

# Reliability and validity of the Haemophilia Joint Health Score in von Willebrand disease



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**Aim**  
To validate the Haemophilia Joint Health Score (HJHS) in Von Willebrand disease.

**Background**  
Joint bleeds are reported by 23% of patients with VWD (VWF activity  $\leq 30$  IU/dL) and can result in arthropathy.<sup>1</sup> To analyze joint function with emphasis on knees, ankles and elbows, the HJHS has been developed for patients with haemophilia. The performance and feasibility of this instrument in patients with VWD is currently unknown.

**Methods**  
One trained physiotherapist conducted the HJHS version 2.1. For the inter-observer reliability a second trained physiotherapist repeated HJHS assessment in 24 patients (67% history of JB). We considered an Intraclass Correlation (ICC) > 0.6 and Limits of Agreement (LoA) +/- 12 as acceptable (HJHS range 0-124). To determine construct validity we calculated Spearman's  $r$  ( $r_s$ ) between the HJHS and joint X-ray (Pettersson score) at joint level and between the HJHS and the Haemophilia Activities List (HAL) questionnaire ( $r_s > 0.6$  acceptable). We also hypothesized that the HJHS score should be higher ( $p < 0.10$  Mann Whitney U) in patients with type 3 VWD (VWF < 5%) and in patients with a history of more than 5 JB.

**Hemophilia Joint Health Score 2.1 - Summary Score Sheet**

	Left Elbow	Right Elbow	Left Knee	Right Knee	Left Ankle	Right Ankle
Swelling	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE
Duration (swelling)	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE
Muscle Atrophy	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE
Crepitus on motion	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE
Flexion Loss	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE
Extension Loss	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE
Joint Pain	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE
Strength	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE	<input type="checkbox"/> NE
<b>Joint Total</b>						

Sum of Joint Totals  NE = Non-Evaluable

Global Gait Score **+**  (  NE included in Gait items)

HJHS Total Score **=**

Figure 1: HJHS score sheet

**Study population**

- 96 adult VWD patients with historically lowest VWF levels  $\leq 30$  U/dl and/or FVIII:C  $\leq 40$  U/dl and a bleeding- or family history of VWD
- 60% male, median age 47 years
- Mean VWF activity 11 U/dL and FVIII:C 28 U/dL
- 49% had a documented history of JB

**Results**

The HJHS score ranged from 0-47 (n=96). We observed a floor effect in the 49 VWD patients without a history of documented JB (35% 0 points compared to 15% of JB patients,  $p=0.025$ ). Agreement between physiotherapists was good with ICC 0.84 (absolute agreement analysis, 95% CI 0.63-0.93) and LoA +/- 10.3 (Figure 2). We found an acceptable correlation between the HJHS and Pettersson scores (all 6 joints  $r_s > 0.60$ ). The HJHS also correlated with the HAL ( $r_s = 0.71$ ,  $n=94$ ). Patients with type 3 VWD and patients with > 5 JB scored significantly higher on the HJHS (Table 1).

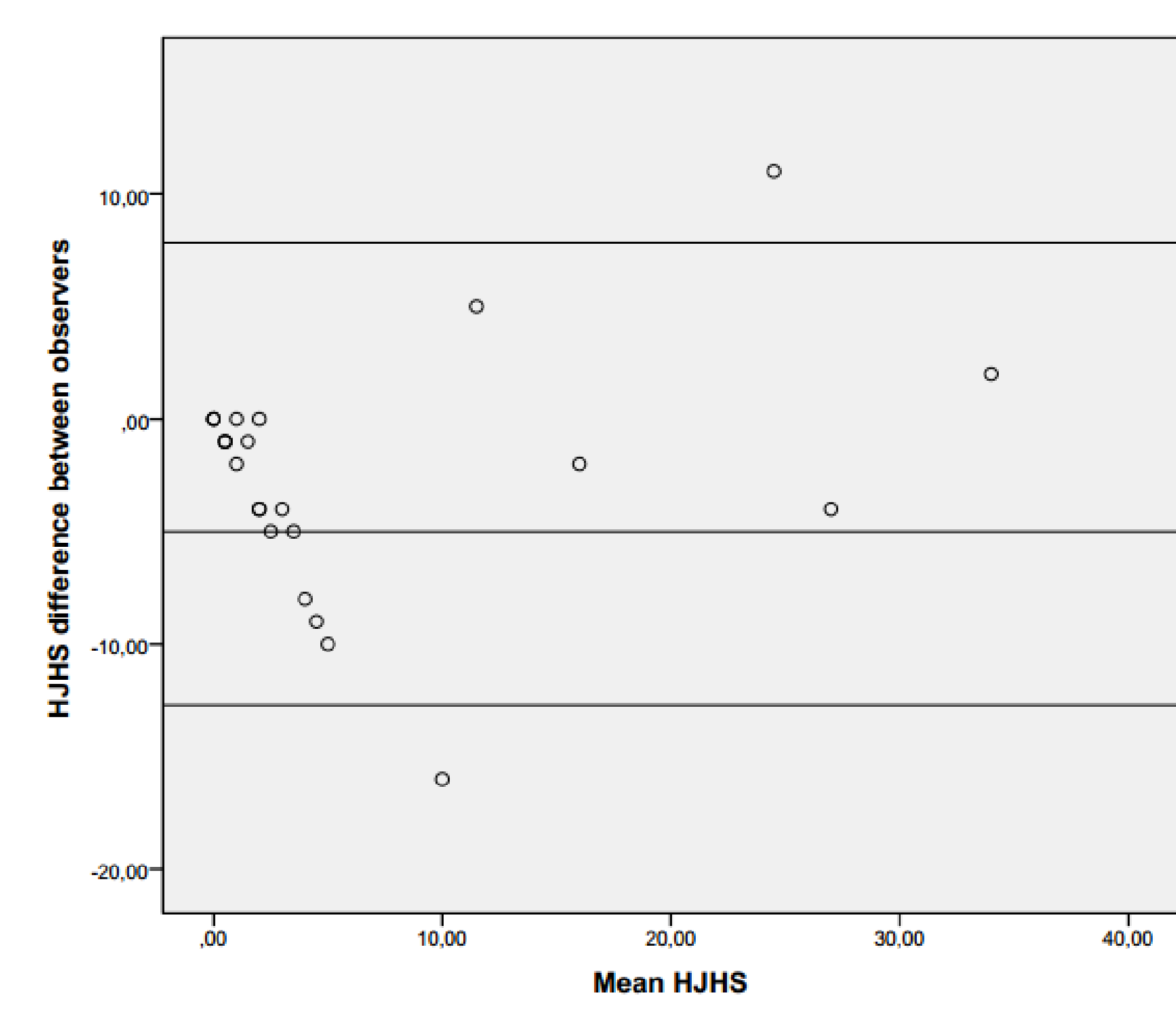


Figure 2: Bland-Altman plot. LoA +/- 10.3 between the two physiotherapists

Table 1: construct validity of the HJHS in VWD

	Type 3 VWD n=23	>5 joint bleeds n=26	Non-type 3 VWD and <5 joint bleeds n=65
HJHS (median, IQR)	12 (3-21)*	10 (2-18)*	2 (0-5)

\*  $p < 0.01$  compared to nogn type 3 and <5 joint bleeds

**Conclusion**  
As in haemophilia, the HJHS appears sufficiently reliable and valid to assess joint function of elbows, knees and ankles in adult patients with VWD and a history of joint bleeds.

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Poster Session Online

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