

Physical Activity of Severe Haemophilia A Patients Receiving Prophylaxis with Helixate NexGen

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Introduction

- Until the mid 70ies sport was discouraged in people with haemophilia (PWH) due to the bleeding risk [1]. Nowadays there is an increasing recognition that physical activity is important for people with haemophilia (PWH) [2,3]. Moreover, prophylaxis is beneficial to avoid bleeds, prevent joint damage and improve HRQoL [4], allowing PWH to practice individualised sport.
- The observational, prospective, non-interventional, SHAPE Study evaluated adherence to long-term prophylaxis by monitoring changes in clinical outcomes and physical activity in 14 centres in Italy over 36 months. We evaluated the impact of prophylaxis on physical activity of severe haemophilia A patients. This Study was sponsored by CSL Behring S.p.A., Italy.

Methods

- Patients of any age suffering from severe Haemophilia A, who have been prescribed a long-term prophylaxis regimen (at least 1 infusion/week for at least 46 weeks/year) with Helixate NexGen were included.
- Physical functioning and physical activity were assessed via self-rated questionnaires (HEP-TEST-Q [5]; EPIC Norfolk Physical Activity Questionnaire [6]). Demographic and clinical data were collected via electronic CRFs; the orthopaedic status was assessed with the WFH Orthopaedic Joint Score (OJS) or the Hemophilia Joint Health Score (HJHS).

Results

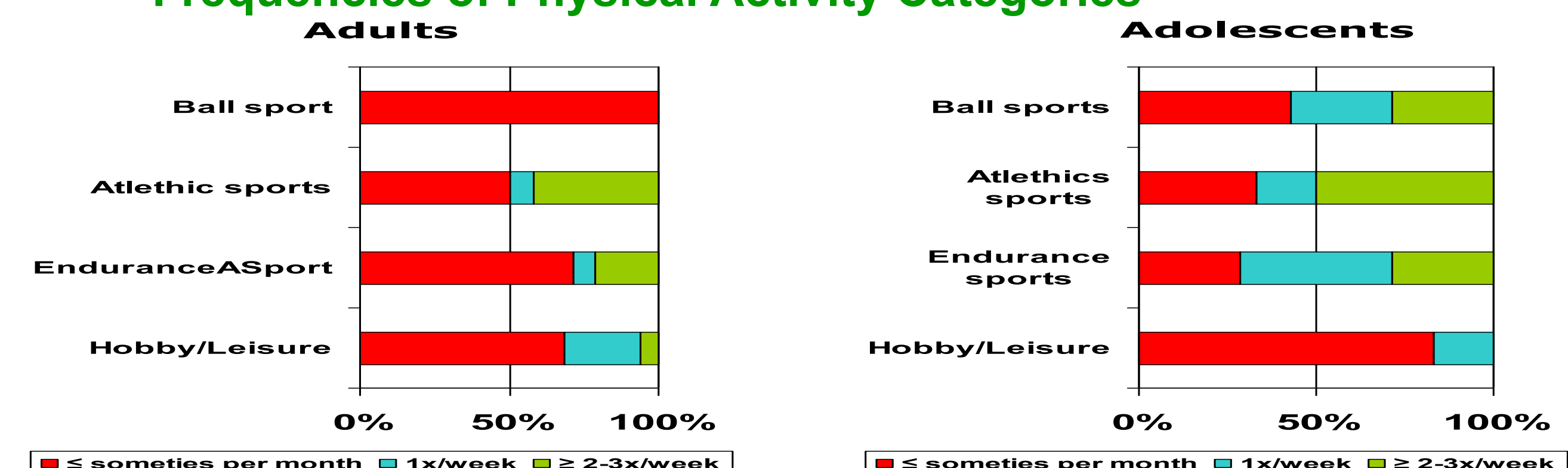
- 42 patients were enrolled; 47.6% adults, 21.4% adolescents (12-17 years) and 31% children (0-11 years).
- Patients had a median number of 1 bleed in the previous year and a mean OJS of ($M_{adults}=10.15\pm 9.8$; $M_{children}=1.63\pm 2.2$).

- Only adolescents (n=9) and adults (n=20) were asked questions about sports.
 - Only 1 adult did not practice sports.
 - Most of the patients were doing endurance sports, followed by hobby activities. Main physical activities in adults were walking for pleasure, Do-It-Yourself (DIY) and floor exercises; none of the adults played football, whereas adolescents were mainly walking for pleasure, playing netball, volleyball or basketball and football.

Categories of Physical Activities Regarding the Type of Activity

	Adolescents (n=7)	Adults (n=16)	P
Endurance sports	7	14	n.s.
Hobby/Leisure	6	14	n.s.
Athletic sports	5	10	n.s.
Ball sports	7	5	.005

Frequencies of Physical Activity Categories

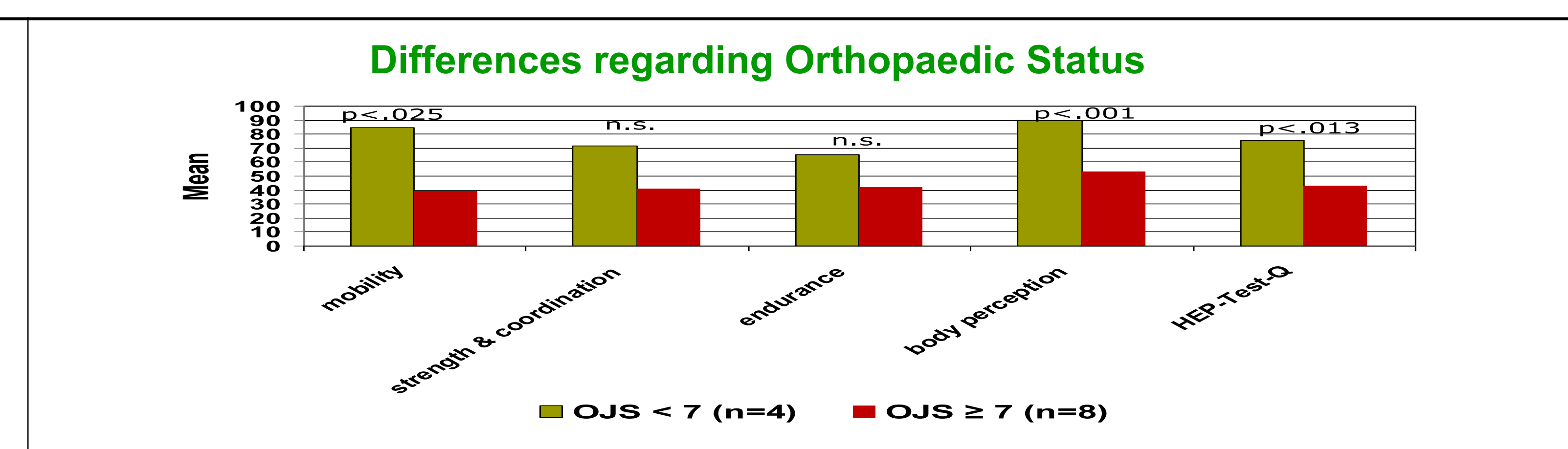
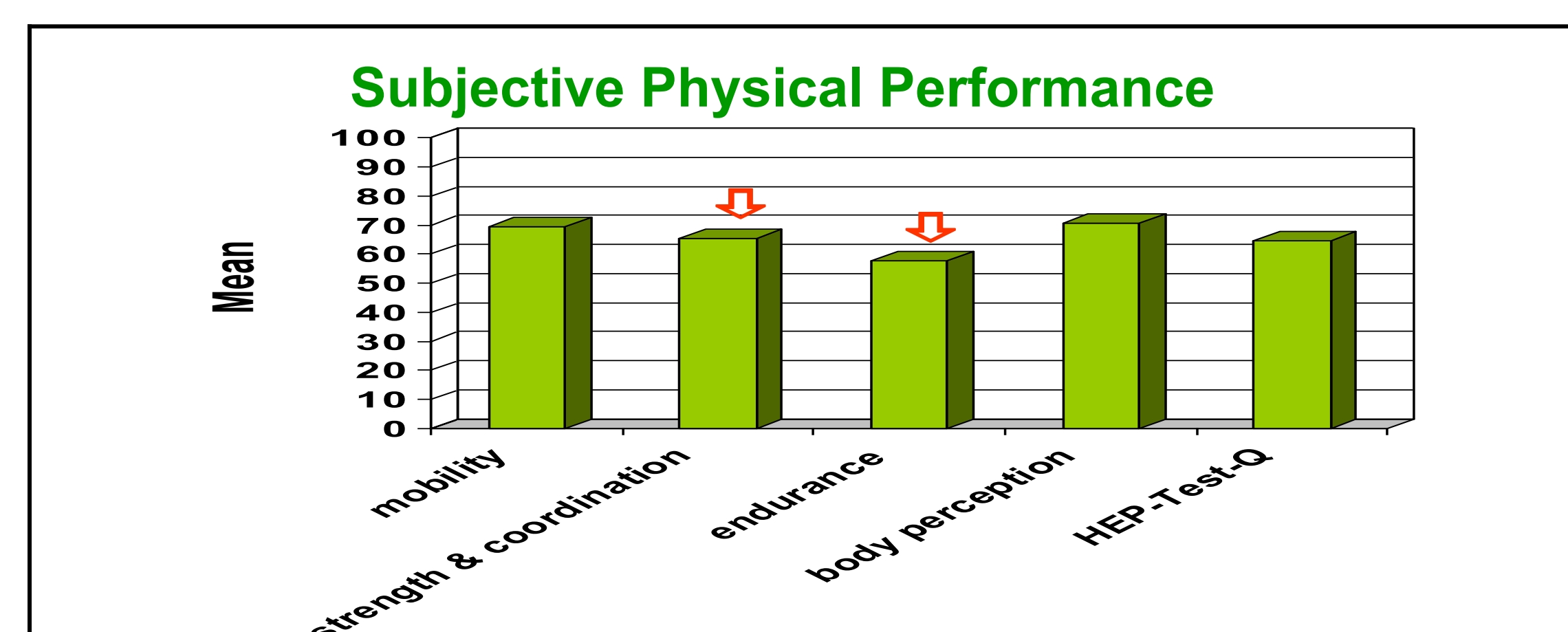


Categories of Physical Activities Regarding their Impact on the Joints of PWH

	Adolescents (n=7)	Adults (n=16)	p
No impact*	6	15	n.s.
Medium impact**	7	13	n.s.
High impact***	6	-	<.0001

* e.g. swimming, walking, bicycling; ** e.g. tennis, rowing, volleyball; *** e.g. football, hockey, boxing

Subjective Physical Performance (HEP-Test-Q)



Conclusion

There was a difference between adolescents and adults in type, frequency and impact of practiced physical activities. Highest impairments in the HEP-Test-Q were found in the domain 'endurance', 32.4% reported a fair/poor actual physical activity and in the domain 'strength & coordination', 47.2% could never/seldom carry out exhausting activities. PWH with a better orthopaedic joint status reported a better subjective physical performance.

References: [1] Weigle N & Carson BR. Physical activity and the hemophiliac: yes or no? *Am Correct Ther J* 1975; 29: 197-205. [2] Negrier C, Seuser A, Forsyth A, Lobet S, Llinas A, Rosas M, Heijnen L. The benefits of exercise for patients with haemophilia and recommendations for safe and effective physical activity. *Haemophilia* 2013;19(4):487-98. [3] von Mackensen S. Quality of life and sports activities in patients with haemophilia. *Haemophilia* 2007; 13 (supp 2): 38-43. [4] Khawaji M, Astermark J, Berntorp E. Lifelong prophylaxis in a large cohort of adult patients with severe haemophilia: a beneficial effect on orthopaedic outcome and quality of life. *Eur J Haematol.* 2012;88(4):329-35. [5] von Mackensen S., Czepa D., Herbsleb M., Hilberg T.: Development and validation of a new questionnaire for the assessment of subjective physical performance in adult patients with haemophilia – the HEP-Test-Q. *Haemophilia* 2010; 16:170–178. [6] Wareham NJ., Jakes RW., Rennie KL., Mitchell J., Hennings S., Day NE. Validity and repeatability of the EPIC-Norfolk Physical Activity Questionnaire. *International Journal of Epidemiology*, 2002, 31, 168-174.

