Hemophilia Joint Health Score 2.1

ADDENDUMS

IPSG International Prophylaxis Study Group

Hemophilia Joint Health Score (HJHS 2.1)

Addendum- Swelling

The evaluation of joint swelling can be complicated by a number of factors.

Acute intra-articular swelling will be more liquid in nature. For example in the knee a "patellar ballottement" (or patellar tap) test will likely be positive. Also, a brush, stroke or bulge test may be positive. Synovial thickening is generally more spongy or boggy on palpation. There is often partial, or full obscuring of the bony landmarks in either situation.

In very young children, it may be difficult to differentiate swelling, especially persistent synovial inflammation, from "baby fat or pudge". The same may be true for obese patients with excess subcutaneous fatty tissue.

Swelling around a joint that occurs from other conditions such as Chronic Venous Insufficiency, Lymphedema (may be "pitting edema"), chronic bursitis, repetitive ankle sprains etc. may be difficult to differentiate from joint swelling.

Bony overgrowth (chronic **bony** changes in the joint) should not be scored as swelling.

Generally, it is recommended that you evaluate and score what you see for the particular joint you are examining. Use the comments section on the work sheets to clarify your findings (to help with scoring and for comparison with subsequent evaluations). Since the HJHS evaluates 8 items per joint, a single finding does not necessarily indicate that a joint is damaged or in trouble. More abnormal items and a higher joint score indicate more joint damage.

Hemophilia Joint Health Score (HJHS 2.1)

Addendum - Range of Motion (Flexion Loss and Extension Loss) : Scoring

1) HOW TO SCORE

To determine the score, use the range of motion values recorded on the HJHS work sheets.

1. First, compare the 2 sides (left and right) against each other.

2. Write down the range of motion measurements on the HJHS worksheet

3. Calculate the numerical difference between the 2 sides and then the score using the scoring chart from chart 1) below (pg. 8 in the HJHS version 2.1 Instruction Manual).
   (Example: if the patient's left knee flexion is 139 degrees and the right knee is 150, the difference is 11 degrees, then the score would be 2.)

4. Next, look at the Normative range table (page 9 in the HJHS version 2.1 Instruction Manual), and calculate the score according to the measurement and age of the patient.
   (Example: If the patient above is 15 years old, normal knee flexion range (with 2 standard deviations) for his age is between 129.6-154.8 degrees. In this case, his knee flexion for both knees is within the normative range. Therefore, the score would be 0, when using the Normative range table. (See chart below, 2) Normative range tables)

5. For recording the score on the score sheet, use the worse (or higher) of the 2 scores that you have calculated.
   (Example: This patient would receive a score of 2 for left knee flexion.)

IMPORTANT NOTE: The scoring chart when comparing the 2 sides is different than when using the normative range table. Since 2 standard deviations were included in developing the Normative range table, any measurement outside of the provided range would result in at least a score of 1.
PLEASE REFER TO THE REVISED HJHS 2.1 WORKSHEETS AND SCORE SHEET (February 2013)

1) Normal Contralateral side
Scoring using the **Normal vs Contralateral side** for the Flexion and Extension Loss categories is as follows:

- ❑ **0 = < 5 degrees**
- ❑ **1 = Loss of 5 to 10°**
- ❑ **2 = Loss of 11° - 20°**
- ❑ **3 = Loss of > 20°**

2) Normative range tables
Scoring using the **Normative table** for the Flexion and Extension Loss categories is as follows:

- ❑ **0 = Within normative table range**
- ❑ **1 = Loss of 1 to 4°**
- ❑ **2 = Loss of 5° - 10°**
- ❑ **3 = Loss of > 10°**

Below are examples for scoring Range of motion:

Example 1a

Assessment	Worksheet details	Score
▪ Age = 12 year old boy ▪ Right ankle dorsiflexion =5 ^{0} ▪ Left ankle dorsiflexion =8 ^{0}	▪ The measurements for ankle dorsiflexion would be recorded on the worksheet ▪ When **comparing the 2 sides against each other**, he would obtain a score of 0 for both sides (loss of less than 5 degrees). (From chart 1) above) ▪ When scoring using the **Normative range table**, he would receive a score of 1 for right ankle DF (since 5 ^{0} is less than the 6.1-26.5 ^{0} range), but a score of 0 for the left ankle (since 8 ^{0} ankle DF is within the 6.1^0-26.5 ^{0} Normative table's range for ankle DF in 12 year olds).	▪ The **worse** (or higher) of the two scores would be recorded on the score sheet. ▪ Therefore, a score of 1 for the right ankle and 0 for the left ankle should be recorded on the score sheet.

Example 1b

Assessment	Worksheet details	Score
▪ Age = 12 year old boy ▪ Right ankle dorsiflexion =5 ^{0} ▪ Left ankle dorsiflexion =5 ^{0}	▪ The measurements for ankle dorsiflexion would be recorded on the worksheet ▪ When **comparing the 2 sides against each other**, he would obtain a score of 0 for both sides (loss of less than 5 degrees). (From chart 1) above) ▪ When scoring using the **Normative range table**, he	▪ The **worse** (or higher) of the two scores would be recorded on the score sheet. ▪ Therefore, a score of 1 for both ankles should be recorded on the score sheet.

	would receive a score of 1 for both ankle DF (since 5^0 is less than the 6.1-26.5 ^{0} Normative table's range for ankle DF in 12 year olds).	

Example 2

*** Remember that extension loss is calculated from hyperextension where applicable.**

Assessment	Worksheet details	Score
▪ Age = 8 year old boy ▪ Right Elbow = 10^0 of hyperextension ▪ Left Elbow = Lacking 6^0 degrees from neutral	▪ The measurements for elbow extension would be recorded on the worksheet ▪ When **comparing the 2 sides against each other**, the difference between the 2 elbows is 16^0 so the score would be 2 for the left elbow. (From chart 1) above) ▪ When scoring using the **Normative range table**, he would receive a score of 0 for both left and right sides, since both left and right elbow extension range of motion values are within the Normative range of -8^0 to $+12^0$ degrees for his age.	▪ The **worse** or higher of the two scores would be recorded on the score sheet. ▪ Therefore, a score of 2 for the left elbow and 0 for the right elbow should be recorded on the score sheet.

Hemophilia Joint Health Score (HJHS 2.1)

Addendum - NE (Non-Evaluable) item : Use & Scoring

1) HOW TO USE "NE"

An HJHS item will be marked as **"NE"** when the physiotherapist does not evaluate it, based on their professional judgment, in certain circumstances.*

Examples include:

- When there is a possible medical risk of bleeding or other injury in assessing that particular item.
- If a child is not developmentally able to perform a particular gait item, then the item must be marked as "NE".

Since one obviously cannot give/attribute an HJHS score to an item or test which was not evaluated (NE), ***that item or test will remain as "NE" consistently throughout that particular HJHS assessment.***

It will be identified as "NE" on both the worksheets as well as the HJHS Summary Score Sheet.

Important Note: The HJHS has been tested for reliability and validity in its integral, complete version. If certain original HJHS items are omitted or modified, one cannot extrapolate that the reliability and validity of the modified score's results are then equally reliable or valid, based simply on the original version's testing.

(Similarly), when "NE" items are introduced and calculated into the HJHS score, that item and the total HJHS score will be "modified" when compared to the original HJHS reliability and validity testing. As such the co-developers recommend any HJHS scores containing "NE"s be clearly identified.

Below are examples for Gait using "NE":

- Gait - This is an item comprised of progressively demanding skills:
  1. Walking (easiest),
  2. Climbing/descending stairs
  3. Running
  4. Unilateral hopping (hardest)

Example 1

Assessment	Worksheet details	Score
▪ If a patient is already limping during walking (ie. an "abnormal" walking gait) – the physiotherapist may choose ***not*** to progress the patient* to perform the other gait skills.	▪ Walking will then be marked as **abnormal** *on the worksheet.* ▪ The other skills (ie. stairs, running, hopping) will be marked as Non- Evaluable **(NE)** *on the work sheet*	▪ The final score on the HJHS Summary Score Sheet for Global Gait would be "4" ▪ ***Check/tick-off the small "NE" box under the Global Gait Score.***

Example 2

Assessment	Worksheet details	Score
▪ If walking and stairs are normal but running is abnormal	▪ Mark "normal" for walking and stairs, mark "abnormal" for running, and mark "NE" for hopping *on the worksheet.*	▪ Give a score of "2" for Global Gait on the HJHS Summary Score Sheet ▪ ***Check/tick-off the small "NE" box under the Global Gait Score.***

Example 3

Assessment	Worksheet details	Score
▪ If a child is not developmentally ready* to perform certain Gait skills	▪ The particular **non-performed gait skill(s) would be marked as "NE" on the worksheet**. ▪ The non-performed gait skills would be specifically marked as **"NE",** rather than "abnormal" on the worksheet.	▪ The Global Gait Score would be scored as **"NE",** regardless of the number of gait skills performed or not. ▪ ***Check/tick-off the small "NE" box under the Global Gait Score.***

Below is an example for Strength using "NE":

Example 1

Assessment	Worksheet details	Score
▪ 4 year old with healthy joints ▪ Holds the strength test position well, but unable to understand the concept of maximal resistance due to young age	▪ Child would be given a manual muscle test grade of 4 out of 5.	▪ The Strength Score would be **"NE",** regardless of the grade given due to developmental immaturity (as opposed to muscle weakness). ▪ ***Check/tick-off the small "NE" box under the Global Gait Score.***

*** Please remember to note the specific reason(s) for the "NE" in the "General Comments" section, provided at the bottom of the HJHS Summary Score Sheet.**

2) SCORING

CALCULATING THE FINAL HJHS SCORE USING "NE":

If "NE" is due to age of development

1) If an "NE" is provided as an actual **final score** of one of the HJHS items for a specific joint or for Global Gait (due to age of development only, and not due to a bleed, arthropathy or other injury), - then the maximum score for that specific item would be subtracted from the total maximum HJHS score. This will account for the NE, without obtaining a total HJHS score that penalizes the young child.

For example:

In the calculations for example 3 above, you would adjust the denominator accordingly and subtract the maximum number for Global Gait from the total HJHS score.

MAXIMUM TOTAL HJHS SCORE THEORETICALLY POSSIBLE = (124) – THE MAXIMUM TOTAL POSSIBLE FOR THE SPECIFIC "NE" SCORED ITEM (Global Gait = 4) = ADJUSTED MAXIMUM HJHS TOTAL SCORE POSSIBLE NOW = (120)

The adjusted maximum HJHS total score (120, instead of 124), will now be used as the maximum HJHS total score possible for that specific patient.

If "NE" is due to a bleed, arthropathy or other injury

2) If an item contains an "NE" for any reasons other than age of development (for example - such as due to bleeds, arthropathy or injuries) then that item must be scored as explained above (examples 1 and 2) and it's score integrated into the final HJHS score. The total maximum HJHS score attainable in this case, would remain unchanged at 124.

*** Once again - Please remember to note the specific reason(s) for the "NE" in the "General Comments" section, provided at the bottom of the HJHS Summary Score Sheet.**

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Hemophilia Joint Health Score (HJHS 2.1)

Addendum – Clinical use of the HJHS 2.1

The HJHS provides a snapshot of the health of a joint at a specific point in time. If the HJHS is being used to inform clinical decisions regarding modifying treatment regimens or referral for surgical intervention, it is important to capture the status of the joint during a period of time when the joint is not bleeding and in a stable state which may take at least 2 weeks following the last joint or muscle bleed (Although the muscles themselves are not actually scored, muscle bleeding impacts joint mobility at adjacent joints and will also affect the strength testing section of the examination.)

The HJHS may be used clinically in any of the following ways:

1. To provide a **global picture** of an individual's total joint health by totaling the 6 individual joint scores.
2. To provide an individual joint score for a single specific joint.
3. To provide a means to track and compare a single, or multiple joints' scores over time using the same standardized measure.

To use the HJHS clinically to monitor change over time in a specific joint, a **single joint score** total should be used.

To monitor several joints which may or may not be showing signs of joint changes an individual score on each affected joint should be recorded. These should not be added together, as this will add confusion as to which joint or joints are showing degenerative joint disease symptoms. For each follow-up evaluation, individual scores should again be recorded on each affected joint to provide a direct comparison over time.

If all 6 joints scores are totaled, the final score will not reflect where the major problem areas are. Although this can be useful in reporting research, this would not be recommended for clinical decision-making, as a 6-joint total score cannot provide the information on which joint or joints are affected.

The category of Global Gait is recorded separately from each individual joint score. This is not added on to the total of an individual joint, but is considered separately. For the knee and ankle joints, the Global Gait score should be reported as a second score. It, too, can then be monitored over time

The following points should be considered when using the HJHS in making clinical decisions:

The minimum clinically important difference (MCID) has not yet been established for the HJHS, so it cannot yet be said that an increase or decrease in the score of a specific amount indicates a true change in the health of the joint. Future studies may help to define this parameter. However, tracking an overall increase or decrease in the scores can show a trend of improvement or further degeneration, and the scores, as they are currently reported, may be useful in informing the following clinical decisions:

1. When to implement or increase frequency of regular factor infusion therapy
2. When referral for imaging studies is indicated (MRI or radiographs)
3. When to refer for physical therapy intervention
4. When referral for full orthopedic evaluation is indicated
5. When to intervene surgically
6. Help track the effect of a certain treatment(s) (such as factor prophylaxis, exercise, bracing, radiosynovectomy, surgery, etc.)

Hemophilia Joint Health Score (HJHS 2.1)

Addendum – Research use of the HJHS 2.1

Over the last years psychometric properties of the different versions of the HJHS have been established. Reliability of the HJHS 1.0 is excellent with an inter-observer co-efficient of 0.83 and a test–retest of 0.89 (ref a,b). Construct validity of the HJHS is strong based on moderate correlation with physician's global joint scores and overall arthropathy impact. Moreover HJHS was 97% more efficient than WFH-score at differentiating severe from mild and moderate hemophilia and 74% more efficient than WFH-score at differentiating subjects treated with prophylaxis from those who were on demand therapy. Redundancy of certain items was noted in the reliability and validation studies of the HJHS 1.0 which lead to the deletion of these items. Although the revised version has not been tested for its psychometric properties, the current version (HJHS 2.1) is expected to have improved on all aspects.

The instructions in the HJHS manual in respect to NE items are based on the experiences of the PT Expert Working Group that tested the HJHS for its psychometric properties. The HJHS has been tested for reliability and validity in its integral, complete version. If certain original HJHS items are omitted or modified, one cannot extrapolate that the reliability and validity of the modified score's results are then equally reliable or valid, based simply on the original version's testing.

References:

a] Manco-Johnson MJ, Pettersson H, Petrini P, Babyn PS, Bergstrom B-M, Bradley CS , Doria AS, Feldman BM, Funk S, Hilliard P, Kilcoyne R, Lundin B, Nuss R, Rivard G, Schoenmakers MA, van den berg M, Wiedel J, Zourikian N, and Blanchette VS. **Physical therapy and imaging outcome measures in a haemophilia population treated with factor prophylaxis: current status and future directions.** Haemophilia 2004 Oct; 10 (S4):88 – 93.

b] Hilliard P, Funk S, Zourikian N, Bergstrom B-M, Bradley CS, McLimont M, Manco-Johnson M, Petrini P, van den Berg M, and Feldman BM. **Hemophilia joint health score reliability study.** Haemophilia 2006 Sept; 12: 518 – 525.

c] Feldman BM, Funk S, Bergstrom B-M, Zourikian N, Hilliard P, van der Net J, Engelbert RHH, Petrini P, van den Berg M , Manco-Johnson M, Rivard GE, Abad A, and Blanchette VS. **Validation of a new pediatric joint scoring system from the International Hemophilia Prophylaxis Study Group: Validity of the Hemophilia Joint Health Score (HJHS).** Arthritis Care & Research 2011 Feb; 63 (2):223-30.