



# Management of knee flexion contracture in haemophilia with the Ilizarov technique

Jiliang Zhai, Xisheng Weng\*, Baozhong Zhang, Huiming Peng, Yanyan Bian

Department of Orthopaedic Surgery, Peking Union Medical College Hospital, Chinese Academy of Medical Sciences and Peking Union Medical College, Beijing, China

## ARTICLE INFO

### Article history:

Received 17 September 2017  
Received in revised form 2 July 2018  
Accepted 14 August 2018

### Keywords:

Haemophilic arthropathy  
Haemophilia  
Ilizarov distraction  
Ilizarov technique  
Knee flexion contracture

## ABSTRACT

**Background:** There are limited reports about management of Knee flexion contracture (KFC) in haemophiliacs with Ilizarov technique. The aim of this study was to retrospectively analyzed the results of Ilizarov technique to treat KFC.

**Methods:** Six patients with haemophilia A and one with haemophilia B were included in this study, with an average age of 17 years old. The mean preoperative KFC and flexion angle of the knee were  $58 \pm 21^\circ$  (mean  $\pm$  standard deviation) and  $127 \pm 12^\circ$ , respectively. Preoperative HSS score (hospital for special surgery knee score) was  $51 \pm 4$ . The average time of follow-up was  $39.3 \pm 23.3$  months.

**Results:** All the patients achieved full correction of flexion contracture at the end of distraction and maintained at the last follow-up. The mean flexion angle at the end of distraction and at the last follow-up were  $41 \pm 35^\circ$  and  $38 \pm 19^\circ$  respectively, which were significantly lower than preoperative flexion angle. The mean HSS score at the end of distraction and at the last follow-up were  $65 \pm 4$  and  $64 \pm 2$  respectively, which were significantly higher than the preoperative HSS score. One patient suffered from transient numbness of left leg, and all the patients had loss of range of knee flexion at last follow-up.

**Conclusions:** Ilizarov is an effective and safe procedure to treat KFC in haemophiliacs. However, loss of knee flexion was the most common complication.

© 2018 Published by Elsevier B.V.

## 1. Introduction

Haemophilia is a genetic bleeding disorder with deficiency in clotting factor VIII or factor IX. Recurrent bleeding of the knee joint often induces chronic haemophilic arthropathy, which presents as pain, chronic swelling, soft tissue contracture, and cartilage and bone destruction [1–3]. Due to soft tissue contracture or cartilage and bone destruction, haemophilic arthropathy will induce a knee flexion contracture (KFC) [4]. A KFC is most common in children and young adults with haemophilia, and is frequent in developing countries [4]. As the severity of haemophilia increases, so does the ratio of KFC; about 50% of severe haemophiliacs have a ratio  $>10^\circ$  [2].

\* Corresponding author at: Department of Orthopaedic Surgery, Peking Union Medical College Hospital, Chinese Academy of Medical Sciences and Peking Union Medical College, Beijing 100730, China.

E-mail address: xshweng@medmail.com.cn (X. Weng).

If not treated quickly and effectively, KFC can cause severe disability and patients to use crutches or a wheelchair [4]. Many surgical procedures have been proposed to treat fixed KFC, including casting techniques, soft-tissue release, skeletal traction, and supracondylar extension osteotomy [5,6].

The Ilizarov technique is a skeletal traction method that slowly and steadily distracts the joint, and can reduce the risk of skin problems or neurovascular injury [7]. It has become an appealing alternative to open surgery [8]. However, there has been only one study describing the management of KFC in haemophilia with Ilizarov distraction [2]. The current study hypothesized that the Ilizarov technique can safely and effectively correct KFC in haemophiliacs. This study aimed to retrospectively analyse the results of the Ilizarov technique to treat KFC arising from soft-tissue contractures.

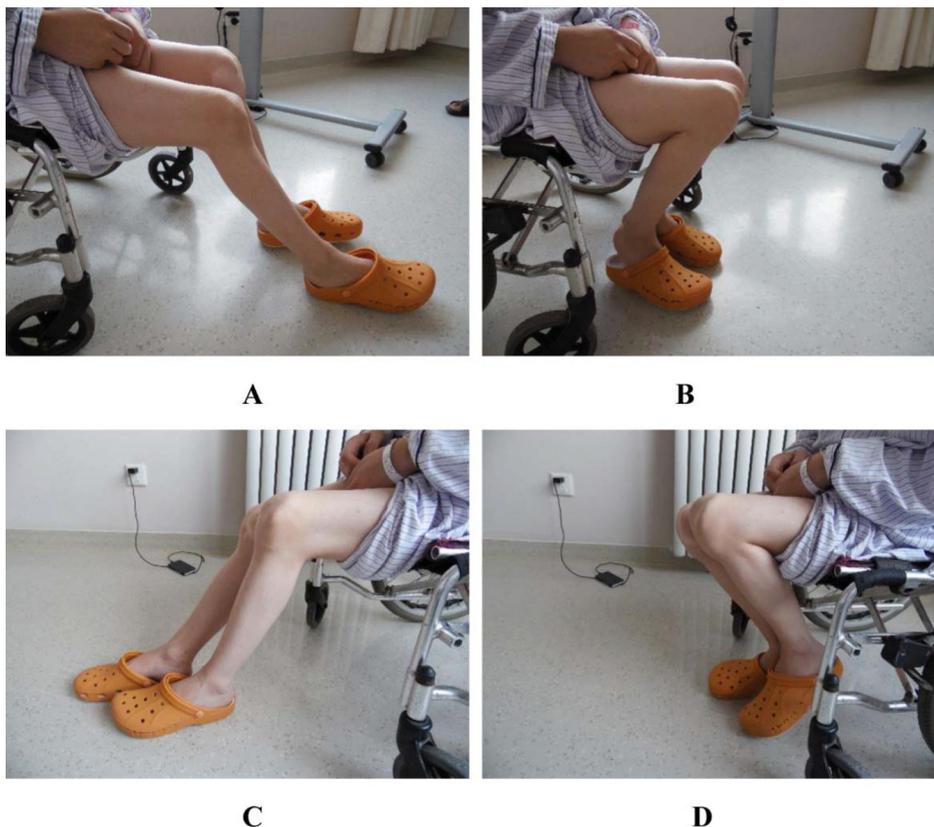
## 2. Materials and methods

Seven patients (10 knees) who underwent continuous distraction with the Ilizarov technique from December 2009 to September 2015 were retrospectively reviewed. There were six patients with haemophilia A and one with haemophilia B. Two of them were classified as moderate type ( $1\% < \text{factor VII:C or factor IX:C} < 5\%$ ), and the other five were severe type ( $\text{factor VII:C or factor IX:C} < 1\%$ ). None was positive for inhibitors against coagulation factors. Two patients were positive of hepatitis C.

The mean pre-operative flexion contracture was  $58 \pm 21^\circ$  (range 30–95). The mean pre-operative flexion angle was  $127 \pm 12^\circ$  (range 100–140). The mean pre-operative range of motion (ROM) of the knee joint was  $64 \pm 29^\circ$  (range 15–110) (Figure 1). The pre-operative Hospital for Special Surgery (HSS) knee score was  $51 \pm 4$  (range 44–60). The average age of these patients at surgery was  $17.0 \pm 4.8$  years (range 11–24). Three patients (Cases 1, 2, 4) underwent bilateral KFC distraction simultaneously. In addition to Ilizarov distraction, one patient (Case 7) underwent supracondylar osteotomy due to genu valgum. Another two patients (Cases 2, 4) had bilateral release of the iliotibial band. The average time of follow-up was  $39.3 \pm 23.3$  months (range 16–85). The patients' demographics are listed in Table 1.

### 2.1. Coagulation factor substitution protocol

Plasma factor VII and factor IX concentrations were measured pre-operatively, and a pharmacokinetics test was performed to determine the protocol for perioperative coagulation factor substitution. Factor VII (plasma-derived or recombinant) or activated prothrombin



**Figure 1.** Case 4 had bilateral knee flexion contractures and was unable to stand up. A–B: Extension and flexion of the right knee showed a 50–130° range of motion; C–D: Extension and flexion of the left knee showed a 60–130° range of motion.

**Table 1**  
Demographics of cases in the series.

Case	Age (years)	Side	Fixation time (days)	Follow-up time (months)	Pre-operative			Postoperative			Follow-up		
					Con	ROM	HSS	Con	ROM	HSS	Con	ROM	HSS
1	14	Left	42	85	80°	40°	47	0°	5°	64	0°	40°	65
		Right	42	85	95°	15°	44	0°	10°	61	0°	20°	62
2	17	Left	30	43	60°	80°	52	0°	30°	64	0°	30°	64
		Right	30	43	70°	60°	50	0°	40°	65	0°	30°	64
3	15	Left	60	42	30°	110°	60	0°	10°	61	0°	40°	65
4	24	Left	40	41	60°	70°	51	0°	35°	64	0°	30°	64
		Right	40	41	50°	80°	52	0°	20°	62	0°	30°	64
5	15	Right	21	31	45°	65°	50	0°	100°	72	0°	90°	71
6	23	Right	31	16	60°	80°	52	0°	100°	72	0°	30°	64
7	11	Right	60	17	30°	90°	53	0°	60°	67	0°	40°	65

Con, contracture; ROM, range of motion; HSS, Hospital for Special Surgery knee score.

complex concentrate (APCC) was infused as a single bolus for haemophilia A and B, respectively. The peak level of factor concentration was maintained at 80–100% on the day of surgery, 40–60% at postoperative days (PODs) 1–3, 20–30% at PODs 4–6, and seven to 15% thereafter until distraction instrument removal.



**A**



**B**

**Figure 2.** A: Ilizarov frames (red arrows show the distraction rods); B: Case 4 stood up with two crutches one week postoperatively.

## 2.2. Surgical and postoperative treatment

Both the femoral and tibial constructs consisted of one and a half rings, and each was fixed with two K wires. The femoral ring was located about 15 cm away from the groin and the half ring was fixed to the metaphyseal part of the lower end of the femur. The tibial ring and half ring were fixed about 15 cm above the ankle joint and 10 cm below the knee joint (almost at the level of tibial tubercle), respectively. Distraction was applied posteriorly by two rods (Figure 2A), and started from the third day postoperatively at a rate of 0.25 mm four times a day (one millimetre per day). The rate of distraction was modified according to the patient's compliance or whether neurological complications presented. Manual passive extension and flexion of the knee started once a day after unlocking the distraction rods. Local antiseptic was applied twice a day for pin care. Gait rehabilitation started one week postoperatively by attempting full weight bearing with two crutches (Figure 2B). When full extension was achieved, the fixation device was removed and intensive physical rehabilitation was conducted, at the local hospital or home, to restore ROM, muscle strength, proprioception and gait pattern. The signs of pin tract infection, peripheral circulation and sensation, and measurement of the knee flexion angle were observed every day.

## 2.3. Statistics analyses

One-sample Kolmogorov Smirnov Test was performed to test the data distribution. Paired *t*-test was used to evaluate the normally distributed parametric data before and after the surgery. Wilcoxon signed-rank test was applied when the data distributed unnormally.  $P < 0.05$  was considered significant.

## 3. Results

The mean duration of distraction was  $40.6 \pm 15$  days (range 21–60) and the average time of hospital stay was  $34.7 \pm 19.9$  days (range 17–78).

All the patients achieved full flexion contracture correction at the end of distraction and this was maintained at the last follow-up (Figure 3). However, the mean flexion angles at the end of distraction and at the last follow-up were  $41 \pm 35^\circ$  (range five to 100) and  $38 \pm 19^\circ$  (range 20–90), respectively, which were significantly lower than the pre-operative flexion angle ( $P < 0.001$ ). The ROM decreased from  $64 \pm 29^\circ$  to  $41 \pm 35^\circ$  at the end of distraction and  $38 \pm 19^\circ$  at the last follow-up, respectively. The mean HSS scores at the end of distraction and at the last follow-up were  $65 \pm 4$  (range 61–72) and  $64 \pm 2$  (range 62–71), respectively, which were significantly higher than the pre-operative HSS score ( $P < 0.001$ ). There were no significant differences in flexion angle and HSS score at the end of distraction and at the last follow-up ( $P = 0.754$  and  $P = 0.699$ , respectively). One patient suffered from transient numbness of the left leg, which vanished two days later after slowing the speed of distraction down (Case 6). No other complications occurred, such as bleeding, vascular injury or bone fracture.

## 4. Discussion

Recurrent bleeding and haematoma in haemophilia usually cause joint swelling and pain. Patients will adopt a flexed position to maximize the intra-articular cavity and alleviate pain, which leads to the development of soft-tissue contractures [3,9]. Soft-tissue contractures and secondary fixed KFC can cause decreased mobility and functional impairment [2,7]. Treatment of KFC is complex and can be very challenging [1,2,10]. Many procedures have been proposed to treat fixed KFC, including casting, soft-tissue release, skeletal traction, and supracondylar extension osteotomy. For the majority of patients in the early stages with mild flexion contractures, serial casting results in significant improvement of ROM [11,12]. However, surgical procedures are required for severe contractures with a flexion deformity  $>30^\circ$  [13]. Posterior soft tissue release, supracondylar extension osteotomy, and mechanical distraction with external fixators are optional; however, none have been accepted worldwide [5,6]. The appropriate procedure should be chosen according to the severity of flexion contracture and arthropathy of the knee [14,15].

The Ilizarov technique continuously and slowly distracts the knee joint, and decreases the complications related to open surgery such as neurovascular injury, bone non-union, internal fixation failure and deep infection [3]. However, there are still some potential complications such as pin tract infection, loss of knee flexion, recurrence of flexion contracture, and nerve injury [2,3,11,15]. In the current study, seven patients underwent continuous distraction with the Ilizarov technique and achieved full correction of flexion contractures at the end of distraction and at an average follow-up of 39.3 months. However, all the patients had loss of knee flexion and one patient suffered from transient numbness.

Pin tract infection is the most common complication with the Ilizarov technique, with a rate ranging from 5.12 to 100% [2,6,7,16–18]. No infection occurred in the present cohort. Compared to previous studies, patients in the current study had longer hospital stays, which means that they had longer regular medical care of the pin tract by the medical staff.

Loss of knee flexion is another common problem using the Ilizarov method. A decrease of  $6.7^\circ$  in ROM was observed in the postoperative period in the study by Balci et al. [2]. The loss of knee flexion showed a rapid fall during the latency period between the operation and start of distraction, due to physical constraints imposed by the fixator and insufficient analgesia, and continued during lengthening, which was due to increased passive tension exerted by the distracted muscles. Another factor influencing the final loss of knee flexion was the poor knee flexion during lengthening. There was a relationship between the ROM achieved during lengthening and the final loss of knee flexion [17]. Intensive physical therapy plays a key role in preserving joint ROM with Ilizarov distraction [17,19,20]. In order to prevent loss of knee flexion, the current study performed manual motion of the knee



A



B



C

**Figure 3.** A–C: Case 4 achieved full correction of knee flexion contractures, but loss of knee flexion at the last follow-up at 41 months.

joint once every day. At the last follow-up, all the patients received full correction of KFC. However, the mean knee flexion angle decreased from  $127^\circ$  pre-operatively to  $38^\circ$  at the last follow-up. It seems that passive motion of the knee joint in the current study did not prevent loss of knee flexion. The reason could have been short duration, low frequency, and less strength of manual passive motion. Flexing and extending the knee gradually using Ilizarov instrumentation at a faster rate than the original rate of distraction could preserve the original degree of knee flexion [6].

Recurrence of a flexion contracture is another complication of the Ilizarov technique at long-term follow-up [21–24]. Long-term follow-up has shown that recurrence of a knee flexion contracture is most likely to occur during the first two postoperative years [2]. To prevent this rebound phenomenon, hyperextension of five to  $10^\circ$  during the external fixation period is recommended so that full extension is maintained, even if recurrence of the flexion contracture occurred during the follow-up [2]. However, in the current study, patients had full correction without hyperextension and no flexion contracture recurrence occurred at an average follow-up of 39.3 months. The reason for the difference was unclear. Compared with the study by Balci et al. [2], the average age and degree of KFC were not significantly different. However, patients in the current study had lower degrees of knee flexion

during follow-up (38° vs. 51.6°), which may have been due to inadequate physical rehabilitation. It was also speculated that this may have been the reason for the difference between the two studies.

The acute nonoperative or operative treatment of severe flexion deformities is associated with relatively high risks of serious complications, especially neurovascular injury. Nerves are known to regenerate at one millimetre per day following axonotmesis [6]. The Ilizarov technique distracts the joint slowly and steadily at one millimetre per day and can reduce the risk of neurovascular complications [7]. One patient in the current study suffered from transient nerve injury and presented with leg numbness. The symptoms vanished after the speed of distraction was slowed down.

In conclusion, the patients in this study suffered from a loss of knee flexion at the last follow-up. However, they had full correction of KFC with the Ilizarov technique and the HSS score improved at the end of distraction and at the last follow-up. Pin tract infection did not occur in this study, which was common in previous studies. The study was limited by the nature of a retrospective review and small number of cases. However, studies on the Ilizarov technique in haemophilic patients are scarce, and this study had its strengths to support its conclusions.

### Conflict of interest

The authors declare that they have no competing interest. This paper has not been published previously, and is not under consideration for publication elsewhere.

### Funding

No funding was obtained for this study.

### References

- [1] Arnold WD, Hilgartner MW. Hemophilic arthropathy. Current concepts of pathogenesis and management. *J Bone Joint Surg Am* 1977;59:287–305.
- [2] Balci HI, Kocaoğlu M, Eralp L, Bilen FE. Knee flexion contracture in haemophilia: treatment with circular external fixator. *Haemophilia* 2014;20:879–83.
- [3] Kiely PD, McMahon C, Smith OP, Moore DP. The treatment of flexion contracture of the knee using the Ilizarov technique in a child with haemophilia B. *Haemophilia* 2003;9:336–9.
- [4] Daffunchio C, Caviglia H, Nassif J, Moretti N, Galatro G. Knee flexion contracture treated with botulinum toxin type A in patients with haemophilia (PWH). *Haemophilia* 2016;22:134–41.
- [5] Dunn AL. Pathophysiology, diagnosis and prevention of arthropathy in patients with haemophilia. *Haemophilia* 2011;17:571–8.
- [6] Hosny GA, Fadel M. Managing flexion knee deformity using a circular frame. *Clin Orthop Relat Res* 2008;466:2995–3002.
- [7] Gaurav K, Vilas J. A new approach to the management of fixed flexion deformity of the knee using Ilizarov's principle of distraction histogenesis: a preliminary communication. *Int J Low Extrem Wounds* 2010;9:70–3.
- [8] Jeong BO, Kim TY, Song WJ. Use of Ilizarov external fixation without soft tissue release to correct severe, rigid equinus deformity. *J Foot Ankle Surg* 2015;54:821–5.
- [9] Greene WB, McMillan CW, Warren MW. Prophylactic transfusion for hypertrophic synovitis in children with hemophilia. *Clin Orthop Relat Res* 1997;19–24.
- [10] Gamble JG, Vallier H, Rossi M, Glader B. Loss of elbow and wrist motion in hemophilia. *Clin Orthop Relat Res* 1996;94–101.
- [11] Fernandez-Palazzi F, Battistella LR. Non-operative treatment of flexion contracture of the knee in haemophilia. *Haemophilia* 1999;5(Suppl. 1):20–4.
- [12] Wallny T, Eickhoff HH, Raderschadt G, Brackmann HH. Hamstring release and posterior capsulotomy for fixed knee flexion contracture in haemophiliacs. *Haemophilia* 1999;5(Suppl. 1):25–7.
- [13] Kale JS, Ghosh K, Mohanty D, Pathare AV, Jijina F. Use of the dual force system to correct chronic knee deformities due to severe haemophilia. *Haemophilia* 2000;6:177–80.
- [14] Solimeno L, Goddard N, Pasta G, Mohanty S, Mortazavi S, Pacheco L, Sohail T, Luck J. Management of arthrofibrosis in haemophilic arthropathy. *Haemophilia* 2010;16(Suppl. 5):115–20.
- [15] Rodriguez-Merchan EC. Therapeutic options in the management of articular contractures in haemophiliacs. *Haemophilia* 1999;5(Suppl. 1):5–9.
- [16] Donnan LT, Gomes B, Donnan A, Harris C, Torode I, Heidt C. Ilizarov tibial lengthening in the skeletally immature patient. *Bone Joint J* 2016;98-B:1276–82.
- [17] Motmans R, Lammens J. Knee mobility in femoral lengthening using Ilizarov's method. *Acta Orthop Belg* 2008;74:184–9.
- [18] Ramos T, Karlsson J, Eriksson BI, Nistor L. Treatment of distal tibial fractures with the Ilizarov external fixator – a prospective observational study in 39 consecutive patients. *BMC Musculoskelet Disord* 2013;14:30.
- [19] Barker KL, Simpson AH, Lamb SE. Loss of knee range of motion in leg lengthening. *J Orthop Sports Phys Ther* 2001;31:238–46.
- [20] Coglianese DB, Herzenberg JE, Goulet JA. Physical therapy management of patients undergoing limb lengthening by distraction osteogenesis. *J Orthop Sports Phys Ther* 1993;17:124–32.
- [21] Herzenberg JE, Davis JR, Paley D, Bhave A. Mechanical distraction for treatment of severe knee flexion contractures. *Clin Orthop Relat Res* 1994:80–8.
- [22] Damsin JP, Ghanem I. Treatment of severe flexion deformity of the knee in children and adolescents using the Ilizarov technique. *J Bone Joint Surg Br* 1996;78:140–4.
- [23] Huang SC. Soft tissue contractures of the knee or ankle treated by the Ilizarov technique. High recurrence rate in 26 patients followed for 3–6 years. *Acta Orthop Scand* 1996;67:443–9.
- [24] Brunner R, Hefti F, Tgetgel JD. Arthrogyptic joint contracture at the knee and the foot: correction with a circular frame. *J Pediatr Orthop B* 1997;6:192–7.