This form guides assessment of individual elbow, knee and ankle joints of persons with hemophilia, providing a systematic method for evaluation, documentation and interpretation of information that has been documented during a thorough musculoskeletal evaluation. Each joint receives a numeric score, which can be tracked longitudinally to monitor joint health impacted by joint bleeding. This instrument took its foundation from the World Federation of Hemophilia (WFH) Musculoskeletal Assessment which was modified to reflect the smaller joint degenerative changes of today's person with hemophilia. These instructions provide a brief explanation of how the joints should be examined within each category.

## **SWELLING**

This instrument is designed to pick up the earliest signs of joint swelling. Evaluation of swelling can be very subjective. Use the unaffected contralateral joint for comparison in cases of very mild swelling. Use the descriptors below as well as circumferential measurements, along with the evaluator's judgment to determine the severity. Palpation around bony prominences can identify swelling. If the joint has mild swelling, a circumferential measure may not indicate the true nature of the swelling but may only measure the bony prominences. If this is the case with the joint being measured, the descriptive definitions of the swelling should be used to give the joint its rating. If measuring the joint or extremity, document exactly where the measurement is taken so this can be duplicated in the future or by another individual. When determining the correct number to assign, consider if a large discrepancy between the two joints is due to true swelling or bony changes. In more advanced joint disease, bony overgrowth may be present. BONY OVERGROWTH SHOULD NOT BE CLASSIFIED AS SWELLING!

## **DESCRIPTORS** – (Preferred method of assessment)

- ☐ Mild The joint looks slightly "puffy", there is slight palpable swelling present and bony landmarks are clearly visible. There may/may not be any measurable difference between the joints.
- ☐ Moderate Joint swelling is easily visible. The swollen area feels firm or boggy on palpation. Bony landmarks are not visible but palpable. There is a measurable difference between the joints.
- □ Severe The joint and surrounding area looks very swollen and can be tense or very boggy to palpation. Bony landmarks are not visible and are difficult to palpate. There is a measurable circumferential difference between the joints.

It's important to palpate around the joint and its bony prominences while moving the joint through range of motion to determine if palpable synovium can be felt.

## **MEASUREMENTS** – (Use only if significant swelling is present)

Circumferential measurement of the **ankle** is difficult to obtain due to the difficulty in accurately locating landmarks for this measurement. A Figure-of-Eight measure is recommended if it covers the area of swelling. Otherwise circumferential measure is indicated.

- **Ankle Joint** Circumferential measurement for the ankle joint should be done over the part of the ankle with the greatest swelling. This may be inferior or superior to the malleoli or directly over them.
- **Elbow joint** Measure around the antecubital fossa with the elbow in as much extension as possible

**Knee joint** — Measure around the medial joint line with the knee in as much extension as possible

FINAL SWELLING SCORE SHOULD BE BASED ON WHICH CATEGORY OF <u>DESCRIPTORS</u> BEST DEFINES THE JOINT.

# **MUSCLE ATROPHY**

Atrophy, or muscle wasting, can indicate that there might be a problem in the adjacent joint. Atrophy can be assessed through visually noting the changes in muscle belly contours as well as circumferential measures. Since individual extremities vary in length and bony structure, not all muscles can be measured at the exact same point. Within one individual, an atrophied muscle can be measured and compared to its normal counterpart. Sequential circumferential measurements at the same identified point can be helpful in monitoring an adult over time. In children, it can be done for short-term follow-up, when normal growth of the child would not affect these measurements.

Visualize the whole extremity, comparing to the contralateral side, looking for symmetry between the two muscle groups. The point where the muscle belly is typically the largest can prove to be a consistent point from year to year. (Note: This can be difficult to accurately assess in someone who is overweight. Make note if this is an issue at the time of the evaluation.)

#### DESCRIPTORS

Usin	g the following definitions to record the muscle atrophy.
	Mild — muscle has slightly less contour than the contralatral side
	Moderate — flattening of the muscle belly
	Severe — severe muscle wasting and depression

#### **MEASUREMENTS**

**Measure with the muscle in a relaxed state.** When noting the circumferential measurement, a 1 or 2 cm difference may be noted. A 1 cm difference on a very small child might indicate moderate atrophy, while the same difference on an older child or adult may indicate mild atrophy. This is why the descriptors should be used in combination with a circumferential measure to score this category.

**Thigh** — Measure up from the knee joint line to the greatest bulk of vastus medialis. If the knee is quite swollen, measure just above the area of swelling and note that the swelling was present.

**Calf** — Mark the calf muscle with the patient in standing. Measure from the popliteal fossa down to the greatest bulk of the gastroc-soleus muscle group. Then have the patient sit or lie down and take the circumferential measurement with the leg relaxed in a non-weightbearing position.

**Upper arm** — Measure up from the antecubital fossa. With the elbow flexed to 90° mark the biceps at its largest point. Then relax the biceps muscle and let the elbow go back into extension. Take the circumferential measure of the biceps in this relaxed position of extension.

**Forearm** – Measure from the antecubital fossa down to the greatest bulk of the forearm.

If there is  $\leq$  .5 cm difference between the two sides there will probably not be any change in the descriptors, therefore neither muscle group will be atrophic compared to its contralateral side. If bilateral joints both demonstrate obvious muscle atrophy in the adjacent muscle groups then the circumferential measure will be invalid. If this is the case, rate the muscle according to the descriptors.

# **AXIAL DEFORMITY**

Axial deformity is defined as the degree of variation in the joint from the midline of the body. It is measured only in the ankle and the knee. The two terms used to measure this deformity are varus and valgus.

Varus is defined as angulation of the part distal to the joint towards the midline (in the knee this would be bowlegged) and valgus is defined as angulation of the part distal to the joint away from the midline (knock-kneed).

Axial angulation measurements should be done in a weightbearing position, standing on a firm surface with body weight evenly distributed on each leg. Legs should be slightly apart (hip-width).

**Measure the ankle from behind**. Bisect the calcaneus, then align one of the arms of the goniometer with the Achilles tendon and the other arm with the line bisecting the calcaneus. Axis is just superior to calcaneus.

**Measure the knee from the front**. Align one arm of the goniometer along the shaft of the femur and the other along the shaft of the tibia. The axis is at the knee.

## **CREPITUS**

Crepitus is the crackling or grinding sound or sensation produced by the rough articulation of the bony surfaces of the joint. Crepitus should not be confused with just a single pop which indicates a release of pressure. This WFH-based category has been modified to include a **Moderate** definition, as there are times when crepitus is just barely perceptible, other times when it is quite severe, but frequently it is more than mild but not bad enough to truly be rated severe.

Palpate and listen for crepitus. The therapist's hands should be placed comfortably on the joint while encouraging the subject to move actively through all ranges of motion.

<b>Mild</b> - Crepitus is slightly present. There is a barely detectable audible or palpable sensation during joint motion.
$\label{eq:moderate} \textbf{Moderate} \text{ - Crepitus is more easily felt and/or heard. There is more pronounced crackling and/or a rough sound or sensation during joint motion.}$
<b>Severe</b> - Crepitus is very pronounced. There is very audible and palpable grinding and crunching during joint motion.

## RANGE OF MOTION

Range of motion should be done using standard Range of Motion (ROM) protocols. We recommend the use of the average ranges of motion values of Norkin and White as recommended by the American Academy of Orthopedic Surgeons. (Measurement of Joint Motion: A Guide to Goniometry. Norkin, CC and White DJ. FA Davis Company. Philadelphia, 1985, revised edition, 1995). ROM should be recorded, and the total loss of motion should be compared to normal standards for that particular joint. Normal ranges of motion used for each joint are labeled on the CAJAS worksheet. Since the elbow, ankle, shoulder, and hip each move in more than one plane of motion, loss of motion can potentially occur in several directions, but for the purposes of this scale only one plane of movement will be the identifying plane.

To simplify the assessment, **elbow data will be recorded in flexion/extension**, and **ankle data will be recorded in dorsiflexion/plantarflexion**. Record the range of motion measures on the worksheet. The Range of Motion score that is recorded on the Joint Tracking Form is determined by dividing the number of degrees of total motion lost in the identified range by the total number of degrees in that range of a normal joint. If the knee or elbow hyperextends, the denominator will increase by the number of degrees of hyperextension.

With both the elbow and the knee, normal extension for an individual may actually be in a hyperextension range. In this situation, an elbow extension measure of  $0^{\circ}$  may actually indicate a loss of extension from the hyperextension baseline. This is probably more common in the elbow joint, but may also be present in the knee joint. A measurement of  $0^{\circ}$  in a case where hyperextension is present may indicate that there has been a problem in the joint. This loss of motion should be accounted for. Example: Instead of having an elbow joint with  $150^{\circ}$  of motion his total may be  $160-165^{\circ}$ . Therefore the number of degrees which he has lost from his fully hyperextended position should be divided by a denominator of 160, or whatever his baseline is

The Norkin & White reference measures knee flexion in the prone position. This ROM evaluation assesses passive, available range of motion, with each joint measured with adjacent muscles in a relaxed state. Measure knee flexion and extension in supine. Ankle motion should be measured with the knee in the flexed position to prevent muscle tightness from interfering with motion. If there appears to be muscle tightness interfering with motion when knee or ankle muscles are put on stretch, it should be noted, as it will probably interfere with normal functional movement and may affect how the subject scores on the **GAIT** category.

## ROM MEASUREMENT POSITIONS

Elbow flexion — Measure the subject in supine.
<b>Elbow extension</b> - Measure the subject in supine. Support the distal humerus off of the table to allow for any possible hyperextension
<b>Knee flexion</b> — Measure the subject in supine. May also be measured in sitting if the subject has difficulty lying flat. If measuring is sitting, make sure that he is able to go through entire flexion range. (NOTE: Norkin and White measures this in prone. <b>DO NOT MEASURE IN PRONE!</b> )
<b>Knee extension</b> – Measure the subject in supine. Support the heel slightly off the table to allow for passive hyperextension.
<b>Ankle dorsiflexion</b> – Measure the subject in supine with a roll under the knee or in sitting with the foot slightly inverted.
<b>Ankle plantarflexion</b> – Measure the subject in supine with a roll under the knee or in sitting.

## CONTRACTURE

Flexion contracture is recorded as the number of degrees the joint is lacking from full passive extension. This category pinpoints exactly where the loss of motion has occurred, rather than just the total loss of motion. It also describes a functionally more involved joint. This will be taken from the directly from the range of motion measure.

The knee and elbow contracture score is easily obtained from the **extension** measurement of the range of motion. In the ankle joint, an **equinus contracture** will be the defined position of contracture.

**NOTE**: If the contralateral side normally has some hyperextension then that loss of hyperextension should be added to the additional loss of full extension to  $0^{\circ}$ . For example, if one elbow has  $10^{\circ}$  of hyperextension but the contralateral side lacks  $15^{\circ}$  from  $0^{\circ}$ , the total contracture would be 10 + 15 or  $25^{\circ}$ .

# **INSTABILITY**

Instability has been taken directly from the WFH Musculoskeletal Assessment. True instability due to hemophilic joint arthropathy and not injury will only be seen in severe joint disease. Instability caused by ligamentous injury may be seen in active patients with mild hemophilia or in active patients with moderate or severe hemophilia who treat prophylactically in order to be able to participate in sports and recreational activities. Standard tests of elbow, knee and ankle ligamentous integrity should be performed.

Muscle weakness should not be recorded as instability!

Instability Scoring:		
	0 - None	
	1 - Noted on exam but doesn't interfere with function or require bracing	
	2 - Instability that creates a functional deficit or requires bracing or orthotics	
STI	RENGTH	
perfo child the st stron	adult strength category is based on standard manual muscle testing (MMT: 1-5 per Kendall), ormed by most physical therapists. Manual muscle testing can be used for strength testing of an older or an adult. A higher muscle strength grade will transfer to a lower CAJAS score, in order to keep trength category in the same format as the other items on the joint assessment scale. That is, a ger (higher) muscle grade indicates a healthier musculoskeletal system and therefore scores a lower AS score, with a lower score indicative of a healthier joint.	
Use t	the following definitions for each strength score.	
	0 - Moves through full range of motion against gravity, takes maximal resistance (Grade 5)	
	1 - Moves through full or available range of motion against gravity. Takes moderate resistance if full range. Takes maximum resistance in available range (Grade 4 to 5-)	
	2 - Moves through full or available range of motion against gravity, takes minimal resistance (Grade 3 to $$ 4-)	
	3 - Moves through full or available range of motion. (Grade 2+ $/$ 3-). This category includes both antigravity as well as gravity-eliminated testing.)	
	4 - Unable to move through full or available range due to weakness ( grades $\leq$ Grade 2)	
Seve	ral of the criteria above list <b>available</b> range of motion. Frequently people with hemophilia can be	

For CAJAS scoring, a score of 0 cannot be given unless the patient has full joint range of motion.

very strong within their available range, which may be decreased due to chronic joint changes.

Each identified muscle group should be tested. In the knee, test flexion and extension. In the elbow, test flexion and extension only, and in the ankle, test dorsiflexion and plantarflexion. Different muscles may

have different strengths. The overall score for the joint will be taken from the direction that is the **WEAKEST.** For example, on the knee joint, both flexion and extension will be assessed. The weaker of these two scores will determine the score that goes onto the worksheet.

#### KNEE FLEXION

☐ TEST POSITION: PRONE KNEE BEND – Break test

With the patient lying prone, have him bend his knee. He should be able to completely bend it against gravity. To provide resistance, have him bend the knee against gravity and hold the knee in flexion while the therapist pushes against him into extension.

This test can be done in sitting if the patient is unable to lie prone.

#### **KNEE EXTENSION -**

☐ TEST POSITION: SITTING KNEE EXTENSION –Break test

With the patient sitting with the hip and knee in 90° of flexion, have him straighten the knee completely. If unable to completely straighten the knee in sitting, hamstring tightness must be ruled out. Have him semi-recline, leaning back on the elbows. Support the distal end of the femur to maintain the leg horizontally. In this position have him fully extend the knee. To provide resistance, have him flex the knee slightly to unlock it, and then push against the therapist's hand as he straightens the knee.

#### ANKLE DORSIFLEXION -

☐ TEST POSITION: SITTING WITH HIP AND KNEE AT 90° - Break test

With the patient sitting with the hip and knee in  $90^{\circ}$ , have him lift the foot up, slightly inverting as he lifts. To provide resistance, push down on the top of the foot as he lifts the foot against gravity.

#### ANKLE PLANTARFLEXION -

☐ TEST POSITION: STANDING ON SINGLE LEG TO BE TESTED – Heel raise test

With the patient standing on one leg with knee in extension have him raise up on his toes, lifting the heel off of the floor. Have the patient hold lightly to the door frame or wall for balance only. He should attempt to do 20 repetitions.

- $\mathbf{0} > 20 \text{ heel raises}$
- 1 10-19 heel raises
- 2 1-9 heel raises
- 3 Partial heel raise against gravity

OR

☐ TEST POSITION: PRONE WITH KNEE FLEXED TO 90° – Break test

With the patient prone with the knee flexed to  $90^{\circ}$ , have him plantarflex, or point the toes toward the ceiling. Support the lower leg at the ankle with one hand and with the other hand on the ball of the foot apply pressure down.

OR

☐ TEST POSITION: SITTING – Break test

Have the patient in sitting with the knee in flexion and the foot resting on the therapist's leg. Hold the ankle just above the malleoli with one hand. Place the other hand on the plantar surface of the foot. Ask the patient to point the toes down and hold, as the therapist gives pressure through the ball of the foot moving the ankle into dorsiflexion.

The heel raise test requires a much stronger gastroc-soleus muscle and a good, healthy ankle joint. This test may not be appropriate for someone with ankle joint disease. If he is unable to do the heel-raise test then the highest score that he can achieve is a 2 + on the muscle test which will be scored as a 3 on the CAJAS score sheet.

# **ELBOW FLEXION -**

☐ TEST POSITION: SUPINE OR SITTING WITH THE ARM DOWN AT THE SIDE —Break test With the patient supine or sitting with the arm at his side, have him bend the elbow. Stabilize and cushion the elbow by placing one hand under the elbow. Give resistance in this position applying pressure against the distal forearm in the direction of extension.

## **ELBOW EXTENSION –**

☐ TEST POSITION: SUPINE WITH THE SHOULDER AT 90° FLEXION WITH THE ARM SUPPORTED PERPENDICULAR TO THE TABLE – Break test

Usually tested in prone with the shoulder in  $90^{\circ}$  of abduction. Some patients may not be able to tolerate prone, so test with the patient **supine.** With the arm supported in the test position, have him fully extend the elbow against gravity. Tell him to unlock and bend it slightly. Give resistance in this position, applying pressure against the distal forearm in the direction of flexion.

## **GAIT**

Four gait skills are tested: walking, ascending and descending stairs, running and skipping. Early ankle or knee joint problems can cause subtle gait changes. An ideal gait should have an equal amount of weight on each leg, with a heel-toe pattern that includes pushing off into plantarflexion. Steps should be of equal length and cadence, and weight-shifting should be equal to each side. The toes should be pointed forward symmetrically. Abnormalities in any of these areas may be slightly evident when walking but may be even more noticeable when ascending or descending stairs, running and skipping.

Ask the patient to walk and, if capable, do all of the other skills as well. If he is able to do all of the required skills he would be given a score of 0. If he is able to do the skill but the movement is uncoordinated for some reason **OTHER** than issues related to the ankle or knee joints then this should not be counted against him. The incoordination should be noted but the score for gait should be based on the movement and strength of the specific ankle or knee joint.

If unable to walk normally due to problems in either knee or ankle joint, the patient may still attempt to ascend and descend the stairs if he has been doing this regularly in the community. If this is too difficult, then performing the stairs should NOT be required. He should NOT progress

to the next level beyond doing the stairs, i.e. running and skipping. The running and skipping tests should be avoided as they would be potentially harmful to the joint.

Tips on Assessing GAIT:

Ascending and descending stairs are mainly a task for the knee joint, however the ankle also must have good dorsiflexion in order to allow the knee extensors to work properly. In addition to good knee strength for concentric motion when ascending stairs, the ankle and knee muscles must eccentrically contract as the person descends to the next step.

If he is able to walk well, he can then be asked to run. Note how each knee and ankle performs the running pattern in relation to the gait components described below. If he is able to run well he can progress to skipping.

Skipping shows primarily ankle function; however if there is weakness in the knees or hips, skipping will be a difficult task to do correctly due to lack of stability. In assessing skipping skills, the subject should have a symmetrical pattern with a distinct step-hop-hold pattern on each foot. Some people try to skip more on a flat foot, which may indicate some sort of ankle problem with either ROM, strength or both. If there is some calf weakness, the subject may exaggerate the hip and knee flexion to compensate for the lack of push-off at the ankle.

Scoring: Give the joint a **0** if all of the gait skills are normal, i.e. walking, ascending/descending stairs, running and skipping. Give a **1** if walking is normal but at least one of the other skills is abnormal. If walking is abnormal and any other higher skills are abnormal then he should receive a **2**.

# **Normal Gait Components:**

## Ankle Knee 1. Equal weight shift 1. Equal weight shift 2. Heel-toe pattern 2. Heel strike with full knee extension 3. Good plantarflexion push-off 3. Good knee extension push-off 4. Steps of equal length 4. Steps of equal length 5. Steps of equal cadence 5. Steps of equal cadence 6. Toes pointed symmetrically forward 6. Toes pointed symmetrically forward If any of the above normal gait components are not being done correctly then the skill (walking, stairs, running or skipping) will be considered abnormal. Score each knee and ankle joint independently of the others. Start with walking first and then do the stairs. If the patient can do each of these, then progress to running and skipping. IF HE CANNOT DO A SKILL, DO NOT PROGRESS HIM TO THE NEXT LEVEL AS IT MIGHT CAUSE PROBLEMS FOR THE JOINT!

# **GAIT Scoring:**

0 - Normal walking, stairs, running and skipping
1 - Normal walking, abnormal stairs, running or skipping
2 - Abnormal walking and stairs – no further skills tested

# OTHER JOINT INTERFERENCE

There are times when GAIT can be impacted by another joint, i.e. the hip, or by muscular issues (tightness or muscle shortening, or weakness related to muscle scarring or nerve damage as in the case of sequelae from iliopsoas bleeding). These situations should be taken into account when scoring each of the ankle or knee joints on GAIT. On the worksheet, mark "YES" to indicate that the problems that are being noted for each of those four joints may be caused by a different problem, or interference, like those described above. If this is the case, the GAIT score for that particular joint should reflect the worst observable score, with the understanding that this joint might score differently (better) if the interference was not occurring and more of the GAIT skills could be safely performed. If the evaluator marks "YES" for Other Joint Interference, the joint being scored cannot score a 0 for GAIT because all skills could not be performed normally.