ORIGINAL ARTICLE

Functional Outcome of Open Relocation of the Congenital Dislocated Knee with and without Distal Arthrogryposis

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ABSTRACT

Objective: To evaluate the functional outcome of surgical relocation of congenital knee dislocation with or without arthrogryposis in age group over six months age.

Methods: A descriptive study conducted in Paediatric Orthopaedic Clinic at Jinnah Postgraduate Medical Centre Karachi from January 2004 to December 2012. The patients with congenital dislocated knees type III (complete dislocation), over the age of 6 months to 5 years, having isolated Knee dislocation, knee dislocations with Hip dislocation and clubfoot were included as group A. The knee dislocations as a part of arthrogrypotic contractures were included as group B. Patients with paralytic disorders, arthrogryposis unable to sit due to poor muscle functions were excluded. Open reduction of Knee through dorsal midline incision and Quadriceps V-Y plasty. Relocated Knees were stabilized with Plaster of Paris cast, in 70° flexion of knee for 5-6 weeks, with scoring system for knee functions described by Ferris and Aichron.

Results: Recurrent stiffness with poor results observed in 6 patients (12 knees) with group B, fair in 5 patients (10 knees) and none behaved good or excellent. In-group A (Knee dislocations with or without hip dislocation and clubfoot) results were Good to Excellent in 22 patients (36 knees), fair in 4 (7 knees) and poor with recurrence of severe stiffness in 1patient (7 knees).

Conclusion: The results of surgery were excellent in isolated congenital knee dislocations, satisfactory in dislocated knees associated with clubfoot and dislocated hips. The outcome was less promising in Arthrogryposis (Amyoplasia) and in older walking age groups.

Key Words: Arthrogryposis, Congenital, Dislocation, Isolated, Knee.

INTRODUCTION

The manipulation and serial casting is the treatment of choice recommended for infants born with Congenital Dislocated Knee (CDK), before the age of than 3 months age [1], and patients over 6 months age often require a percutaneous quadriceps tenotomy[2]. Whereas, after the age 6 months manipulative treatment is not usually recommended[3]. The forceful manipulation in later age group and in resistant cases like dislocation in Arthrogryposis Multiplex Congenital (AMC) leads to high risk of pressure necrosis of skin and separation of the cartilaginous epiphysis[3,4]. In these cases of CDK, the surgical reduction is the only way to way to achieve optimum results. The surgical relocation of CDK however, needs to be accomplished at least before commencement of walking[4], to avoid flattening of femoral condyle and subsequent poor results of surgery. The patient with multiple deformities and Syndromes are most resistant to treatment. They often require multiple secondary procedures and prolonged immobilization in casts /braces and thereby lowering the functional status of the limb[5,6,7]. The purpose of this study was therefore, to evaluate the functional outcome of surgical relocation of congenital knee dislocation with or without arthrogryposis in our circumstances. And to strategize the future directions for these two conditions to prevent adulthood disabilities.

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METHODS

This descriptive, case series study was carried out at the department of orthopedic surgery, Jinnah postgraduate medical centre, Karachi from January 2005 to December 2012. Non-probability purposive sampling technique was used for selection criteria of the patients of either sex, and age group over 6 months age to 5 years. The patients with isolated, Congenital

Dislocated Knee (CDK) of hyper extension type of Grade III severity (complete dislocation)[3,5], CDK grade III associated with Talipes Equino Varus (TEV) and / or Developmental Dysplastic Hip (DDH), without significant arthrogrypotic contractures were included as group A. Patients with CDK as a part of AMC were included as group B. Patients with paralytic disorders, hyper-laxity syndrome, CDK type I (Subluxation) and CDK Type II (Parital dislocation, where knee can be flexed to full range from 15^{o}hyper-extention deformity)[3,5] and arthrogrypotic patient with poor muscle function unable to sit, were excluded. Patients were registered after taking informed written consent, counseling with parents and approval from Institutional Review Board.

The surgical treatment included open reduction of Knee with Quadriceps V-Y plasty. In patients with associated DDH, TEV and Vertical Talus (VT), The TEV and VT were operated with CDK under same sitting. Whereas; DDH was relocated with surgery as the last one, 6-12 months after correction of CDK, TEV or VT. Surgical procedure of CDK included a midline incision starting from tibial tuberosity to mid-thigh. Quadriceps apponeurosis was separated from Muscles with inverted V incision, vastus medialis and lateralis were mobilized with taking care for neurovascular bundles on either side. Medial and lateral dissection were made to relocate anteriorly displaced hamstrings[1,5,8,9] back to their original position posterior to axis of knee flexion[3]. Supra-patellar transverse arthrotomy made, knee flexed to over 120 degree to visualize anterior cruciate. In some older age patients with gross knee valgus deformity, Iliotibial band was also released by maintaining its insertion on Gerdy'stubercle[3]. After securing haemostasis, quadriceps V-Y plasty re-suturing was made at 50^{0} flexion. Relocated Knees were stabilized with Plaster of Paris cast, applied in 70° flexion of knee for 5-6 weeks, followed by gentle mobilization exercise to achieve full flexion and extension and quadriceps strength. The weight bearing was allowed 8-12 weeks after operation when optimal strength and full ROM was achieved[8]. Radiographs of the knee were taken before and after reduction of the knee joint, every three months during the first year of follow-up and then annually. A predesigned proforma was used for each patient, to record demographic information, clinical findings, procedures performed and the outcome were recorded in predesigned Proforma on each visit. Clinic-radiological evaluation was made at minimum follow-up duration of one year, from findings in available proforma, clinical photographs and Xrays. The Scoring system for knee function described by Ferris and Aichroth[1] modified by Alock[5] and Tarek Hassan Abdelaziz[9] as referred in table1, was adopted to evaluate the results at last follow-up.

Table 1: Modified, Knee function scoring system[3,12,13]

Grade	Flexion (range)	Extension (power)	Instability	Pain
Excellent	Full	G5	Nil	Nil
Good	$>90^{0}$	>G3	Mild	Mild
Fair	$45\text{-}90^{0}$	G2	Mild	Mild
Poor	$<45^{0}$	<G2	Gross	Sever

Mild / Gross depends on whether the activities of daily living are affected or not. To qualify for certain grade, all four criteria need to be fulfilled. If any single criterion was not fulfilled, the result was downgraded according to this criterion.

RESULTS

Out of the 38 patients with 66 Congenital Dislocated Knees, 15 patients had isolated knee dislocations, 12 had knee dislocation associated with DD Hand TEV without arthrogrypotic contractures. 11 dislocated knees were part of arthrogryposis, two of them had Larsen's syndrome. The patients with unilateral dislocation, on other side had grade 1 or 2 CDK, correctable subluxations, that were stable on extension and nor requiring surgery. The age range of the patients at the time of surgery was 6 months to 4 years (Table 2).

Table 2: Deformity by age and laterality

Deformity		**No. of Patients**		**6-12 months age**		**13-24 months age**		**25-36 months age**		**>36 months age**	
		UL	**BL**	**UL**	**BL**	**UL**	**BL**	**UL**	**BL**	**UL**	**BL**
CDK	n=15	7	8	6	8	1	0	0	0	0	0
CDK+CDH	n=4	0	4	0	2	0	2	0	0	0	0
CDK+TEV	n=4	2	2	0	2	1	0	0	0	0	0
CDK+CDH+TEV	n=4	1	3	1	2	0	1	0	0	0	0
AMC:CDK, CDH+TEV	n=6	0	6	0	2	0		0	2	0	2
AMC:CDK, CDH+VT:	n=1	0	1	0	0	0	1	0	0	0	0
AMC:CDK:VT	n=4	0	4	0	0	0	1	1	1	0	2
Total	**n=38**	10	28	7	16	2	5	1	3	0	4

CDK = Congenital Dislocated Knee, CDH= Congenital Dislocated Hip, TEV= Telipes Equino Varus, VT= Vertical Talus, Mon= Months, UL = Unilateral, BL = Bilateral, Excel=Excellent

Table 3: Outcome of Open Relocation of CDK

Deformity		**No. of Patients**		**Laterality**	**Outcome**			
		UL	**BL**		**Excel**	**Good**	**Fair**	**Poor**
CDK	n=15	7	8	UL	3	2	1	1
				BL	4	2	2	0
CDK+CDH	n=4	0	4	UL	0	0	0	0
				BL	2	1	1	1
CDK+TEV	n=4	2	2	UL	2	0	0	0
				BL	2	0	0	0
CDK+CDH+TEV	n=4	1	3	UL	0	1	0	0
				BL	1	2	0	0
AMC:CDK, CDH+TEV	n=6	0	6	UL	0	0	0	0
				BL	0	0	3	3
AMC:CDK, CDH+VT:	n=1	0	1	UL	0	0	0	0
				BL	0	0	1	0
AMC:CDK:VT	n=4	0	4	UL	0	0	0	0
				BL	0	0	1	3
Total	n=38	10	28	UL	5	3	1	1
				BL	9	5	8	6

CDK = Congenital Dislocated Knee, CDH= Congenital Dislocated Hip,
TEV= Talipes Equino Varus, VT= Vertical Talus, Mon= Months,
UL = Unilateral, BL = Bilateral, Excel=Excellent.

Recurrent stiffness with poor results was observed in 6 patient (12 knees) with group B (arthrogrypotic patients), fair in 5 patient (10 knees) and none behaved good or excellent. Comparatively in the group A patients (Knee dislocations with or without hip dislocation and clubfoot) results were Good to Excellent in 22 patients (36 knees), fair in 4 (7 knees) and poor with recurrence of severe stiffness in 1 patient (1 knees). The reason of poor result in a patient with isolated knee dislocations was his failure to comply with directions to have continuous mobilization exercises after removal of cast (table 3).

The patients with fair results, were able to walk with support, squat with support, had some degree valgus deformity but no instability and 30^0 extension lag. The patients having good results of CDK surgery, who were also operated for associated DDH&TEV, had good range of movement insignificant extension lag, no valgus deformity or instability, able to squat easy and were able to walk independently. Whereas; the patients with excellent result of isolated CDK with or without surgeries for associated DDH and TEV, were normal in every parameter of assessment. They were pain free and enjoying normal life as other kids. Skin slough around incision occurred in two patients (3 knees) having AMC; one of that needed skin graft, other just needed multiple dressings and antibiotic cover. Patients with poor and fair results had significant Quadriceps wasting, whereas; patients with good results redeveloped a good Quadriceps mass.

DISCUSSION

Two types of congenital contracture are identified; one isolated and another multiple contracture commonly known as Arthrogryposis Multiplex Congenital (AMC). Among isolated contractures the isolated congenital dislocated knee is the most rare one[7]. Prevalence reported for isolated CDK as a part of AMC is 1:3000 live births[5,7]. Generally the CDK exhibit a strong familial relationship[5,10], with genetic abnormalities (trisomy), that is significantly influenced with mechanical and environmental factors; which include decreased fetal movements and uterine crowding[7,11]. However, in isolated CKD the principal factors are mechanical, traumatic dislocation during parturition and primary contracture of Quadriceps muscle and to some extent genetic abnormalities[9,12]. Clinico-radiologicaly three grades of CDK are defined according to severity. Grade III have complete dislocation, tibia is displaced anteriorly to long axis of femur, and there is no articular contact. The lateral view radiograph shows tibial plateau severely inclined posteriorly up to 35° (Normal adult 10°), and inter-condylar X-ray projection show markedly reduced height of intercondylar notch, rounded femoral condyles compared to normal, which is elliptical with horizontal diameter more than vertical diameter[–6]. Grade III is the least common in occurrence (20%). In grade II knee is subluxation, longitudinal articular contact between tibia and femur is at least partially maintained. Knee is in hyperextension of 45^0, however it can be passively flexed to neutral. Grade II is less common deformity (30%). Whereas, in Grade I there is hyperextension deformity with maintained longitudinal relationship, joint can be passively flexed to 45^o-90^0. Grade I is the most common (50%)[3,8]. The study includes type III dislocation only. The patho-anatomic changes in type III, CDK include: shortening of the extensor apparatus, tight anterior articular capsule[4], displacement of hamstrings anterior to the axis of knee flexion[2] and abnormal hypoplastic anterior cruciate ligament in some cases[5]. The fibrotic quadriceps femoris does not allow any flexion rendering a highly unstable and useless knee joint[5,6]. We found tiny patella, hardly palpable through skin, atrophic, shortened anterior cruciate particularly in older age group. The menisci were intact and hamstrings were displaced anteriorly, blended with vasti, needed meticulous dissection but no lengthening of Iliotibialtract[9] iliotibial lengthening in this study was required in CDK of AMC group, where patients presents over age two years and have sever muscle contracture. Similar to observation of Ali Al Kaissi[3], the other common finding in older age group with isolated CDK was flattening of lateral condyle[1]. Over to that, the extensive extra articular release, quadriceps lengthening, and intraarticular releases, leads to incomplete quadriceps mechanism, extensor weakness, instability, persistent valgus deformity and stifness[1] and poor ambulatory function. This was also reported by T. H. Abdelaziz[9] and Ferris[1].In older age group of CDk without AMC, to avoid quadriceps lengthening and extensive dissection, many investigators[9,12,13] have recommended the femoral shortening and patelo-femoral re-alignment with lateral retinaculum release[3], to accomplish the reduction and flexion of knee and to achieve the optimum functional knees. Fractional lengthening of hamstrings in grade III CDK to overcome occurrence of

extensor lag has been another procedure recommended by Tachdjian[14]. We have not yet tried this strategy in our cases. Persistent valgus knee, residual / recurrent contracture and restricted function requiring an assistance for daily living activities was the common behavior in our cases of CDK with AMC. For these cases of AMC Michael Bamshad[7] and others[9,12], therefore sets the objective of initial treatment as to mobilize the joints, achieve improved position and function, with the long term objective would be of increased joint mobility, muscle strength and development of adoptive use pattern, that allow an independence in walking ability and accomplishing daily living activitieswith or without assistance in the adulthood[7].Similar to Alock[3]the findings in table 3 in this study reflects excellent to good results in isolated CDK and in younger age group compared to good / fair results in CDK associated with DDH and TEV and poor in CDK associated with AMC and in older age groups. The residual flexion deformity and extensor lag was the common occurrence in syndromic (AMC) patients but very less in isolated CDK and non-syndromic patients. The shortened hamstrings and gastrocnemius in CDK associated with DDH and club feet respectively, that exerts strong flexor influence, has been cited as the main dynamic factors behind hindrance in development of quadriceps power, extensor lag and residual contractures in syndromiccases[4,5,15]. Ooishi et al[4] elaborates further, on scientific basis that the good hip position cannot be maintained as long as the knee is not sufficiently flexed, because the hamstrings are under continuous tension while the knee remains dislocated, and the femoral head is pulled superiorly and laterally[9]. We therefore, followed the recommendations of Alok[5], Ooishi[4], Abdelaziz[9] and Roth[15] correct CKD deformity 4-6 months before embarking on the relocation of DDH.

CONCLUSION

Good function to achieve in CDK therefore significantly relates to age at initiation of treatment, concomitant deformities or syndrome. The results of surgery were excellent in isolated congenital knee dislocations, satisfactory in dislocated knees associated with clubfoot and dislocated hips of nonarthrogrypotic patients. The outcome was less promising in Arthrogryposis (Amyoplasia) and in older age groups. The mobilization of hamstrings to natural position behind axis of the knee and post surgical care with continuous mobilization and quadriceps strengthening exercises over the long duration was the mainstay of treatment.

Open relocation earlier than walking age leads to best results without residual genu valgus.

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