Early Reduction for Congenital Dislocation of the Knee within Twenty-four Hours of Birth

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Background: Congenital dislocation of the knee (CDK) is a very rare condition that comprises a spectrum of deformities from subluxation to complete dislocation. The incidence of CDK is estimated at 1 per 100,000 live births, which is 1% of the incidence of developmental dysplasia of the hip (DDH). Moreover, 40–100% of patients with CDK have additional musculoskeletal anomalies, the most common being DDH and clubfoot. In general, the diagnosis is established immediately after birth according to the position of the knee recurvatum. Treatment with conservative methods at an early stage is most likely to yield successful results. We report here successful treatment of a series of CDK patients with early reduction.

Methods: From July 1990 to June 2007, 19 patients with CDK (affecting 25 knees) were treated with early reduction. Of these, 6 knees had dislocation, and 19 had subluxation. Since 1990, treatment has been guided by a protocol that considers patient age and the severity of the condition. In patients examined within 24 hours of birth, early, direct reduction under gentle, persistent manual traction was attempted. Birth history and perinatal course were obtained from medical records. Associated musculoskeletal anomalies were observed and treated after reduction of the knee joint. A Pavlik harness was used for at least 4 months in the concomitant treatment of DDH and CDK. Knee function was graded as excellent, good, fair, or poor. Radiographs were used to assess DDH during follow-up.

Results: After an average follow-up duration of 4.3 years, 18 patients showed an excellent or good outcome. One patient, whose knee could not be reduced, had severe multiple anomalies and died 16 days after birth. Fifteen of the nineteen patients had associated musculoskeletal anomalies, including DDH and foot deformity. Two cases of residual hip dysplasia after Pavlik harness application required an acetabular osteotomy.

Conclusions: For CDK patients, early and direct closed reduction within 24 hours of birth affords outcomes graded as either excellent or good.

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Key words: congenital dislocation of the knee (CDK); developmental hip dysplasia (DDH)

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Congenital dislocation of the knee (CDK) was first described by Chatelaine in 1822. It is a rare condition that comprises a spectrum of deformities from subluxation to complete dislocation.[1-10] The incidence of CDK is estimated at 1 per 100,000 live births, which is 1% of the incidence of developmental dysplasia of the hip (DDH).[2,11] Moreover, 40–100% of patients with CDK have additional musculoskeletal anomalies,[2-4] the most common being DDH and clubfoot.

Although there are some reports of occurrences within families, most cases of CDK are sporadic.[3-6] The deformity may be unilateral or bilateral, and predominantly affects girls. In general, the characteristic presentation of genu recurvatum facilitates the recognition of CDK (Fig. 1A).[7-11] A dimple or deep crease may be present over the anterior aspect of the knee. Many modalities can be utilized for conservative treatment, including serial casting, a Pavlik harness, skin traction, and skeletal traction.[1,3,6,7,11-14] Haga et al. advised waiting 1 month for spontaneous reduction of CDK in cases not associated with clubfoot or Larsen's syndrome.[12] However, Laurence claimed that the prognosis of CDK is adversely affected by a delay in treatment,[7] and by the presence of certain associated deformities or generalized joint laxity. This difference of opinion can make the treatment of CDK confusing. We report here on our experience in treating a series of CDK patients with early reduction, and their outcomes.

METHODS

We retrospectively reviewed 19 infants with CDK (affecting 25 knees) who had been treated with early reduction at our institution between July 1990 and June 2007. There were 5 boys and 14 girls. The birth history and perinatal course were obtained from medical records.

The severity of knee deformity was classified as either subluxation or dislocation according to the tibiofemoral articular relationship.[1,3,7] The diagnosis was dislocation if the tibia was displaced anterior to the long axis of the femur (Fig. 2A), and subluxation if longitudinal contact was at least partially maintained (Fig. 1B). There were 6 knees with disloca-

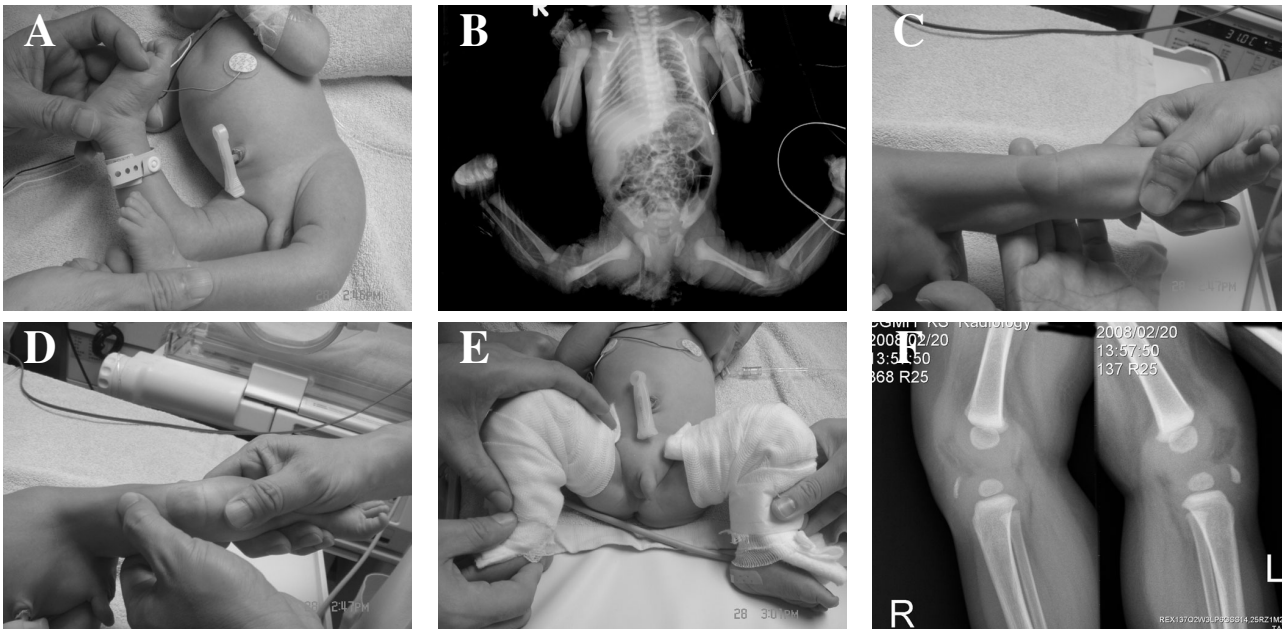

Fig. 1 Case 19 (A) The characteristic appearance of bilateral CDK at birth. (B) Radiograph before reduction demonstrating anterior subluxation in the bilateral knees. (C) The closed reduction starts with gentle, persistent manual traction. (D) During traction, an anteriorly directed force is applied to the distal femur, and a posteriorly directed force to the proximal tibia. (E) Reduction of the dislocated knee is achieved with flexion > 90°, and the extremity is immobilized with a splint. (F) Follow-up radiograph showing normal alignment of both knees at 9 months of age.

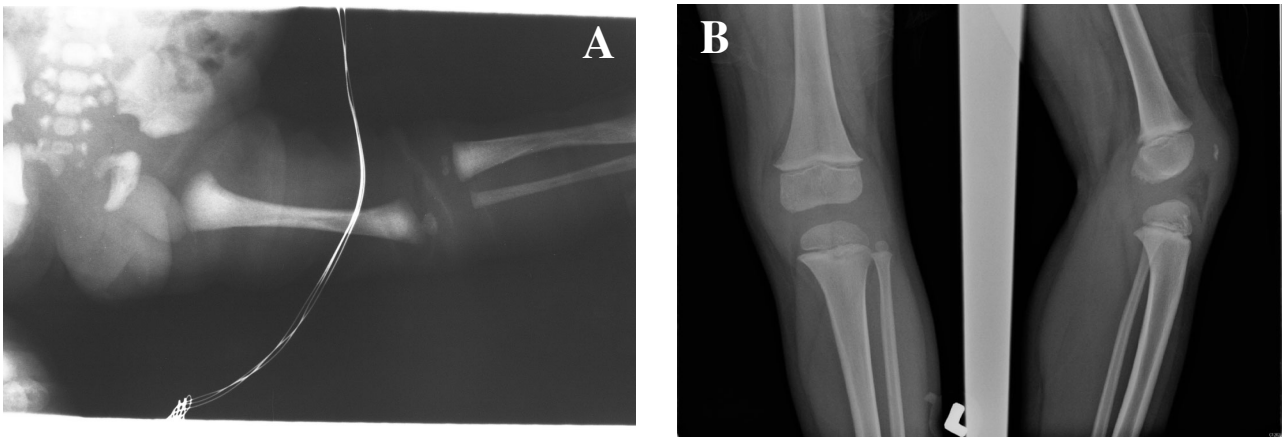

Fig. 2 Case 11 (A) Radiograph demonstrating anterior dislocation of the left knee. (B) Follow-up radiograph showing normal alignment of the left knee at 3 years of age.

tion, and 19 knees with subluxation. Of 6 patients with bilateral knee problems, bilateral knee subluxation was noted in 5, and bilateral knee dislocation in 1. Associated musculoskeletal anomalies were noted in 15 patients.

A general treatment protocol that considers patient age and condition severity was established by our senior author (J.Y. Ko).[1] For patients examined within 24 hours of birth, early and direct reduction under gentle, persistent manual traction was attempted (Fig. 1C). During this approach, an anteriorly directed force was applied to the distal femur, and a posteriorly directed force to the proximal tibia, in an effort to effect reduction of the dislocation or subluxation (Fig. 1D). Reduction of the dislocated knee was achieved with flexion > 90°. The extremity was then immobilized in a dorsal long leg splint or cast, with knee flexion > 90° (Fig. 1E). The reduction of CDK was confirmed following radiographic examination. Toe circulation was checked to avoid compartment syndrome. The casting or splinting was changed every 2 weeks over a 6 to 8-week period if there were no associated musculoskeletal anomalies. All patients were monitored for any unexpected sequelae or complications. If there were no associated anomalies, the patient was examined every 2 months until of walking age, and then examined annually. Radiographs of the knee were taken before and after reduction of the knee joint, every 4 months during the first year of follow-up, and then annually. No specific rehabilitation was administered unless there was an associated musculoskeletal anomaly that influenced motion or stability of the knee; this was the case for 1 patient with arthrogryposis multiplex congenita. Fourteen patients received concomitant treatment using a Pavlik harness for associated DDH; the treatment was initiated from the age of 6 weeks and was maintained for at least 4 months. The follow-up period ranged from 9 months to 14 years (average, 4.3 years).

Knee function was graded as (A) excellent, a full range of stable, pain-free movement; (B) good, flexion to ≥ 90°, slight instability or pain; (C) fair, flexion 45–90°, mild instability or pain; or (D) poor, flexion < 45°, gross instability, or severe pain.[1] Knee function, radiographs, and the outcomes for any associated anomalies were all assessed in the final follow-up session.

RESULTS

Pertinent data for each patient are noted in the Table 1. All 19 infants (25 knees) with CDK were treated with this method. There was a predominance of girls (14/19, 74%). Six patients had bilateral involvement, with bilateral knee subluxation in 5 patients and bilateral knee dislocation in 1. Breech presentation was noted in the perinatal history of 3 patients (patients 3, 6, and 8). One infant (patient 12) had the combined problems of premature birth, breech presentation, and premature rupture of the membranes.

Fifteen patients (79%) had associated musculoskeletal anomalies, including DDH, arthrogryposis,

Table 1. Clinical Data for Congenital Dislocation of the Knee

Case	Gender	Age at Treatment	Side	Classification	Associated Anomalies	ROM (°) Before (hyperext./flex.)	ROM (°) After (hyperext./flex.)	Pain	Instabiltity	Years of follow-up	Result
1	Female	6 h	L	Dislocation	–	80/0	0/130	–	–	11	Excellent
2	Male	6 h	L	Subluxation	–	45/5	0/125	–	–	4	Excellent
3	Female	8 h	L	Subluxation	R DDH	60/0	0/130	–	–	14	Excellent
4	Female	20 h	R	Subluxation	Bil DDH	50/5	10/140	–	–	7	Excellent
			L	Subluxation		50/0	0/140	–	slight		Good
5	Female	10 min	R	Dislocation	Bil DDH	90/-10	5/140	–	–	3	Excellent
			L	Dislocation		90/-10	5/140	–	–		Excellent
6	Male	2 h	R	Subluxation	AMC, Bil. clubfeet	60/0	0/135	–	–	8	Excellent
			L	Subluxation	Bil DDH*	60/0	0/135	–	–		Excellent
7	Female	5 h	R	Subluxation	Bil DDH	70/-5	0/140	–	–	8	Excellent
8	Female	1 h	R	Dislocation	Bil DDH, bil clubfeet, hydronephrosis, ASD, corpus callosus agenesis	80/-10	Failure				Expired 16 days later
9	Female	8 h	R	Subluxation	L DDH	60/20	0/135	–	–	2	Excellent
10	Female	4 h	R	Dislocation	Bil DDH	75/-5	0/120	–	–	2	Excellent
11	Male	16 h	L	Dislocation	Bil DDH, bil hindfoot valgus	80/-5	0/130	–	–	3	Excellent
12	Male	24 h	R	Subluxation	Bil DDH	60/10	5/125	–	–	4	Excellent
			L	Subluxation		70/10	10/120	–	–		Excellent
13	Female	2 h	L	Subluxation	R calcaneal valgus	50/5	0/140	–	–	4	Excellent
14	Female	24 h	R	Subluxation	L DDH*	70/5	5/135	–	–	3	Excellent
			L	Subluxation		80/0	10/120	–	–		Excellent
15	Female	24 h	L	Subluxation	–	60/10	0/140	–	–	1.5	Excellent
16	Female	24 h	L	Subluxation	L DDH	90/10	-10/130	–	–	2	Excellent
17	Female	24 h	L	Subluxation	L DDH	70/10	0/140	–	–	3	Excellent
18	Female	24 h	R	Subluxation	R DDH	50/5	0/140	–	–	1.5	Excellent
19	Male	21 h	R	Subluxation	–	80/10	0/130	–	–	9 months	Excellent
			L	Subluxation		60/10	0/140	–	–		Excellent

Abbreviations: R: right; L: left; Bil: bilateral; DDH: developmental dislocation of the hip; AMC: arthrogryposis multiplex congenita; hyperext./flex.: hyperextension/flexion; ROM: range of motion; *: Patients 6 and 14 underwent left acetabular osteotomy at age 3 and 2 years respectively.

and foot deformity. Twelve patients (16 knees) had a combination of ipsilateral CDK and DDH, and 2 patients had contralateral hip dysplasia. Three patients (patients 6, 8, and 11; 4 knees) had a combination of CDK, DDH, and foot deformities. Of the 6 patients with bilateral CDK, bilateral DDH was noted in 2 with knee subluxation, and in 1 with knee dislocation. One patient had unilateral DDH, and another with arthrogryposis multiplex congenita also had bilateral DDH and bilateral clubfeet.

The difficulty in achieving early reduction of CDK was found to be related to the duration between birth and treatment. Dislocated knees could be reduced in 5 minutes or less within 8 hours of birth (10 knees), while more than 20 minutes was needed for reduction in patients treated 20 hours after birth (7 knees). There was only one failure due to severe knee contracture in 1 patient (patient 8) who had severe multiple anomalies (hydronephrosis, atrial septal defect, and corpus callosus agenesis); this patient died 16 days after birth. The other 24 knees treated all had an excellent (22 knees) or good outcome (1 knee) as of the latest follow-up (average follow-up duration = 4.3 years). Knee function outcome was not influenced by the severity of deformity (subluxation or dislocation), laterality (unilateral or bilateral), or concomitant DDH. Gait and growing problems were not influenced by knee reduction in these patients. However, hindfoot valgus and residual talipes equinovarus were noted in 1 patient each. A Pavlik harness was used for at least 4 months in the concomitant treatment of DDH and CDK. Two of the 14 patients with associated DDH (2 hips, patients 6 and 14) had residual hip dysplasia (Fig. 3A), and underwent acetabular osteotomy at 3 and 2 years old, respectively. Good coverage and stability of all hips were obtained in the final follow-up session (Fig. 3B).

DISCUSSION

The incidence of CDK is estimated to be 1 per 100,000 live births, an occurrence 100 times less common than congenital dislocation of the hip.[11] There are numerous hypotheses concerning the etiology of CDK, and both intrinsic and extrinsic causes have been suggested. The intrinsic causes are genetic abnormalities, whereas the extrinsic causes are mechanical factors. In a review of 200 cases by Provenzano,[5] 7 families had a history of CDK. Mac Farland[4] reported a case of a family in which a mother and her three children from three different fathers all had CDK. Curtis and Fisher have described "heritable congenital tibio-femoral subluxation,"[3] a genetically transmitted syndrome where CDK is combined with some abnormalities of the face and spine. The familial occurrence suggests a possible genetic basis for CDK, whereas a non-genetic dysplasia etiology is supported by the sporadic occurrence of most cases. The latter etiology is more in keeping with the present series of patients, who all lacked a positive family history.

The extrinsic causes of CDK are considered to include a lack of amniotic fluid, lack of intrauterine space, malposition of the fetus, fibrotic contracture of the quadriceps, and traumatic dislocation during

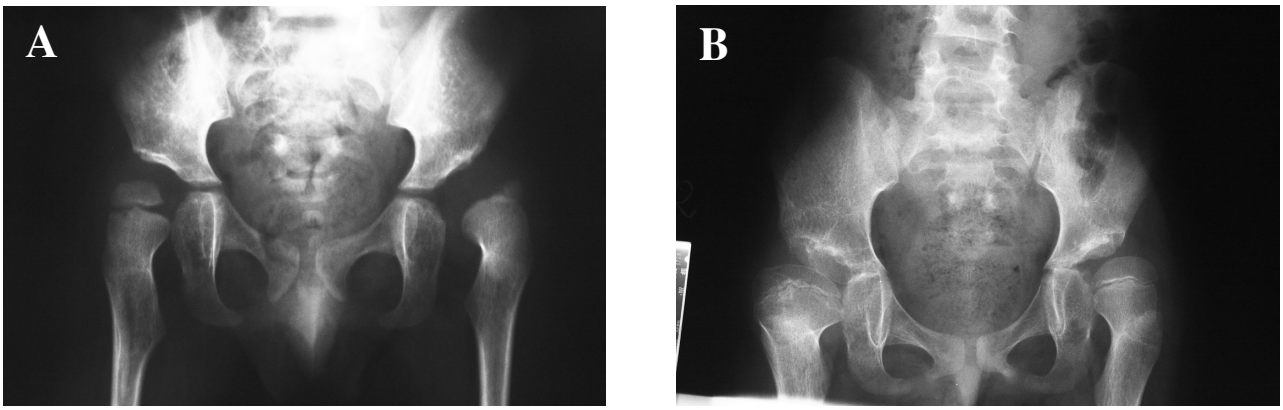

Fig. 3 Case 6 (A) Radiograph demonstrating residual dysplasia of the left hip. (B) Follow-up radiograph showing good coverage at 8 years of age.

birth.[3,6,8,10] Abnormal intrauterine positioning is implicated by observations of hyperextended knees being associated with a breech presentation. Fetal molding due to oligohydramnios or extended breech position was suggested as a cause by Shattuck, and supported by Niebauer and King.[8] The incidence of breech positioning at delivery was 21% in our patients. Other factors proposed as contributing to CDK include quadriceps contracture and hypoplasia of the anterior cruciate ligament.[6,10] Although many factors contribute to the development of CDK, a dislocation that cannot be treated by closed reduction within 24 hours of birth by our method may be considered teratogenic, and other congenital anomalies should be investigated in these cases.

There are a large number of associated musculoskeletal conditions in CDK, the most common of which are DDH, club foot, arthrogryposis, and Larsen's syndrome. In our series, 15 patients (78.9%) had associated musculoskeletal anomalies. Because the incidence of CDK is far less common than DDH, it is difficult to consider DDH as a causative factor of CDK. Rather, we propose that CDK contributes, at least in part, to the development of DDH, with the contractive quadriceps femoris muscle and dorsally displaced hamstrings potentially rendering the hip joint unstable. This is in agreement with the majority of studies that suggest that CDK should be treated first.[2,3,7]

The mainstay of treatment for CDK is early non-operative reduction. Many modalities have been utilized, including serial casting, Pavlik harness, skin traction, and skeletal traction. Haga et al. suggested waiting 1 month for spontaneous reduction of CDK not associated with clubfoot or Larsen's syndrome.[12] Mayer reviewed 68 patients, and found that treatment was successful in 81% of patients if performed before the age of 3 months, but only 33% if performed between the ages of 3 and 6 months.[13] Similar results were reported in a review of 10 knees by Stern.[9] Laurence suggested that treatment should be serial splinting for 2 weeks, followed by 2 months of traction if this fails.[7] We found that patients who responded to early closed treatment generally had normal knee function, without any complications, at follow-up. These results compare favorably to previous reports of knees reduced 2 days after birth.[1] In our previous report, traction followed by casting or use of a spica cast with or without general anesthesia was needed in all patients treated 2 days after birth. Serial casting was needed in patients treated from 24 to 48 hours after birth. Serial casting increased the complexity of reduction. The present report appears to be the first to recommend early, gentle reduction of CDK within 24 hours of birth. The difficulty in reducing CDK increases with the increase in the number of hours after birth. Indeed, reduction is especially easy in patients treated less than 8 hours after birth. In addition, delayed reduction may require traction or anesthesia, which increases hospitalization costs and surgical risks. When there are risk factors for CDK such as oligohydramnios and breach presentation and CDK is impressed on prenatal l ultrasonographic examination,[15,16] a paediatric orthopaedic surgeon should be consulted so that CDK can be reduced as soon as possible.

It has been reported that the Pavlik harness can simultaneously reduce both CDK and DDH.[14] If not treated early, the associated DDH may be resistant to closed reduction, and surgical reduction with or without acetabular osteotomy may be indicated for these patients. Two of our patients had residual hip dysplasia after concomitant treatment with a Pavlik harness for 4 months; this is consistent with other reports where treatment of DDH was not started early.

In conclusion, early reduction of CDK within 24 hours of birth affords excellent or good outcomes for all CDK patients not presenting with severe associated congenital anomalies (teratogenic CDK).

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出生二十四小時之內以早期整復治療先天性膝關節脫臼

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背 景： 先天性膝關節脫臼是一個非常罕見的疾病，比髖臼發育不良還少見一百倍。我們回溯性地探討在出生二十四小時內，利用早期整復來治療先天性膝關節脫臼，並評估其治療結果。

方 法： 在 1990 年 7 月至 2007 年 6 月間，共計十九位病人 (二十五個膝關節) 接受此種治療。評估膝關節功能包括活動角度，疼痛及穩定度。

結 果： 在平均 4.3 年的追蹤中，總共有十八位病人 (二十四個膝關節) 達到滿意結果，可以達到正常的膝關節功能，而且沒有併發症發生。

結 論： 對於先天性膝關節脫臼的患者在出生二十四小時內，利用早期整復可以成功矯正患者膝關節變形，並且長期追蹤結果可以達到正常的膝關節功能。

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關鍵詞： 先天性膝關節脫臼，髖臼發育不良

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