Management of ACL Elongation in the Surgical Treatment of Congenital Knee Dislocation

KEVIN E. KLINGELE, MD; SCOTT STEPHENS, MD

abstract

Congenital knee dislocation is a rare anomaly associated with a variety of neuromuscular diseases and deformities. The etiology of this condition remains unclear, but it is usually associated with a variety of disorders, such as Larsen’s syndrome, arthrogryposis, spondyloepiphyseal dysplasia, Ehlers-Danlos syndrome, Down syndrome, and Streeter’s dysplasia. It is rarely an isolated entity, and 60% of patients with congenital knee dislocation had additional congenital anomalies, most commonly hip dysplasia. The ideal method of treatment is debated. No current treatment algorithms address anterior cruciate ligament (ACL) elongation and its role in recurrent deformity or hyperextension.

This article describes 2 patients who underwent open reduction of the knee for recurrent and neglected congenital knee dislocations. An ACL shortening and reinforcement technique is described. Both patients’ treatment consisted of V-Y advancement of the extensor mechanism, soft tissue release, anterior capsulotomy, and posterior capsulorrhaphy. Anterior cruciate ligament shortening and reinforcement using an iliotibial band physeal-sparing technique was performed. The technique improved maintenance of reduction and prevented hyperextension of the knee. Anterior cruciate ligament elongation is an underemphasized anatomical feature associated with congenital knee dislocation. Due to its role in the prevention of anterior subluxation of the tibia and its effect on knee stability, incompetence should be addressed at the time of open reduction. The presence of an intact ACL with a congenital knee dislocation does not preclude management of anterior instability. Competence of the intact ACL should be addressed following reduction.

Dr Klingele is from the Department of Orthopaedics, Nationwide Children’s Hospital, and the Department of Orthopaedics, The Ohio State University, and Dr Stephens is from Mount Carmel West Medical Center, Columbus, Ohio.

Drs Klingele and Stephens have no relevant financial relationships to disclose.

The authors thank Anthony Baker from The Ohio State University, who created the illustrations for this article.

Correspondence should be addressed to: Kevin E. Klingele, MD, Department of Orthopaedics, Nationwide Children’s Hospital, 700 Children’s Dr, Columbus, OH 43205 (kevin.klingele@nationwidechildrens.org).

doi: 10.3928/01477447-20120621-29
Congenital dislocation of the knee is a rare disorder initially described by Chatelaine in 1822 and Bord in 1834. It is seen in approximately 1 of 100,000 newborns and is less common than congenital hip dislocations. The etiology of this condition remains unclear, but it is usually associated with a variety of disorders, such as Larsen’s syndrome, arthrogryposis, spondyloepiphyseal dysplasia, Ehlers-Danlos syndrome, Down syndrome, and Streeter’s dysplasia. It is rarely an isolated entity, and 60% of patients with congenital knee dislocation were found to have additional congenital anomalies, most commonly hip dysplasia.

Although no clear pathogenesis has been found, a variety of anatomical patterns are consistently found in congenitally dislocated knees. The lateral quadriceps and fascia lata are typically fibrosed, with a scarred suprapatellar pouch. The hamstrings and collateral ligaments are displaced anteriorly to their normal position. The patella may be absent or hypoplastic. Hypoplasia of the intercondylar notch and tibial spines, increased posterior slope and proximal tibial bowing, and valgus alignment at the knee may exist. Posterior capsular redundancy is significant. The anterior cruciate ligament (ACL) may be absent, elongated, or hypoplastic, and the posterior cruciate ligament may be shortened and tight. Variable involvement of the menisci may include discoid menisci or hypoplastic menisci.

Treatment modalities range from serial stretching or closed manipulations to open reduction. Outcomes vary depending on the severity and type of treatment required, but several residual problems have been identified, including extensor mechanism weakness, recurrent hyperextension, and progressive valgus instability and angulation. The cause of the deformity is not well understood and may be related to failure to address posterior capsular redundancy at the index operation. Operative techniques now emphasize extensor advancement with or without femoral shortening, associated soft tissue release and transposition to obtain reduction, and the addition of a posterior capsulorrhaphy. Few descriptions address the competence of an often elongated ACL and the role this pathology may have in preventing or treating recurrent or chronic deformity.

This article presents 2 cases of congenital knee dislocation associated with significant ACL elongation. Both patients underwent open reduction in addition to ACL shortening and physeal-sparing reconstruction. This as an important variable during the management of congenital knee dislocation. The parents of both patients were advised that information regarding their cases would be submitted for publication and provided consent.

**CASE REPORTS**

**Patient 1**

A 2-year, 3-month-old girl presented following international adoption with an isolated left knee hyperextension deformity. No history of treatment prior to adoption was found. On physical examination, the patient had 60° of hyperextension and was unable to flex beyond neutral (Figure 1A). Anteroposterior and lateral radiographs revealed anterior subluxation of the tibia with hyperextension and with forced flexion to neutral (Figure 1B). Preoperative magnetic resonance imaging (MRI) confirmed the anterior tibial subluxation and identified significant elongation of an intact ACL (Figure 1C).

The patient underwent open reduction of her left knee consisting of anterior arthrotomy, extensive lateral release, V-Y quadriceplasty, posterior transposition of the pes anserinus attachment, and posterolateral capsular imbrication. On reduction of the anterior subluxation, significant laxity of the elongated ACL was noted. This allowed for continued anterior subluxation and residual 30° of hyperextension despite posterior capsulorrhaphy. Using the technique discussed below, an acute shortening of the native ACL was augmented with a physeal-sparing ACL reconstruction using an autogenous iliotibial band, which prevented subluxation and hyperextension. At the time of arthrotomy, the patient was also noted to have a complete, stable discoid lateral meniscus that was managed with an open 1-piece saucerization.

Postoperatively, the patient was placed in a long-leg cast in 70° of flexion for 4 weeks. On cast removal, she was transitioned to a hinged knee–ankle–foot orthosis with a neutral extension stop for 3
months. At 1-year follow-up, the patient exhibited full and symmetric passive knee flexion and passive extension to neutral (Figure 2). She demonstrated no anterior subluxation of her tibia throughout passive and active range of motion and had no active extension lag.

Patient 2

A 2-year, 6-month-old boy presented with an undetermined genetic disorder resulting in multiple joint contractures. He had undergone bilateral open reductions of his knees at age 5 months due to failure of nonoperative treatment. Surgery consisted of acute femoral shortening, V-Y quadricepplasty, transposition of the pes anserinus attachment, and posteromedial and lateral capsulorrhaphy. He had an intact ACL at the index procedures. At 2-year follow-up, the patient’s left knee showed a recurrent hyperextension deformity of approximately 45° with forced flexion only to neutral. Radiographs confirmed anterior subluxation of the tibia, and follow-up MRI confirmed an elongated ACL (Figure 3).

The patient underwent revision open reduction with ACL shortening, reinforced with a physeal-sparing ACL reconstruction. Repeat quadricepplasty required the use of autogenous semitendinosis and gracilis grafts to reinforce the extensor mechanism. This resulted in a knee range of motion of 0° to 90° intraoperatively. He was placed in a long-leg cast for 4 weeks and was then transitioned into a hinged knee–ankle–foot orthosis for 3 months. At 1-year follow-up, the patient exhibited 0° to 90° of flexion without hyperextension and minimal quad lag.

SURGICAL TECHNIQUE

After wide exposure via a longitudinal anterior incision, the anterior and central portion of the iliotibial band is harvested with maximum length and left attached to Gerdy’s tubercle. The graft is tubularized with a heavy suture and the open reduction is performed in the usual fashion. The graft harvest site can be used as a lateral release. Following anterior arthrotomy, a V-Y quadricepplasty with or without femoral shortening and posterior transposition of the pes anserine structures is performed. Posterior capsulorrhaphy following tibial reduction is achieved via posteromedial and posterolateral capsular imbrication. If intact, the length of the ACL is then examined and the amount of laxity determined. If anterior tibial subluxation is seen with extension to or beyond 10° to 15° of neutral, the redundant ligament is excised and the shortened ligament repaired to the tibial epiphysis footprint (Figure 4). The iliotibial graft is brought around the posterior lateral condyle and over the top of the native ACL.12 The graft is placed under the intermeniscal ligament and sewn underneath the periosteum of the proximal medial tibia. Tenodesis to the posterolateral femur and intermuscular septum is reinforced with suture attachment to the native ACL (Figures 5, 6). The graft is tensioned to prevent more than 1 to 2 mm of anterior tibial translation at full extension of the knee. Adequate tensioning prevents hyperextension.

DISCUSSION

Congenital knee dislocation is a complex disorder associated with a variety of syndromes and additional comorbidities that can complicate treatment and function. Because of its variable presentation, ranging from flexible hyperextension to frank dislocation, no consensus exists on the ideal treatment of this condition. Although the pathogenesis of this disorder is unclear, typical patterns of anatomic deformity exist that surgeons must consider when attempting to gain reduction of the knee. Surgical management includes extensor mechanism lengthening with or without femoral shortening, soft tissue releases, posterior transposition of the pes anserinus, and posterior capsulorrhaphy. The elongation and incompetence of the ACL has limited discussion in the literature and is not incorporated in current surgical algorithms.

Mayer,3 McFarland,6 and Odell and Holt12 described the ACL as being attenuated, elongated, and poorly developed. Katz et al12 reported that the cruciate ligaments of 5 patients with congenitally dislocated knees were either absent or hypoplastic. The ACL was reconstructed with the use of a retinaculum and a portion of the patellar tendon. They reported that the
primary cause of the congenital dislocation was the absence or hypoplasia of the cruciate ligaments.\(^5\)

Sud et al\(^7\) reported 17 knees in 10 patients who underwent V-Y lengthening of the quadriceps tendon and anterior capsulotomy and reported that no patients had clinically apparent sagittal plane instability and that the ACL was intact visually but was long and atrophic.\(^4\)

Soyuncu et al\(^13\) reported on 2 adolescents with congenital knee dislocation. One patient had an elongated ACL after reduction and was tightened with the use of a Bunnell suture technique at the base of the tibia and suture tunnels to properly tension the ACL. They recommended advancement of the ACL to prevent recurrences, especially in delayed cases, and noted decreased anterior laxity on knees that were tightened.\(^7\)

Oetgen et al\(^10\) performed a retrospective review of 7 patients treated surgically and compared quadriceplasty and femoral shortening. Patients were followed for an average of 12 years, and each patient underwent a clinical examination, functional evaluation, and gait evaluation. Seven (78%) of 9 knees had some degree of instability on examination, but no patient wore a brace. Although good functional results were demonstrated with both surgical approaches, they scored significantly lower in the sports and physical functioning aspects, demonstrating an underlying instability during more demanding activities. This instability was noted even in patients who achieved early successful knee reduction, indicating that ligamentous structures may be abnormal.

On open reduction of the knee in the current cases, the ACL was intact but functionally incompetent due to elongation. The authors identified residual anterior instability and a tendency toward hyperextension following open reduction, quadriceps lengthening, lateral release, posterior transposition of the pes anserine, and posterior capsulorrhaphy. The described technique of ACL shortening and reinforcement helped maintain reduction and prevented anterior tibial subluxation and residual hyperextension at surgery and at last follow-up.

Long-term follow-up studies of congenitally deficient ACL knees and congenital knee dislocations describe progressive valgus angulation. Much of this angulation may be related to the anterior instability and subsequent rotatory instability seen with cruciate deficiency. Treatment of anterior instability may prevent progressive valgus angulation. Nonetheless, the presence of an ACL in management of congenital knee dislocation does not equate to a stable knee. The surgeon must account for ACL elongation following reduction.

**CONCLUSION**

Although good results have been reported with a variety of treatment techniques, ACL incompetence due to elongation is an underreported anatomic feature that warrants consideration during initial surgical treatment of congenital knee dislocation. In addition to assisting in maintenance of reduction, stabilizing and reinforcing this ligament may prevent long-term anterior instability, hyperextension of the knee, and recurrent deformity.

**REFERENCES**


