training, and suggested using Boston's brace for scoliosis.

Post-Operative Evaluation

After receiving the complex therapy, including operative, medication, and physical therapy, the left patella's stability was evaluated. Clinical appearance after six months post-operation, the surgical wound was healed, felt the least pain when moving the left knee, able to stand and walk without walking aids, and no complaint while sitting with a flexed knee. In addition, the physical examination shows re-dislocation and subluxation of the left patella revealed to J-sign positive in 20° flexions of the left knee, which shows a better result from the first examination. Also, the Merchant view of X-Ray examination through radiological assessment was evaluated. This assessment identified the malposition of the articular surface patella outside of the patellofemoral surface, which is located in the lateral side of the femoral condyle. The absence of medial facet patellar and medial femoral condyle could develop the displacement of the patella while such maneuvers that are flexion or extension were applied. The medial plication was assumed from the caps and the retinaculum, which were not strong enough to fixate the patella. However, the operation succeeded in reducing the pain and increasing the patient's mobility, one of which is no complaint in climbing up and down the stairs.

DISCUSSION

Patients with cerebral palsy (CP), Agenesis of MPFL, and scoliosis are rare cases. Meanwhile, this is the only case reported in the literature. The objective of this study was to determine the holistic examination and management of habitual patella dislocation in adolescent patients with congenital disease.

Based on the anamnesis and examination, this patient complains of knee pain due to patellar dislocation, although it has been given nonoperative management. However, nearly all patellar dislocation is treated by the conservative procedure. Besides, the surgery will be needed if adequate bracing and muscle strengthening do not reduce pain.[6]

For more than decades, a different surgical technique has been described for patellar dislocation treatment. The various procedures that have been described are anatomical and non-anatomical reconstruction. MPFL reconstruction improves the anatomical repair of recurrent and habitual patellar dislocation. MPFL plays a role in the patella as a soft tissue stabilizer and prevents patellar dislocations. The successful patellar stabilization with MPFL reconstruction depends on several factors. The first is appropriate patient selection, and the second is a proper surgical technique.[6] In this case, the main problem was the absence of MPFL, and no MPFL remnants were discovered. Therefore, the surgery was performed by medial plication and lateral retinacular release,

which many authors previously described, which is not a new technique.[6–8]

In this case, the holistic examination was conducted and diagnosed with spastic diplegia type of cerebral palsy with idiopathic structural scoliosis, habitual left knee dislocation, and the absence of MPFL on the left knee. By discovering these abnormalities, treatment in pharmacotherapy, physical therapy, and decision in operation procedures could be decided and benefit the patient. On the other hand, patellar stabilization in habitual patellar dislocation will not be successful if multi-pathologies are present. Hence, a holistic examination should be performed to determine the abnormality and other pathologies.[6]

There is a correlation in each of the patient abnormalities that have been described in many studies. The children who had CP were born with an immature brain which consequential musculoskeletal deformities.[9] About 50%–75% of patients with quadriplegic CP have severe motor involvement that leads to spinal deformities such as scoliosis and impairment in the development of movement muscle tone and posture.[9] The etiology of scoliosis in CP has yet to be well defined. Spinal deformity is associated with muscular imbalance around the spinal axis from either spastic or flaccid muscular weakness. In studies focusing on CP, the factors contributing to the development of spinal deformity have been suggested to include spasticity, muscle weakness, and poor muscle control.[10] Previous studies of 70 children (aged 12-18 years) with severe spastic and/or dystonic cerebral palsy showed that the present history of previous hip surgery, intractable epilepsy, and female gender are significant predictors of developing severe scoliosis in children with CP.[11] Hagglund et al., who used the Gross Motor Function Classification System (GMFCS) was discovered scoliosis signs start to appear from the age of 5 years old. By the time they would have a risk of ≥40° Cobb angle (75%), which means the motoric cannot be function normally (GMFCS I) (0%).[12] The mechanism of spine deformity in CP patients began from the early phase of postnatal child development. The development of scoliosis in CP patients is thought to result partly from persistent primitive reflex patterns and asymmetrical tone in the paraspinous and intercostal muscles. Placing patients with a weak trunk and total body involvement into artificial upright positions without appropriate spinal support may encourage gravity-related kyphosis and scoliosis. Lack of neuromuscular control prevents proper head alignment, which means these patients do not develop compensatory curves to bring the shoulders and head over the pelvis.[13] This distortion is also affected by anatomical factors such as muscle tone, laxity of ligamentous structures, joint position and mobility, gravitation, motor damage, and postural factor.[12,13] Physical treatment for scoliosis, including bracing, is currently not fully known whether there is a difference in effectiveness between brace types. According to a study by Costa, Schlosser, Jimale, et al., rigid braces have a high success rate compared to soft brace for Adolescent Idiopathic Scoliosis (AIS) 73.2% (95% CI 61-86%). Brace types included in rigid braces are Boston, Cheneau brace, Progressive Action Short Brace (PASB), etc.[14] Therefore, it was suggested that the patient take the Boston's brace for her scoliosis.

Furthermore, recurrent dislocation of the left knee among the patient is caused by two factors, namely an anatomical abnormality and ligament laxity. Since patellofemoral stability depends on multifactorial tissue stabilizers, osseous structures, and lower limb alignment, the MPFL is a major tissue stabilizer for knee mobility.[6] Unfortunately, an advanced examination under anesthesia revealed the agenesis of MPFL. Hence, decision-making for surgery was medial plication of the caps and the retinaculum and the release of lateral retinacular. This technique is previously described by Fonseca for patellofemoral joint surgery.[8] The basic theory for this technique is the imbalance of the knee caused by excessive tension of the lateral retinaculum, which leads to patella instability and knee pain. Releasing the

lateral retinaculum reduces pressure on the lateral surface during flexion.[8,15] Meanwhile, the absence of medial facet patellar and medial femoral condyle is no contraindication for MPFL reconstruction. Instead, it indicates the medial retinaculum plication as a major technique in such conditions. The medial retinaculum plication (MRP) is less invasive, has a cosmetic advantage, and does not reconstruct the medial patellofemoral ligament.[16] Therefore, this case study revealed the agenesis of MPFL and stated that MRP is the best choice for the surgical technique.

CONCLUSION

A deep anamnesis and holistic examination assist in determining the diagnosis and deciding the appropriate treatment. For example, recent challenges in examining a patient with knee pain and cerebral palsy are not limited to the subjective and neurological condition of the patient. However, the patient needs to include functional, morphological, and anatomical changes to determine the objective side. Therefore, these aim to discover the abnormality which becomes a confounding factor in patient treatment.

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