Original Article

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Musculoskeletal Complications and Surgical Needs of Patients with Haemophilia

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Abstract

Background: Musculoskeletal problems in haemophiliaoccur due to delayed therapy or lack of detection, leading to joint damage and other complications Orthopaedic surgery is one option to improve deformity of haemophilic patients caused by repeated bleeding.

Materials and Methods: This study includes 85 patients in which all musculoskeletal survey was done .

Observation and Results: Surgery was recommended in 51 (60%) patients (37 severe haemophilia, 12 moderate and 2 mild). Minor surgery was most common in younger patients, while major surgeries increased proportionately with patient age.

Conclusion: Insufficient use of factor replacement and lack of regular supply of factor are major cause for joint damage and other musculoskeletal problems in haemophilic patient.This study is beneficial for assessment of the grade of musculoskeletal problem and identification of the surgical needs of the haemophilic patients.

Keywords: Haemophilia, Musculoskeletal

Introduction

The objective of our study was to study the various musculoskeletal problems in haemophilics and the need for surgical intervention in these cases.

Material and Mathod

The study (Prospective and retrospective) was conducted in the Orthopaedics Department, from July 2013 to August 2015. The haemophilics were categorised as haemophilia A and haemophila B, and within each of these types the degree of the disease is classified as severe, moderate or mild. We studied an evaluation of patient orthopaedic status and the requirement for orthopaedic intervention. Arnold-Hilgartner radiological classification of haemophilic arthropathy(1) was used to grade arthropathy. Three definitions of surgical intervention were used: minor surgery (synoviorthesis); Group 1 major surgery (synovectomy, osteotomy or arthrodesis); and Group 2 major surgery (joint replacement with a prosthesis)(2). All major joints were examined clinically. The gross configuration was studied and any deformity, increase in breath of epiphyses, capsular thickening or muscular atrophy was noted. The range of passive motion of the joints was measured. Loss of range of motion wasestimated, where possible, by comparison with the mobility of the contralateral joint.

The affected joints were examined radiologically. CT scan, MRI, USGwere done as per requirement.

Result

Out of 85 patients 84 were male and 1 was female. Overall 211 joints were having affection of haemophilic arthropathy. Knee joint(95/211) involvement was more followed by elbow (68/211) and ankle joint(40/211).Wedid not find shoulder joint involvement in our study but Hip joint(6/211) and wrist joint(2/211) involvement were noted.. According to grading of arthropathy (Arnold-Hilgartner classification of haemophilic arthropathy),grade-1 arthropathy was more(103/211),followed by grade-3 (57/211),surprisinglygrade-2 arthropathy (36/211) was less than grade-3 haemophilic arthropathy. Hip arthropathy occurred predominantely in patients with severe haemophilia, all 6 patients of hip involvement were severely haemophilic. Patients had knee joint arthropathy, which was bilateral in many cases and was found in 4 cases out of every 5 patients with severe haemophilia, in every alternate patient with moderate haemophilia. Grade-3 or 4 arthropathy was predominantly seen in severe and moderate haemophilia. 40 case showing ankle affection, 20 patients have bleeding episode but no x-rays finding. grade-3or 4 arthropathy seen only in severe haemophilic with age more than 25yrs. The arthropathy of elbow and wrist joint was equally common in moderate and severe haemophilia, and was seen in about one tenth of these patients. It was not seen in patients with mild haemophilia. 1 patient having grade -2 knee arthropathy, had fracture both bone leg Rt side due to trival trauma. 1 patient with grade-3 knee arthropathy with malunited fracture mid shaft femur Rt side was also seen. In the present investigation 2 patients who had severe haemophilia showing ankylosis features believed that the joint had stiffened due to lack of treatment. 20 patient had muscular bleed, 1 female patient had soft tissue bleed over thigh following slip while walking,2 patients had femoral nerve palsy with haematoma in illiacus muscle confirmed by USG. Most of the patients (61%; n=52/85) required surgical intervention for joint disease. Out of 52, 38 had severe haemophilia (<1% factor activity), while 13 had moderate (1– 5% factor activity) and one mild haemophilia (5– 40%

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factor activity). The patients age for surgery was relatively young: approximately 3% were aged ≤5 years; 9% were aged 6–10 years; 32% were aged between 11–20 years and 35% were aged 21–30 years. Less than one-quarter (21%) were aged ≥31 years. 14% of all required procedures were Group 2 major surgeries(prosthesis), 56% were Group 1 major surgeries (synovectomy, osteotomy or arthrodesis) and 30% minor surgeries (synoviorthesis). Minor surgery was the predominantly recommended option in younger patients, while the need for Group 1 and Group 2 major surgery increased with increasing age.

Disucssion

Assessment of joint health has been the most important goals in haemophilia care.Ideally, joint health should be preserved and bleeds prevented so that surgery is not needed. There has been improved use of on-demand therapy in recent years(8). knee was the commonest joint involved as in other studies. It is well established that early intervention in haemophilia is important in order to manage and prevent lasting joint damage, and priority should be given to minor surgery in patients under the age of 11 years, followed by Group 1 major surgery in patients aged 11–20 years. The majority of surgeries in patients ≤5 years and 6–10 years were minor and the majority of surgeries in patients aged 11–20 years and 21–30 years were Group 1 major surgery. As surgical needs of the patients with haemophilia assessed but we are not able to implement it in our institution due to lack of suitable haematological infrastructure and resources however minor surgery with use of chemical agent like Rifampicin is very well indicated in haemophilic synovitis(5). Availability of injectable rifampicin is a big issuein India,same results may be obtained by using very freely available Tetracycline or by phonopheresis(7). There are some limitations associated with this study, including the lack of data on patient outcomes. Primary prophylaxis should be started before the age of 2 years(8).Surgery (as a method of slowing disease progression and improving joint health), may reduce the need for more major and costly surgery in later life. Fear associated with surgery for haemophilic patients is major hindrance in treatment; whatever it is from anaesthetic or orthopaedic surgeon. Proper evaluation, encouragement and boldness in our approach should be the key for surgical procedure which is recommended in this study. On demand factor availability is another hindrance, to cope up, steps should be taken for its easily availability.

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