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Integrated postural analysis in children with haemophilia

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Summary. The maintenance of a correct posture in haemophilic boys might contribute to prevent joint bleeds, chronic pain and dysfunction. This single-centre study was aimed at evaluating whether or not postural alterations are more common in haemophilic than in non-haemophilic boys and whether they are related to the orthopaedic status. Posture and balance were investigated in boys with severe/moderate haemophilia (cases) and in age-matched non-haemophilic peers (controls). Thirty-five cases (89% with haemophilia A: 74% with severe disease) were included in the study and compared with 57 controls. Posture was evaluated on digital pictures of anterior, lateral and posterior views of the habitual standing position. Balance was examined with a portable force platform with eyes open and closed. The trajectory of the total body centre of force (CoF) displacement over the platform was computed by multiple planes obtaining different measures: sway area, velocity, acceleration and body loads. The joint status of cases was assessed with the Haemophilia Joint Health Score. Cases were more disharmonic than controls (52% vs. 26% in controls; $P = 0.04$), swayed significantly less and more slowly than controls ($P < 0.05$ for several parameters of CoF displacement) revealing stiffness of the musculoskeletal system. However, they were able to maintain their stance within a similar sway area. Haemophilic boys have more postural disharmonies than non-haemophilic peers, hence a global evaluation of the orthopaedic status should include also balance and posture examination to identify early dysfunction and establish a tailored physical or rehabilitation programme.

Keywords: balance, children, haemophilia, joint score, orthopaedic status, posture

Introduction

Boys with haemophilia are susceptible to clinical and subclinical joint bleeding that may cause irreversible joint damage. Some degree of damage may already occur after the first haemarthrosis or even in children who never experienced clinically evident joint bleeds [1]. Joints are mechanical systems with a structure strictly related to functioning. Therefore, any alteration in structure may have an impact on function (starting from the primary level of posture, balance and podalic support), which might in turn stress the joints and increase the risk of bleeding. Posture is a strategy implemented by the human body to maintain balance in response to gravity, motion and environmental stimuli in the most comfortable, harmonious and economical fashion, so that stress exerted on the musculoskeletal system is minimal [2]. The regulation of posture involves the balance system [3], which is aimed at maintaining the projection of the gravity centre within the maintenance polygon; and the fine postural system, which is aimed at maintaining the projection of the gravity centre in the actual centre of the maintenance polygon. Posture is not a synonymous of balance; the difference may appear subtle, however, the former is something more refined and complex than the latter. For instance, the more correct is posture, the narrower is the projection on the maintenance polygon, while the more correct is balance, the broader is the aforementioned projection. The postural system is a complex one, because it implies continuous changes and adaptations with the involvement of different regulation mechanisms. Besides a feedback control, a feed-forward control is involved in the regulation of fine postural adaptive mechanisms [4]. The essential components of the postural system are

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peripheral receptors that are sensitive to environmental and somatic inputs (i.e. sensorial, proprioceptive and esteroceptive).

With this as background, the maintenance of a correct posture, and hence the correction of any musculoskeletal imbalance in boys with haemophilia, might contribute to prevent joint bleeds and/or chronic pain and dysfunction [5–7]. An integrated postural analysis is needed to objectively evaluate the presence of any functional imbalance that, stemming or not from structural changes, may facilitate the onset or progression of joint damage. This study was designed to evaluate whether or not postural alterations are more common in boys with haemophilia than in a control group of age-matched healthy boys, both evaluated with traditional and innovative methods of postural analysis. Moreover, additional aims of the study were to determine the relationship of posture and balance with clinical orthopaedic scores (as the Haemophilia Joint Health Score, HJHS) in children with haemophilia and to compare haemophilic boys without joint involvement with controls.

Materials and methods

Subjects

Posture and balance were investigated in a group of boys with severe or moderate haemophilia (referred to as cases) regularly followed up at the Angelo Bianchi Bonomi Hemophilia and Thrombosis Center of Milan, and compared with a group of age-matched healthy boys (referred to as controls). Overall 35 cases (31 with haemophilia A and 4 with haemophilia B; 26 with severe and 9 with moderate haemophilia) were included in the study and compared with 57 healthy controls matched for age and gender. The control group included schoolfellows of the cases that accepted to participate as volunteers in the study. Parents or carers gave their written informed consent for the participation of boys in the study and all examinations were performed in conformity with the Principles of the Declaration of Helsinki on clinical research.

Physical examination

The musculoskeletal system was assessed in boys with haemophilia by using the Haemophilia Joint Health Score (HJHS, version 2.1) developed by the International Prophylaxis Study Group [8] and validated as a sensitive tool able to ascertain the presence of joint damage even at an early stage and at very young ages [9]. HJHS was calculated by the same operator (G.P.) in all cases. The total joint score (the sum of the six joint scores) and the global gait score, when combined, provide an overall total score ranging from 0

(healthy joint status) to 124 (severe haemophilic arthropathy in multiple joints) [8,9].

Postural examination was performed during habitual standing from the anterior, lateral and posterior views. To quantify global alignment characteristics, *post hoc* analysis of digital images taken by a digital camera and a clinical screening protocol were used. Participants were instructed to stand in their usual, comfortable and relaxed posture, with resting arms along the sides, feet apart at their shoulder width and equally balanced on both feet. They were asked to wear only a bathing suit or underpants to adequately expose body areas useful to perform the postural examination by using the following anatomical landmarks: the head, the earlobe, the seventh cervical vertebra, the acromial process, the thoracic spine, the sacrum and the lateral malleolus [10]. To standardize head posture, participants stared at a visual target (i.e. a 6-cm-diameter circle) set 1.5 m in front of them at eye level. As already reported, measures were obtained by means of serial pictures of the anterior, lateral and posterior views obtained by a 12.40 megapixel digital camera. The camera was fixed on a tripod and maintained at a height specific for each participant so that his entire spine length was in view. A level was used to maintain camera position between sessions. [11,12].

As previously reported, *normal posture from the anterior view* was defined by the alignment of the four main body functional units (head, shoulders, pelvis and ankles); *normal posture from the lateral view* was defined by looking at a vertical line touching the earlobe, the seventh cervical vertebra, the acromial process, the greater trochanter (just anterior to the midline of the knee and slightly anterior to the lateral malleolus); and *normal posture from the posterior view* was defined by looking at a vertical line passing through the head, the seventh cervical vertebra, the thoracic spine and the sacrum [10,13]. Postural abnormalities were operationally defined on a 3-point scale as follows: normal, presence of one abnormality or presence of more than one abnormality (referred to as disharmonies).

Balance examination

A portable force platform plus pedobarography (BioPostural System, AXA Srl, Vimercate, Italy) was used for data acquisition during postural evaluations. Participants were instructed to maintain an upright standing position for 51.2 s with their arms along their sides and feet positioned over footprints traced on the platform with an angle of 30° respect to the antero-posterior (AP) direction as already reported [14]. Data were collected in the morning in two different conditions assessed one after the other during the same visit: first with open eyes, staring at the aforementioned 6-cm-diameter circle, and then by keeping

eyes closed in order to avoid possible learning effects or effects of fatigue. All subjects were evaluated after one attempt only under each condition. A 60 s rest was allowed between the two assessments. Plantar pressure and upper limb dominance (i.e. right- or left-handed) were registered as well.

The trajectory of the total body centre of force (CoF) over the support surface was computed from the vertical force of the force platform and the body load measured as well. A total of four CoF measures were computed from the CoF displacement in the horizontal plane: CoF displacement that evaluated the CoF trajectory over the support surface; Sway Area that estimated the area covered by the CoF; velocity and acceleration that estimated the velocity and the acceleration of body sway over the support surface [15].

Statistical analysis

Continuous variables, expressed as median values and interquartile ranges (IQR), were compared by the Student t-test or the Mann–Whitney U-test. Categorical variables, expressed as frequencies and percentage values, were compared by chi-squared or Fisher's exact test. All reported P values are two-sided and values <0.05 were considered statistically significant. All analyses were performed by using the SPSS software (release 16.0, SPSS Inc., Chicago, Illinois).

Results

The median age at the time of examination was similar in cases and controls (12 and 13 years; IQR: 9–14 and 9–15 respectively). At the time of postural evaluation 25 cases (71%) were on regular prophylaxis (i.e. FVIII/FIX concentrates were regularly infused 2–3 times per week).

Physical examination

The HJHS score was zero in 25 (71%) haemophilic children, 18 (72%) of whom were on regular prophylaxis. The proportion of patients with a zero score did not differ with respect to haemophilia treatment regimen (i.e. on demand or prophylaxis). In the remaining 10 cases, the median HJHS score was 3 (IQR: 2–8), all of them having at least one target joint while only two cases of those cases with a zero score had target joints (8%).

Postural analysis

Postural analysis was performed in 29 cases (83%) and in all the 57 controls, because the digital camera was not available during the assessment visit of six cases. The evaluation of the alignment of the four main functional units (head, shoulders, pelvis and ankles) by anterior view showed a higher prevalence

of disharmonies in cases than in controls. In particular, 15 children with haemophilia (52%) had more than one disharmony in comparison with 15 controls (26%; $P = 0.04$). By posterior and lateral views a similar prevalence of alignment alterations and/or changes of spinal curves was found in the two groups (data not shown).

Force platform analysis

Balance assessment was performed in all cases and controls, and the main parameters of balance and loading in the two groups are summarized in Table 1. Children with haemophilia swayed significantly less and more slowly than controls, however, they were able to maintain their stance within a similar sway area (Table 1). Unexpectedly, children with haemophilia showed a greater body load on the left side than controls (Table 1) and this difference remained statistically significant also considering right- and left-handed subjects separately (data not shown). The main parameters of pedobarography measured in two groups of haemophilic children, distinguished also on the basis of the HJHS, are shown in Table 1. Although not statistically significant, the median values of acceleration and velocity of CoF displacement were higher in haemophilic boys with a HJHS zero score than in those with clinical signs of muscoloskeletal changes, rendering the former cases more similar to healthy controls.

Discussion

This study, focused on postural control problems in children with haemophilia, is to our knowledge the first to have evaluated the posture and balance in children with haemophilia and their correlation with a clinical score. In our cohort, no significant association between the clinical score (HJHS) and balance was shown in children with haemophilia. At variance with previously reported data in adults with haemophilia without arthropathy in whom a balance defect had been reported [16], our haemophilic boys with no joint involvement had balance similar to their age and sex peers. These results may be explained by an increased level of physical activity in young generation of haemophiliacs as well as by early implementation of prophylaxis regimens that allowed maintenance of a zero clinical score in the vast majority of our haemophilic boys.

The significant increase in load on the left foot independently from the hand side dominance should be explained by the effect of the dominant leg [17]. Indeed, leg dominance and its correlation with balance have been previously investigated [18,19], however, data on this aspect were not available. No significant association between the presence of signs of joint damage (assessed by HJHS) and body load was found in the patient population.

Table 1. Main parameters of force platform and pedobarography in 35 children with haemophilia (cases) distinguished on the basis of orthopaedic clinical score and 57 age-matched healthy boys (controls).

Parameter	Cases (n = 35)	Cases with HJHS > 0 (n = 10)	Cases with HJHS = 0 (n = 25)	Controls (n = 57)	P value*
Median velocity, mm/s (IQR)					
Eyes open	1.6 (1.4–2.6)	1.6 (1.4–1.9)	1.8 (1.4–2.7)	2.3 (1.8–3.5)	0.001
Eyes closed	1.9 (1.4–3.1)	1.8 (1.5–2.8)	2.0 (1.4–3.5)	2.5 (1.9–5.0)	0.02
Median acceleration, mm s^{-2} (IQR)					
Eyes open	9.4 (8.2–16.1)	8.4 (7.8–10.3)	10.2 (8.2–17.8)	13.0 (9.6–23.5)	0.01
Eyes closed	11.3 (7.9–18.8)	10.8 (8.8–15.7)	11.3 (7.7–25.6)	14.8 (9.8–29.6)	0.05
Median CoF displacement, mm (IQR)					
Eyes open	84.6 (74.2–131.8)	79.9 (73.9–98.9)	92.1 (74.1–139.5)	119.8 (90.8–182.0)	0.001
Eyes closed	98.6 (74.2–158.3)	92.0 (74.2–143.1)	103.1 (73.4–180.8)	127.6 (96.5–259.3)	0.02
Median sway area, mm² (IQR)					
Eyes open	8.4 (3.8–16.0)	9.0 (4.0–11.8)	8.4 (2.6–24.3)	11.0 (4.6–40.4)	ns
Eyes closed	8.2 (3.7–24.4)	8.4 (4.4–14.5)	8.2 (3.3–32.4)	11.4 (4.6–52.3)	ns
Median body load,% (IQR)					
Left load, eyes open	56.4 (52.8–58.2)	56.9 (52.8–58.5)	55.9 (52.8–58.1)	50.8 (49.4–53.1)	<0.001
Left load, eyes closed	56.3 (52.9–59.0)	56.2 (53.0–59.5)	56.4 (52.9–58.8)	50.1 (49.0–52.4)	<0.001
Right load, eyes open	43.6 (41.7–47.2)	43.1 (41.5–47.2)	44.1 (41.9–47.2)	49.1 (46.9–50.6)	<0.001
Right load, eyes closed	43.6 (40.9–47.1)	43.8 (40.5–47.0)	43.6 (41.2–47.1)	50.0 (47.6–51.0)	<0.001

HJHS, Hemophilia Joint Health Score; IQR, interquartile range; CoF, Centre of Force.
*P values are calculated comparing all cases with controls.

In haemophilic children posture was often abnormal but alterations were not associated with the clinical orthopaedic status nor with balance measures. This finding may be explained considering that posture is the result of a complex interactions between multiple variables, some of which are dynamic and should be also assessed with different tools (i.e. motion analysis). Moreover, the absence of association with the clinical score may be explained by the fact that HJHS was designed to evaluate signs of joint damage even at an early stage, but that the score is not sensitive enough to ascertain subtle postural changes that may occur also in the absence of arthropathy. Moreover, a larger sample size of children with clinically evident joint damage might provide more data on this possible correlation. Functional disturbances may occur in early childhood before any structural changes are diagnosed: unbalance and postural disharmonies may cause uneven joint loading as well as reduced mechanical stimulation, which on the long-term would reduce cartilage nutrition, and produce tissue degeneration leading to haemophilic arthropathy [20]. Similar pathogenetic mechanisms have been already described in haemophilia as, for instance in case of haemarthrosis, being in that case a biochemical signal of the early steps of the degenerative pathway (i.e. inflammation induced by presence of blood within the joint) [21]. It can be assumed that posture and balance changes may trigger mechanically similar degenerative pathway.

The main limitation of this study is the lack of a dynamic evaluation of posture, as early changes in function can be further analysed by means of motion analysis [22]. Unfortunately, such analysis could not be performed as the dedicated equipment is not available at our Center.

In conclusion, on the basis of these results indicating that haemophilic children have more postural disharmonies than their non-haemophilic peers, a global evaluation of the orthopaedic status of children with haemophilia should include not only the assessment of a clinical orthopaedic score (i.e. HJHS) but also balance and posture examination, to identify early dysfunctions and establish a tailored rehabilitation programme. In particular, posture should be part of the routine assessment of haemophilic children, because it can be considered an expression of the static and dynamic adaptations of the body to physical problems, even in the absence of clinical signs of arthropathy. Because proprioception, coordination and stretching are necessary tools to prevent the establishment of potentially harmful compensatory strategies, the identification of posture and/or balance alterations in boys with haemophilia may prompt the implementation of corrective physiotherapeutic interventions.

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Author contributions

EB, GP and MEM conceived the study and were involved in study design, acquisition of data, analysis of data and manuscript preparation; ES, AS, FP, PMM, MEM and LPS were involved in the interpretation of data and revision of the manuscript. All the authors read and approved the final version of the paper.

Disclosures

The authors stated that they had no interests which might be perceived as posing a conflict or bias.

References

1 Manco-Johnson MJ, Abshire TC, Shapiro AD *et al.* Prophylaxis versus episodic treatment to prevent joint disease in boys with severe haemophilia. *N Engl J Med* 2007; 357: 535–44.

2 Woollacott M, Shumway-Cook A. Attention and the control of posture and gait: a review of an emerging area of research. *Gait Posture* 2002; 16: 1–14.

3 Andriacchi TP, Strickland AB. Gait analysis is a tool to assess joint kinetics. In: Berme N, Engin AE, Correia Da Silva KM eds. *Biomechanics of Normal and Pathological Human Articulating joints.* NATO ASI Series, Martinus Nijhoff: Dordecht, 1993: 83–102.

4 Winter DA. Human balance and posture control during standing and walking. *Gait Posture* 1995; 3: 193–214.

5 Filho J, Battistella LR, Lourenco C. Computerized pedobarography in the characterization of ankle-foot instabilities of haemophilic patients. *Haemophilia* 2006; 12: 140–6.

6 Buzzard BM. Proprioceptive training haemophilia. *Haemophilia* 1998; 4: 528–31.

7 Hilberg T, Herbsleb M, Gabriel HHW, Jeschke D, Schramm W. Proprioception and isometric muscular strength in haemophilic subjects. *Haemophilia* 2001; 7: 582–8.

8 Hilliard P, Funk S, Zourikian N *et al.* Haemophilia joint health score reliability study. *Haemophilia* 2006; 12: 18–25.

9 Feldman BM, Funk SM, Bergstrom BM *et al.* Validation of a new paediatric joint scoring system from the International Hemophilia Prophylaxis Study Group: validity of the haemophilia joint health score. *Arthritis Care Res* 2011; 63: 223–30.

10 Kendall FP, McCreary EK, Provance PG. *Muscle Testing and Function with Posture and Pain,* Vol. 103. Philadelphia, PA: Williams & Wilkins, 1993.

11 Dunk NM, Lalonde J, Callaghan JP. Implications for the use of postural analysis as a clinical diagnostic tool: reliability of quantifying upright standing spinal postures from photographic images. *J Manipulative Physiol Ther* 2005; 28: 386–92.

12 Dolphens M, Cagnie B, Coorevits P *et al.* Sagittal standing posture and its association with spinal pain: a school-based epidemiological study of 1196 Flemish adolescents before age at peak height velocity. *Spine* 2012; 37: 1657–66.

13 Gagey PM, Weber B. *Régulation et dérèglements de la station debout.* Paris: Elsevier Masson, 2005.

14 Galli M, Rigoldi C, Celletti C *et al.* Postural analysis in time and frequency domains in patients with Ehlers-Danlos syndrome. *Res Dev Disabil* 2011; 32: 322–5.

15 Prieto TE, Myklebust JB, Hoffmann RG. Measures of postural steadiness: differences between healthy young and elderly patients. *IEEE Trans Biomed Eng* 1996; 43: 956–66.

16 Gallach JE, Querol F, Gonzalez LM, Pardo A, Aznar JA. Posturographic analysis of balance control in patients with haemophilic arthropathy. *Haemophilia* 2008; 14: 329–35.

17 Sadeghi H, Allard P, Prince F, Labelle H. Simmetry and limb dominance in able-bodied gait: a review. *Gait Posture* 2000; 12: 34–45.

18 Alonso AC, Brech GC, Bourquin AM, Greve JM. The influence of lower-limb dominance on postural balance. *Sao Paulo Med J* 2011; 129: 410–3.

19 Hoffman M, Schrader J, Applegate T, Koceja D. Unilateral postural control of the functionally dominant and nondominant extremities of healthy subjects. *J Athl Train* 1998; 33: 319–22.

20 Seuser A. Haemophilia and cartilage – the role of movement. *Hamostaseologie* 2012; 32: 52–61.

21 Roosendaal G, Lafeber FP. Pathogenesis of haemophilic arthropathy. *Haemophilia* 2006; 12: 117–21.

22 Seuser A, Wallny T, Schumpe G, Brackmann HH, Kramer C. Motion analysis in children with haemophilia. In: Rodriguez-Merchan EC ed. *The Haemophilic Joints; New Perspectives.* Oxford: Blackwell 2003; 155–62.