Musculoskeletal pain and functional ability in haemophilia A and B. Physiotherapy and rehabilitation in haemophilia patients

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Abstract The present study was carried out to evaluate the progress of physical musculoskeletal ability and pain in Finnish haemophilia A and B patients during a given period of time and to find out the role of rehabilitation and physiotherapy in this process. In the physiotherapy/rehabilitation group, pain index by visual analogue scale (VAS) improved through time by 26%/4.6%. The non-treatment group showed slight aggravation. Regression analysis revealed that age, pain intensity, and functional disability in 1994 were the only significant explanatory factors influencing musculoskeletal functional ability at the end of the study. The current study was not planned as an intervention study and yet the results show some evidence, although not strongly, of the positive effect of physiotherapy and inpatient rehabilitation courses in reducing pain and functional disability for haemophilia patients. One of the important contributions of physiotherapy and rehabilitation is their educational role.

Keywords Haemophilia · Pain · Disability · Physiotherapy · Inpatient rehabilitation

Introduction

Most patients with severe haemophilia suffer from haemarthroses in the ankle, knee, and elbow joints [1, 2, 3, 4]. The hip and humeroscapular joints are less frequently affected. The reason for intra-articular bleeding is the low tissue content of thromboplasticin articular structures. Acute bleeding is painful and patients tend to seek positions in which intracapsular pressure is at its lowest [5]. Long and repeated intra-articular bleeding cause haemophilic synovitis. The consequences are derangement of the articular cartilage, reactive degenerative bone formation and sometimes severe deformity, and functional disability [3]. Musculoskeletal complications of bleeding diseases are by far the most prominent.

Pain may be acute or chronic. Medical and preventive treatments have become very effective [6]. However, treatments are very expensive and not easily available in less developed countries [7]. Also, 15% to 25% of patients with severe haemophilia develop antibodies to therapeutic and prophylactic factor replacement.

In Finland, the care of haemophilia patients is well organised. The national patient association is educationally active and supervises rehabilitation courses, which are mainly centralised in one institution. Thus the therapists are experienced in the disease and the treatments and programs are well standardised.

The main aim of the current study was to evaluate changes in physical ability and pain over a period of years and to discern the role of physiotherapy in this process.

Materials and methods

Subjects

The sample was drawn from the registry of the Finnish Red Cross Blood Transfusion Service. Subjects were eligible if they were between 13 and 75 years of age. A total of 93 male patients with haemophilia A and B disorders agreed to participate in the study. They completed postal questionnaires sent in spring 1994 (time 1) and autumn 1997 (time 2).

The mean age of the patients at follow-up was 41 years (SD 16 years, range 16–73). Various degrees of haemophilia were represented: severe type A (n = 45), severe type B (n = 4), moderate type A (n = 17), moderate type B (n = 7), mild type A (n = 10), and mild type B (n = 10).
Measures

Pain intensity

Pain intensity was assessed by a 10-cm, double-anchored visual analogue scale (VAS) in which 0 cm = no pain and 10 cm = very severe pain. Subjects were asked to indicate the average intensity of their pain during the previous 4 weeks. The intensity of acute and chronic pain was assessed separately at follow-up.

Functional disability

Functional disability was measured using the Stanford Arthritis Center Health Assessment Questionnaire (HAQ) at time 1 and the physical dimension score of the Sickness Impact Profile (SIP) at time 2.

The HAQ [8, 9] includes two or three questions regarding each of eight areas of activities related to daily living: rising, dressing and toilet, eating, walking, hygiene, reach, grip, and miscellaneous activities. For each area, the possible scores are 0 (no difficulty), 1 (some difficulty), 2 (much difficulty or with assistance), and 3 (unable to perform). Any activity requiring assistance from another individual or with an assistive device receives a score of 2. The scores for each of the eight areas are added and divided by 8 to yield a 0–3 functional disability index (FDI) on a continuous scale. HAQ FDI is valid and reflects the subject’s objective impairment status due to structural changes in joints [10, 11, 12]. Cronbach’s alpha value for the studied sample was 0.91.

The SIP physical dimension score [13, 14, 15] is a standardised list of 45 statements describing a possible effect of the sickness on body care and movement, mobility, and ambulation. Respondents were asked to tick health-related statements appropriate to their situation. Each statement had a weighted score. The weighted total score for dysfunction and percentiles for an SIP physical dimension were calculated. Scores can range from 0 to 100, with higher scores representing more severe dysfunction. The SIP is known to be valid, reliable [16, 17], and responsive in measuring functional status and dysfunction for cross-sectional studies. It has been applied in a number of samples of chronic pain patients [14, 18, 19, 20]. Cronbach’s alpha for this studied sample was 0.92.

Physical activity level

Physical activity was measured on a five-grade scale with 1 (no difficulties at all), 2 (difficulties only when performing very heavy physical exercise), 3 (difficulties in moderate physical activities), 4 (difficulties even in light physical activities), and 5 (difficulties even when resting).

Demographic data, disease-related variables (frequency of bleeding episodes during the previous 12 months, amount of clotting factor medication, number of ice packs used during last 12 months, number of operations, pain medication usage, development of antibody inhibition of clotting factor, other diseases), and questions concerning frequency of sports activities, smoking habits, and alcohol consumption were included in the questionnaire.

Statistical analysis

The normality for each variable was tested to decide whether to use parametric or nonparametric methods. A sign test was applied for evaluating changes in disease-related parameters within groups. One-way analysis of variance (ANOVA) with repeated measurement and a set of variables as covariants were used for determining intensity of pain and the physical activity level. One-way analysis of covariance (ANCOVA) was used for the group differences in SIP. Stepwise multiple linear regression analysis was used to examine predictors of pain intensity and physical functioning at time 2. Statistical significance was assumed if P < 0.05. The 1993 BMDP data processing program was used for analysing the data (Statistical Software, Los Angeles, Calif., USA).

Results

On average in 1994, the patients had five soft tissue bleedings (median 2, range 0–62) in 12 months and eight bleedings (median 4, range 0–53) in 1997 (P = 0.015). Intra-articular bleeding occurred an average of 19 times (median 11, range 0–119) in 1994 vs 19 times (median 6, range 0–120) in 1997 (NS, P = 0.48). Thirty-eight of the patients (41%) did not use home treatment (factor replacement), whereas 55 (59%) did. Eighty-one (88%) showed no antibodies to bleeding factors, while high levels of antibodies made normal replacement treatment impossible for 11 (12%). The highest annual cumulative dose of replacement treatment per patient was more than 200 bottles. Of the patient sample studied, 53% used prophylactic factor treatment; this did not statistically change during the study. At the beginning of the study, 40% of patients never used analgesics and 48% did so occasionally, while 12% used painkillers on a frequent basis. The use of analgesics did not change significantly during the study. At the end of the study, the average number of joint operations was 1.0 (median 0), varying from 0 to 6. The use of ice packs to treat bleeding-related pain for one patient varied from 13 to 19 times a year, with a maximum of more than 130 times a year.

Physiotherapy was experienced by 17 of the 93 patients only after surgery, by 36 some time in their lives, and never by the remaining 40. During 1994–1997, 20 patients had undergone physiotherapy for the first time during the period under investigation, eight of whom were new patients. Six patients received physiotherapy regularly once a week with ice packs, transcutaneous stimulation with interferential current (TNS), lymph massage, and electrotherapy, including pulsed short waves.

Of the study group, 25 patients had attended inpatient rehabilitation courses at some time and 68 had never done so. These courses consisted of exercises to prevent disabilities, such as swimming, walking, hydromassage, stretching reflexes that do not produce pain, muscle coordination exercises, enhancement of coping skills, and psychosocial support.

Regarding pain, the average VAS value was 3.8 in both 1994 (median 3, range 0–10) and 1997 (median 3.2, range 0–9.3). The P value of 0.39 was not significant. In 1997, the patients also reported on the effects of pain; disturbances of sleep, physical functional ability, and mental concentration were the most common.

During the study period, musculoskeletal function (scale of 1–5) improved slightly but significantly from 2.3 to 2.0 (P = 0.005). These changes can not be considered clinically significant. The median of functional ability was 2 throughout the study.

In 1994, functional disability was measured using the HAQ index from 0 (perfect function) to 3 (advanced disability). The scores for 44 of the 93 individuals tested ranged from 0 to 0.2, reflecting good functional ability,