Don't Let Hemophilia Slow Them Down

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Abstract

At hemophilic centre in Merjan Teaching Hospital a (56) patients with severe hemophilia were investigated using a pre-structured questionnaire,were measuring mainly (4) parameters: Factor availability , academic achievement and work attendance , immediate and long term rehabilitation and participation in sport and physical activities.

Due to perennial and then subsequent shortages of replacement therapy, and lacking of immediate and long term rehabilitation measures , most of patients developed various form of chronic arthropathies and disabilities with poor school and work attendance and most of them confined to their home with no employment , and were not participated in sport or physical activities.

الخلاصة

في مركز الميموفيليا – مستشفى مرجان التعليمي اجريت دراسة على(56) مريضا مصابون بمرض النزف الوراثي الهيموفيليا وكان التحري يستهدف بالاساس(4) محاور هي: مدى توفر العامل التخثري في المركز ومدى اجراء التاهيل الطبـــي الاولـــى والبعيد المدى ومدى التحصيل العلمى والمواظبة على الدراسة . كذلك مدى المشاركة في الفعاليات الرياضية. نظرا لتكرار عدم توفر العامل العلاجي في المركز ولعدم وجود تاهيل طبي آني وطويل الامد مما ادى الى اصابة معظم المرضى بالتهابات وتحدد في المفاصل مسببة كثرة الغيابات وقصور في التحصيل العلمي والعزوف عن الدراسة والوظيفة لمعظم المرضى وكذلك العزوف عن المشاركة في الفعاليات الرياضية .

Introduction

Deficiencies of coagulation factors have been recognized for centuries[1,2].

Patients with genetic deficiency of plasma coagulation factor exhibit life long recurrent bleeding episodes into joints , muscles and closed spaces, either spontaneous or following an injury[1,3].

The most common inherited factor deficiencies are hemophilia which is X-linked disease caused by deficiency

of Factor VIII(Hemophilia A)or Factor IX (Hemophilia B)[1,3-7].

The major morbidity of recurrent bleeding in severe hemophilia is musculoskeletal system .If early treatment is not to arrest bleeding, a hot swollen and very painful joint or muscle hematoma develop[4]. Recurrent bleedings into joints lead to synovial

hyper trophy , destruction of cartilage, inflammation and secondary osteoarth-ritis, contractures and loss of mobility [3,4,6].

As recently as the early 1960s people with hemophilia routinely confined themselves .to their homes, not because they were too ill to go out , but for their own safety , kids couldn't walk to school and adult couldn't work [8].

Till 1970s , with advent of Factor-replacement therapy and moreover prophylactic therapy (house-hold Factor infusion), there were marvelous improvement in hemophilia manage-ment[8,9].

Today , most people with this condition can lead active-even adventurous –lives[8,10].

Although activities with significant trauma risk should be avoided , the modern therapy option offer more and more possibilities for participation in ,enjoy and even excel in variety of physical activities , regular school. attendance , improvement in academic achievement and non dangerous sports with high degree of life qualification and not have to put the hemophiliacs on the side line[8,11-13].

These mentioned facts about marvelous progress in management of hemophilia are well known in hemophilic centers of developed countries and even in some mounting-up developing countries. But what about people with hemophilia ,in present time, who had frequent bleedings and perennial shortage of Factor availability, difficulty in transporting the injured patient with hemophilia to the centre, lack of immediate and long term rehabilitation , no hemophilic society to deal with them, parents ,and school staff, with no financial support all making the hemophiliacs either confined to home with poor school performance or no schooling at all and no work attendance .

Unfortunately these tragic facts that generally not though about or prepare for, are concerning the Iraqi hemophiliacs in Babylon .

The purpose of this study is to investigate the impacts of the perennial and/or frequent shortages of replacement therapy and no or inadequate rehabilitation measures, on the academic achievement or work attendance and participation in sport and physical activities.

Patients and Method

At hemophilic centre in Merjan teaching hospital , a cross sectional study was conducted in period between Feb.2009- Sep.2009., in which (56) people with hemophilia A or B, attended the single centre in the period of the study because of new bleeding episode , were included in the study and investigated by a pre-structured questionnaire which measures four main parameters : Factor availability ,academic achievement and school or work attendance , immediate and long term rehabilitation with subsequent musculoskeletal deformities, and the attitude of patients , their parents , school staff toward participation in sport or other physical activities. Interrogations regarding financial support, education of patients, family, and school staff were done.

Pre school age cases were excluded from the study.

The diagnoses of the type and severity of hemophilia were done in specialized haematological centre in Baghdad based on clinical and Factor assay technique.

The cases were considered severe when the clotting factor activity was less than 1% of the normal range

In addition to full medical history and physical examination , psychological assessment were done by psychologist . Our cases excluded other form bleeding disorders , any co morbid diseases , cerebral palsy or other neurological disorders and mentally retarded cases.

Musculoskeletal examinations included site and number of affected joints, range of motion and any deformities of joints were noted, muscle status or contractures and ambulation of patient were included in examination.

As the clinical pictures of hemophilia A and B do not so much different , the data presented combined for both.

Result

The age of patients ranging between(8-51) years with mean age of (20.17 ± 10.11) years.

People with severe hemophilia were included in the study, (50) cases were hemophilia A and (6) cases were hemophilia B, with average of bleeding episode(1-2 episode)per month.

A (50) patients had age (equal or more than 10 years) faced with perennial shortage of factor supply during bleeding episodes , in addition to frequent shortages in factor supply in the following years .The remaining (6) patients with age below (10) years all faced only with frequent shortage of factor supply during their bleeding episodes.

A (36) patients lived in rural area or vicinities of governorate with average more than (30) Km far from the centre.

Fig.I. shows the patients distribution according to their ages , in which most of patients (30 cases) were aged between (11-20) years.

A (45) patients had no academic achievement or school attendance all

without employment i.e family-dependant, where as (4) cases has been already graduated from college, two of them were employed and the other two graduated people waiting for employment

(7)cases they were still in academic or school attendance (5 cases at primary school and 2 cases at secondary school) Fig.2.

Fig. 3 shows the number of joints with chronic hemophilic arthropathy increases with age which means more frequent attacks of bleeding.

Only(10) cases had occasional participation in non contact sports in leisure time and were house-hold only, where as(46) cases had no any sort of sport Fig.4

Fig.5 shows the frequency of joint involvement due bleeding episodes in form of chronic arthropathy, deformities ,limitations of movement and contractures. the knee joint coming first followed consequently by elbow, ankles, hips. and shoulders.

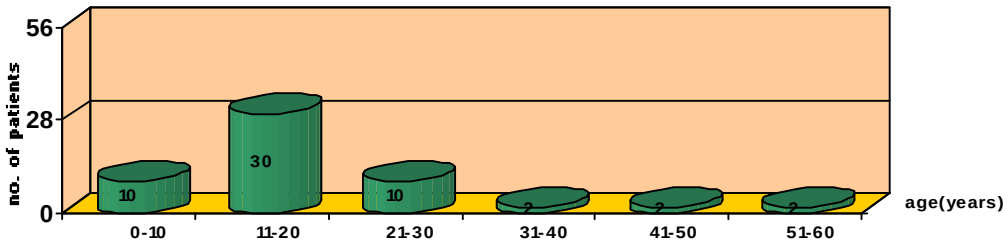
(6)cases had received immediate simple rehabilitation measures in form of (Rest, Ice applications, Compression, Elevation)(RICE)in each bleeding episode, where as 50 cases had not,Fig.6

Fig.7. shows that only(4) cases had got regular attending long term rehabilitation program in the out patient department of rehabilitation and rheumatology unit in the hospital , where as (52) cases had not.

No any people with hemophilia in current study registered in or receiving a fund from any medical society or federation of hemophilia..

Families as well as the school staff were not educated by medical staff about the nature of disease , how to minimized bleeding and proper first aid management during bleeding episode, the importance of long term rehabilitation ,the importance of non dangerous non contact sport, and there was no active link between the centre, parents and the school staff.

Parents as well as school staff were not supplied with published simple medical articles regarding hemophilia, or simple guide line for first aid measures during bleeding episode as well as other important , valuable and useful information. Similarly all cases didn’t supplied with special card informing about the diagnosis and blood group.



Mean age : 20.17± 10.11 years

Figure 1 Distribution of patients with hemophilia according to the age groups.

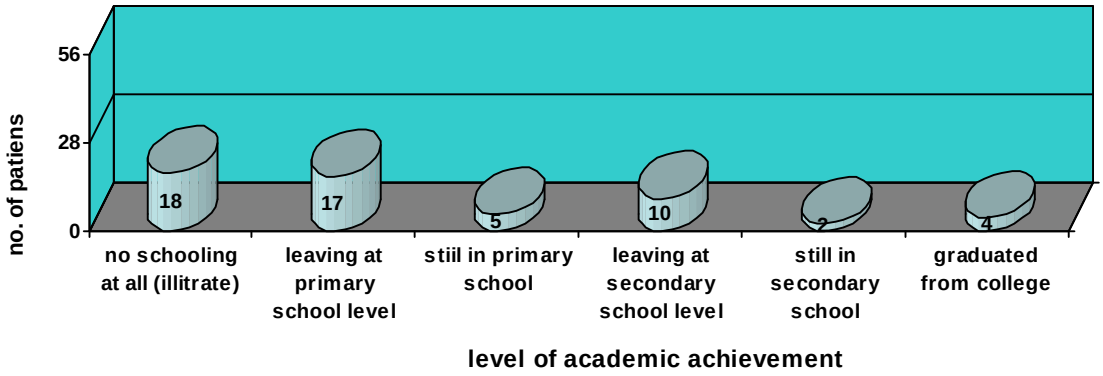


Figure 2 Distribution of patients with hemophilia according to the levels of academic achievement and the level of education..

Figure 3 Grouping and number of patients with chronic hemophilic arthropathy according to number of joints involved in each patient and their mean ages.

No. of joints involved	No. of patients	Mean ages in years
One joint	4	10.7
Two joints	24	15.9
Three joints	6	18.8
Four joints	6	19.7
Five joints and more	16	32.9

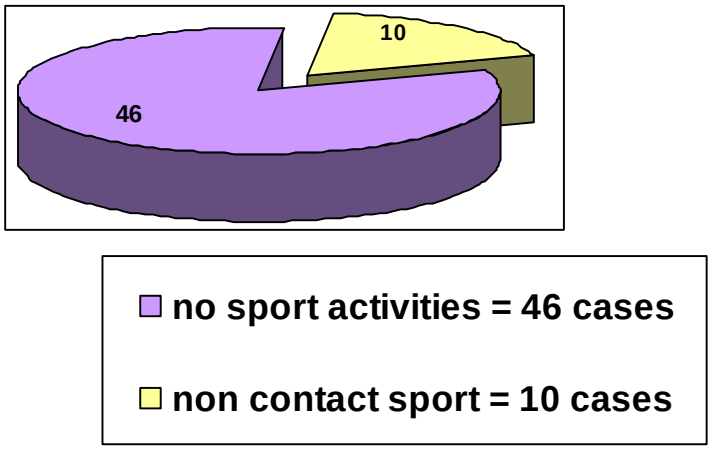


Figure 4 Number of patients with hemophilia according to participation in sport activities.

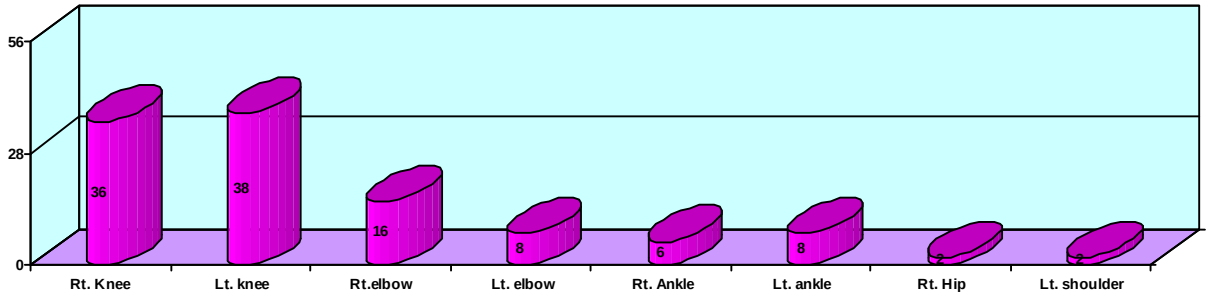


Figure 5 Frequency of type of joints involved in chronic hemophilic arthropathy.

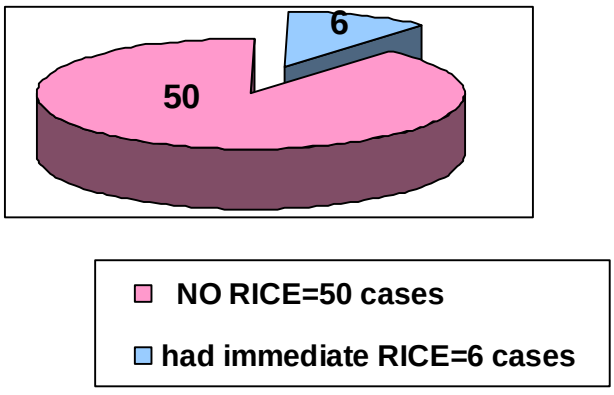


Figure 6 Number of patients with hemophilia were distributed according to reception of immediate rehabilitation measures (RICE).

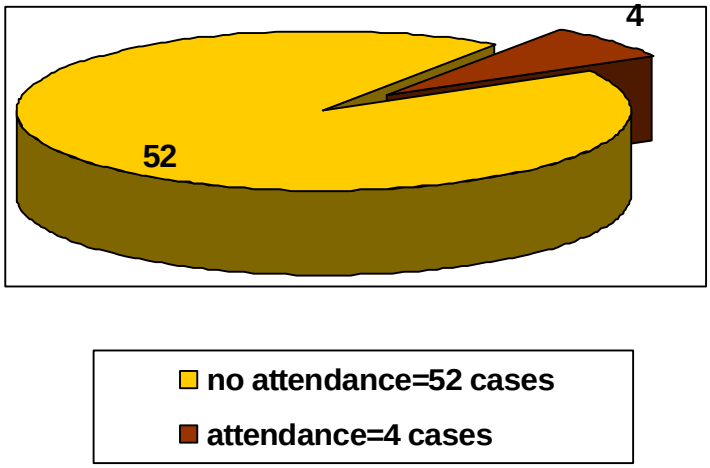


Figure 7 Number of patients with hemophilia distributed according to attendance to long term rehabilitation measures.

Disscussion

Although an uncommon disease, hemophilia is a life –long condition that places a considerable burden on parents , health care system and society [14].

Education is one of the most important aspect and resource of development. Poor school performance and failure of academic achievement not only results in the child having a low self – esteem , but also causes significant stress to parents[15]and each child should have the chance to achieve his potential [16]

On data basis of the study and due perennial shortages and further subsequent shortages of Factor-replacement therapy in following years, resulted in badly controlled bleeding attacks making most of our patients had frequent school absence which result in failure of academic achievement or school attendance. This poor school attendance, may also, contribute to other possible reasons , of which, parents preferred to keep their hemophilic son at home to avoid any possible trauma and bleeding episode they consider their child as(fragile one) , in addition to that , the presence of chronic arthropathies and or joint contracture as a sequel of previous badly treated and rehabilitated bleeding episode resulting in difficult ambulating especially speaking that some of schools are so far need the boy with hemophilia walk long distance in usual situation as most of Iraqi schools has no special school buses. All abovementioned lead to school absenteeism making boys with hemophilia under-achiever especially when no active home-school link making the subject teachers did not compensate for missing days during absence periods during bleeding episodes.

In most instances the school staff isolate the boy with hemophilia from peer and resented him from playing in recess periods. Some teachers" scared to death", and feel anxious about having a pupil with hemophilia in their school i.e hemophiliac were not welcomed at school registration.

The low socioeconomic level of some hemophilic families may also influence the continuation of education and

school attendance .un fortunately the exact statistics regarding the incidence of school escape among healthy individuals in Iraq making difficult to compare or to measure the ratio of non school attendance of healthy boys to that with hemophilia.

The situation is so far different in developed countries and even in some of developing countries , where there is a link between home, school , and hemophiliac centre , and continuous education regarding immediate rehabilitation (RICE) and immediate access to Factor replacement therapy . more over now days the parents or and teaching staff are able to infuse the factor in home or at school either on demand basis or as prophylaxis .[9,17-20].

The mode of prophylaxis therapy means fewer bleeding episodes , better academic achievement[13]. And looking to the hemophilic as a person first, then a person with hemophilia and should be looked as normal student " with access the pluses and disregard the minuses" as can as possible., and most of hemophiliacs in such centre usually climbing up in education hoping an employment , that carry minimal risk of trauma as managerous or academic jobs.

Regarding various sport activities , physical activities , camping, school trips , in our vision and on data basis , most, if not all, hemophiliacs attending Merjan teaching hospital hemophilic centre were informed by their parents , to avoid any sort of school and house hold physical activities , even the school staff also were informed to not engaged the ;fragile son to any form of school sport or/ and physical activities, fearing of trauma and bleeding episode which mean in present situation ,a difficulty of stopping bleeding due to possible shortage of factor, more deformed joints, further school absenteeism.

In comparison to the most of hemophilic centers of developed countries , where the hemophliacs are engaged in various non vigorous sport activities and with advent of prophylaxis therapy, home infused therapy in morning on day of physical or sport activity enable those with severe hemophilia to participate in non rough ,safe sport including recess and physical education times [7,13,21].

In the setting of regular prophylaxis and adult coaching and supervision, significant bleeding complications are uncommon, giving potential health benefits in an increasingly overweight pediaetrics population [20].

Because sport activities include not only physical and functional aspect, but also have an impact on an individual self-esteem and social interaction , now days, the attitude toward sport for people with hemophilia has changed , and the World Federation of Hemophilia has formulated recommendations concer-ning physical activities for people with hemophilia[21,22]. Awareness of the importance of sport activities for these patient groups has increased , and several sports projects are ongoing, for example, a twice-annual sports camp for adult hemophiliac had been held in Germany for the years 2005 and 2006 [21].

Sport activities and consistent participation in quality physical activity at any age is central to managing hemophilia and equally important to achieving over all heath and well-being [11,21]

In addition to enhance motivation and sense of well being , regular exercises strengthen muscle and protecting joints

which can help in reducing spontaneous bleeding,[8,23].

hemophiliacs in the current study in addition to new bleeding episode, they presented with chronic joint involvement due to lack of treatment of previous attacks , lack of immediate rehabilitation measures(RICE), inadequate or no long term rehabilitations leading to joint deformities , decreased mobility, chronic pain , increased family dependence , difficulty in completing education or finding employment . therefore the maintenance of healthy joints and muscles is crucial to quality of life of a people with hemophilia.[24]

replacement therapy may stop or even prevent bleedings, but it does not restore joint or muscle function., only regular movement and rehabilitation and exercises can do[24]

Repeated bleeding into a joint can result in permanent changes (chronic hemophilic arthropathy [24].the knee joints were on the top of the list of joints affected and this result is consistent with Shapiro et al study [13].

With frequent shortage of replacement therapy , or delayed in accessing the therapy and lack of proper immediate and long term rehabilitation ,patient with severe hemophilia had five or more damaged joints at the age of 20 and this fact is consistent with study of Heijnen et al.[25]

Today in the west and some developing countries , grossly deformed joint are a novelty[26], thank to availability of prophylaxis or home infusion therapy, to immediate proper therapy , early and rapid accessed to the centre, immediate and long term rehabilitation .to societies and hemophilia federation education .program of family and teaching staff ,and financial support all making the people of hemophilia have not to slowdown and become on sidelines.

Conclusions and Recommendations

The consequence of improper management of hemophilia can affect abroad range of physical, social, and academic activities. Signifying ideal updated comprehensive treatment modalities that can reduce recurrent bleeding episode and consequent musculoskeletal complications has an important target for keeping our people with hemophilia to high degree of participation in social activities, high levels of academic achievement, well qualified life and employment and becoming an active member of society and don't let hemophilia to slow them down ,putting them on sidelines, and it is never too late to start.

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