



## **Orthopedic Complications and Treatment Related to Chronic Hemarthrosis**

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### **CHRONIC JOINT ARTHROPATHY**

The term “target joint” is used frequently in hemophilia care to describe a joint that has had recurrent bleeding episodes. The definition of target joint varies, but a common definition is four bleeds into the same joint in a six-month time period. The most common target joints are the knees, ankles, and elbows, although patients can develop targets in other joints including the hip, wrist, and shoulder. Once a target joint is established, there usually is persistent synovial thickening and joint effusion with limited range of motion. (1) Recurrent hemarthrosis usually results in some type of permanent damage to the joint. This damage can occur in a single joint or in multiple joints; the number of joints involved may determine the best course of treatment for a particular patient. Permanent joint damage caused by chronic hemophilic arthropathy can lead to decreased work capacity and may eventually lead to permanent disability.

Hemophilic arthropathy can develop rapidly or over the course of many years. In the early stages some of the symptoms include joint swelling or bogginess, muscle atrophy and weakness, and limited range of motion. Early in the course of the disease, it may be difficult to distinguish between chronic synovitis and an acute hemarthrosis. In the end stages of hemophilic arthropathy there is increased pain, joint destruction, and loss of range of motion. Treatment is dependent on the stage of arthropathy and the location of the affected joint. (2)

### **ASSESSMENT**

#### **HISTORY AND PHYSICAL**

The assessment of chronic hemophilic joint arthropathy begins with a good history of the involved joint including when problems with that joint began, the frequency of hemorrhage in the past, current bleeding history during the previous six to twelve months, loss of motion, pain, and gait abnormality. The pain associated with hemophilic arthropathy is similar to pain experienced by a patient with chronic arthritis and does not change with factor infusions. The history should also include a functional assessment of the patient’s ability to perform activities of daily living.

A musculoskeletal evaluation by a physician, nurse, or physical therapist is essential to determine loss of range of motion and decreased muscle strength in the joint being evaluated. Serial measurements of range of motion (ROM) over time are very helpful in determining when an intervention is necessary. See Table 1 for the most common changes in target joints of persons with hemophilia.

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**Table 1: COMMON CHANGES IN TARGET JOINTS (3)**

<b>JOINT</b>	<b>TYPICAL CHANGES</b>
<b>KNEE</b>	<ul style="list-style-type: none"><li>• knee flexion deformity with quadriceps atrophy</li><li>• posterior subluxation, valgus, and external rotation of the tibia</li><li>• hypertrophy of the medial femoral condyle</li><li>• squaring of the patella</li><li>• widening of the intra-condylar notch</li></ul>
<b>ELBOW</b>	<ul style="list-style-type: none"><li>• enlarged olecranon fossa with decreased flexion and extension</li><li>• enlarged radial head resulting in decreased forearm rotation</li><li>• subchondral bone cysts especially on the proximal ulna</li></ul>
<b>ANKLE</b>	<ul style="list-style-type: none"><li>• flattening of talus</li><li>• valgus deformity</li><li>• osteophyte formation</li><li>• calf atrophy</li><li>• loss of dorsiflexion</li></ul>
<b>HIP</b>	<ul style="list-style-type: none"><li>• avascular necrosis looking like Perth's disease</li><li>• cystic changes in the superior weight-bearing area of the femoral head</li><li>• osteophyte formation</li></ul>
<b>SHOULDER</b>	<ul style="list-style-type: none"><li>• pronounced osteophyte formation</li><li>• small atrophic head of humerus with varus deformity</li><li>• rare subchondral cysts</li></ul>

### **RADIOGRAPHIC EVALUATION**

In the past, the most frequent form of radiologic evaluation of hemophilic joint arthropathy was the conventional radiograph. Today a wide variety of radiologic imaging studies are used to evaluate hemophilic arthropathy, including ultrasound, CT scan, and magnetic resonance imaging (MRI).

Conventional radiographs are most helpful in outlining bone and joint changes. These include the development of bone cysts and erosions, presence of osteophytes, and narrowing of the joint space. It is also possible to visualize overgrowth that has taken place at the bony epiphysis resulting in a “knobby”-looking joint.

There are two radiograph classification systems used to grade the joint changes on plain X-rays, the Arnold-Hilgartner scale and the Pettersson Radiologic Classifications scale. The Arnold-Hilgartner classification is based on radiographic and clinical findings and attempts to separate the joint changes into stages that have surgical significance (Table 2). (4) The Pettersson classification is based on the presence or

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absence of radiographic changes that can be assessed quantitatively (Table 3). (5) The degree of X-ray changes may help to determine appropriate orthopedic intervention.

**Table 2: ARNOLD-HILGARTNER CLASSIFICATION OF HEMOPHILIC ARTHROPATHY (4)**

<b>STAGE</b>	<b>CHARACTERISTICS</b>
<b>I</b>	<ul style="list-style-type: none"><li>• No visible skeletal abnormalities</li><li>• Soft tissue swelling secondary to hemarthrosis or bleeding into the soft tissues in and around joint</li></ul>
<b>II</b>	<ul style="list-style-type: none"><li>• Osteoporosis, especially in epiphyses and their overgrowth</li><li>• Joint integrity maintained, with no narrowing of cartilage space and no bone cysts</li></ul>
<b>III</b>	<ul style="list-style-type: none"><li>• Presence of disorganization of joint but no significant narrowing of cartilage space so that articular cartilage is usually preserved</li><li>• Synovium may be opacified, with hemosiderin deposits present</li></ul>
<b>IV</b>	<ul style="list-style-type: none"><li>• Narrowing of joint space and evidence of cartilage destruction in addition to stage III changes</li></ul>
<b>V</b>	<ul style="list-style-type: none"><li>• Fibrous joint contracture, loss of joint space, extensive enlargement of epiphyses, and substantial disorganization of joint structures</li><li>• Marked restriction of joint motion</li><li>• Little recognizable synovial tissue found when joint is opened</li><li>• Absence of articular cartilage</li></ul>

Adapted from Hilgartner et al (4)

**Table 3: PETERSSON RADIOGRAPHIC CLASSIFICATION OF HEMOPHILIC ARTHROPATHY (5)**

TYPES OF CHANGE	FINDINGS	SCORE *
<b>Osteoporosis</b>	Absent	0
	Present	1
<b>Enlarged epiphysis</b>	Absent	0
	Present	1
<b>Irregular Subchondral surface</b>	Absent	0
	Surface partially involved	1
	Surface totally involved	2
<b>Narrowing of joint space</b>	Absent	0
	Present: Joint space >1 mm	1
	Joint space <1 mm	2
<b>Subchondral cyst formation</b>	Absent	0
	1 cyst	1
	> 1 cyst	2
<b>Erosions at joint margins</b>	Absent	0
	Present	1
<b>Gross incongruence of articulating bone ends</b>	Absent	0
	Slight	1
	Pronounced	2
<b>Joint deformity (angulation and/or displacement between articulating bones)</b>	Absent	0
	Slight	1
	Pronounced	2

Adapted from Pettersson, et al.(5) \* Possible joint score of 0-13 total points per joint X 6 joints = 78 possible score, where 0 is best and 78 is worst.

MRI is being used more frequently to facilitate the evaluation of early hemophilic arthropathy. This technique has improved soft tissue contrast discrimination and tissue characterization, both of which have been difficult to achieve with any other modality. (6) The MRI enables visualization of the synovial lining as well as the amount of hemosiderin deposit present in the lining. (7)

## **PAIN MANAGEMENT**

Patients with chronic hemophilic joint arthropathy may have difficulty distinguishing arthritic pain from the acute pain of hemarthrosis in a target joint. If there is no relief of pain following aggressive factor treatment of an acute hemarthrosis, the pain may be related to the arthritic changes in the joint. The use of nonsteroidal anti-inflammatory drugs (NSAIDs) in the treatment of chronic arthritic pain in hemophilia is controversial, because these drugs inhibit platelet function. Use of medications that inhibit platelet function must be considered on an individual basis by the patient and his/her physician. NSAIDs that block only prostaglandins at sites of inflammation (COX-2 inhibitors) may

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be safer than traditional NSAIDs as they do not affect platelet function (7) and can be used in persons with hemophilia under physician supervision. In some of the more debilitating cases of chronic hemophilic arthropathy, narcotics may be needed to control pain. Often referral to a pain management specialist may be helpful in these severe cases.

## **EARLY INTERVENTION**

### **COMPREHENSIVE CARE**

The best way to prevent hemophilic arthropathy is by establishing a good relationship between the patient and the hemophilia treatment center (HTC) or hematologist. Yearly comprehensive care visits help to identify problems early and assist patients in getting optimal care.

### **PROPHYLAXIS**

Hemophilic arthropathy can be prevented if recurrent hemarthroses are avoided. One strategy to decrease the likelihood of a single joint bleed leading to recurrent bleeds in the same joint is to administer factor prophylactically after the first joint hemorrhage.

A multi-center clinical trial was conducted in 65 boys with severe hemophilia A. The children were randomized between an aggressive on-demand therapy and prophylaxis. This study showed that prophylaxis with recombinant factor VIII can prevent joint damage in young boys with severe hemophilia A. (8)

See MASAC Medical Bulletin #193 (9) and the chapter on prophylaxis for more information.

### **PHYSICAL THERAPY**

Physical therapists (PTs) are an integral part of the hemophilia team. They are trained in the assessment and treatment of musculoskeletal issues using a wide variety of modalities. Not only are PTs helpful in the management of patients with target joints or those with hemophilic arthropathy, they are also instrumental in the prevention of new target joints in persons with hemophilia by working closely with the patient and their family.

There are several tools that are currently being used to assess joint function. The most widely used instrument is the World Federation of Hemophilia (WFH) Physical Joint Examination. These were expanded to a new instrument termed the Child PE Instrument (Colorado PE-1, Colorado PE-0.5) that was more appropriate for young children. (10) These scoring systems then led to the development of the Hemophilia Joint Health Score (HJHS) that is now being used internationally to assess joint health. (11, 12)

### **SPLINTING, ORTHOTICS, AND ASSISTIVE DEVICES**

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There are several types of adjunctive devices that can be used in persons with hemophilia who have chronic joint arthropathy. These can provide limb stability and often decreased pain.

Splints are used for the following reasons and are sometimes used in conjunction with other modalities:

- \* To immobilize a bleeding or painful joint
- \* To prevent deformity in chronic bleeding joints and/or to prevent postsurgical deformity
- \* To correct joint deformity if a contracture is present
- \* To maintain range of motion
- \* To improve joint function
- \* To provide support for weakness or instability (3)

Some examples of splints for the ankles are air or gel stirrups and lace-up supports. Knee splints include neoprene or elastic supports for mild problems, dynamic or passive tension splints for mild flexion contractures, and custom-made desubluxation hinged splints for severe flexion contractures. There are a wide variety of other splints available on the market today for knee joints, as well as for elbows and shoulders. Long-term splinting may be contraindicated, as restricting joint movement for lengthy periods of time can cause muscle wasting and weakness.

Orthotics or lifts in the shoe may be useful in positioning the foot properly to avoid undue stress on a joint. They often can help to decrease pain for patients with chronic ankle problems. The patient should be evaluated by a physical therapist to determine if the orthotic device changes the patient's gait pattern or causes stress on another joint.

Hemophilic patients with chronic joint arthropathy often use other assistive devices including crutches, a cane, or a walker. Care should be taken to make sure that they have been fitted properly for the patient and that the patient has had proper instruction in the use of these devices.

## **RADIONUCLIDE SYNOVECTOMY**

Radionuclide synovectomy is a nonsurgical approach to the treatment of a target joint in persons with hemophilia. It has been performed for many years in the United States and Canada as well as in other parts of the world. This procedure is best done early in the course of chronic hemophilic synovitis, prior to any significant degenerative changes. This procedure is done on an outpatient basis at most institutions and does not require large amounts of factor replacement. In the United States, colloidal <sup>32</sup>P chronic phosphate is injected into the target joint, where this agent acts to destroy the synovial tissue. (13, 14) Radionuclide synovectomy may be the only suitable treatment for those patients who cannot undergo conventional surgical procedures without unacceptable risk, such as inhibitor patients and those with significant liver disease. This procedure is done

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on an outpatient basis and is therefore cost-effective. It usually results in a decrease in the number of bleeding episodes; however, this procedure does not stop degeneration of the joint in patients with hemophilic arthrosis.

### **ARTHROSCOPIC/OPEN SYNOVECTOMY**

Arthroscopic and open synovectomy have been performed for many years in persons with hemophilia. Like the radionuclide synovectomy, arthroscopic synovectomy is a procedure that is best done early in the course of chronic hemophilic synovitis. An arthroscopic synovectomy is a surgical procedure that may be done as an outpatient or with a minimal hospitalization. Arthroscopy allows direct visualization of the intra-articular structures. During this procedure, a fiber-optic instrument is introduced through small incisions into the joint, and the damaged synovial lining is removed. By removing the hypertrophied synovial lining, bleeding is decreased. The joints most amenable to the use of the arthroscope are the knee, ankle, elbow, and shoulder. (15)

An open synovectomy is a similar procedure but is done through an open arthrotomy by opening up the joint capsule and then removing the synovial lining. This almost always requires an overnight hospitalization and intensive physical therapy afterwards.

Both the arthroscopic and open synovectomies require prolonged treatment with factor concentrate throughout the postoperative and rehabilitation period. These procedures usually decrease the number of bleeding episodes into a target joint but do not necessarily improve range of motion. Physical therapy is imperative after both of these procedures to achieve maximal joint motion and muscle strength.

## **LATER INTERVENTIONS**

### **HYALURONIC ACID**

Intra-articular injection of hyaluronic acid in the treatment of hemophilic arthropathy of the knee has been used. Hyaluronic acid is a natural substance consisting of the building blocks of cartilage and synovial fluid. One published study, Wally et al, concluded that treatment with hyaluronic acid can be recommended in patients with hemophilia in certain circumstances. (16)

### **ARTHRODESIS/TOTAL JOINT REPLACEMENT**

Once hemophilic arthropathy is advanced and significant degenerative changes in the joint have occurred, more extensive orthopedic surgical procedures are required. These include arthrodesis (fusion) and total joint replacement (arthroplasty). Pain is usually the symptom that brings patients to consider surgery. By this advanced stage of disease, acute bleeding into the joint is unusual.

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Typically arthrodesis, or fusion of the joint, is the surgery of choice for someone with end-stage ankle problems. This procedure involves fusing some or all of the bones of the joint. It requires skeletal fixation with screws and/or pins, and its objective is to decrease the pain by eliminating “bone on bone” motion. Most hemophilia patients undergoing fusion report decreased pain and do not feel that there is a significant loss in overall mobility, even though the joint itself does not move. Fusion can also be used for joints other than the ankle in extreme situations.

Total joint replacement is done in end-stage arthropathy, for example, in a patient with an Arnold-Hilgartner stage V X-ray score. Total joint replacement is by far the most complex of all the orthopedic procedures done on persons with hemophilia. These procedures are most successful when done on knees and hips; however, there are anecdotal reports of successful total elbows. Because total joints are often done in persons with hemophilia at an earlier age than in the general population, these total joints may loosen or wear out, often necessitating a revision several years after the initial replacement.

There are several new innovations in total joint replacement such as ceramic total hips and the Birmingham Hip. There are also new total ankles on the market; STAR has recently been released in the United States. There are no published studies on the use of these in hemophilia patients.

## **PREPARATION FOR SURGERY**

No surgery can be considered minor in a person with hemophilia. Preoperative planning is essential and should include not only the patient but his or her family as well. The goal during and after surgery is to provide the patient with adequate factor replacement therapy to maintain hemostasis and promote healing. Many orthopedic procedures require intense rehabilitation on the part of the patient. Commitment to a rehabilitation program is essential to a good surgical outcome.

Five conditions should be present before surgery should take place:

- 1) a hematologist and diagnostic coagulation laboratory should be available
- 2) the surgeon should feel comfortable handling a patient with a bleeding disorder
- 3) there should be access to a pharmacy that can supply adequate factor replacement products
- 4) an appropriate rehabilitation team should be available for postoperative management
- 5) no inhibitor should be present.

Laboratory studies to assess the presence of an inhibitor should be performed prior to surgery. In addition, a dose of factor may be given ahead of time to check the half-life of the infused concentrate. (19)



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Clotting factor concentrate should be given immediately prior to the beginning of a surgical procedure. A level of between 80% and 100% should be achieved, and adequate factor should be given throughout the postoperative period to maintain hemostasis. This will vary based on the patient's baseline level, the procedure being done, and each individual treatment center's protocols. Factor can be delivered either by continuous infusion or by bolus infusion. Keep in mind that some hemophilia patients have liver disease related to hepatitis C and may have a prolonged prothrombin time requiring the administration of other blood products in addition to specific factor replacement.

Prior to surgery, plans must be made regarding post-operative factor replacement and care upon discharge from the hospital. This may involve the use of a rehabilitation center or home health agency to provide nursing care and to administer factor. If venous access is of concern, a venous access device such as a PICC line may be required.

## **OSTEOPOROSIS**

Osteoporosis has become a concern for persons with hemophilia; Hepatitis C and prolonged rest during bleeding episodes prior to the use of prophylaxis may be contributing factors. Exercise plays a key role in reducing the risk of osteoporosis in hemophiliacs, as does adequate dietary intake of Calcium and Vitamin D. The occurrence of osteoporosis raises the question whether routine screening for osteoporosis should be done in persons with hemophilia. (20)

## **CONCLUSIONS**

Going forward, the hope for persons with hemophilia is a significant decrease in orthopedic complications related to bleeding. That said, chronic synovitis and arthropathy are manageable complications in hemophilia today. With advances in radiologic evaluation and orthopedic interventions, patients today are more likely to have a better functional outcome than ever before.

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