

# Reliability of Hemophilia Joint Health Score (HJHS) version 2.1 in person with Hemophilia



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## Background

- The HJHS was developed by the International Prophylaxis Study Group (IPSG), primarily to detect the early joint changes in children with hemophilia and hence its reliability [1] and validity [2] has been assessed in those with minimal joint disease.
- Whether the same would hold true if this instrument is applied to both pediatric and adult patients with more advanced joint disease remains to be evaluated.
- The psychometric analysis was assessed on the initial versions of the HJHS. There is no similar data on the current version.

## Objectives

To assess the inter-rater and intra-rater reliability of the HJHS version 2.1 in children and adults with varying degrees of joint damage.

## Patients and Methods

### Study Participants

- 21 Persons with severe hemophilia (PWH), between the ages 4 and 38 years.

### Study Setting

- Physiotherapy outpatient unit of Christian Medical College, Vellore.

### Outcome measure

- HJHS 2.1 is an 8-item tool, that scores the six joints most affected by hemophilia: ankles, knees and elbows, with an additional gait score (Maximum score of 124)

### Assessment

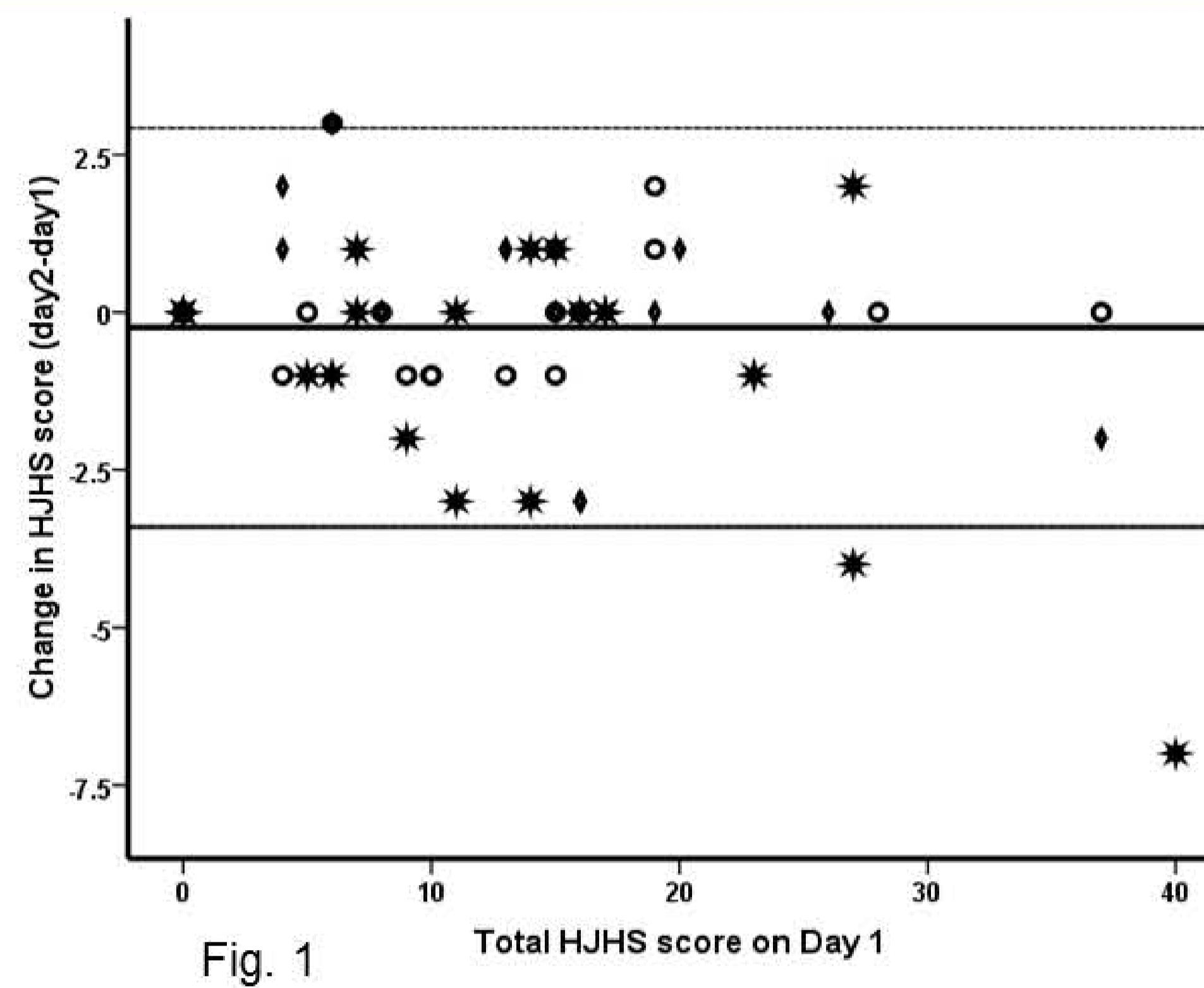
- All PWH were assessed by three experienced physiotherapists on 2 consecutive days, using the HJHS version 2.1.

### Analysis

- Inter-observer reliability using intraclass correlation coefficient (ICC)
- Test re-test reliability (ICC)
- Internal Consistency (Cronbach's Alpha)
- Bland-Altman Limits of agreement
- Subgroup analysis by age (**Age ≤10**, **Age>10**)

## Tables and Figures

The Bland-Altman limits of agreement (Figure. 1)



- The horizontal lines represent the mean change in HJHS score and 2 SD from the mean.
- The symbols are different for each of the three physiotherapists.

Reliability (ICC) of the HJHS scores by joint (Table 1)

Joint	Inter-observer	Test-retest
Elbow	0.97	0.98
Knee	0.98	0.97
Ankle	<b>0.63</b>	0.90
Total HJHS	0.98	0.98

Reliability (ICC) of the HJHS scores by item (Table 2)

Items	Inter-observer	Test-retest
Swelling	0.93	0.95
Duration of swelling	0.99	0.99
Muscle atrophy	<b>0.69</b>	0.85
Crepitus	<b>0.75</b>	0.88
Flexion loss	0.87	0.88
Extension loss	0.97	0.94
Joint pain	<b>0.74</b>	0.83
Strength	0.97	0.98
GAIT	1.00	1.00
Cronbach's Alpha	0.87	

Reliability (ICC) of Range of motion (Table 3)

Joint	Inter-observer	Test-retest
<b>Elbow</b>		
Flexion	0.96	0.97
Extension	0.96	0.97
<b>Knee</b>		
Flexion	0.94	0.94
Extension	0.96	0.97
<b>Ankle</b>		
Plantar flexion	<b>0.47</b>	<b>0.56</b>
Dorsiflexion	<b>0.39</b>	<b>0.43</b>

## SUBGROUP ANALYSIS BY AGE (Table 4)

Item	Age ≤10 (n=12)		Age>10 (n=9)	
	Inter-observer	Test-retest	Inter-observer	Test-retest
Swelling	0.87	0.93	0.95	0.95
Duration of swelling	1.00	1.00	0.98	0.98
Muscle atrophy	<b>0.60</b>	0.87	<b>0.69</b>	0.81
Crepitus	0.79	0.91	<b>0.66</b>	0.83
Flexion loss	0.95	0.95	<b>0.70</b>	<b>0.73</b>
Extension loss	0.97	0.86	0.97	0.97
Joint pain	0.86	0.93	<b>0.52</b>	<b>0.65</b>
Strength	0.85	0.96	0.85	0.95
Gait	1.00	1.00	1.00	1.00
Cronbach's Alpha	0.88		0.85	

ICC >0.75 is excellent,  
 ICC 0.40-0.75 is fair to good  
 ICC <0.40 is poor.

## Results and Discussion

- The median HJHS score was 10.5 (range 0 - 40).
- The inter-rater and intra-rater reliability was excellent (ICC = 0.98, Table 1).
- The internal consistency was excellent (Cronbach's Alpha = 0.87, Table 2).
- Reliability of HJHS measurement was excellent in all joints, **except in the ankle (ICC =0.63, Table 1)**.
- The **poor reliability of HJHS measurement in the ankle** is partly due to the inconsistency in the measurement of the ROM of the ankle, (ICC ranging from 0.39 to 0.56, Table 3) & also due to differences in the scores for
  - crepitus,
  - joint pain, &
  - muscle atrophy. (Table 2)
- Subgroup analysis showed an excellent internal consistency in both children and adults (Table 4).

## Conclusions

- This study confirms that inter-rater and intra-rater reliability of HJHS Version 2.1, in children and young adults with haemophilia, with varying degrees of joint damage is very good.
- Factors affecting the reliability of HJHS assessment in the ankle need to be addressed.

## References

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