

AWARENESS BOOKLET ON
JOINT AND MUSCLE ISSUES AND
HEALTHY LIFESTYLE
IN HAEMOPHILIA

FOR

PATIENTS, THEIR FAMILIES, MEDICAL STAFF AND
GENERAL PUBLIC

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Bleeding into joints is the biggest problem in haemophilia

Part-I

1. Care and treatment of joints in Haemophilia

1.1. Introduction

Haemophilia is a bleeding disorder in which one of the factors, Factor VIII or factor ix is missing. This can cause external or internal bleeding anywhere in the body as a result of injury. People with haemophilia bruise easily and bleed for longer time after cuts, tooth removal, surgery or injury. Frequency and severity of bleeds depends on factor levels, age and physical activities. Even a small amount of factor in blood is enough for daily activities.

Most of the bleeds are a result of trauma. In severe haemophilia (F<1%) bruising or bleeding can start without a cause. This called spontaneous bleeding. Babies bruise easily when they start walking. When the child grows, bleeding episodes become more common. **Bleeding into joints is the biggest problem in haemophilia.**

Joint is a place where two bones meet, edges of the bones are covered with a soft bone (cartilage) which performs the function of a shock absorber. Bones are joined with each other by a Joint Capsule which is covered by a Synovial Membrane which contains lot of blood vessels. Rupture of these vessels cause bleeding. Synovial membrane produces a fluid which helps the joint in its mobility. The space between the two bones is called Joint Space. Bleeding into joint space causes swelling, pain and difficulty in moving the joint. These symptoms first occur in pre-school years.

1.2. Common joint bleeds

Knees, elbows and ankle joints are the most affected joints but shoulder and hip joint can also be affected. This is due to many factors.

- Knee, elbow and ankle joints move only in two directions, bent and straight, while hip and shoulder joints can move freely are often not affected by bleeds.
- Knee, elbow and ankle are not protected by muscles while hip and shoulder joints have strong muscles covering them.

1.3 The date of Haemophilia Center, PIMS shows the following distribution of bleeds:

Knee - 65.5%

Elbow - 23.6%

Knees, elbow and ankle joints are the most affected joints but shoulder and hip joint can also be affected

Ankle -	5.5%
Hip -	1.8%
Shoulder-	3.6%

1.4. Identification of Joint Bleeds

Before any visible effects, burning and tingling can be felt. Skin appears red and warm. When the joint fills with blood, it starts to swell. There is pain on movements of the joint.

1.4.1. Affects of Joint Bleeds

Acute bleed occurs in a healthy joint. Blood is absorbed by an enzyme produced by synovial membrane within two weeks. This enzyme also damages the cartilage. More than 2-3 episodes of bleeding per month can reduce the capacity of the synovial membrane to absorb blood completely and blood can stay in joint for a long period. This is called Chronic Haemarthrosis. Synovial membrane swells in case of chronic haemarthrosis. Swollen synovium can burst due to the movement of joint and cause bleeding. Thus the joint becomes target joint. With repeated bleeding, the synovial membrane becomes chronically inflamed causing the joint to appear extremely swollen. Swelling usually gives the feeling of tension in the muscle and is not very painful but muscles surrounding the joint bleeds can cause irreversible damage to the joint cartilage (haemophilic orthopathy) with loss of motion, limb deformity and pain.

1.5. Different stages of chronic haemophilic orthopathy

Synovial information

Chronic haemarthrosis

Treatment, management and prevention of bleeds are the key to improving health and quality of life for a patient with haemophilia.

Chronic Synovitis

Invasion of joint cartilage

Chronic Haemophilic Orthopathy

1.6. Prevention of joint bleeds

This can be done by:

- Recognizing bleeds early
- Stopping bleeds as quickly as possible with treatment
- Exercising, when bleeding has stopped

1.7. Managing joint bleeds

Treatment, management and prevention of bleeds are the key to improving health and quality of life for a patient with haemophilia. Simple and easy management of bleeds at home are first-aid and RICE (Rest, Ice, Compression, Elevation).

1.8. Home management

- **Rest** – It is very important to give rest to the bleeding joint e.g. use crutches or wheel chair in case of bleeding in leg.
- **Ice** – constricts blood vessels and decreases pain.
- **Ice treatment** – wrap crushed ice or ice cubes in a wet towel and leave it in place for 5 minutes. Repeat the procedure 3-4 times daily with 10 minutes interval between the treatments.
- **Gel packs** – treatment by frozen gel packs is same as the treatment with ice packs. Avoid contact with skin.
- **Four stages of sensation in ice treatment** – cold, pain, burning, slight numbness. Leave the ice on the affected area until you feel numbness.
- **Immobilization** – immobilization can be done with a splint or half cast. The joint should be kept immobile for a limited period of time otherwise it can weaken the muscle.
- **Compression** – joint can be wrapped in an elastic bandage. If the area is properly wrapped, it can minimize swelling and the joint feels comfortable. Gentle compression can limit bleeding.
- **How to use the elastic bandage** – start from one extreme of the limb or body part, wrapping towards the body. Overlap the bandage by 1/3 of its width. Increase the overlap to 1/2 its width with each turn around the limb. As you proceed, start wrapping the bandage less tightly. Repeat the procedure 2-3 times.
- **Elevation** – raising the injured part above heart level will decrease pressure in the area and help minimize bleeding and swelling.

1.9. Treatment at hospital

It is important to give treatment as quickly as possible to prevent long term damages.

1.9.1. Blood clotting products

- **Factor concentrates**

Treatment is done by replacing missing factor. Factor replacement should be done daily or on alternative days until the joint returns to normal. This is the most appropriate and safe treatment.

- **Cryoprecipitate**

It is prepared from plasma and consists of FVIII and vWF. Therefore it is used for treating deficiencies of FVIII and vWF. It should not be used in FIX deficiency.

- **FFP**

It is prepared by removing WBC's, and platelets and then freezing the plasma. **It contains all clotting factors and can be used in treating all kinds of bleeding disorders.**

- **Whole blood**

Not effective in haemophilia treatment.

1.9.2. Aspiration of haemarthrosis

Aspiration should be considered under following circumstances only:

- When the joint is extremely swollen and painful.
- A joint which has not improved even after 2-3 days of treatment.
- Aspiration should always be done under aseptic conditions.
- Aspiration from elbow, knee and ankle can be done in OPD.
- Aspiration of hip and shoulder joints should be done under radiographic control in the operation theatre by an orthopedic surgeon.
- Whenever aspiration is done, the factor levels should be 30-50% for at least 2-3 days.
- If factors are unavailable, aspiration should not be done.
- Aspiration should not be done if there is an infection.

Do not use aspirin or brufen. Use paracetamol instead

1.9.3. What other treatments might help

Joint with numerous bleeds is termed a target joint. Following treatments might be recommended.

- **Synovectomy**

It is surgical removal of synovial membrane to prevent future bleedings. Synovectomy should be done before any irreversible damage is done to the joint. An ideal candidate for synovectomy is a person who has frequent haemarthrosis in a target joint, factor treatment has failed and there is no visible damage to the joint in X-rays.

- **Synoviorthesis**

A substance is injected directed into the joint to cause the synovium to make scar tissue which stops bleeding. 70-80% patients show excellent result.

- **Steroids**

If a joint is extremely swollen, your doctor can give you steroids in form of tablets or injections which can help reduce swelling.

1.9.4. Prevention of joint deterioration

The goal of treatment is to prevent joint deterioration. Following are the ways to do so:

- Prompt treatment of each bleed
- 4-5 days of rest to the affected joint
- Ice therapy to relieve pain and reduce extent of bleeding
- Exercises to keep joint moving and build the strength of muscles

Regular exercises help prevent bleeds and joint damage

Part-II

2. Muscle bleed and its treatment

- Muscle bleeds can happen anywhere in the body as a result of a direct blow, stretching of a muscle, sprain or intramuscular injection.
- Usually a group of muscles is involved.
- During the bleed, muscle feels stiff and painful.
- Bleeding causes swelling, warmth and pain in the muscle. It can also press nerve and the patient feels tingling and numbness. If treatment is not given on time, it can cause permanent muscle and nerve damage and can lead to disability.
- Bleeding into knee, elbow and hip is common.

Prevention of muscle wasting

- Rehabilitation to prevent problems.
- Physical therapy to maintain good function. This should be gentle and consistent so that muscle can stretch and move normally again.
- Early treatment with factors.
- Restrengthening of muscle by different exercises.

2.1. Muscles which should be treated immediately in case of bleeding

Following muscles are affected badly if treatment is not given on time.

- Iliopsoas
- Gastronemius
- Fore am flexors

2.1.1 How to recognize a muscle bleed

- Redness
- Warmth
- Bruising (not present in deep blees)

2.1.2 Long-term effects of bleed

- Scarring
- Weakness
- Permanent damage to muscles, joints, nerves and deformity

2.2 Treatment

- Clinical evaluation and monitoring
- Immediate factor replacement
- Rest, ice treatment, elevation etc

2.3 Pain management

- Use of analgesics can reduce pain
- Do not use use aspirin or brufen. Use paracetamol instead.

If pain is not relieved, following medicines can be used:

Drug	Adult Dose	Paediatric Dose
Co-Proxamol	2 tab 3-4 times daily	Not recommended for children
Codeine	30-60 mg every 4-6 hours	0.5-1mg/kh every 4 hour
Temgesic	0.8 mg every 6 hours sub-lingual	Not recommended for children
Tramadol	50-100 mg every 6 hours	Not recommended for children
Pentazosine	30-60 mg I/V 3-4 hours 50	

	mg P.O. 3-4 hours	
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Consult your haematologist before starting any sport

Part-III

3. Healthy Lifestyle

In the life of haemophilic, apart from medical treatment and vaccination, exercise, physical activities, good nutrition etc. are very important for good health. It is good for a person to stay active because it has good affects on a person, physically and psychologically. There are numerous benefits of doing regular exercises.

- Strengthens muscles, joints and bones
- Builds self-esteem
- Increase participation in team work
- Increases good cholesterol
- Improve performance at school
- Reduces weight
- Reduces depression
- Reduces the probability of diabetes, blood pressure, cancer, bleeding and heart diseases

3.1 Exercises and physical activities in haemophilia

Some patients with haemophilia avoid Exercises because they think that it may cause bleeding. Whereas regular exercises help prevent bleeds and joint damage. It is good for building strong bones and strengthening muscles that support and protect joints. This strengthens the joints and reduces the probability of bleeding.

It is important to restart exercise as soon as the pain is reduces in the joint. This improves the mobility of the joint.

Recommended Sports:

Swimming, table tennis, walk, badminton, cycling etc.

3.2. Benefits of sports for patients with haemophilia

Physical activities nourish the child physically, socially of parents of not letting the child to participate in activities because of bleeding. For example, in case

of ankle bleed, running is not a good option but swimming is. Activities in which a child is involved should be determined by the child's condition and if bleeding starts, the child should stop playing. It is important to give factor treatment within 24-48 hours of bleeding. Following RICE can also help in reduction of swelling and pain.

Regular checkups are necessary (children every 6 months and adults annually)

3.3. Continue exercises after bleeding

There are no particular instructions on starting exercises after bleeding. All patients are different from each other and before making any decision regarding exercises, consult your haemophilia centre. Some people take less time than others to get their muscles mobile. Any bleeding episode which can be treated by one treatment only and does not hinder daily activities is not a very worrisome thing. But if multiple treatments are needed and muscles start to get numb, consult your haemophilia centre before starting any kind of exercise or sport. If a joint is a target joint, repeated bleeding may hinder the treatment and can cause permanent damage to the joint. Regular bleeding episodes can make the joint immobile and limit involvement in physical activities.

3.4. Sports recommended for patients with haemophilia

- Swimming
- Table tennis
- Walking
- Badminton
- Cycling

3.5 Sports not recommended for patients with haemophilia

- Football
- Wrestling
- Boxing
- Motorcycling
- Cricket
- Hockey
- Karate

Consult your hematologist before starting any other sports.

3.6. Good nutrition

Regular exercise and a healthy diet is the key to building strong muscles and stable joints. Healthy body weight is important so as not to put extra strain on joints. Consult your haemophilia centre for proper and healthy diet.

Contact your Haemophilia Treatment Centre (HTC) in case of injury. Do not wait for swelling or bleeding to stop

Sports teach team work to children

3.7. Medical checkups

Regular medical checkups including examination of joints and muscles are essential to maintaining good health. **Regular checkups are necessary (children every 6 months and adults annually).**

3.8. Prevention is better than cure

Patients with haemophilia should take part in different physical activities depending upon their age and under supervision. After factor treatment, it becomes easy for haemophiliacs to take part in different activities but the probability of bleeding after injury is still there. Contact your haemophilia centre immediately after injury without waiting for the bleeding to stop.

3.9. How can parents help?

Parents of children with haemophilia should encourage children's natural abilities and help adapt to the realities of having chronic disorders. This can help in living a normal life. Activities in which a child is involved should depend on his/her age because it has a number of benefits:

- Child learns to discipline himself
- These activities prepare child for the coming future
- Builds self-confidence

3.10. For parents of infants

Child starts learning from his environment when he is young. Therefore it is necessary that parents provide him with soft toys. When the child starts crawling provide support to him so he can learn to walk. When he starts walking, encourage him to run and jump and provide him with toys with wheels. Keep them under your supervision. Put cloth or foam on edges of the table, carpet the floor and put a gate on the stairs. When a child starts playing with other kids, teach him how he can take his own care and supervise him. **Keep the child under supervision when he is playing.**

3.11. For parents of school going children

Sports are important for school going children and provide him with opportunities. Research shows that strong bones and muscles help in minimizing bleeding episodes and sports and exercises nourish the child socially. **Sport teach team work to children and they understand the difference between winning and losing.** Appreciation of child's success encourages the child and builds self-confidence. As children get emotionally affected with bleeding, as his self-confidence develops bleeding episodes reduce. When children start going to school, they should know that in case of bleeding, they have to tell their teachers who will then contact the parents.

All patients should have Hepatitis B vaccination

3.12. Vaccination and medication

- All patients should have Hepatitis B vaccination
- This should be given sub-cutaneously and not intramuscularly
- All family members should also have hepatitis B vaccination
- Consult your haematologist before taking any drugs

3.13 Prepare yourself and make wise decisions

Patients with haemophilia should be encouraged to take part in sports and exercises. Following points are important:

- People with bleeding disorders have a problem with their coagulation system and bruising is common. People with severe haemophilia can bleed spontaneously in joints and muscles.
- Parents should provide their child with such an environment in which he can grow normally and take part in different activities. Children who don't take part in sports and whose parents are very protective bleed more often. Self-esteem and independent lifestyle affects the patient positively and occurrence of bleeding episodes is reduced.
- Parents are the biggest hurdle for a child who wants to play. Parents need to talk to their children about the dangers they face and should make sure that the child must take safety precautions.
- Parents with bleeding disorders should talk to their parents, haemophilia center's team, school staff etc. about the disease they have so they know about their disease and provide immediate treatment in case of injury.
- It is very important for the patient of haemophilia to be physically fit. Strong muscles have the capability to resist injuries. Exercise and sport makes muscles strong.
- There are numerous benefits of exercises and sports. Therefore children should participate in sports from their childhood.