Rehabilitation in Haemophilia

The role of the rehabilitation specialist in the comprehensive care team

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Introduction

Functional disorders caused by the sequelae of haemarthroses and muscle-bleeds can cause long-lasting disability and may handicap the haemophiliac, because he cannot perform the role that is normal according to his social circumstances.

Within the comprehensive care team the different members all have the same aim: to allow the haemophiliac to live a normal life. They advise and treat the haemophiliac with the specific knowledge of their own profession.

Haemophilia is a disease which in itself cannot be cured, therefore the first-level of prevention—apart from genetic counselling—starts with the treatment of the clotting defect by the haematologist.

Definitions

- Impairment = any loss or abnormality of psychological, physiological, or anatomical structure or function (in the case of a haemophilia patient—any sequela of any bleed).
- Disability = any restriction or lack of ability to perform an activity in the manner or within the range considered normal for a human being.
- Handicap = is a disadvantage for a given individual, resulting from an impairment or a disability, that limits or prevents the fulfilment of a role that is normal (depending on age, sex, and social and cultural factors) for that individual.
- First-level prevention = reducing the occurrence of impairments.
- second-level prevention = limiting or reversing disability caused by impairment.
- Third-level prevention = preventing the transition of disability into handicap.

Implications

The task of the rehabilitation specialist is second- and third-level prevention.

Rehabilitation of haemophilia patients includes all measures aimed at reducing the impact of disabling and handicapping conditions, and at enabling the disabled and handicapped haemophiliacs to achieve social integration.

The rehabilitation diagnosis is prognostic and defines the plan of treatment. It consists of the description of impairments and disabilities in five areas (Physical (Ph), Activities of daily living (A), Social (S), Psyche (P), Communication (C)).

Physical: impairments concerning, for example, muscle strength, mobility pain, coordination, sensibility.

Activities of daily living: disabilities concerning eating and drinking, personal hygienic care, walking, cycling and driving a car.

Social: disabilities concerning fulfiling one's social role, like living in one's own house, doing housework, taking part in family life, going to school, social life and employment.

Psyche: impairments of consciousness, intelligence, memory, behaviour disabilities concerning orientation, concentration, spirit, mood, awarness and acceptance of illness, disability and handicap.

Communication: impairments concerning speech, hearing, vision; disabilities concerning making oneself understood, comprehending spoken or written language and reading or writing.

Assuming that a physical problem is always present one can make 16 different rehabilitation diagnoses by combining the areas in which problems are found. For example: Ph, Ph-A, Ph-A-S, Ph-A-S-Ps and so on. In the case of haemophilia patients we expect to find that most diagnoses consist of combinations of the first four areas.

The rehabilitation diagnosis made in this way is complementary to the current medical diagnosis and is in fact especially aimed at the social consequences of the causal ailment, and thus largely prognostic. In combination with the medical diagnosis the rehabilitation diagnosis-prognosis defines the plan of treatment, and which members of the comprehensive care team are to be active in treating the haemophiliac and whether the Physiotherapist and orthopaedic surgeon (Ph), Occupational therapist and rehabilitation technician (A), Social Worker (S), Psychologist or Psychiatrist (P), speech therapist and dentis (C) should be involved.

It must be evident that the earlier one can make such a rehabilitation diagnosis in the event of post-haemorrhagic functional disorders, the more it may be possible to take adequate measures to limit or reverse disability and to prevent handicaps. This implies that the rehabilitation specialist should be a member of the comprehensive care team and should see the haemophilia patients regularly from the start, when they visit the centre for their check-ups.

Practical Applications

1. Babies and small children: prevent extreme risk of trauma, but let them play with as many different materials as possible to teach them differencies between soft/hard/sharp etc. Let them move around freely so they can learn to use their limbs and get a normal idea of their own motor abilities. Let them use normal eating tools at an age so that they can learn to use them properly. A normal psychomotor development will enable the boy to handle his body and to manage bleeds later on.

2. School boys: Teach them to use scissors, knife, hammer and saw. Let them ride bicycles and participate in all school and playground activities which are not extremely dangerous, and also gymnastics. Stimulate swimming and joining a club where activities and or sports are supervised; and try to involve them in those sports in which they can enter a competition if they wish. If you have to restrict an activity after a bleed, try to do it in the functional way, for example by taping the ankle.

3. Teenagers: should have an important say in when to receive clotting factor and if it is worthwhile to risk a bleed which will impede normal activities and might further damage joints. Choosing schooling and jobs in relation to haemophilia in general and specific musculoskeletal problems is another important item. When participating in more vigorous sports, spend money on the proper equipment: good sport shoes have excellent shockabsorbing and stabilising properties (but some sports, like tackle-football are always dangerous even for non-haemophiliacs with the proper gear).

4. Grown-ups: often have the sequelae of old bleeds (limited Range Of Movement (ROM), muscle wasting and/or pain). Try to give practical advice

for adaptations in the work situtation and at home. The height of the desk and chair are important (with limited ROM and not enough muscle power of hips and knees, the arms will be used when sitting down and getting up, which may lead to elbow bleeds or pain). Cycling may be possible with a high saddle. In some countries visiting the toilet requires maximal flexion of hips and knees, so an "English toilet" or a wooden stool with a hole in it as makeshift is necessary.

It depends on the local situation within the family and culture where the haemophiliac lives, whether some limitation is a problem or not (90 degrees of flexion in hip and knee are enough to sit in the cinema or to use most forms of public transport, but not enough to sit normally at home in India or Japan). So one must act according to the local circumstances which are best known to the local team treating haemophilia patients and of course to the patient and his family.

Conclusion

The rehabilitation specialist should be a member of the comprehensive care team. The rehabilitation diagnosis especially aims at the social consequences of the post-haemorrhagic functional disorders, and defines which members of the comprehensive care team are to be active in treating the haemophiliac to limit or reverse disabilities and to prevent handicaps.

Physical Activity and Sport for Haemophilic Patients

Introduction

Physical prophylaxis for the musculoskeletal system of the haemophiliac is a must from early childhood on. Joints which are protected by well developed muscles can withstand the traumas of daily life better and patients with a strong musculoskeletal system recover quicker from a haemarthrosis and have less "spontaneous" bleeds (which are in my opinion bleeds caused by a micro truma). The idea that exercises help to protect joints is supported by a study in which the left knee and ankle of a group of "left handed" haemophilic patients are in a significantly better condition (as assessed by the range of movement and the Pettersson score) than those of the non-dominant right leg; this was also the case with the right knee of right-footed haemophilic patients. This led to the conclusion that the dominant leg, which is used in dynamic activity and in situations which require more skill, is protected better against the sequelae of haemarthroses, than the other leg which gets a static load with the body weight.

General Remarks

A daily exercise programme for all persons with haemophilia, beginning in childhood, is advised, because strong muscles stabilise and thus protect the joint. In the past only "safe" sports like swimming, table tennis and golf were advised. Nowadays not only the risk of an injury, but also wish of the haemophiliac to be one of his social-age group and to compete with his peers, next to the benefit of the exercise are taken into consideration when choosing a sport.

Physical Activities

The haemophiliac should be enouraged to participate in all activities normal for his age group and culture, as long as the risk of getting injured is small. So walking, riding a bicycle and participating in gymnastics at school, carrying shopping or jogging with collegues at work should be normal if there is no acute bleed or a chronic joint problem.

For the young child who starts standing up and learning to walk, this inevitably means falling and bruising. Parents should be aware of the fact that this is the only way for the child to learn to stand on his own feet!! And as long as the bruises are superficial and the child is not limping there is no need to pay any more attention to it than one glance.

Of course the child should ride a tricycle or other vehicle propelled by manpower. Roller-skates look dangerous, but if all children in the neighbourhood are doing it the haemophiliac will no doubt borrow some if he does not get them; so why not give him good material and teach him to do it the right way?

Special exercises to improve the general condition, to increase muscle strength and range of motion should be done in a programme which starts with warming-up and next to active muscle strengthening should also include stretching especially of the psoas muscle, hamstrings and calf muscles. The programme should be adapted to any specific joint and muscle problems to prevent the triggering of bleeds. It should be built up gradually. If there is arthropathy in one or more joints, the active muscle strengthening exercises should preferably be done without the bodyweight pressing down on elbows, knees or ankles.

Sports

We are all convinced that participation in sports is good for haemophiliacs but as to which sports are safe and which are too risky the ideas of parents, doctors and patients differ. For the schoolboy the feeling that he is good at a game or sport which is done by his age group is important, whereas the teenager needs the opportunity to compete with his peers. So their choice will be influenced by these motives. If possible a sport should be chosen in which there is a good chance of successful participation by the child; and this success will be more probable if the preparation is adequate. Attempting a new sport carries a risk of failure for each child and a haemophiliac has to learn this like anyone else; and he also has to learn to decide for himself if this activity is worth the risk of trauma; on the other hand he might find that due to this activity he gets less bleeds in other situations.

The sports we consider best are of course those with the least chance of trauma combined with activity for all joints and muscles: Swimming for all ! But not all like it. More generally speaking individual sports have less risk than team sprots, in which there is a chance of "bodily collision" and also because the team decides where the limit lies, which might be too high for the individual.

A. Recommended sports in which most haemophiliacs can participate safely are: Badminton, Ballroom dancing, Billiards, Fishing, Golf, Rowing, Swimming, Table tennis, Walking.

B. Sport in which the physical, psychological and social benefits often outweight the risk: Archery, Athletics, Baseball, Basketball, Bodybuilding, Bowling, Canoeing, Cricket, Cross-countryskiing, Curling, Cycling, Horseback riding, Ice-skating, Jogging, Roller-skating, Running, Sailing, Skiing, Softball, Tennis, Volleyball, Weightlifting, Wheelchair basket-ball, Windsurfing. When supervised by experts and with proper equipment Moutain climbing and Water skiing also might be done by individuals in good condition.

C. Dangerous sports (with a high risk of trauma for all individuals): Boxing, Competition-Football (soccer/tackle), Handball, Hockey, Ice hockey, Jiu-jitsu, Squash (Racquetball), Wrestling.

D. Sports which we do not encourage but cannot be avoided in some countries are Football and Skateboarding.

Some Remarks on the Different Sports

Archery good shoulder and elbow function required; can also be done seated.

Athletics (running, jumping, throwing, steeplechase) participating on all subjects asks a good condition of all muscles and joints and puts a strain on all of them, so competition is not advised, unless the boy is in "perfect condition" and provided injuries can be treated at once.

Badminton asks some movement of wrists as well as good shoulder and elbow function; it also can be done from a wheelchair or sitting on a mat.

Basketball not advised, knees and ankles are in danger; if the boy wants to play it in the driveway or at school, shock-absorbing shoes are advise (see also athletics).

Baseball favourite with American children; it should be supervised (at school), helmets are recommended and base-sliding and pitching should be avoided.

Bodybuilding a good way to do otherwise dull exercises; a gradually built-up programme should be supervised by a trainer.

Bowling strain on elbow wrist and fingers; with hip or knee arthropathy the walk technique should be adapted.

Boxing Strongly not advised, will hurt anyone!!

Canoeing in calm waters good arm exercise and a nice way to travel, in wild waters dangerous (not advised and at least helmets are a must).

Cricket popular in some countries e.g. England, India, risk similar to baseball.

Cross-country skiing good exercise, use high shoes to protect ankles, less chance to collide with other than in downhill skiing.

Curling may cause strain on elbow (seasonal sport in wintersport resorts).

Cycling can be done to school and work; but also as a recreational sport or in competition. As a recreational activity it is also a good exercise for the quadriceps, and the distance can be adapted according to the condition of the participant, the roads and the weather; competition has a greater danger of falls.

Fishing in some situations good strength of the arms is requried.

Football neither tackle football nor soccer are advised as the risk of injuries is big, especially when playing competition.

Golf can be done until old age, but in some countries an elite sport.

Handball not advised, too much body contact causing trauma.

Hockey not advised, trauma due to hard ball and stick.

Horseback riding has a risk of falling off and hurting one's head; if the whole family or community does it, the haemophiliac should get lessons on a reliable horse and wearing a hard well-fitting helmet; jumping should be discouraged.

Ice-hockey not advised even more traumatic than hockey.

Ice-skating if ankles are weak with high shoes (ice-hockey skates) no jumps.

Jiu-jitsu not advised.

Jogging good sportshoes with shockabsorbing material and providing stability are a must as is a gradually built up programme.

Judo if started at a young age a way to learn the proper way of falling, but not without risks.

Mountainclimbing not advised. (When supervised by experts and with proper equipment it might be done by individuals in good condition).

Roller-skating controlled, when the child is young it might only cause some bruising; when done more vigorously by older children serious falls may occur.

rowing with two sculls it is a good symmetrical exercise which also asks for knee and hip flexion (some knee flexioncontracture is no problem).

Running see jogging.

Sailing depending on the kind of boat and weather, a leisurely activity or strenuous.

Skiing (down hill) if the haemophiliac lives in a country, where it is a common sport, he should get serious lessons using good material; and he should avoid the "Sunday rush hours" when chances of collision with fanatics are high.

Skateboarding not advised (severe ankle fractures).

Softball ball softer than for baseball (see Baseball).

Squash (racquetball) not advised: rapid movements, hard ball and rackets.

Swimming this is much encouraged, as it is considered a safe sport which exercises both arms and legs symmetrically.

Tabletennis safe, can also be done from a wheelchair.

Tennis a good chance if one gets proper training and plays with players of the same level.

Volleyball risk increased in competition.

Walking preferably with good sport shoes.

Water-skiing not advised because of the risk of head trauma, and stress on joints and muscles of legs. (When supervised by experts and with proper equipment it might be done by individuals in good condition).

Weightlifting within reasonable weight-limit no problem for adults.

(Wind-)surfing depending on the kind of water and weather, a leisurely activity or strenuous.

Wrestling not advised.

Conclusion

When advising about participation in sports, the past clinical history and the present condition of the joints and muscles of the haemophilic patient should be considered, but his ideas and wishes concerning a special activity are most important. Whatever sport is chosen, "controlled" participation with supervised training and, if necessary, a preparatory conditioning programme, with special exercises to strengthen weak muscles and diminish contractures, should precede the actual sport. The possibilities of participating regularly (not just one week per year) should also be considerd, as well as the demands of family, and schoolmates and cultural and geographic circumstances.

When asking 160 Dutch and British haemophiliacs about their preference I found that "A" sports like swimming and tabletennis were quite popular, but so was "C" sport, football (= soccer). This last sport, however, was played only with friends at school or in the street and not in a club competition; but knees and ankles are nevertheless at risk. Football (= soccer)—like baseball in the USA and cricket in India—is an example of sport that is almost a "must" within a certain age group, but can never be played seriously later on. Whereas sports like golf, swimming, badminton and sailing can be enjoyed well into old age.

SUMMARY

Exercises and sports are very important to keep haemophilic patients in good condition, and help to prevent recurrent haemarthroses and arthropathy.

These activities should be maintained lifelong; therefore the choice should be the patient's, although advantages and risks are to be considered before the final decision is made.

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