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ORIGINAL ARTICLE

Knee flexion contracture in haemophilia: treatment with circular external fixator

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Summary. Haemophilia, a bleeding disorder, causes recurrent intra-articular bleeding of the joints resulting in chronic haemophilic arthropathy with fixed knee flexion deformity. Mid-long-term results (between 2002 and 2006) of deformity correction in haemophilic patients with Ilizarov type circular external fixators were retrospectively evaluated. There were six patients (five haemophilia A and one haemophilia B). The mean age was 14.7 years (range, 8–22 years) at the time of initial surgery. The mean knee flexion contracture was 45 degrees (range, 30–75 degrees). The mean arc of motion was 58.3 degrees (range, 40–100) before the surgery. The mean duration of follow-up was 8 years (range, 5.5–10 years). The mean duration of external fixation was 4.4 months (range, 2.5–10.5 months). Full extension of the knee joint was obtained in all patients in the early postoperative period. No bleeding, neurological or vascular complications were encountered. The mean amount of recurrence in knee flexion contracture was 10 degrees (range, 0–15 degrees). The amount of the correction was significant ($P = 0.0012$) and the mean arc of motion was 51.6 degrees (range, 25–90 degrees) that show a decrease of 6.7 degrees ($P = 0.04$) at the end of follow-up. The circular external fixator is an important, safe and less invasive alternative surgical treatment modality with low recurrence rate. Using the external hinges and distraction during the correction has a protective effect on the joint. It requires a team-work consisting of a haematologist, an orthopaedic surgeon and a physical therapist.

Keywords: external fixator, haemophilia, knee flexion contracture

Introduction

Haemophilia is a genetic bleeding disorder consisting of a partial or total absence of clotting factor VIII or factor IX. Its distribution is uniform in the world [1,2]. Articular contractures in patients with haemophilia are the result of recurrent intra-articular and intra-muscular bleeding episodes. Recurrent intra-articular bleeding of the knee joint in haemophiliacs often results in chronic haemophilic arthropathy with localized pain, chronic swelling, soft tissue contracture, muscular imbalance, cartilage and bone destruction which in turn could lead to a fixed flexion contracture of the knee because of destructive arthropathy [1]. Approximately, 50% of severe haemophiliacs have articular contractures of more than 10 degrees. These contractures can be disabling, causing decreased mobility and functional impairment [3]. In developed countries, orthopaedic problems of haemophiliacs have decreased after the introduction of continuous prophylaxis from ages of 2 to 18 years [4]. Primary prophylaxis that provides the coagulation factor level over 1% for several years [5], can transform a severe form of haemophilia into a moderate one. However, the tremendous cost of prophylaxis prevents 70–80% of the haemophiliac world population to access to such prophylaxis [4].

Conservative treatment methods, which include serial casting [6], reversed dynamic sling [7,8] and extension de-subluxation hinges [9] should be tried first to correct the deformity of the knee in the early stages. We advise the use of an extension brace (Fig. 1) for mild and moderate flexion contractures. However, in severe contractures where flexion deformity is more than 30 degrees, these measures may not suffice and corrective surgical procedures are required [1,6–9]. Posterior soft tissue procedures, supracondylar extension osteotomy, correction of contractures during total knee arthroplasty and mechanical distrac-

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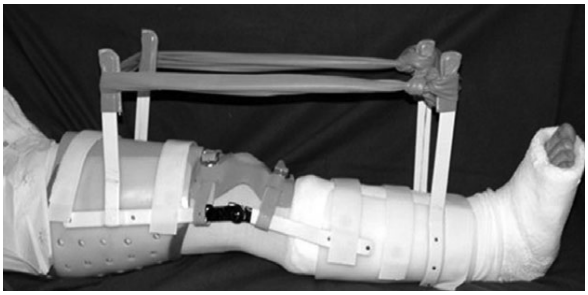

Fig. 1. Extension brace that used for mild flexion contracture.

tion using external fixators are treatment options and the appropriate procedure should be chosen according to the severity of flexion contracture on the knee [3,10]. There is no single operative treatment modality that is accepted worldwide. Circular type external fixator and gradual correction of the fixed knee flexion contracture was described in a case report for haemophiliacs [11].

In our study, we retrospectively evaluated mid-long-term results of fixed knee flexion contractures of haemophiliac patients that were corrected with circular type external fixators. We evaluated early and late results of flexion contractures and range of motion on the knee.

Material and methods

Six patients (five patients with Haemophilia A – one patient with Haemophilia B) with knee flexion contracture underwent surgical treatment between 2002 and 2006. The mean age was 14.7 (range, 8–22) years at the time of surgery. The mean flexion contracture was 45 degrees (range, 30–75 degrees). The mean arc of motion was 58.3 degrees (range, 40–90 degrees) before the surgery (Table 1). In all patients, posterior soft tissue including posterior capsular and posterior cruciate ligament release and gradual arthrodiastasis with correction of flexion deformity on the knee joint using an Ilizarov type circular external fixator were performed. Posterior soft tissue release includes biceps tendon lengthening, release of gastrocnemius head, Z-lengthening of medial flexor tendons, dorsal capsule and posterior cruciate ligament release. The circular external fixator is prepared with two rings above and two rings below the knee joint and external hinges at the intersection of anterior cortices on lateral view with maximum extension (Fig. 2). Motor unit of the system was on the posterior aspect of the ring. The aim of using the external hinges and arthrodiastasis was to protect the articular surface of the joint from any high load developing on the knee cartilage during the correction. Acute correction was not performed during the operation. On the first postoperative day distraction was initiated with a rate of 1 mm per day. After 1 cm of distraction, correction of the flexion deformity was initiated with a 1 mm in (1 mm per day) four times per day distraction at the posterior neurovascular structures at risk, which is the sciatic nerve [12]. Safe distraction for neurovascular structures was carried out in accordance with the equilateral triangular rule calculations [12] (Fig. 2). Patients were monitored by antero-posterior and lateral X-rays every week to rule out knee subluxation. At the end of the correction, the external fixator was fixed and maintained at full extension of the knee joint for at least as long as the correction time (Fig. 3). An over-correction was not carried out for haemophilic patients. After removal of the external fixator under general anaesthesia, we applied a long leg cast with a mean duration of 43 days to prevent rebound phenomena that can be seen after correction of severe flexion deformities. During the external fixator and casting period, physical therapy was not utilized as active or passive range of motion exercises could easily lead to bleeding episode following the initial distraction. Only after removal of the cast, physiotherapy was started and a dynamic knee extension brace was applied (Fig. 4).

Blood factor replacement therapy was initiated preoperatively, the dose was reduced daily and was stopped at the end of the physical therapy according to the advice of the haematology department. The serum factor level of the patients was maintained at 80–100% during the perioperative period and was decreased to 20% during the physical therapy. We did not have a bleeding problem from the irritation of the pins. Patients were followed up every 6 months after the completion of physical therapy with X-rays and physical examination.

Paired T test is used to evaluate the parametric data before and after the surgery. A P value <0.05 is considered significant.

Table 1. Demographics of patients with external fixator time, initial and last follow up knee flexion contracture, maximum knee flexion and arc of motion of the knee.

					Preoperative			Last follow-up		
No	Age	Follow-up Years	Haemophilia Type	External Fixator time	Contracture	Max flexion	Arc of motion	Contracture	Max flexion	Arc of motion
1	11	10	A	2.5	30	130	100	7.5	95.0	90
2	13	8	A	10.5	75	120	45	15	55.0	40
3	14	9	B	4	45	80	35	12.5	37.5	25
4	22	8	A	3.5	40	80	40	10	50.0	40
5	15	7	A	3.2	40	90	50	10	60.0	50
6	8	5.5	A	3	40	120	80	5	70.0	65

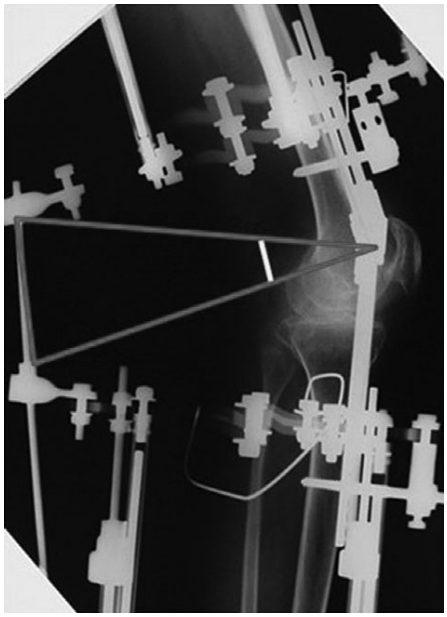

Fig. 2. Placement of the hinges of the Ilizarov frame are shown on this lateral X-ray. The rate of distraction is calculated according to the equilateral triangular rule (triangle in red, neurovascular structures at risk is shown in yellow, which is the sciatic nerve).

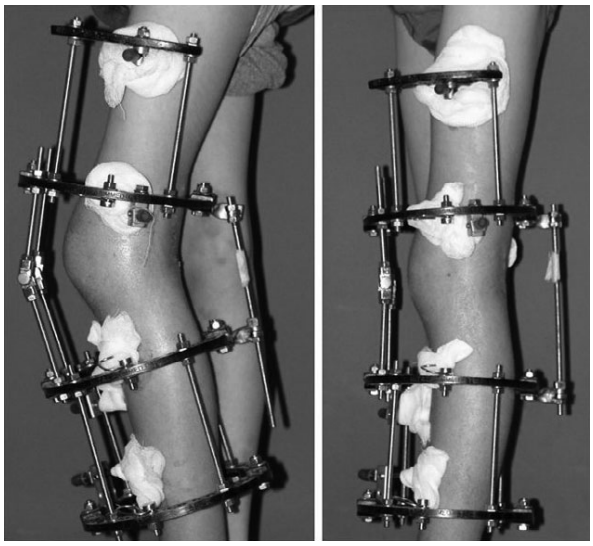

Fig. 3. Patient no 6, position of the leg during the correction and at the end of the correction.

Results

The mean duration of follow-up was 8 years (range, 5.5–10 years) (Table 2). The mean duration of external fixation was 4.4 months (range, 2.5–10.5 months) and the mean duration of physical therapy was 191 days (range, 60–364 days) in the postoperative period. Earlier during the follow-up, full extension was obtained in all patients. No major complications were encountered such as bleeding, neurological and vascular injury. There was one pin tract infection as a minor complication, treated with daily dressing and oral antibiotics. Subluxation of the knee, occurred in one patient during the correction of the flexion contracture, which was monitored with weekly X-rays and corrected with a minor change in the localization of the hinge on the circular frame.

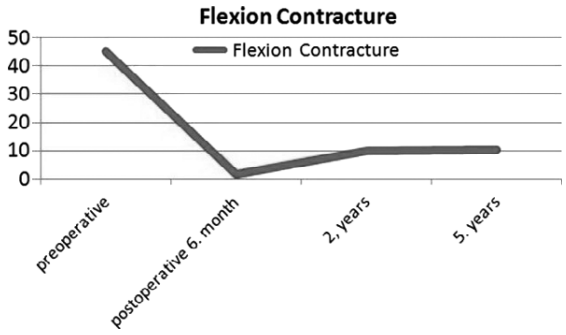

Fig. 4. The mean flexion contracture of the patients. Note that there is no increase in flexion contracture after 2 years of operation.

There was an average of 10 degrees (range, 5–15 degrees) of knee flexion contracture at the end of 8 years follow-up. It was significantly different from the preoperative flexion contracture ($P = 0.0012$). Most of the flexion contractures occurred within the first 2 years of the postoperative period (Table 2). In one patient, the flexion contracture continued to increase (an increase of 5 degrees was noted at the end of follow-up) (patient no 2 at Table 2). There was no change in recurrence of flexion contracture after 2 years of follow-up for the other five patients (Fig. 4). Mean arc of motion was 51.6 degrees (range, 25–90 degrees) at the end of 8 years follow-up (Table 1). According to the preoperative value there is significant decrease in arc of motion ($P = 0.04$). The mean recurrence of knee flexion contracture is provided in Table 2. A decrease of 10 degrees in arc of motion was observed in the postoperative period. However, the achieved extension of the knee joint provided the ability of walking in all patients (Figs. 5,6).There was no knee extensor strength loss after the physical therapy period. Although we had transected the posterior cruciate ligament, there were no unstable knees.

Discussion

As there are new factor prophylaxis regimens for haemophiliacs, we came across bleeding complications less frequently. On the other hand, as life expectancy increases, long-term complications must be overcome such as joint contractures. Delayed treatment of haem-

Table 2. Flexion contracture of the patients according to the years of follow-up

		Postoperative					
No	Preoperative	6 month	1 year	2 years	3 years	5 years	Last follow-up
1	30	0	5	7.5	7.5	7.5	7.5
2	75	0	5	10	12	15	15
3	45	0	10	12.5	12.5	12.5	12.5
4	40	0	5	10	10	10	10
5	40	0	5	10	10	10	10
6	40	0	0	5	5	5	5

Fig. 5. Patient no 6 at last follow-up clinical and radiologic view.

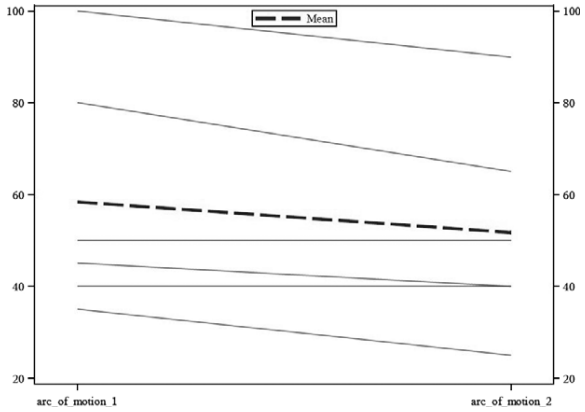

Fig. 6. Pre and postoperative arc of motions, there is a decrease at the end of treatment ($P = 0.04$).

arthrosis may lead to progressive joint contracture which results in re-bleeding episodes and progressive articular surface destruction and alteration in joint biomechanics [6]. The knee joint is more dramatically involved than any other joints in the lower extremity.

Treatment of knee joint contracture has positive long-term effects on the hip, lumbosacral region and the ankle joints [13]. Modern arthroplasty had provided an alternative to joint preservation surgery. However, with the increase in life expectancy of haemophiliacs, joint preservation surgeries have a greater importance now than in previous times [13].

Serial casting and extension de-subluxation devices result in significant improvement in the majority of cases in range of motion with restoration of physiological weight bearing, especially for the mild flexion contractures [6,13]. Hamstring release and dorsal capsulotomy have the advantage of an improvement in muscular imbalance, however, intensive physical therapy is needed to avoid instability [13]. In cases of moderate and severe deformities, soft tissue only procedures, with no further intervention, have a high recurrence rate, because of the nature of the disease.

Although supracondylar femoral osteotomies are an alternative option for the treatment of moderate to severe deformities, they do not change the range of motion of the knee, but they do change the arc of the motion. Soft tissue procedures and serial castings are inadequate for full recovery, especially for fixed flexion contractures that are greater than 30 degrees. Supracondylar extension osteotomy may provide an extended knee, but does not increase the range of motion of the knee [14]. Moreover, acute correction of the knee flexion deformity can cause neurovascular problems with traction injury to the nerves and vessels, and also skin necrosis leading to an increase in the contracture [15,16]. In our study the total arc of motion changed ($P = 0.04$). We think it is because of late start of physical therapy because of the risk of bleeding during the correction period. The use of external fixation is well known and safe in haemophiliacs especially for fracture healing and arthrodesis [16,17]. In our clinic, complex knee flexion contracture correction consists of tendon lengthening with capsular release, extension osteotomies of the distal

femur or gradual arthrodiastasis using circular external fixator with the Ilizarov technique.

Complete correction of the knee flexion contractures in arthrogryposis has been reported [14,18–20], however, at the long-term follow-up, rebound phenomenon and recurrence of the knee flexion contracture occurred. The recurrence of 10 degrees of flexion deformity on average in our series after mid-long-term survey is a good opportunity for the young patients whose arthroplasty operation can be delayed. To prevent the rebound phenomenon we could provide a hyperextension of 5–10 degrees during the external fixation period. Currently, we overcorrect our new patients about 10 degrees so that full extension is maintained even after loss of correction during the follow-up.

The main difference of our method from the previous ones is the use of an external hinge and distraction in the joint before the correction (Fig. 2). These measures protect the chondral surface of the joint and prevent chondropathy that can occur during the correction because of high compressive forces on cartilage tissue. Close survey of the patients is obligatory with this technique because an elongated joint becomes more prone to luxation. Mid-long-term follow-up of the patients in our series showed that recurrence in knee flexion contracture is most likely to occur during the first two postoperative years, beyond this period loss of extension remains uncommon.

We suggest treatment of knee flexion contractures of haemophiliac patients according to the severity of the disease. In fixed flexion contracture of the knee after the soft tissue procedures, gradual correction of the deformity with external hinge can be applied just with good cooperation of a team that involve a haematologist, a physical therapist and an orthopaedic surgeon.

Author Contribution

HIB, MK, IE and FB performed the research, designed the research study, contributed essential reagent and tools, and wrote the paper. HIB and FB analysed the data.

Disclosures

The authors stated that they had no interests which might be perceived as posing a conflict or bias.

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