

# Management of Congenital Dislocation of the Knee

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## **Abstract:**

Congenital knee dislocation is a rare disorder and the published literature largely consists of small case series. Congenital knee dislocation represents a broad spectrum of severity from hyperextension to frank dislocation. It can be isolated or be present in syndromic infants with concomitant joint disorders such as clubfoot or developmental dysplasia of the hip. Hyperextended knees can be reduced with serial casting alone in the neonatal period and do not require further treatment. Surgical treatment of the congenital knee dislocation results in satisfactory outcomes in the majority of reported cases, although outcomes are highly dependent on the degree of concomitant joint involvement. Treatment decisions are often based on radiographic findings and passive range of motion of the knee. Described procedures include percutaneous or mini-open quadriceps tenotomy, VY quadricepsplasty, and femoral shortening. The current review summarizes the most up-to-date literature on the surgical management of congenital knee dislocation.

## **Key Concepts:**

- Congenital knee dislocation is a rare disorder affecting approximately 1 in 100,000 children.
- Patients should be evaluated for syndromes and concomitant musculoskeletal deformity affecting other joints.
- Serial casting should be attempted in most cases of congenital knee dislocation as successful reduction with serial casting obviates the need for surgical treatment.
- Surgical procedures to facilitate reduction of irreducible knees include percutaneous or mini-open quadriceps tenotomy, V-Y quadricepsplasty, and femoral shortening.

## **Introduction**

Congenital dislocation of the knee (CDK) is a rare abnormality with an estimated incidence of 1 in 100,000 births; in contrast, developmental dysplasia of the hip (DDH)<sup>1</sup> is 100x more common. It is a diagnosis that is recognized either on prenatal ultrasound or immediately after birth and is characterized by hyperextension deformity at the knee. Females are more commonly affected than males, and patterns of inheritance within families suggest a possible genetic basis.<sup>2,3</sup> CDK is associated with other musculoskeletal anomalies in many

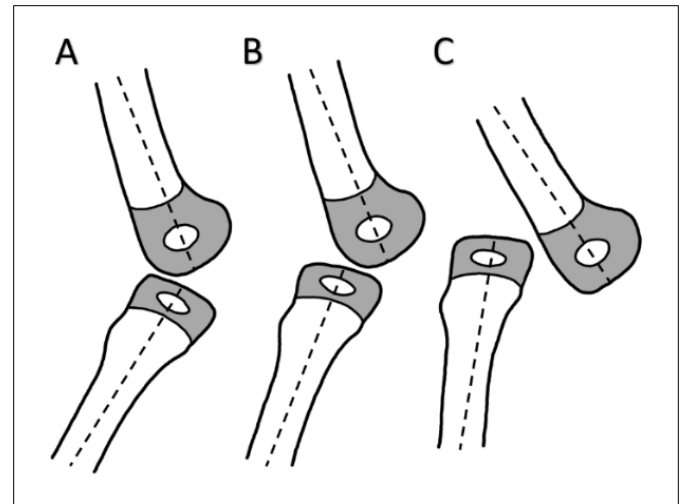
cases, the two most common being DDH and clubfoot. It has also been observed concomitantly with congenital vertical talus (CVT), forefoot adductus, calcaneovalgus (CV) foot deformity, cleft palate, spina bifida, hydrocephalus, myelomeningocele, and genitourinary abnormalities.<sup>2,4</sup> CDK can be idiopathic or associated with syndromes such as Larsen, Ehlers-Danlos, arthrogryposis multiplex congenita (AMC), Beals, syndrome and Down syndrome.<sup>2</sup>

### Classification

Leveuf and Pais published the most widely used classification for CDK in 1946.<sup>5</sup> The classification is based on radiographs and defines Grade I (GI) as simple hyperextension or recurvatum of the knee with a preserved anatomical relationship between the tibia and femur.<sup>3</sup> Grade II (GII) represents subluxation of the joint with partial longitudinal contact between the tibia and femur, and Grade III (GIII) is defined as complete dislocation (Figure 1).<sup>1,3,6,7</sup> In 2011, the Tarek CDK grading system (Table 1) was introduced by Abdelaziz and Samir based on their observations that passive knee flexion was an important indicator of severity and could also help guide treatment.<sup>8</sup> In this classification, GI is defined as >90 degree range of passive flexion with simple recurvatum on radiographs. GII is passive flexion between 30 degrees and 90 degrees with subluxation or dislocation on radiographs. GIII is passive flexion less than 30 degrees with dislocation on radiographs.<sup>8</sup> They proposed that percutaneous quadriceps tenotomy (PQT) should be reserved for GI and GII knees that have failed serial casting and that VY quadricepsplasty (VYQ) be performed for GIII knees.

### Conservative Management

It is widely supported that initial recognition and conservative management consisting of passive stretching and serial casting/splinting in flexion is associated with the good outcomes in those patients with less severe deformity.<sup>2,3,7,9</sup> In a 2010 study of 19 patients (25 knees) over 18 years in a single hospital, reduction within the first 24 hours of life demonstrated excellent outcomes.<sup>9</sup> Serial casts should be placed to hold the knee in flexion. Casts may be changed every few days in neonates and then weekly as the position of the knee improves. Transition to a removable splint or a Pavlik harness to maintain flexion can be performed once 90 degrees of flexion has been achieved with serial casting. Attempted closed treatment should be abandoned if knee flexion is not improving through the knee joint as seen on lateral radiographs as it is possible to cause physeal injury or plastically deform the proximal tibia.



**Figure 1.** The Leveuf and Pais classification of congenital knee dislocation based on radiographs.<sup>5</sup> Grade I (A) is defined as a simple hyperextension or recurvatum, Grade II (B) represents subluxation of the joint, and Grade III (C) represents a complete dislocation. Adapted from *J Bone Joint Surg Am.* 1969; 51:255-269.<sup>3</sup>

Grade	Passive Flexion	Radiology
GI	>90°	Simple recurvatum
GII	30–90°	Subluxation/dislocation
GIII	<30°	Dislocation

**Table 1.** The Tarek grading system uses passive knee flexion to determine the severity of dislocation and guide management.<sup>8</sup>

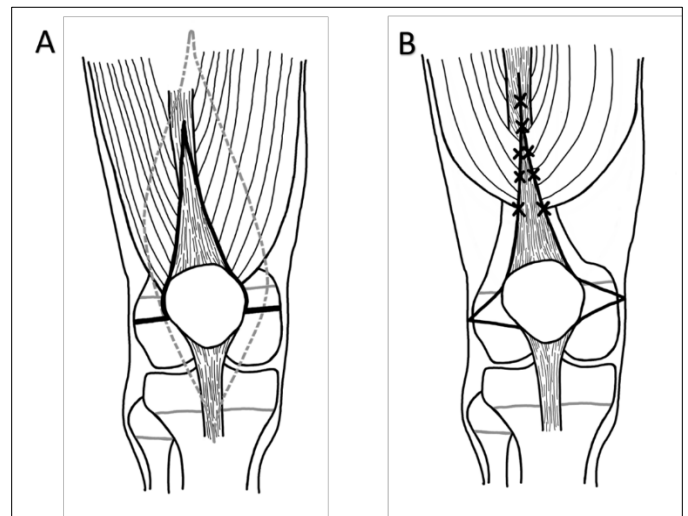
In the event of failure of closed reduction, there is no clear consensus on how to proceed surgically, which is the focus of this review.<sup>10</sup> The primary aim of any surgical approach is to elongate or effectively elongate the shortened and fibrosed quadriceps tendon to make useful knee flexion possible.<sup>11</sup> There are three main procedures reported for the surgical management of CDK: percutaneous or mini-open quadriceps tenotomy (PQT), the VY quadricepsplasty (VYQ), and the femoral shortening osteotomy (FS), which will be described below.

### Percutaneous or Mini-Open Quadriceps Tenotomy

The idea of a less extensive surgery to treat congenital knee dislocation was originally described by Roy and

Crawford and has since been modified by several authors.<sup>12-14</sup> The PQT described by Roy and Crawford consists of three stab incisions (Figure 2). With the knee held in maximal flexion, the first incision is midline and is made one to two patella lengths proximal to the patella. The rectus fascia is released percutaneously. Two additional percutaneous incisions are made just medial and lateral to the patella in the retinaculum to allow for knee flexion. A similar “mini-open” quadriceps tenotomy was described by Shah et al.<sup>13</sup> This procedure is performed if 90 degrees of knee flexion is not achieved after six or seven casts. The procedure consists of a 2-cm longitudinal midline incision over the superior pole of the patella. The main quadriceps tendon is transected 1 cm proximal to the superior pole of the patella. If the knee is still unable to be reduced, an anterior capsulotomy and lateral retinacular release is performed through the same incision. Youssef described a similar mini-open procedure.<sup>15</sup> Similarly, Patwardhan et al. described percutaneous needle tenotomy using a 16-gauge needle.<sup>14</sup>

Advocates of PQT prefer this procedure to VYQ as it minimizes extensive scarring of the quadriceps which

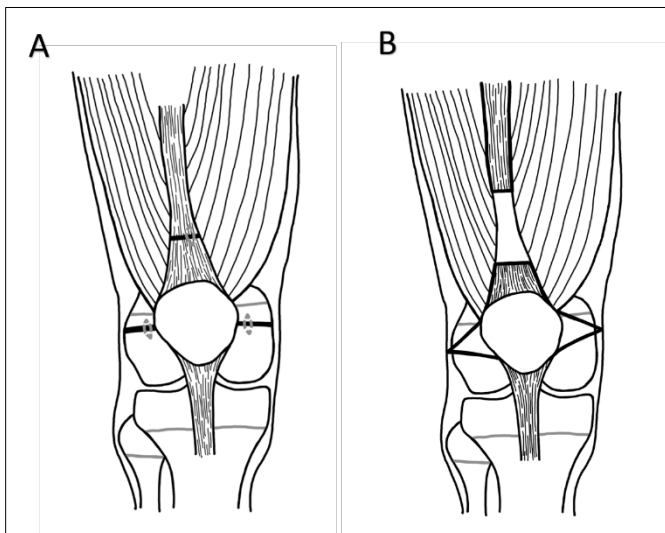


**Figure 3.** The VY quadricepsplasty (VYQ) described by Curtis and Fisher.<sup>3</sup> A) The long anterolateral incision with the division of the central portion of the quadriceps tendon in an inverted “V” as well as incision of the retinaculum medially and laterally as far as the collateral ligaments. B) The knee is flexed and reduced and the VY advancement is completed by suturing the proximal aspect of the rectus tendon to the distal medial and lateral vastus tendons. Adapted from *J Child Orthop.* 2012. 6; 397-410.<sup>16</sup>

can cause limited range of motion.<sup>7</sup> PQT can also be performed in younger infants with limited anesthetic and decreased blood loss. However, extensor lag has been reported with PQT.<sup>13,15</sup> Currently, there is no consensus on which patients should have this procedure. Abdelaziz and Samir advocate for the use of PQT only in less severe GI or GII cases which have failed serial casting.<sup>8</sup>

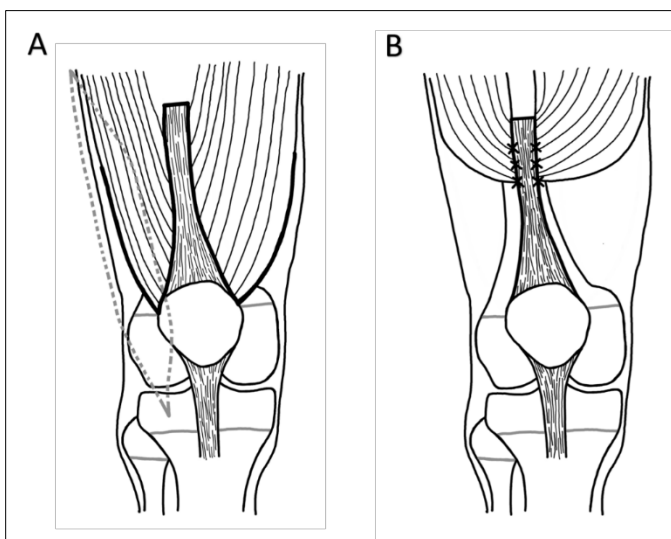
### VY Quadricepsplasty

VYQ was originally described by Curtis and Fisher in 1969 and involves a long anterolateral incision with division of the central portion of the quadriceps tendon in an inverted “V” to allow V-Y advancement (Figure 3).<sup>3</sup> An anterior arthrotomy, incision of the scarred suprapatellar pouch and tight anterior capsule as far as the collateral ligaments, and mobilization of the quadriceps allow for knee reduction and maximal flexion. Tercier et al. described a modification of the Curtis and Fisher VYQ, citing wound dehiscence of anteriorly placed incisions, insufficient lengthening of the quadriceps, and instability in the coronal plane following release of the retinacula up to the



**Figure 2.** The percutaneous quadriceps tenotomy (PQT) described by Roy and Crawford.<sup>12</sup> A) The three stab incisions proximal, medial, and lateral to the patella are illustrated. B) The lengthening that occurs in the rectus tendon and medial and lateral retinacula when the knee is flexed and reduced. Adapted from *J Child Orthop.* 2012. 6; 397-410.<sup>16</sup>

collateral ligaments as weaknesses of the original technique.<sup>16</sup> They describe a lateral incision from the mid-thigh to the knee. The rectus femoris tendon is separated from the vastus medialis and lateralis and the rectus is divided transversely at the musculotendinous junction. The rectus and patella are then reflected distally, and the knee is reduced. The medial and lateral retinacula are not released. The knee is extended and the distal limbs of the vastus medialis and lateralis are sutured to the proximal aspect of the divided rectus tendon (Figure 4).



**Figure 4.** The VY quadricepsplasty (VYQ) described by Tercier et al.<sup>16</sup> A) A lateral incision is used and the rectus femoris tendon is separated from the vastus medialis and lateralis. The rectus is divided transversely at the musculotendinous junction. B) The rectus and patella are then reflected distally, and the knee is reduced. The medial and lateral retinacula are not released. The knee is extended and the distal limbs of the vastus medialis and lateralis are sutured to the proximal aspect of the divided rectus tendon. Adapted from *J Child Orthop.* 2012. 6; 397-410.<sup>16</sup>

Advantages of the VYQ include addressing the primary pathology in congenital knee dislocation which is thought to be shortening and fibrosis of the quadriceps extensor mechanism.<sup>2,7,11,16-18</sup> Disadvantages include reports of wound dehiscence, extensor lag, extensor weakness, and flexion contracture.<sup>19</sup>

## Femoral Shortening

Femoral shortening has been proposed to effectively elongate the quadriceps and facilitate reduction of the knee without causing extensive scarring of the quadriceps. This procedure has been described by Johnston et al. and Sud et al. and has been used both in the management of isolated CDK in addition to simultaneous reduction of CDK and congenital dislocation of the hip (CDH).<sup>19-21</sup> Sud et al. describe a femoral osteotomy in the distal meta-diaphyseal region using a 5-cm longitudinal incision over the lateral aspect of the thigh. An average shortening of 2.35 cm is performed based on the femoral overlap that occurs when the knee is reduced. The fragments are then fixed with a single K-wire and the end is left buried under the skin. The patient is placed into a spica cast for 6–9 weeks depending on callous formation. The spica and K-wire are then removed after radiographs demonstrate healing at the osteotomy site. Johnston et al. use a 4-hole plate as opposed to a K-wire for fixation.<sup>19,20</sup> The femoral shortening osteotomy may also be performed proximally in the subtrochanteric or proximal diaphyseal regions if accompanying open reduction of the hip (Johnston).

Potential advantages of FS over VYQ include minimal damage to and scarring of the quadriceps which may decrease extensor lag, extensor weakness, and flexion contracture.<sup>21</sup> Additionally, FS may also facilitate simultaneous reduction of an ipsilateral CDH. Disadvantages include need for likely a second procedure for removal of hardware.

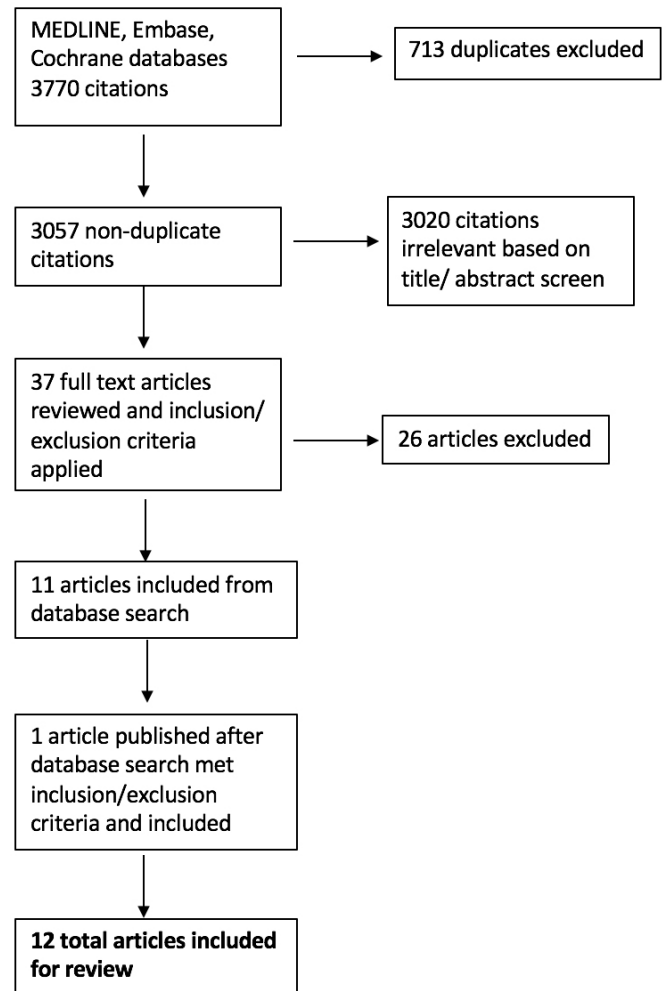
## Literature Review

While this report was not intended to be a formal systematic review given the limitations of the mostly small case series published on this rare disorder as well as variations in reported outcomes of CDK, a comprehensive literature review was performed to evaluate the reported outcomes of the above surgeries to address CDK. Studies were included that reported surgical outcomes for

CDK with a minimum of 1-year follow-up. Studies were excluded that reported on fewer than five knees that underwent surgical treatment, had less than 1-year follow-up, did not report clinical outcomes, or were not PubMed indexed.

In January 2020, we used Covidence to search MEDLINE, Embase, and Cochrane databases for all relevant studies (Figure 5). There was a total of 3770 citations returned, 713 of which were duplicates. The remaining 3057 studies were screened by two of the authors using title and abstract, and 3020 were found to be irrelevant. Thirty-seven full-text articles were reviewed and inclusion/exclusion criteria applied. Twenty-six of these articles were excluded, leaving 11 articles to be included in the study based on the initial search. Additionally, one article which met inclusion/exclusion criteria was published in December 2020 after the initial search and was also included in this review.<sup>17</sup>

Detailed results of the literature review are shown in Tables 2-4 broken down by surgical treatment.



**Figure 5.** A study selection flow chart describes the process by which studies were screened, reviewed, and included.

**Table 2. Results After Percutaneous or Mini-Open Quadriceps Tenotomy (PQT) for CDK**

ROM: range of motion, US: ultrasound, GII: Grade II, GIII: Grade III, ACL: anterior cruciate ligament, VYQ: VY quadricepsplasty

Study	Study Level	Outcomes measured	Average age at surgery in months (range)	Average follow-up in years (range)	Number and grade of knees treated (if specified)	Summary of Outcomes	Complications
Abdelaziz and Samir 2011 <sup>8</sup>	IV	Excellent, good, fair, poor designations based on ROM, quadriceps power, instability, and pain	2 (1-4)	3 (2-4)	12 (11 GII, 1 GIII)	7 excellent, 4 good, 1 fair	1 failure (GIII) with VYQ performed 8 weeks later complicated by recurrent dislocation. Revision VYQ performed complicated by procurvatum and mild anterior subluxation of tibia
Patwardhan et al. 2015 <sup>14</sup>	IV	Knee evaluation score (based on pain, motion, function), extensor lag, stability, and knee flexion deformity. Integrity of quadriceps mechanism assessed using US	1 (0-2)	1 (1-3)	20 (GII)	- good in 9 patients, fair in 3 patients - ROM mean 0°-135° - no patient with extensor weakness or flexion deformity - 1 patient with instability due to ACL aplasia	None
Shah et al. 2009 <sup>13</sup>	IV	Excellent, good, fair, poor designations based on ROM, stability, and pain with movement	3	3 (1 - 6)	13 (GII or GIII)	- 8 excellent, 3 good, 2 fair - 2/13 knees clinical instability - 5/13 knees extensor lag 10°	-2/13 knees plastic deformation fractures during therapy -2/13 knees required additional VYQ for re-dislocation
Youssef 2017 <sup>15</sup>	IV	Excellent, good, fair, poor designations based on ROM, power, instability, and pain with movement	2 (2-4)	2 (1-3)	16 (GII or GIII)	- 6 excellent, 10 good - ROM mean 8.5°-110° - quad power 5/5 in all knees - extensor lag 10/13 children - 13/13 children ambulated, 7 without support/bracing - 6/16 with mild instability	1 knee with intraoperative distal femur greenstick fracture

**Table 3. Results After VY Quadricepsplasty for CDK**

ROM: range of motion, GII: Grade II, GIII: Grade III, FS: femoral shortening, NR: Not recorded

Study	Study Level	Outcomes measured	Average age at surgery in months (range)	Average follow-up in years (range)	Number and grade of knees treated (if specified)	Summary of Outcomes	Complications
Abdelaziz and Samir 2011 <sup>8</sup>	IV	Excellent, good, fair, poor designations based on ROM, quadriceps power, instability, and pain	4 (1-11)	4 (1-6)	4 (GIII)	3 good, 1 fair	- 3 deep infections and wound breakdown - 2 genu valgum - 1 recurrent dislocation
Bell et al. 1987 <sup>11</sup>	IV	ROM, stability, quadriceps power, and function	9 (2-17)	3 (1-17)	9 (GIII)	- All patients able to walk - Flex: 96° +/- 37° - Flexion >75° in 8/9 knees - Extensor lag in 7/9 knees, average 18° +/- 9° - quad power ≥ 4/5 in all knees - 1 patient requiring B/L quad reefing due to extensor lag	1 skin necrosis and breakdown
Ferris and Aichroth 1987 <sup>7</sup>	IV	Excellent, good, fair, poor designations based on ROM, stability, and pain	6 (1-15)	9 (2-15)	7 (4 GII, 3 GIII)	-GII knees with 2 excellent and 2 good results - GIII knees with 1 excellent and 2 good results - ROM mean -3°-100° -all knees flex to at least 90°	NR
Johnston 2011 <sup>20</sup>	III	Excellent, good, fair, poor designations based on radiographic congruity, stability, ROM, and function/gait	9 (4-11)	11 (8-15)	6 (GIII)	2 fair, 4 poor	None
Oetgen et al. 2010 <sup>19</sup>	III	stability, ROM, PROMs (Lysholm and PODCI), gait analysis	7	11	6 (GII or III)	- ROM mean -2.5°- 104° - 1 with extensor lag - 5 with clinical instability -mean Lysholm 76 - PODCI comparable to normal knees except for "sports and physical functioning" domain, which CKD knees scored lower -less flexion instance, less peak knee flexion during swing than FS group	NR

**Table 3. Results After VY Quadricepsplasty for CDK (continued)**

ROM: range of motion, GII: Grade II, GIII: Grade III, FS: femoral shortening, NR: Not recorded

Study	Study Level	Outcomes measured	Average age at surgery in months (range)	Average follow-up in years (range)	Number and grade of knees treated (if specified)	Summary of Outcomes	Complications
Raj et al. 2020 <sup>17</sup>	IV	ROM, stability, quadriceps power, and function	8 (6 - 16)	7 (3 - 16)	45 (Grade NR)	<ul style="list-style-type: none"> <li>- ROM mean 101°</li> <li>- mean quad power 4/5</li> <li>- mean knee extensor lag 4°</li> <li>- 5/45 knees with clinical instability</li> <li>- 8 patients require braces for ambulation</li> <li>- 22 community ambulators, 2 household ambulators</li> </ul>	None
Sud et al. 2009 <sup>18</sup>	IV	Excellent, good, fair, poor designations based on ROM, stability, pain with movement, and ability to ambulate without support. Deductions were made for presence of extensor lag or flexion contracture	10 (4-26)	6 (3-9)	17 (2 GII, 15 GIII)	<ul style="list-style-type: none"> <li>- 4 excellent, 5 good, 10 fair</li> <li>- ROM mean 8°-122°</li> <li>- 16/17 able to walk independently, 1 required brace</li> <li>- 0/17 instability</li> <li>- all flex to 90°</li> <li>- 11/17 knees with extensor lag of 12° +/- 3°</li> <li>- 1/17 knee with flexion deformity of 15°</li> </ul>	NR
Tercier et al. 2012 <sup>16</sup>	IV	ROM, stability, quadriceps power, function, use of braces & Lysholm score (for select patients)	11 (5-26)	5 (2-9)	33 (3 GII, 30 GIII)	<ul style="list-style-type: none"> <li>- ROM mean 4°-104°</li> <li>- 12/33 knees with instability</li> <li>- 28/33 knees with extensor lag (mean 21°)</li> <li>- 13/33 knees with flexion deformity (mean 13°)</li> <li>- 18 community walkers, 2 household walkers, 7 used braces for ambulation</li> <li>- quad power <math>\geq</math>4 /5 in 28/33</li> <li>- Lysholm (n=13) mean 91+/- 8.6</li> </ul>	1 wound dehiscence

**Table 4. Results from Femoral Shortening for CDK**

ROM: range of motion, GII: Grade II, GIII: Grade III, LS: Larsen syndrome, ACL: anterior cruciate ligament, VYQ: VY quadricepsplasty, CDK: congenital dislocation of the knee, FS: femoral shortening, AMC: arthrogyriposis multiplex congenita, NR: Not recorded

Study	Study Level	Outcomes measured	Average age at surgery in months (range)	Average follow-up in years (range)	Number and grade of knees treated (if specified)	Summary of Outcomes	Complications
Johnston 2011 <sup>20</sup>	III	Excellent, good, fair, poor designations based on radiographic congruity, stability, ROM, and function/gait	17 (11-32)	12 (5-16)	5 (GIII)	3 excellent, 1 good, 1 fair	1 knee (LS) with proximal tibia growth arrest after ACL reconstruction performed simultaneously with FS
Oetgen et al. 2010 <sup>19</sup>	III	stability, ROM, PROMs (Lysholm and PODCI), gait analysis	25	12	3 (GII or III)	<ul style="list-style-type: none"> <li>- ROM mean 2.5°-122°</li> <li>- 1 with extensor lag</li> <li>- 2 with clinical instability</li> <li>- mean Lysholm 94</li> <li>- PODCI comparable to normal knees except for "sports and physical functioning" domain, which CDK knees scored lower</li> <li>- more flexion in stance, more peak knee flexion during swing than VYQ group</li> </ul>	NR
Sud et al. 2013 <sup>21</sup>	IV	Excellent, good, fair, poor designations based on ROM, stability, and pain with movement	6 (5 - 8)	4 (3-5)	10 (GIII)	<ul style="list-style-type: none"> <li>- 6 excellent, 4 fair</li> <li>- ROM mean -0.5°-107°</li> <li>- 3/10 mild instability</li> <li>- no extensor lag or flexion deformity</li> <li>- quad power ≥ 4/5 in all knees</li> <li>- no brace required for ambulation</li> <li>- all except 2 with AMC able to squat</li> </ul>	NR

## Summary of Literature Review

There is a paucity of data in the literature to support the use of one procedure over another. All of the studies were level III (2) or IV (10) and mostly consist of small case series. Most studies designated outcomes as “excellent, good, fair, or poor” based on range of motion (ROM), stability, and pain. Few studies reported patient reported outcomes. Functional outcomes were reported

for 61 total knees after PQT, 127 after VYQ, and 18 after FS in the combined included studies. Many patients in the included studies had additional deformities and/or syndromes associated with CDK. These included AMC in 41 patients, clubfoot in 30 patients, congenital dislocation of the hip (CDH) in 26 patients, Larsen syndrome (LS) in 12 patients, CVT in 11 patients, nonspecific foot

deformity in 10 patients, DDH in 10 patients, CV in 8 patients, Down syndrome in 2 patients, trisomy 8 in 1 patient, syndactyly in 1 patient, and achondroplasia in 1 patient.

Although many patients with CDK have other musculoskeletal deformities that undoubtedly influence functional outcome at final follow-up, the majority of patients treated for CDK are able to ambulate independently in the community regardless of procedure chosen. However, they do have limitations in sporting activities.<sup>19</sup>

PQT is a less extensive procedure that has been performed at an earlier age than FS and VYQ in these studies, with potential advantages of reduced blood loss and anesthesia time. Reported outcomes after PQT seem to be favorable; however, extensor lag was reported in two of four studies. As mentioned previously, Abdelaziz and Samir advocate for a PQT in GI and GII knees that are unable to achieve flexion beyond 90 degrees after serial casting.<sup>8</sup> They liken the PQT to the achilles tenotomy after serial clubfoot casting using the Ponseti method.<sup>8</sup> They do not recommend PQT in GIII knees as a result of a poor outcome observed in a patient with a GIII CDK who was treated with PQT and subsequently required VYQ which was complicated by recurrent dislocation.

VYQ and FS are more extensive procedures that are reserved for severe GIII cases of CDK which have failed conservative treatment. While the outcomes of FS were only reported in three studies, each of these studies reported satisfactory outcomes after FS for CDK.<sup>19-21</sup> In fact, in one of these studies the authors directly compared FS with VYQ.<sup>19</sup> While the outcomes were not statistically significant, the authors found that FS had a slightly higher Lysholm score, flexion, and total arc of motion than VYQ. However, the authors conclude that there is insufficient evidence to recommend FS over VYQ in the treatment of CDK.

## Conclusion/Summary

Based on this literature review, it is reasonable to consider PQT in treatment of GI and GII knees that have

failed serial casting. GIII knees which are dislocated with minimal passive flexion will likely require either VYQ or FS. We feel that both FS and VYQ can be considered for GIII CDK based on surgeon experience and preference. High-quality prospective studies comparing these treatments should be considered in the future. However, this may be unrealistic given the low incidence of CDK; thus, small case series will likely continue to guide management.

## Additional Link

POSNA Study Guide: <https://posna.org/Physician-Education/Study-Guide/Congenital-Knee-Dislocation>

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