The Treatment of Flexion Contracture of the Knee Joint in Hemophilic Patients Using Gradual Manipulation and Serial Casting

Ali Bakir Al-Hilli

ABSTRACT:

BACKGROUND :

Hemophilia is a bleeding disorder caused by deficiency of clotting factor VIII or IX; it is inherited as sex linked recessive pattern. Musculoskeletal complications in haemophilic patients are disabling. Bleeding in to the large joints and soft tissues are associated with arthritic changes in the joints and contractures in the soft tissues. Deformities of the large joints especially flexion contractures are commonly seen in knee joint. It is important to prevent deformities, treat existing contractures and prevent recurrence to improve the patient's quality of life.

OBJECTIVE:

This is a retrospective study analyzing the effectiveness of gradual manipulation and serial casting for treatment of knee joint flexion contracture in Hemophilic Iraqi patients.

PATIENTS AND METHOD:

We included ten Hemophilic patients type A, treated between 2006-2008, six with unilateral and four with bilateral knee joint flexion contractures, excluding patients with multiple joint contractures other than knees, have severe arthritic changes, have subluxated unstable knees or have acute bleeding. Their age is between 16-22 years, the duration of deformity is 4-9 months, knee flexion contractures between 20-80 degrees and all of them don't have significant pain but their daily life is severely restricted by the deformity. Under factor cover, general anaesthesia and haematological care, we started serial manipulations and plaster of Paris casting, the number of casts is between 2-6 casts, till we get straight knees. After that splints are prescribed for the patient and followed up for a minimum 6 months, physiotherapy to strengthen the muscles around knee is initiated from the start and gradually increasing after the application of the removable splints.

RESULTS:

We have ten patients with fourteen deformed knees, after manipulations and serial casting 8 knees get full correction, and 6 knees still have less than 10 degrees of flexion which is acceptable for the patients, all the patients have improvement of their quality of life and walk free of pain after 2 months.

CONCLUSION:

Serial manipulation and casting under general anaesthesia is a very good maneuver to correct knee joint flexion contractures in hemophilic patients.

KEY WORDS: hemophilia, knee flexion contracture, serial manipulation and casting.

INTRODUCTION:

Hemophilia is a life long condition characterized by factor VIII (Hemophilia A) or XI (Hemophilia B) deficiency , it is inherited by a sex linked recessive pattern[1] , with a high risks towards disability if not properly treated. Von Willebrand disease is a moderate bleeding disorder caused by quantitative or qualitative protein deficiency of von Willebrand factor and is inherited in an autosomal dominant or recessive manner[1,2] .Hemophilia A and B are classified as mild, moderate, or severe by the scientific and standardization committee of the international society on thrombosis and haemostasis[3].Severe hemophilia A is the most common form, occurring in 60% of patients, and indicated by active FVIII levels of less than 1% to 2% with serious bleeds occurring more than 20 times per year spontaneously, without trauma or injury[1,3], Moderate hemophilia is indicated by active FVIII levels of 2% to 5% and a lower, but variable, frequency of spontaneous bleeds, while mild hemophilia is indicated by active FVIII level of 5% to 25%[1]. Factor deficiency leads to bleeding

College of Medicine/ Baghdad University.

tendency and recurrent episodes of joint and soft tissue bleeding. Recurrent episodes of hemarthrosis causing severe limitation of joint function, and repeated intra-articular hemorrhage produce synovial membrane hyperplasia, hemosidrin deposition and fibrous scaring[4] ,the synovial tissue becomes hypervascular, villous, and friable, which in turn makes it more susceptible to bleeding[5] . Lack of good factor cover and poor cooperation between hematologist, orthopaedic surgeon and physiotherapist is the primary problem which leads to a long lasting suffering and disability to hemophilic patients.

Bleeding usually affects the large joints such as knee, ankle, elbow, shoulder and others, and with recurrent bleedings intra-articular adhesions result in reduction of the joint cavity and limitation of movement, in the knee the recurrent hemorrhages cause progressive atrophy of the quadriceps muscle and a fixed flexion contracture[4].

The primary goal of rehabilitation is to diminish the disabilities and prevent handicaps caused by impairments, physiotherapy aims at pain reduction by means of manual traction then intensive physiotherapy program including mobilization techniques, muscle strengthening exercises and stretching, joint stability training, postural and gait training and functional training[6].

Hemophilic contractures is seen most commonly as an equinus deformity of the ankle or at the knee or elbow in the form of a flexion deformity and treatment options are varied and decision making is based on the degree of contracture, its chronicity, the presence of articular subluxation, the patients ability to participate in the treatment and the available medical facilities[7].

The treatment options available for joint contractures are physiotherapy, orthotics, corrective devices and the late or severe cases may require surgical correction in the form of soft tissue procedures such as hamstring release of the flexion contracture of the knee or lower femoral osteotomy[7].

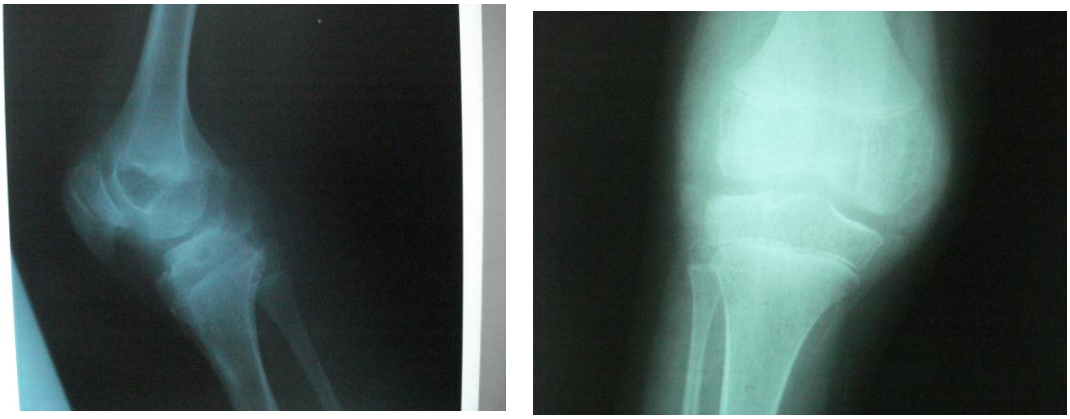
PATIENTS AND METHOD:

Ten Iraqi Hemophilic male patients were included in this study. All of them have hemophilia A, presented and treated during 2006-2008, Six with unilateral (three left side and three right side), and four with bilateral knee joint flexion contractures, a total of 14 knees (Picture 1 and 2). Exclusion criteria include patients with multiple joint contractures other than knees, have severe arthritic changes diagnosed by X-ray and clinical symptoms and signs, have subluxated unstable knees or have acute bleeding (picture 3).

Picture 1: Severe bilateral flexion contractures in a hemophilic patient.

Their age is between 16-22 years, the duration of the deformity is between 4-9 months, knee flexion contractures are between 20-80 degrees and all of them don't have significant pain but their daily life is severely restricted by the deformity.

Picture 2: Another patient with right knee flexion contracture.



Picture 3: a: Lateral and b: Anteroposterior, X-ray shows no arthritic changes.

Under haematological care, factor cover (up to minimum 40%), and general anaesthesia, we started serial manipulations and plaster of Paris casting, the manipulation is gentle to start with increasing gradually by repeated flexion and extension of the contracted knee, after manipulation a well padded plaster of Paris cylinder is applied from the groin till the malleolli or in severe contractures the foot is included in the cast, then the patient is encouraged and taught to do passive physiotherapy to strengthen the muscles around the knee, there is a special exercise program done under the supervision of the physiotherapist , in the same day the patient can discharge home to be seen two weeks later, the cast is opened for checking of the skin condition, in the same day another manipulation under general anaesthesia is done and followed up in the same way.

The number of casts is 2-6 casts, till we get a straight knee. After that a day walking splint and a night sleeping splint is prescribed for the patient and followed up for a minimum 6 months, physiotherapy to strengthen the muscles around knee is initiated from the start and gradually increasing after the application of the removable splint, The patient use crutches for the next month during walking then gradually increase his level of activity(picture 4 and 5).


Picture 4: A: Post manipulation POP cast. B: Post correction removable splintage.

RESULTS:

We have ten patients with fourteen deformed knees. The mean age is (16.6 years) (16-22years), mean duration of deformity is (6.4 months) (4-9 months), and the mean knee flexion contractures range (41.5 degrees) (20-80 degrees). The mean number of casts is 3 casts (2-6 casts), the time between each manipulation is 2 weeks. Physiotherapy to strengthen the muscles around knee is initiated from the start and gradually increasing after the application of the removable splints. After manipulations and serial casting eight knees get full correction, and six knees still have less than 10 degrees of flexion deformity which is acceptable for the patients, all the patients have improvement of their quality of life and walk free of pain after 2 months, a fellow up for six months show that our fourteen knees are doing well, free of pain and without recurrence of the deformity.


Picture 5: A: Patient with straight knees after 6 months. B: Walking free of pain with splint.

DISCUSSION:

Hemophilia is a hereditary haematological problem associated with factor VIII (hemophilia A) or factor XI(hemophilia B) deficiency, and a 75 % of hemophilic patients are living in the underdeveloped countries[8]. Von Willebrand is a moderate bleeding disorder with same clinical picture to haemophilia and treated in the same unit[1].

Treatment of hemophilic patients should be performed in a comprehensive basis with in a multidisciplinary hemophilic unit[9]. In 1960s factor replacement become reality and the challenge of the next century is to prevent any musculoskeletal complications from ever occurring[10]. Liability to bleed and prolonged bleeding make the patients prone to a number of musculoskeletal complications like joint contractures and muscle weakness, wasting and soft tissue psudotumours, it is in hemophilia that the orthopaedist is asked to straighten the bent limbs of the child[10].

Physiotherapy and rehabilitation is tried to improve the quality of life for the patients also to prevent impairments by strengthening, stretching

and gait and postural training[6]. It is essential that those with hemophilia are taught the importance of physical fitness at an early age as a means of preventing articular contractures[11,12,13,14].

It is important to know that once arthropathy develops the functional prognosis is poor although hemophilic patients use to tolerate very well their tremendous joint destruction[9].

Orthotics and corrective devices are also used as conservative measures[7] but in late or severe cases surgery may be the only option to get a straight, and pain free limbs, soft tissue releases or osteotomies may be used[7,13].

A study by F. Fernandez-Palazzi et al used serial casting and wedging in 58 patients and extension/ de-subluxation orthosis in 13 patients, shows both methods have been resulted in significant improvement in joint contractures by 4 weeks time[15]

The reversed dynamic slings for knee contractures had been used in 1975 by H.Stein et al arguing that serial casts is a time consuming and the ultimate degree of correction is incomplete and some times unsatisfactory[4].

The flowtron intermittent compression system has been used by P. Yates et al to treat knee joint contractures for hemophilics and published a prospective study in 1992 were no need to use prophylactic factor, and their results show that this at home method is safe effective and cheap[16] . The use of dual force system to correct chronic knee deformities due to severe hemophilia has been used and published by J.S.Kale et al, were 10 patients with long standing knee deformities included in their study and the result shows that the dual force system is easily affordable and effective[17]

Ilizarov technique has been used in a child with hemophilia B by P. D. Kiely et al to treat a knee joint contracture with on demand early replacement therapy, this technique shows reduction of the deformity from 50 degrees to 5 degrees in a 3 months time and after 4 months the boy walks freely with out pain and there was no peri or post operative bleeding or joint swelling[18].

In our patients we tried to use a simple method to address the problem of knee joint flexion contractures, using serial manipulations and casting under general anaesthesia, as the other methods are not familiar in our country. We have ten patients with fourteen deformed knees, the mean knee flexion contractures range (41.5 degrees) (20-80 degrees). The mean number of casts is 3 casts (2-6 casts). After manipulations and serial casting eight knees get full correction, and six knees still have less than 10 degrees of flexion deformity which is acceptable for the patients. As compared to other studies using serial casts[13,15] **,** our results show that this method is easy, effective and not so expensive if done by an experienced orthopaedic surgeon specialized in dealing with hemophilic patients in association with haematologist and physiotherapist, the follow up for six months shows no recurrence of the deformities and all the patients are satisfied and free of pain.

In cases with severe pain and instability of the knee joint, arthrodesis is a successful option[19].

Radiosynovectomy using radioisotopes[1,20] or Synovectomy using arthroscopy or open method could be used to decrease bleeding episodes, the amount of factor replacement and knee pain[13,14,21].

In severe cases and when facilities are available total joint replacement for the knees in haemophilic patients done in specialized centers show a good results and functional outcomes[14,22,23].

CONCLUSION:

Serial manipulation and casting are effective methods to treat flexion contractures of the knee joint in hemophilic patients; it is safe, easy and can be applied for regions with limited resources and facilities.

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