REVIEWS

Adv Clin Exp Med 2009, **18**, 1, 93–98
ISSN 1230-025X

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Physiotherapy of Children with Hemophilia

Fizjoterapia u dzieci chorych na hemofilię

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Abstract

Hemophilia is a chronic disease that starts in childhood and lasts into adulthood. Apart from many ailments, it causes secondary restriction of the range of movement, usually in the elbow and knee joints. The patients' restricted range of movement is a priority in the physiotherapeutic strategy. The objective of the therapy, depending on the patient's condition, should be to prevent deformation, treat existing deformations, work toward the most effective compensation, and improve motor control. Treatment with physical activities supported by physiotherapy is the most effective. The physiotherapeutic procedures serve to prepare patients for exercises and should have analgesic and anti-edematous effects. The duration of the procedures plays a role here; they should be short. The procedures should not raise the body temperature at the application site. The physical activities should be individualized and complex, evolving as the child makes progress and with his or her psychomotor development. The therapist's attitude, approach to children, and ability to make contact with the child are significant as well. The therapy will be effective if there is cooperation on the part of the parents. Without this it is difficult to adhere to the therapeutic strategy and achieve its objectives (**Adv Clin Exp Med 2009, 18, 1, 93–98**).

Key words: hemophilia, physiotherapeutic procedure, kinesitherapy, restricted range of movement in joints, deformation prevention.

Streszczenie

Hemofilia jest chorobą przewlekłą rozpoczynającą się w wieku rozwojowym i trwającą aż do dorosłości. Oprócz wielu dolegliwości powoduje wtórnie ograniczenia ruchomości, najczęściej w stawach łokciowych i kolanowych. Najważniejszym w postępowaniu fizjoterapeutycznym w pracy z takimi pacjentami jest utrzymanie przez nich ograniczonego zakresu ruchu. Terapia w zależności od jego stanu powinna być nakierowana na profilaktykę deformacji, leczenie istniejących zniekształceń, znalezienie i wypracowanie najbardziej skutecznej kompensacji oraz poprawienie sterowania ruchem. Najlepsze rezultaty daje leczenie ruchem wsparte zabiegami z zakresu fizykoterapii. Mają one za zadanie przygotować pacjenta do ćwiczeń, zadziałać przeciwbólowo i przeciwobrzękowo. Niezwykle ważny jest czas ich trwania, który powinien być krótki. Równie istotne jest to, że zabiegi te nie powinny podnosić temperatury ciała w miejscu ich działania. W zakresie leczenia ruchem należy zwrócić uwagę na indywidualny dobór ćwiczeń, ich kompleksowość ewoluującą zarówno wraz z postępami dziecka, jak i jego rozwojem psychomotorycznym. Ogromną rolę w całym procesie usprawniania spełnia też sam terapeuta i jego postawa, podejście do dzieci i umiejętność nawiązania z nimi kontaktu. Osiągnięcie pozytywnych wyników terapii jest związane z dobrą współpracą z rodzicami małego pacjenta, bez których trudno jest działać zgodnie z przyjętą strategią terapii i uzyskać zamierzony jej cel (**Adv Clin Exp Med 2009, 18, 1, 93–98**).

Słowa kluczowe: hemofilia, zabiegi fizykalne, kinezyterapia, ograniczenie ruchomości w stawach, zapobieganie deformacjom.

Hemophilia is an inherited hemorrhagic diathesis which last the whole life and in which a genetically determined deficiency of coagulation factor VIII or IX in plasma impairs the coagulation pathway. Sufferers do not bleed faster, but the bleeding episodes may last longer and be more intense. The patients are prone to internal bleeding into organs, joints, and muscles. Bleeding may be

caused by trauma, surgical procedures, tooth extraction, or spontaneous injury [1].

The Course and Profile of Afflicted Individuals

The severity of the disease depends on the blood level of coagulation factors. The normal level of the coagulation factors is 50–150%. In mild hemophilia, associated with 5–30% of normal factor activity, bleeding episodes are mild and usually result from injury. In moderate disease, associated with 1–5% of normal factor activity, there are long bleeding episodes after surgery. Bleeding rarely occurs for no apparent reason and the frequency is once a month. In severe hemophilia the factor value is below 1%. The person experiences frequent bleeding into joints, usually the knees, tarsal joints, elbow, and muscles [2].

Hemophilia usually affects young people. However, advances in replacement therapy have changed the demographic picture, leading to a higher prevalence of the disease among people over 20 years old. In moderate and severe hemophilia the first symptoms occur in the first year of life, as the child is prone to injury while learning to walk. In mild hemophilia the symptoms occur later in life, usually following surgical procedures.

In hemophilia A, hemorrhagic diathesis is usually manifested by bleeding in organs, although the most typical are subcutaneous hematomas and muscle hematomas as well as hemarthrosis. Equally common are epistaxis, gastrointestinal hemorrhage, and hemorrhage from the urinary tract. Life-threatening hemorrhages into the central nervous system may also occur, but they are rare in hemophilia. Bleedings into the posterior wall of the nasopharyngeal cavity due to the closure of respiratory tract may also occur. It should be noted that hemorrhages, especially profuse ones, may apply external pressure on nerves and blood vessels. This may lead to their obstruction and necrosis [3].

Changes in joints should also be noted in the severe form of hemophilia. These are a consequence of hemarthrosis and peri-arthritic bleeding, which lead to typical arthropathy. As seen in radiological examination: 1) early changes affect mostly peri-articular soft tissue and the articular capsule, with slight osseous erosion; 2) subsequent changes, which lead to arthropathy, initially present as changes in soft tissue, marked osteoporosis, and articular cartilage defects; at a later stage they assume the form of filling defects on the articular surface, epiphysis deformation with trabecular bone structure changes, peri-articular false cyst, reduced articular space, and increasingly marked soft elements due to blood pigment deposition; 3) late changes include irreversible degenerative changes of joints and fibrous ankylosis.

Usually the knee, elbow, and tarsal joints are involved. Nevertheless, bleeding into other joints may also occur. Typical of hemophilia is recurrent hemarthrosis into the same joint [3]. Depending on the type of changes, there are acute hemarthrosis, recurrent hemarthrosis, and chronic hemophilic arthropathy [4, 5]. The bleedings can occur once or twice a week. The most common manifestations of the disease are ecchymosis, spontaneous hemorrhage, prolonged hemorrhage, pain, stiffness of the area, difficulties in moving [6–8], and bleeding into the muscles and joints. As a result, the range of movement of the joints becomes restricted, the limbs become uneven, and there is angular deformity [9, 10].

The Treatment of Hemophilia

Until recently, hemophilia was treated only by supplying the missing coagulation factor, isolated from human plasma. Lyophilizate preparations of coagulation factors in powder form are dissolved in water and injected intravenously. These preparations must be stored at a temperature of 2–8ºC. Coagulation factors are very perishable; factor VIII has a half-life of about 10–15 hours and factor IX of 20–24 hours. Therefore exogenous coagulation factors work for a dozen or so hours (in hemophilia A) or for about a day (in hemophilia B). Recently, factors are being produced by genetic engineering. Unfortunately, some patients produce inhibitors, which makes applying the standard therapy impossible. Preventive treatment should also be applied in children with hemophilia. This consists of systematic administration (three times a week for hemophilia A or two times a week for hemophilia B) of the suitable coagulation factor. This prevents the formation of degenerative changes in the joints, and the patients' comfort increases considerably. In patients with the mild form of hemophilia A, treatment is also possible with the help desmopressin, a synthetic hormone which increases serum levels of factor VIII. Therapy with desmopressin is not possible in the severe and moderate forms of hemophilia A or in hemophilia B [11, 12]. Patients with hemophilia receive the deficient clotting factor through intravenous injections, fresh blood transfusions, antihemophilic globulins, and plasma to treat the disease.

The treatment of complications is a prolonged process. Apart from medical intervention, it is of

paramount importance that the patient receives physiotherapeutic help as well as help from auxiliary medical personnel [13]. Comprehensive treatment is of the utmost importance because; as discussed by Seyfried [14], hemophilia is a progressive disease. Functional impairment affects many parts of the motor system, with concomitant pain. Lack of acceptance for one's own disability affects the patient's ability to integrate with society [14]. It should be noted that boys suffering from hemophilia radically limit their physical activity out of fear of more bleeding. Also, overprotective parents may hinder the child's progress towards independence [15].

The Tasks and Aims of Physiotherapy

Depending on the patient's condition, the physiotherapist's job is to prevent deformations, improve or maintain the range of movement, treat existing deformations, facilitate compensation that will not strain the organism, and improve motor control. The physiotherapist should also ensure that appropriate forms of activity are chosen [5]. Early treatment of bleeds, disease awareness, body awareness, and awareness of his or her capability can help the patient accept the condition and adapt to life with the disease. The disease's management includes pharmacological treatment (usually the administration of the deficient clotting factor), physiotherapy, and kinesitherapy [16]. It is vital that all the elements of the therapy run smoothly.

Physical Treatment

The aim of physical treatment is to prepare the patient for kinesitherapy. The range of appropriate activities to chose from is limited by the disease profile typical of the rheumatologic disease, i.e. arthritis and a hereditary tendency for bleeding episodes. Activities that can affect stiff joints or tense muscles may have to be excluded because of the risk of bleeding. With hemophilia, some rules have to be adhered to as far as physical activities are concerned. The activities should have analgesic and anti-edematous effects. They should be short and complex and, what is most important, they should not raise the temperature of the treated site.

The patient's age should be considered in all physiotherapeutic activities. With children the procedures have to be adapted to the patient's individual needs with regard to frequency, quality, and intensity. An excessive amount of procedures may affect the child's tolerance threshold and their condition may automatically deteriorate. Precautions have to be taken when using electrotherapy due to the high conductivity and low tissue resistance. Nevertheless, although it is not contraindicated, electrotherapy is rarely used.

The suggested procedures include biostimulation with a laser beam. Its mechanism is not fully known, but it is claimed that it has an effect on vessel metabolism, increases DNA, RNA, and ATP synthesis, and intensifies protein production. It increases the transfer of calcium ions in the inner membranes and quickens collagen synthesis and enzymatic processes. The effect of laser light on tissue depends on the wavelength, power, radiation volume administered, duration, the number of procedures, and the impulse frequency. Tissue condition may affect the therapeutic outcome [16]. The tissue temperature should not rise more than 1°C. The procedure can be done during the acute, subacute, and chronic phases of the disease. The dose for children should be reduced by 30–60%. The most commonly used frequency is in the range up to 3000 Hz.

Another procedure that works on different levels is a low-frequency magnetic field. It permeates the whole body. According to reports it affects cell membranes and ion concentrations (there is a change in ion transport through protein pathways of cytoplasmic membranes and intercellular calcium ion concentration), oxygen utilization of the cell, some enzymatic activities, free-radical processes, DNA synthesis, and cell proliferation. It accelerates the process of wound and fracture healing. The shape of the impulses is chosen depending on the indications and the treated area. Rectangular impulses are used when bones are affected by the disease process, triangular in the case of articular cartilage, tendons, and ligaments, and a sinusoidal shape within nerves and muscles. This is another heatless method and can be used even at the acute stage of the disease process. In addition, it can be applied through a plaster cast, bandages, and clothing and when there are metal implants (not electronic). What is important is that a pulsating magnetic field of low frequency can be applied in children [16].

Cryotherapy is usually associated with the application of cold in a temperature range of ≤ 0°C. Local therapy is the most prevalent. Its actions include stimulation of cold and pain receptors, reduction of thermoreceptor activity, local vascular stenosis, changes in vasomotor activity, changes in sensorimotor nerve conductivity, reduced rate of metabolism, reduction in vascular activity, and a slowing of wound healing processes. The local application of low temperatures has an effect on blood vessels in the skin in which two phases can be distinguished. In the first phase it causes the ves-

sels in the skin and subcutaneous tissue to narrow. This is a defense mechanism protecting the organism from heat loss. However, the tissue temperature becomes somewhat reduced and metabolic processes slow. After 2–3 minutes of cold therapy the second phase commences, in which blood vessels distend. Congestion of the treated tissue then occurs. As a result, the following can be found:

a) an analgesic effect, as there is a reduction in nerve fiber conduction velocity, inhibition of nociceptors in the skin, partial or complete inhibition of C fibers, and a reduction in the release of pain mediators. The effect occurs after 3 minutes of therapy with nitrogen vapor in a temperature range of –180 to –110°C. It lasts for 3–4 hours;

b) an antihemorrhagic effect, reduction of bleeding, which prevents or limits the occurrence of hemotomas following trauma or inflammation; there is reduced lymph secretion;

c) lower tissue temperature, which in various, mainly inflammatory, processes becomes heightened;

d) altered muscle tone.

All these are important factors in the rehabilitation of children with hemophilia. In addition, as the procedure is short, it is well tolerated by patients. Local cryotherapy has another benefit in that it has no impact on the circulatory system [8].

Another group of procedures is electrotherapy. They are not usually applied to children as they carry some risk and also due to young patients' fear of the current. As patients grow older, TENS (transcutaneous electrical neural stimulation) and interferential current are employed. The analgesic effect is based on the gate mechanism within the spinal cord (gate control theory of Wall and Melzack) as well as the theory of endorphin release. To suppress or reduce pain, the conduction of pain impulses by peripheral nerves and those at the spinal cord level must be suppressed (the posterior horn gray matter). In the procedures, units with two or four electrodes are used. If the pain is localized, for example in the knee joint, the cauda equine which supplies the area has to be found. The positive electrode should be placed over it, in a paraspinal position. The negative electrode should be placed on the pathway of the nerve innervating the site where the pain is localized, between the site and the spinal cord. In the case of localized pain that is not radiating to the limbs, transversal positioning (through the joint) or symmetrical on both sides of the spine is acceptable.

Interferential currents are sinusoidal alternating currents of medium frequency, with the amplitude modulated at low frequency. They are induced when two currents of medium frequency flowing in separate therapeutic circuits interfere. Currents of medium frequency have a range 3500 to 10,000 Hz. As currents with slight differences in frequency overlap, a therapeutic stimulus of low frequency is induced. Two electric circuits and four electrodes are used in the procedure. The more the electrodes deviate from rectangular, the weaker the interference or there is no interference. The procedure's duration depends on the condition and usually lasts from 10 to 20 minutes. One or two frequencies can be used during one procedure. Using two (no more than that) frequencies helps to eliminate the risk of the tissue becoming used to the stimulus. It is one of the more pleasant electrotherapeutic procedures. With hemophilic patients the procedure should not last for too long as a hyperemic reaction should be avoided. Although the electrotherapeutic procedures are not contraindicated, they are rarely used in children. This may be because of children's fear of the current or the fact that the therapist cannot guarantee adequate safety. The child may move and the electrode could cause harm.

In general, phototherapy, which relaxes tissue, is contraindicated in people with hemophilia. Tissue overheating at the site of application might lead to further bleeding or extravasation. The only accepted phototherapeutic procedure is therapy with polarized light. The spectrum starts above the ultraviolet range. Therefore the skin tans and does not turn red. Polarized light is used which is polychromatic, non-coherent, low in energy, and UV-ray free. The soft light acts on various levels. It poses no risk to the eyes. The actions of polarized light are that it boosts the energetic activity of the cell membrane and mitochondria produce ATP. This causes cell activation and the cells' energetic potential increases, which in turn activates regenerative processes. In addition, regeneration of the tissue is observed after the therapy as well as improved healing rates and an analgesic effect. Polarized light can permeate into deeper levels of the organism. There it has a specific effect on the molecular order of cell membranes; it enhances cell metabolism and significantly accelerates regenerative processes and healing, produces pain relief, and inhibits inflammatory processes in the tissue. The procedures are usually performed once a day for 5–15 minutes. The light should fall at a right angle, 5 cm from the surface. With chronic joint pain, the light can be placed directly on the treated site. The skin temperature rises by about 1–3°C.

Kinesitherapy

Physical treatment can be performed at every stage of the disease. The choice of kinesitherapy depends on the child's condition. When planning

kinesitherapy one should adhere to general rules. The most desired are fun and descriptive activities adjusted to the patient's age. From age 1, manipulating, imitating, constructing, and didactic activities are fun activities which are introduced. Appropriate toys and equipment should be chosen. The activities should reflect individual needs and the rehabilitation program should change as the child makes progress and along with its psychomotor development. When a young patient lacks motivation to exercise, more attractive activities should be introduced. They should motivate the child to make independent controlled movements. In young children, nice toys can serve the purpose, and in older children a wide variety of activities performed during the therapy. The therapist's attitude, personality, and ability to make contact with the child are important factors. Gaining the child's trust is of paramount importance in facilitating progress in rehabilitation. The parents' cooperation is significant. Without it there will be no positive effect, especially in young children (infants). Parents who undergo training can continue the activities at home. Being systematic is essential [17].

At an early stage of disease, general rehabilitation of the upper and lower limbs can be performed. Special attention should be paid to the restricted mobility range in the elbow and knee joints that develops during the disease process. Props can be used as well, such as sandbags, dumbbells, and others. The activities should be varied as the child may grow weary of them. They should be fun and short, but frequent. The older the child is, the longer and less frequent the exercises can be.

Another example of exercises performed at every stage of disease are isometric activities. These are performed with muscle strengths of –1, +1 on the Lovett scale – the right exercises, and short isometric exercises at levels of 3, 4, and 5 on the Lovett scale. In this type of exercises, muscle attachments must be at maximal proximity to enable isometric contraction 5–7 s long, with the interval twice as long, and the number of contractions should depend on the functional state of the muscle (no more than three series of 10 movements each). The patient should be instructed how to tense the muscles on the healthy limb and then perform the exercises on the affected one. These sorts of exercises are usually suggested for patients as they are easy to perform at home and in the hospital. Pressing the popliteal fossa into the mattress while lying on the back or simulated lifting of the lower limbs in the same position are usually suggested. In this case, the quadriceps femoris, the extensor muscle of the knee joint, is trained. For the elbow the therapist puts resistance on the forearm and the patient tries to extend the elbow joint. The child should learn how to do the isometric exercises properly, as it is one of the easiest ways to prevent muscle atrophy. The trained muscle is better nourished, its strength is increased, and it is more active [17].

Both at the early stage of disease and later on, the exercises should reduce weight bearing. The therapist's hand or a slippery surface can serve this purpose, but the most popular are exercises with the UGUL system. The recommended muscle strength is –2 to +2 on the Lovett scale. Depending on the objective, one can distinguish 1) movement with a pause at the end of the movement range (increasing strength) and 2) a swinging movement (increasing the range of movement). This allows for joint movement in its full mobility range, which supports the correct flexibility of the muscles controlling the joint, ligament, or other. Axis and off-axis suspension can be employed. Axis suspension reduces weight bearing by the joint and off-axis helps or hinders movement, depending on the desired effect. The pain barrier is usually not crossed. With children it is more effective to measure the exercise output by the number of repetitions rather than the time as it is more motivating for the child in this way.

With children it is a good idea to introduce fun activities in the water. The water should not be too warm; it can be 30–36°C. Water is a good environment for the physical therapist to work in. It reduces weight bearing, relaxes muscles, and patients experience less pain. It reduces friction, which is perfect for weak muscles, and helps painful joints to move. It reduces irritability of the nervous system and improves nervous and muscular coordination. First the patient should become familiar with water, then there should be breathing exercises, learning to walk in the water, and swimming lessons if possible.

In the home environment the patient can use a stationary bicycle. This improves efficiency of the pelvic girdle, knees, and feet without putting excessive strain on them. The height of the saddle should allow flexion of the hip and knee joints. The load should be increased after about five minutes. In the case of the upper limbs it also possible to perform the exercises on the rotor, usually fixed to the wall of an UGUL cabinet or a ladder. In both cases the rule is the same: every movement should involve extension of the knee or elbow joint.

Diet

Apart from physical therapy, adequate nutrition is also essential. There is no ready diet for hemophilic patients. The diet should include lots

of fresh vegetables, fruit, wheat, fish, and lean meat. Nevertheless, arthropathy can be an obstacle in keeping a proper diet. The patients often experience fatigue and pain, which weakens appetite. In addition, swollen joints and lack of stability of the posture may discourage patients from preparing meals. In such case, simple meals have to be prepared whose preparation will not overburden the motor apparatus.

Hemophilic child should be under special care. Parents should undergo training on how to deal with trauma and bleeding and to minimize risk. The safety of a child learning to walk is paramount; as there will be many falls, there should always be a soft landing. Also, toys should pose no harm. As the child grows older it should learn to keep order and put away all toys not in use. Knee and elbow protectors are a solution, but children do not want to wear them [5].

Patients and Their Families Education

Parents and caregivers must be vigilant, but also make the child's life as carefree as possible. Once the caregivers learn to live with the disease, it is easier for them to instill in the child certain rules it has to adhere to. Everyone involved in the care of the child should know how to deal with difficult situations. There are workshops for parents and children aimed at providing them with support. Parents can learn how to make their children independent. The name of the organization is HELP and it can be found in all major Polish cities. Children who systematically adhere to the prophylactic program can be active in their peer groups, go on excursions, and not have to miss lessons due to bleeding. Parents do not have to give up work to provide round-the-clock care for the children.

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Conflict of interest: None declared

Received: 1.12.2008
Revised: 5.02.2009
Accepted: 18.02.2009