

Haemophiliac Knee: Role of Physiotherapy

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Objective: To evaluate the outcome of “on demand” factor VIII replacement with physiotherapeutic intervention in the prevention and treatment of recurrent knee hemarthrosis.

Methods: Seventeen patients, aged 7-13 years with median age of (10) years, were prospectively included in this study, between June 1996 and June 1999, at King Hussein Medical Center (KHMC). Patients are known cases of moderate to severe haemophilia A, complicated with knee hemarthrosis. All patients received Factor VIII concentrate at dosage of 30- 40 IU/ kg I .V daily for 5 days, followed by physiotherapy (PT) program including the quadriceps strengthening exercise. They were followed up for fifteen months, focusing on function, range of motion (ROM), and number of bleeding episodes in the target joint.

Results: Fourteen patients showed significant positive correlation between (ROM) and duration of P.T program ($r = 0.96$, $p < 0.01$). They also showed modest significant negative correlation between the bleeding episodes into the target joint and the duration of therapy ($r = - 0.55$, $p = 0.04$). Two patients were not compliant with PT program and one patient with severe flexion contracture showed poor response to therapy and continued to have frequent bleeding episodes into the target joint, and they were excluded from the study.

Conclusion: Prompt “on demand” therapy with F VIII concentrate with quadriceps strengthening exercise will reduce frequency and complications of knee hemarthrosis and allow full recovery of function. So we recommend this therapeutic protocol especially in developing countries where unfortunately giving prophylactic F VIII concentrate is not currently possible.

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Haemophilia A is an X-linked bleeding disorder. It results from a deficiency of a specific plasma coagulation factor and produces its greatest morbidity in the musculoskeletal system.

It is a life long condition with a high potential towards disability, handicap, and impairment if not adequately treated¹. Patients with haemophilia A are classified as mild, moderate or severe depending on the level of factor VIII being 5-20%, 1-5% and less than 1% respectively.

Patient with severe haemophilia may bleed spontaneously, whereas one with mild or moderate haemophilia generally has a history of trauma, which produces hemorrhage into a joint.

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Three categories of joint bleeding- acute, subacute and chronic- may lead to extravasation of blood into the joint ie. hemarthrosis. Delayed or inadequate treatment can trigger a series of pathological changes within the joint, leading to a painful and disabling arthropathy². Chronic synovitis is a predisposing factor for repetitive episodes of hemarthrosis, a leading cause of joint deformity and severe muscle atrophy³, that is why proper treatment is fundamental to prevent disability.

Treatment modalities for chronic knee hemarthrosis are controversial, results of orthotic management are unsatisfactory, and may result in severe quadriceps atrophy and recurrent hemarthrosis⁹.

In this study we evaluate the outcome of management with “on demand” F VIII concentrate, followed by intensive physiotherapy (P.T) program with respect to frequency of further bleeding episodes and improvement of range of motion (ROM) in patients with chronic knee hemarthrosis.

METHODS

Seventeen children, aged 7-13 years, with median age of 10 years, with moderate to severe haemophilia, complicated with chronic knee hemarthrosis were prospectively enrolled in this study at (KHMC) between June 1996 and June 1999. All patients had history of bleeding episodes into the target joint at median frequency of three times per month; their mean ROM at enrollment was 70. All of them received F VIII concentrate intravenously in a dosage of 30-40 IU/kg body weight, daily for five days, then they were referred to our Farah Royal Rehabilitation Center (FRRC) for intensive physiotherapy program.

The PT Program

The program used the following rules.

1. Passive range of motion is contraindicated.
2. The isometric technique of alternation contraction and relaxation is beneficial.
3. When the patients quadriceps is in the fair range, the patient is encouraged to achieve full knee extension in the sitting or supine position.
4. Active resistive exercises may be started when the patient has 90⁰ of ROM in the knee and has less than a 15⁰ flexion contracture. The active resistive exercise is initiated with 1-lb weight twice daily with 10 repetitions for 1 week, which is then advanced to a 2-lb. weight, and then after another week to 3- lb. If hemarthrosis develops the patient must return to isometrics, and again go through the process of advancing from active antigravity to active resistive exercise starting with the 1-lb. weight. .

Once the ROM and strength improve, patient can use stationary bicycles, or other isokinetic exercise machines to strengthen muscles. PT was continued during the next six months at frequency of twice weekly and patients were also encouraged to do PT at home and to have some recreational activities like swimming whenever possible.

Follow-up was continued for 15 months focusing on number of bleeding episodes in the target joint, range of motion (ROM), and strength of quadriceps muscle.

RESULTS

Seventeen patients were enrolled in the study with mean age of 9.91 ± 1.7 years. Two patients were excluded as they failed to continue with the PT program. One patient showed poor response to the therapy and continued to have frequent bleeding episodes into the target joint, he has been referred for possible synovectomy.

The remaining 14 patients showed significant positive correlation between ROM and duration of PT program ($r = 0.96$, $P < 0.01$), this effect started at the third month of therapy, with the highest cumulative effect was at the eleventh to thirteenth month of therapy (Fig 1). They also showed modest significant negative correlation between the bleeding frequency into the target joint and the duration of PT program ($r = -0.55$, $P = 0.04$) as shown in (Fig 2).

Figure 1. Effect of PT program on ROM

Figure 2. Effect of PT program on bleeding frequency

DISCUSSION

The patient with severe haemophilia continues to have significant musculoskeletal problems, with particular morbidity in the knee joint. Hemophilic arthropathy used to be a cause of marked to severe disabilities⁴.

Joint deformity usually results from either subacute or chronic bleeding. Initially there is loss of extension leading to a knee flexion contracture. This is followed by posterior subluxation of the tibia with external rotation and valgus deformity⁵. Such patients may become severely handicapped.

Treatment modalities are controversial while conventional approach to the hemophilic knee is protection against avoidable trauma by restriction of physical activity, external support with braces and energy saving devices^{6,8}.

Some physicians treat subacute hemarthrosis with factor concentrate and immobilization with a plaster splint for 3- 4 weeks⁷. Knee flexion contracture from chronic hemarthrosis can be treated to improve range of motion (ROM) with orthoses to serially extend the knee^{6,8}, or with type of traction device such as a dynamic sling⁹. There are two problem aspects to be noted when treating the hemophilic knee: -

The first is weakness, which aggravates the tendency of the patient to bleed into his joint; and the second is the use of an arthrosis, which results in disuse atrophy compounding the first problem.

Quadriceps atrophy causes instability of the joint, producing repeated bleeding episodes, and hastens the development of serious joint destruction. Based on this many authors advocate, that treatment aims at restoring the original muscle strength, as joint stability is dependent on the uniform distribution of muscle power over joint surfaces^{3,5,9}. Muller et al¹⁰ also has emphasized the importance of joint mobilization techniques in advanced hemophilic arthropathy.

In the developed countries, many studies advocate life-long prophylactic treatment with F VIII concentrate. Even a small number of joint bleeds seems to cause irreversible osteoarthropathic alteration leading to hemophilic arthropathy, which once apparent, further progression of joint damage cannot be arrested despite prophylactic treatment¹¹. In order to prevent hemophilic arthropathy effective prophylaxis should be started before or after the first joint bleeding in severe haemophilia¹¹. Unfortunately this is not possible in developing countries where F VIII concentrate is in short supply, so the aim was to provide a counsel regarding appropriate management to minimize the risk of further bleeding episodes.

Our patients present evidence suggests that PT program has almost normalized the ROM and decreased bleeding frequency, emphasizing the importance of physiotherapy in treatment of hemophilic knee, being concordant with what was reported by different authors^{7,9,12}.

CONCLUSION

The outcome of this study suggests an alternative treatment program to improve ROM as well as decrease the bleeding frequency in the hemophilic knee in the developing countries.

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