

Hemophilia Joint Health Score v2.1 in US Adults With Hemophilia in the Pain, Functional Impairment, and Quality of Life (P-FiQ) Study

P-W-124

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Objective

- To evaluate Hemophilia Joint Health Score v2.1 (HJHS) scores across joints and assess components contributing to overall scores

Introduction

- HJHS is a standardized instrument for measuring joint health that has been validated in children with hemophilia but is infrequently used in the clinical setting in the United States
- Content validity of HJHS was assessed along with 5 patient-reported outcome (PRO) instruments measuring pain, function, and quality of life as part of the P-FiQ study in adults with hemophilia

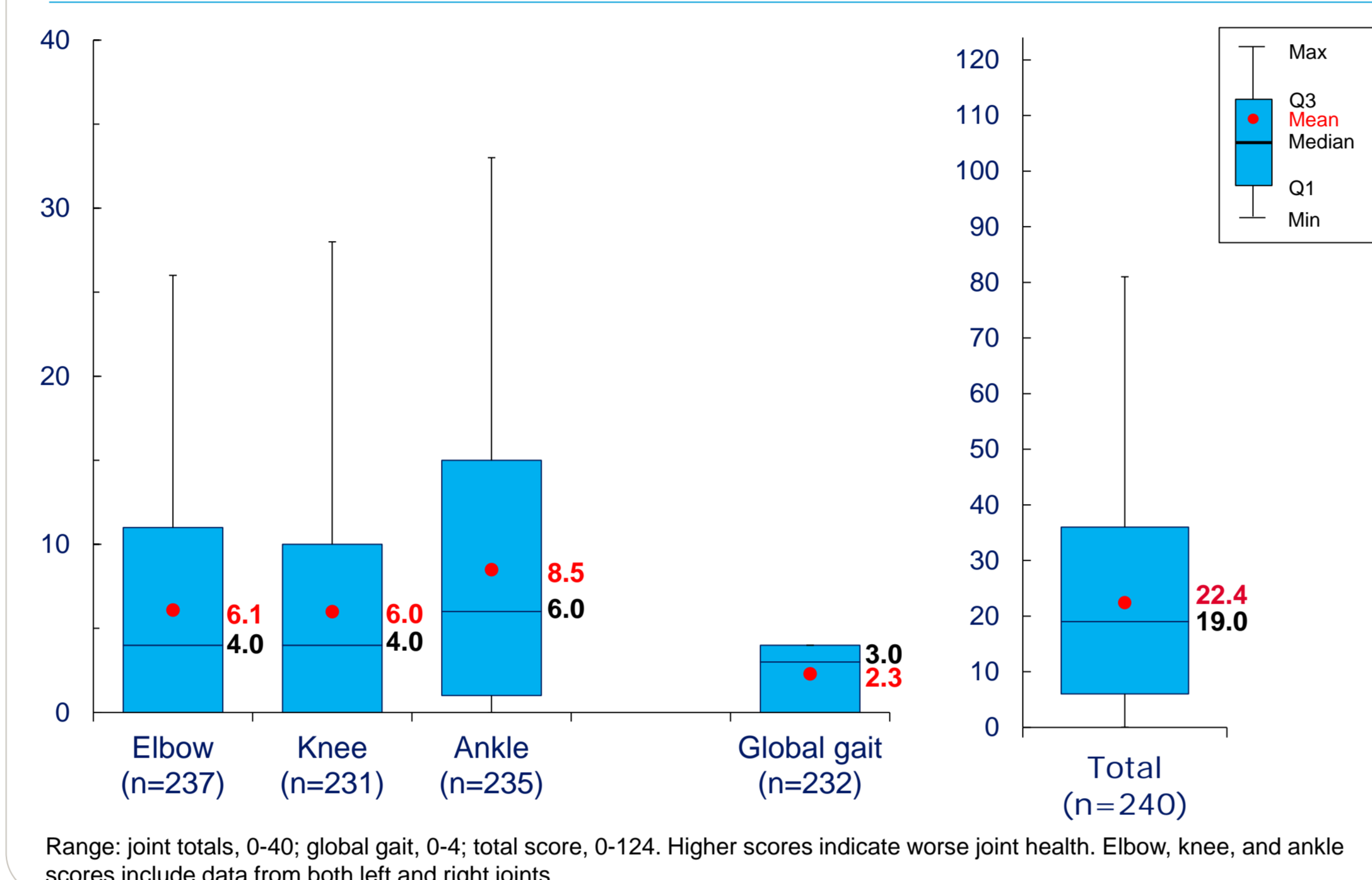
Methods

- Adult males with mild to severe hemophilia and a history of joint pain or bleeding were enrolled during routine comprehensive care visits and completed 5 PRO instruments
- All participants underwent evaluation of range of motion of 6 index joints (elbows, knees, and ankles)
- Optional HJHS evaluation was completed by a trained physical therapist
 - Joint scores were recorded for each elbow, knee, and ankle, and comprised the following domains:
 - Swelling (0=none; 1=mild; 2=moderate; 3=severe)
 - Duration of swelling (0=none or <6 months; 1= \geq 6 months)
 - Muscle atrophy (0=none; 1=mild; 2=severe)
 - Crepitus on motion (0=none; 1=mild; 2=severe)
 - Flexion loss (0= $<5^\circ$; 1= 5° to 10° ; 2= 11° to 20° ; 3= $>20^\circ$)
 - Extension loss (0= $<5^\circ$; 1= 5° to 10° ; 2= 11° to 20° ; 3= $>20^\circ$)
 - Pain (0=none through active range of motion; 1=none through active range, only pain on gentle overpressure or palpation; 2=pain through active range)
 - Strength (0=holds position against gravity with maximum resistance [gr.5]; 1=holds position against gravity with moderate resistance, but breaks with maximal resistance [gr.4]; 2=holds position with minimal resistance [gr.3+] or against gravity [gr.3]; 3=partially completes range of motion against gravity [gr.3-/2+] or moves through range of motion with gravity eliminated [gr.2] or through partial range of motion with gravity eliminated [gr.2-]; 4=trace [gr.1] or no muscle contraction [gr.0])
 - Global gait scores assessed walking, stairs, running, and hopping on 1 leg skills (0=all skills normal; 1=one skill not normal; 2=two skills not normal; 3=three skills not normal; 4=no skills normal)
 - Total scores were calculated from joint total and global gait scores

Results

- Overall, 381 patients enrolled with a median age of 34 years; 65% self-reported arthritis/bone/joint problems, and 50% had a history of joint procedures/surgeries
- HJHS was completed for 240 patients
- HJHS joint totals, global gait score, and total score are shown in **Figure 1**; joint scores were higher (more impaired) for ankles than for elbows and knees

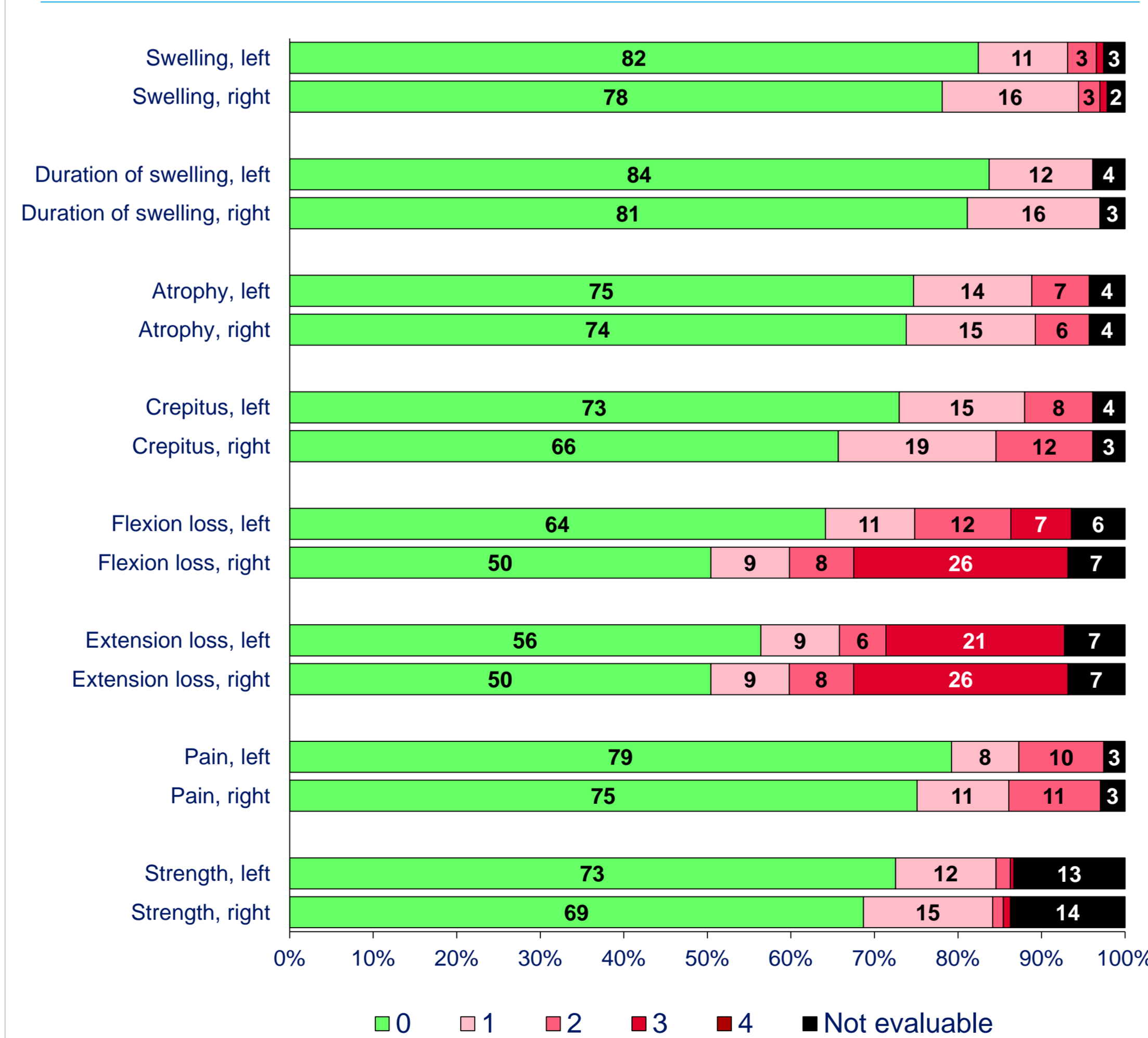
Figure 1 Joint totals, global gait score, and total score



Elbows

- The most affected elbow domains (those with most scores ≥ 1) were extension loss (loss of $\geq 5^\circ$ for 36% of left joints and 43% of right joints) and flexion loss (loss of $\geq 5^\circ$ for 29% of left joints and 43% of right joints) (**Figure 2**)
- The least affected domains were swelling and duration of swelling

Figure 2 Elbow scores^a

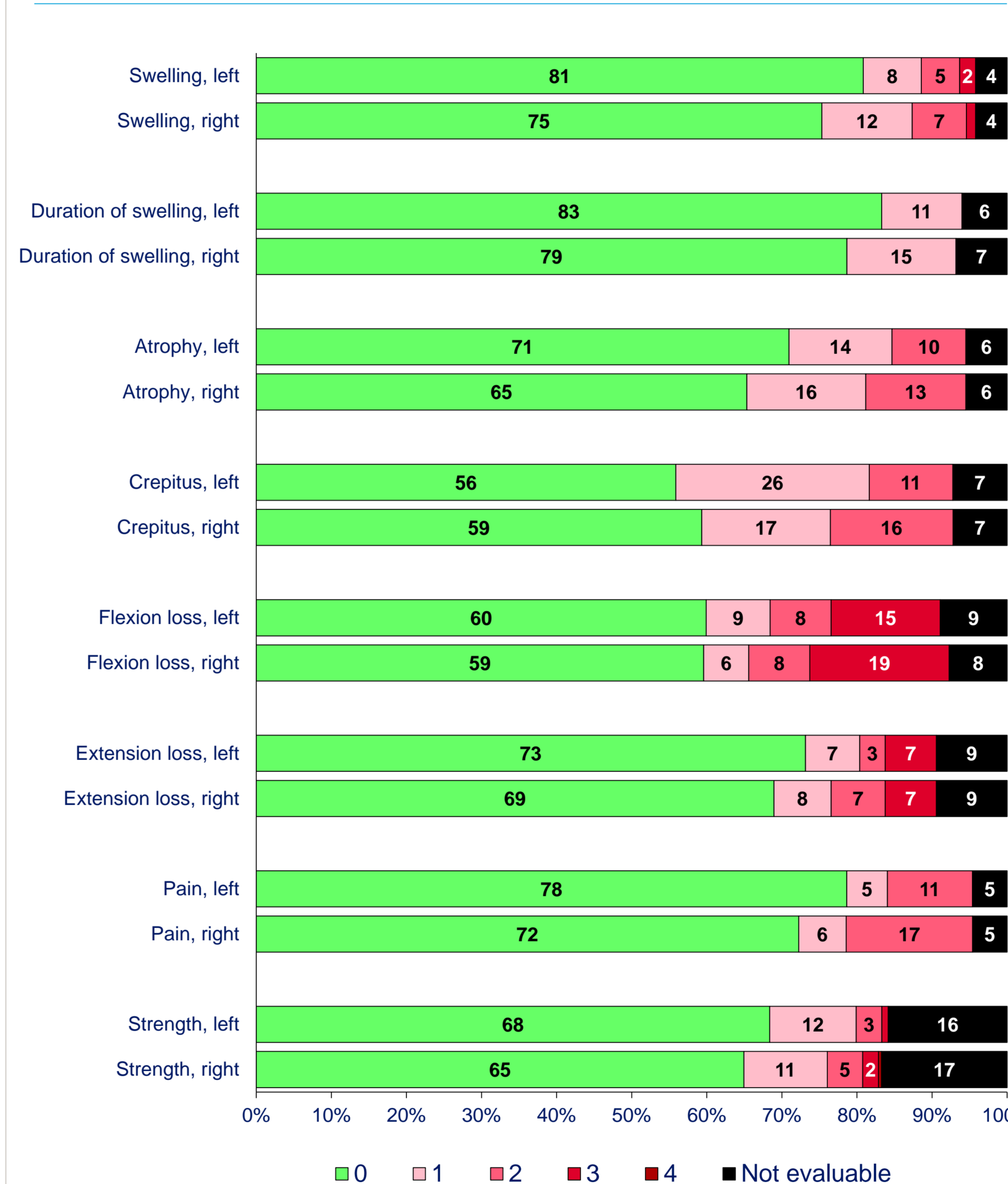


^aDomains differ in assessment ranges, as described in Methods.

Knees

- The most affected knee domains (those with most scores ≥ 1) were crepitus on motion (mild to severe for 37% of left joints and 33% of right joints) and flexion loss (loss of $\geq 5^\circ$ for 31% of left joints and 33% of right joints) (**Figure 3**)
- The least affected domains were swelling and duration of swelling

Figure 3 Knee scores^a



^aDomains differ in assessment ranges, as described in Methods.

Ankles

- Ankle domains were generally more affected than those of other joints (**Figure 4**)
- The lowest reported score (least affected domain) was for pain (pain on palpation or through active range of motion for 28% of left joints and 30% of right joints)

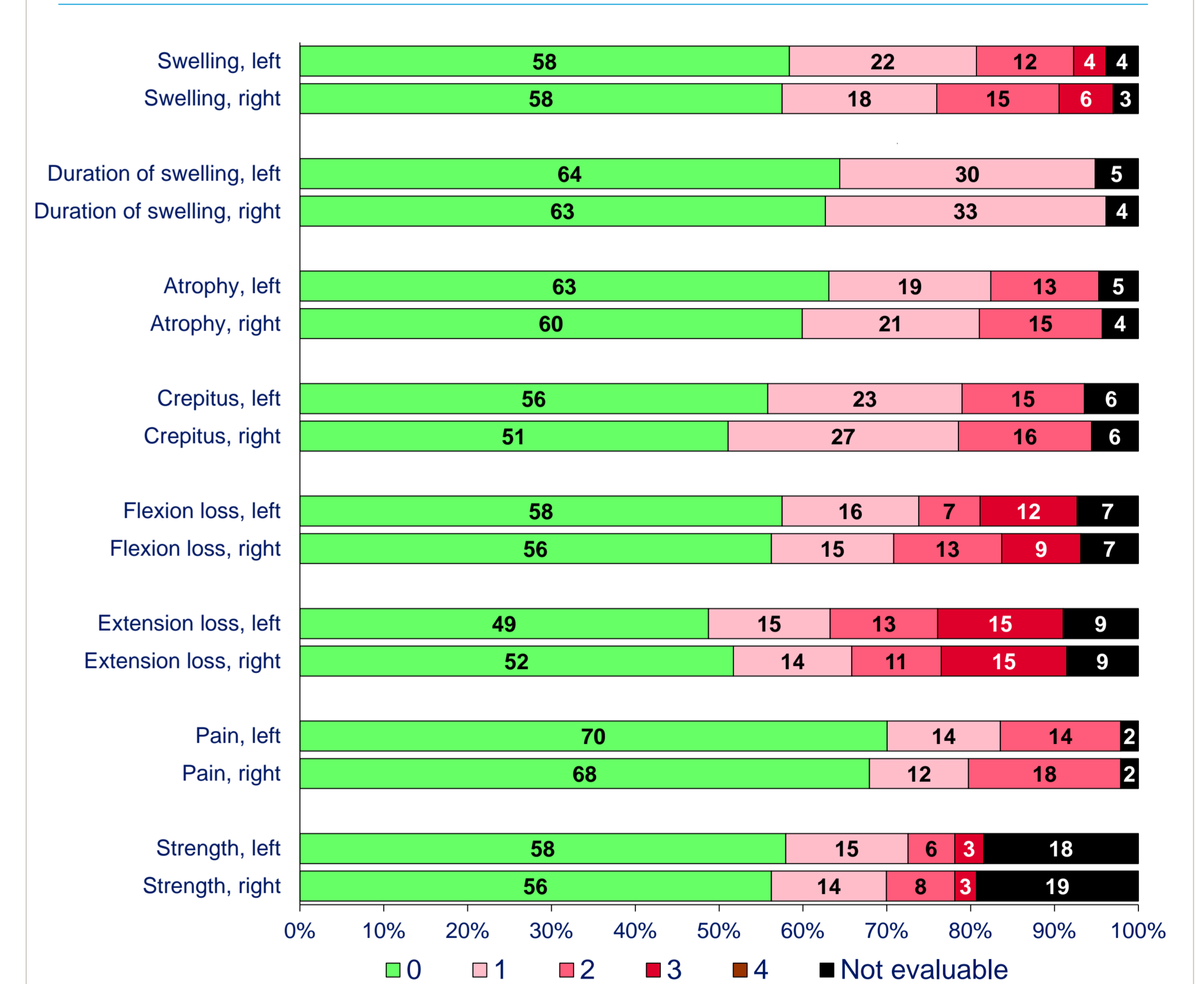
Global gait

- Most participants (68%) exhibited functional limitations within the global gait domain (walking, stairs, running, or hopping on 1 leg) (**Figure 5**)

Conclusions

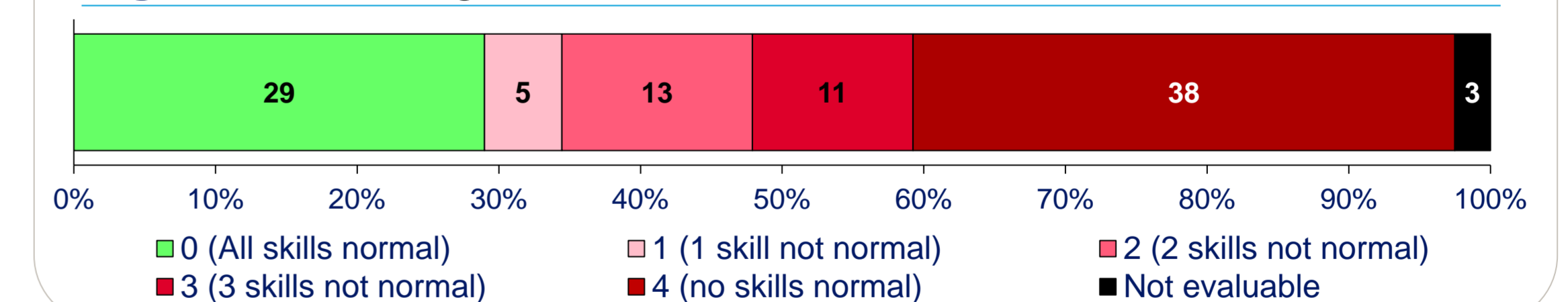
- Ankles were more severely affected by arthropathy than elbows and knees on HJHS; loss of range of motion (flexion and/or extension) was one of the more impacted domains across all 3 joints
- Detailed investigation of HJHS component scores could potentially lead to a simplification of this tool for greater research and clinical use in adults

Figure 4 Ankle scores^a



^aDomains differ in assessment ranges, as described in Methods.

Figure 5 Global gait scores



Conflict of interest disclosure

GH has received grant/research support from Bayer and Novo Nordisk and has served on advisory boards for Biogen Idec and on the speakers bureau for Emergent BioSolutions. KB has served as a consultant for Bayer and Novo Nordisk and as a speaker for Baxter, Bayer, and Novo Nordisk. MR has received grant/research support from Baxter, Biogen Idec, Novo Nordisk, and Pfizer and served as a consultant for Kedron and Novo Nordisk. AN has served on advisory boards for Kedron and Novo Nordisk. DC is on advisory boards for Baxter, Bayer, Biogen, and Novo Nordisk and speakers bureaus for Baxter, Biogen, Grifols, and Novo Nordisk. CK has served as a consultant for Baxter, Bayer, Biogen, Genentech, Grifols, Novo Nordisk, Octapharma, and Pfizer and received grant/research support from Bayer, Novo Nordisk, and Octapharma. MW has served as a consultant for Baxter, Biogen, CSL Behring, and Novo Nordisk. TS has served as a consultant for Baxter and Novo Nordisk. LB is a consultant for Baxter, Bayer, CSL Behring, Novo Nordisk, and Octapharma and has received research support from Baxter, Bayer, CSL Behring, Novo Nordisk, Octapharma, OPKO, and Selezys. RG is an employee of Novo Nordisk Inc. DC is an employee of Novo Nordisk Inc. CK has served as a consultant for Baxter, Biogen, CSL Behring, Hoffman La-Roche, and Kedron and received grant/research support from Novo Nordisk.

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