MUSCULOSKELETAL PROBLEM&STRESS IN PARENTS AND PERSONS WITH HEMOPHILIA

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INTRODUCTION:

"Let me die, I would like to die instead of tolerating heart bursting pain"

These horrifying words spill from the lips of almost every person with hemophilia due to huge is an inherited lifelong disorder with repeated episodes of bleeding. The bleeding commonly occurs in big joints like knee, elbow and hips. A child is born with it. One of the clotting factors in the blood is almost absent named factor VIII or IX. It means, whenever there is bleeding does not stop. Bleeding can also happen anywhere inside the body spontaneously even without any injury and mostly in the joints. Frequent bleeding makes the joint deformed. In short Hemophilia is a lifelong incurable disorder, but fully manageable even in its severest form. In case of any bleeding, Anti Hemophilic Factor/Christmas factor is required, which is very expensive and imported from abroad.

Rationale:

Hemophilia is a rare inherited bleeding disorder in which the blood does not clot normally, because of low level of clotting protein called factor VIII & factor IX. Hemophilia affects one in every 5,000 males across the world. Out of these 60% are severe Hemophilic. In this concern, even the upper class families cannot afford the cost of treatment. It is a lifelong X chromosome linked genetic bleeding disorder in which patients bleed excessively. The Person suffering from hemophilia has prolonged bleeding either internally or externally. The most common internal bleeding sites are joints and muscles whereas intracranial bleeds, G.I. bleeds, brain bleeds etc. are life threatening. The external bleeds include small cuts, wounds, abrasions and bruises.

Types of Hemophilia:

There are three types of hemophilia-

1)	**Deficiency of Factor**	**-**	**VIII is called**	**-**	**Hemophilia**	**A**
2)	**Deficiency of Factor**	**-**	**IX is called**	**-**	**“**	**B**
3)	**Deficiency of Factor**	**-**	**XI is called**	**-**	**“**	**C**

Causes of Hemophilia:

Person with hemophilia has one or more of the 10 (I, II, V, VII, VIII, IX, X, XI, XII, and XIII). These clotting factors either missing or in less amount into the blood. The deficiency of any clotting factors in the blood usually Factor VIII or Factor IX cause hemophilia. Because “X” chromosome governs both of the proteins (clotting factors) VIII / IX.

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Sufferers of Hemophilia-

Hemophilia affects males almost exclusively and is found in all populations irrespective of caste, creed, religion or geographical variations. Thus, males are sufferers of Hemophilia and females are the carriers who can transmit affective gene of hemophilia to the next generation.

Hemophilia is manageable through regular infusion of AHF. The treatment is in the form of intravenous injections of a life saving medicine called "Anti Hemophilic Factor". It is extracted from human blood, hence it is very expensive. Now recombinant factor is also available. It is not manufactured anywhere in India. HFI is only the organization, which imports AHF from abroad countries and distributes to PWH at subsidized rates (and even free) through structured chapters. Unfortunately, there is no financial support from the Government to make it affordable for the PWHs.

When a patient of hemophilia comes to the hospital with major bleed..., his survival depends on immediate administration of the life saving Anti Hemophilia Factor (AHF). For an intracranial bleed, a patient may require as much as 25,000 Units of AHF costing Rs. 2-3 Lakh and this many even go up to 50,000 units plus of AHF costing Rs. 4-6 lakh for any major surgical interventions.

A few of the facts of Hemophilia…

- It is mostly found in male child only.
- The mothers are carrier and transmit affected X chromosome into the new born baby.
- There is one more cause of Hemophilia: Spontaneous mutation.
- It may be inherited to next generation, if not properly managed.
- It's found in all cast, community and religion.
- Occurrence: On every 5000 male populations, a child with Hemophilia is born.
- Everyday, 4 (four) children with Hemophilia are born is India.
- Symptom: Colourful bruises, non-stop prolonged bleeding, swelling into the joints/muscles followed by capacious pain.
- It's related to X-Chromosome and its containing genes.
- The affected genes do not produce adequate amount of factor VIII or IX to make the fibrin net to stop bleeding.
- There are 13 clotting factors in the blood, which are responsible to check to bleeding during the bleeding episodes.
- The factor VIII or IX is very expensive and imported from abroad.

Physical condition:

***** Rate of total disability (in percentage) in hemophilia-
* Sever disability- 15%-20% (Hip, Knee, Elbow, Ankle)
* Moderate disability- 40%-50% (Knee, Elbow & Hip)
* Mild disability- 20%-25% (Knee & Elbow)

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* Without disability- <05% (in case of mild or moderate)
* Rate of Mortality in hemophilia- 10%-15% (In the lack of adequate treatment within the time frame)

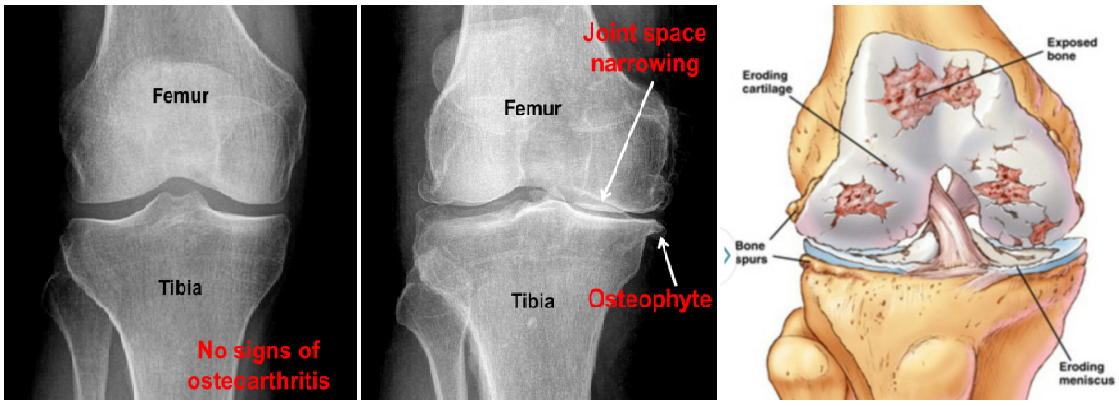
* Approx. 70% Hemophilia occurred due to inheritance.
* Approx. 30% Hemophilia is acquired called Spontaneous mutation.

Treatment: Application of RICE therapy, Replacement of factor, Physiotherapy. What not to do? Avoiding muscular injection, aspirin or NSAIDs as well, Inadequate physical exertion to avoid bleeding.

Prevention of Hemophilia : Get each pregnancy tested named prenatal/antenatal diagnosis of carrier mother. A Hemophilic must avoid the birth of female child.

HOW DIVYANGATA(MUSCULOSKELETAL PROBLEMS)

comes in HEMOPHILIA…Due to the frequent bleeding inside the joints and muscle, it makes the joint stiff/deformed and muscular contracture as well.



The most important clinical strategy for management of patients with hemophilia is the avoidance of recurrent hemarthrosis by means of continuous, intravenous hematological prophylaxis. When only intravenous on-demand hematological treatment is available, frequent evaluations are necessary for the early diagnosis and treatment of episodes of intra-articular bleeding. The natural history of the disease in patients with poorly controlled intra-articular bleeding is the development of chronic synovitis and, later, multi-articular hemophilic arthropathy. Once arthropathy develops, the functional prognosis is poor. Treatment of these patients should be conducted through a comprehensive program by a multidisciplinary hemophilia unit. Although continuous prophylaxis can avoid the develop.ent of the orthopedic complications of hemophilia still seen in the twenty-first century, such a goal has not, so far, been achieved even in developed countries.

Therefore, many different surgical procedures such as arthrocentesis, radiosynoviorthesis (radiosynovectomy) (yttrium-90, rhenium-186), tendon lengthenings,

alignment osteotomies, joint arthroplasties, removal of pseudotumours, and fixation of fractures are still frequently needed in the care of these patients.

The Acute Joint Bleeding (Acute Hemarthrosis):

The most common sites of bleeding in hemophilia are the joints and muscles of the arms and legs. The vast majority of bleeding occurs into the joints of the extremities, although bleeding may occur into most any joint. There appears to be a predilection for large joints, namely the ankles, knees, hips, elbows and shoulders. Spontaneous bleeding into joints is limited to persons with severe hemophilia. Bleeding may occur following trauma in patients with mild and moderate hemophilia. Clinical manifestations of factor VIII and factor IX deficiency are indistinguishable. I would like to point out why bleeding occurs into the joint cavity and the reasons that the characteristic arthritis termed "hemophilic arthropathy" develops following recurrent bleeding into the joint. König pointed out that there were three clinical stages to the development of hemophilic arthropathy. He termed the first the “recurrent bleeding” stage. Following this, there is an inflammatory response and he termed this the panarthritis stage. The third and last stage he called the fibrosis stage and contracture commonly known as arthritis. He was able to point out that bleeding is not a common manifestation of the last stage.

The clinical picture of joint bleeding is characterized by pain, swelling and limitation of motion. Frequently the patient states that he knows he is bleeding prior to any of these findings. This period has been termed the “aura”. It may be accompanied by mild stiffness and sometimes the patient describes a feeling of tightness or tingling. It is believed that this corresponds to the time when the bleeding is limited to the synovium. Once the bleeding fills the joint, it becomes warm, swollen and tense. Limitation of motion and secondary muscle spasm follow. When treatment is started early, the bleeding will stop quickly and the symptoms may recede quickly. However, it is very common for the symptoms to resolve slowly and at times the bleeding may recur despite what seems to be adequate treatment. A joint that displays a tendency towards recurrent bleeding has been termed a **"target joint"** by Aronstam. Once a target joint is established, complete resolution is possible, but more commonly there is a slow response to treatment and arthritis will develop.

The Chronic Swollen Joint (Recurrent Bleeding and Synovitis)-

If joint bleeding is not adequately treated, it tends to recur. The inflamed, swollen synovium bleeds more easily than normal synovium and causes further swelling and inflammation. This vicious cycle must be broken to prevent the iron within the blood and the enzymes from destroying the cartilage, leading to the development of arthritis. Synovitis or recurrent bleeding can be differentiated from an acute hemarthrosis in that the swelling does not respond to a single infusion of factor. The joint is less painful than with an acute hemarthrosis and the range of motion is frequently not limited. It must be remembered that cartilage has a limited ability to repair itself. The synovitis may not be painful, but the destruction is insidious and cumulative and, therefore, the condition must be treated as

vigorously as a- painful hemarthrosis. Prophylaxis, or preventative replacement of the missing clotting factor, for three to six months is indicated. The intermittent use of steroids helps reduce the Inflammation within the joints. A dose of 1 mg/kg/day of Prednisone for one week should be followed by a dose of .5 mg/kg/day for a second week. If the synovitis is recalcitrant, the course maybe repeated in three to six weeks. Aspiration may be useful initially to control the swelling with the joint and should be undertaken after prophylaxis has been started but before steroids are used. Immobilization is useful at times but must be used in conjunction with physical therapy to prevent atrophy and loss of range of motion. A lack of response of the recurrent bleeding of synovitis after three to six months of prophylaxis and treatment is usually considered an indication for more aggressive intervention. Open surgical synovectomy, arthroscopic synovectomy and radioactive synovectomy should then be considered. The indications and management of these procedures will be considered in a later monograph.

Purpose:

The purpose of this study is to discuss the level of stress in Hemophilia children and their parents due to musculoskeletal problems.

Review of related literature:

There has been increased attention in the literature about stress among persons with Hemophilia and his parents due to the musculoskeletal problem. It has been evident that musculoskeletal problem is the most stressful experience in Hemophilia. Just as adults feel depression and stress, children and teenagers do too. Children with a bleeding disorder may feel sadness about their bleeding disorder if they aren't participating in activities in the same way their peers do. Of course, depression and stress may not be related to the bleeding disorder at all. Children with bleeding disorders undergo the same stress that other children do, such as experiencing their parents' divorce, moving, or bullying. Parents and caregivers who notice a change in a child's behavior, such as crying or withdrawal, should open a dialogue with the child and seek professional help, if needed. Children and teens that are experiencing depression or stress should be reassured that they are not alone. There are people who can help and steps they can take to feel better.

WHAT IS STRESS:

Stress is the body's natural defense against predators and danger. It flushes the body with hormones to prepare systems to evade or confront danger. This is known as the **"fight-or-flight" mechanism**. When we are faced with a challenge, part of our response is physical. The body activates resources to protect us by preparing us either to stay and fight or to get away as fast as possible. The body produces larger quantities of the chemicals cortisol, adrenaline, and nor adrenaline. These trigger an increased heart rate, heightened muscle preparedness, sweating, and alertness. All these factors improve the ability to respond to a -

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hazardous or challenging situation. Factors of the environment that trigger this reaction are called stressors. Examples include noises, aggressive behavior, a speeding car, scary moments in movies, or even going out on a first date. The more stressors we experience, the more stressed we tend to feel.

HOW MUSCULOSKELETAL PROBLEM AND STRESS IS CONCERNED IN PERSONS WITH HEMOPHILIA AND PARENTS-

When person with hemophilia starts becoming physically disabled (Musculoskeletal problem) due to the frequent bleeding inside the joint,they gradually become stressed. Some time, it has also been noticed that the stress during the exam periods enhances the internal bleeding into the joints. Almost in every internal or external bleeding, they are scolded and harshly treated. Due to the frequent ill treatment by most of the parents, they become stressed.Their daily physical life becomes hampered gradually. Their daily routine activity gets bored. They start understanding themselves unproductive and load on the family & society as well. Their survival becomes tougher.

IMPORTANCE OF STRESS MANAGEMENT IN HEMOPHILIA AND ITS FAMILY MEMBERS:

Stress can be brought on easily with life changes. For this reason, people suffering from chronic diseases like hemophilia may suffer from more stress than others on a regular basis. Patients and/or patients' families may suffer from emotional, mental and physical stress. Stress can come from the diagnosis of the disease, financial woes, physical discomfort and fear of bleeding, among other things. t has been suggested that patients compose a list of whatever causes stress in a typical day or week. They should then categorize the listed items into two groups: stressors they can change and stressors they cannot change. There are things in their lives they will not be able to control, but they can learn to manage the stress allowing them to function most effectively and normally. With the stresses they cannot control, patients should try having a positive perspective and understand that some circumstances are beyond their control. While this is difficult, letting go of the things they cannot control will help them release their burdens.

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Identify:

Common negative stress reactions include anger, depression, negativity, pain, smoking, crying and overeating. The clinic suggests that you try management strategies to reduce the chances of these reactions by taking up a hobby, scaling back on work, being prepared should a bleed occur, reaching out to others when stressed, getting enough sleep and seeking professional help if needed.

Simplify:

A few of its helpful hints include for reducing stress are:

• Try to exercise and eat healthy. This can help you deal with stress more effectively.
• Learn to balance the stress with relaxation. Come up with a few relaxation techniques, ones that fit your lifestyle best.
• Face life with a positive outlook; try laughing at yourself sometimes, and laugh in general.
• Try asking for help from friends and family instead of doing everything yourself; learn to say no to things you do not have enough time to accomplish or work on.

Untreated chronic stress can result in serious health conditions including anxiety, insomnia, muscle pain, high blood pressure and a weakened immune system. Research shows that stress can contribute to the development of major illnesses, such as heart disease, depression and obesity. But by finding positive, healthy ways to manage stress as it occurs, many of these negative health consequences can be reduced. Everyone is different, and so are the ways they choose to manage their stress. Some people prefer pursuing hobbies such as gardening, playing music and creating art, while others find relief in more solitary activities: meditation, yoga and walking.

The Role of Psychosocial aspect for Stress of persons with Hemophilia:

Psychologists, social workers, and counselors havedifferent expertise and strengths. In HTCs with theresources for a multidisciplinary team, there areindividual staff members who specialize in these areas of expertise. However, in centres with limitedresources, other healthcare professionals may be theones to provide psychosocial support. Therefore, his monograph does not differentiate between psychologists,social workers, and counselors, and

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HT Chealthcare professionals who take on the psychosocialsupport role in their absence (often nurses).Ideally, a hemophilia treatment centre should havea multidisciplinary team with a specialist for eachaspect of care. While this is not always possible wheresources are scarce, there are still opportunities forpsychosocial care by other healthcare providers.Regardless of the resources available, all HTChealthcare professionals should be aware of the psychosocialissues that may arise with hemophilia andother bleeding disorders. To provide optimal care,healthcare professionals need to be able to identifyissues and challenges related to having a bleedingdisorder that may be affecting their patients' cognitive and emotional development.A bleeding disorder is a chronic condition thatimposes limitations, but it can also represent anopportunity to bring about positive change through learning and self awareness. There are differentways of experiencing and coping with pain andchronic illness. Some people with hemophilia mayfocus on the emotional challenges, while others findways to better cope with their situation. Short-termpsychotherapy, alternative therapies, and socialservices can help individuals with hemophilia copewith symptoms and limitations and develop ahealthy sense of self.

The role of psychosocial support:

Psychosocial support in the medical setting helpsindividuals gain a personal understanding of hemophilia.Healthcare professionals provide psychosocialsupport by helping patients and their families developstrategies to cope with physical, mental, emotional,and social challenges related to having a bleedingdisorder. This includes providing education, individualand family counseling, resources, and communityreferral services. The ultimate goal is to empowerpeople affected by hemophilia and other bleedingdisorders to manage their circumstances and challengesautonomously. Quality of life depends large Lyon the ability to adjust to having a chronic disorder and the challenging circumstances that may arise.In psychosocial care, it is as important for healthcareprofessionals to learn from their patients as it is forthem to provide guidance. Cultural values and socioeconomicfactors affect how psychosocial issues areexperienced and addressed. Other important issues forpeople with bleeding disorders include accessibilityof the HTC, availability of factor replacement productsand prophylactic therapy, access to physiotherapy and corrective surgery, and support from hemophiliaassociations.

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