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# Frequency of postural alterations in haemophiliacs

# Frequência de alterações posturais em hemofílicos

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**RESUMO | INTRODUÇÃO:** A hemofilia é uma doença crônica de origem genética causada por uma mutação dos genes que codificam os fatores de coagulação sanguíneos. Uma das consequências dessa alteração é o acometimento do sistema musculoesquelético, o que pode influenciar na postura desses indivíduos. **OBJETIVO:** Estimar a frequência de alterações posturais em portadores de hemofilia, e identificar possíveis fatores associados. **MÉTODOS:** Estudo descritivo de corte transversal, conduzido com indivíduos com idade ≥ 18 anos, com diagnóstico de hemofilia, em acompanhamento na Fundação de Hematologia e Hemoterapia da Bahia (HEMOBA). Os participantes foram submetidos à avaliação postural, por meio da análise visual, teste de Adams e aplicação de um formulário semi-estruturado. Empregou-se o Epi Info® (v.3.5.2) para análise dos dados e, para verificar a existência de associações entre as variáveis do estudo foram utilizados os Testes exato de Fisher, Qui-quadrado (bicaudal) e Qui-Quadrado com a correção de Yates. Consideradas como estatisticamente significantes associações com p <0,05. **RESULTADOS:** Participaram deste estudo 29 hemofílicos, com média de idade de 34,9 ± 12,6 anos, 25 (86,2%) informaram ter artropatia hemofílica. Dos participantes da pesquisa 13 (68,4%) apresentaram postura escoliótica detectada pelo teste de Adams. A alteração foi mais frequente nos indivíduos com mais de uma articulação afetada pela artropatia hemofílica, com valor estatisticamente significativo para esta variável (p= 0,039). **CONCLUSÃO:** As frequências de alterações posturais em indivíduos com hemofilia são elevadas com associação significativa para os indivíduos com idade superior a 40 anos e com mais de uma articulação afetada pela artropatia hemofílica.

**PALAVRAS-CHAVE:** Hemofilia. Postura. Hemartrose. Hematoma.

**ABSTRACT | INTRODUCTION:** A hemophilia is a chronic disease of genetic origin caused by a mutation of the genes which encode blood clotting factors. One of the consequences of this alteration is the involvement of the musculoskeletal system, which may influence the posture of these individuals. **OBJECTIVE:** To estimate the frequency of postural changes in hemophilia patients, and identify possible factors associated. **METHODS:** A cross-sectional descriptive study, conducted with individuals aged ≥ 18 years,with diagnosis of hemophilia,in follow-up at the Hematology and Hemotherapy Foundation of Bahia (HEMOBA).The participants were submitted to postural evaluation,through visual analysis, Adams test and the application of a semi-structured form.Epi Info® (v.3.5.2) was used for data analysis and to verify the existence of associations between the variables of the study were used the Exact Fisher, Qui-Square and Qui-Square Tests with the Yates correction and considered as statistically significant associations with p <0.05. **RESULTS:** Twenty-nine hemophiliacs participated in this study, with mean age of 34.98 ± 12,6 years, 25 (86.2%) reported having hemophilic arthropathy. Of the 13 research participants (68.4%) presented scoliotic posture detected by the Adams test.The change was more frequent in individuals with more than one joint affected by hemophilic arthropathy, with a statistically significant value for this variable (p = 0.039). **CONCLUSION:** The frequencies of postural changes in individuals with hemophilia are elevated with a significant association for individuals over 40 years of age and with with more than one joint affected by haemophilic arthropathy.

**KEYWORDS:** Hemophilia. Posture. Hemarthrosis. Bruise.

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## Introduction

Haemophilia is a chronic disease of genetic origin caused by a mutation of genes encoding coagulation factors VIII (haemophilia A) and IX (haemophilia B)[1-3]. One of the consequences of this alteration is the deterioration of the musculoskeletal system2,3. There are about 400 thousand people affected by this disease in the world, being more frequent in males due to its hereditary character attached to the "X" chromosome. Women carry the gene of the disease, but rarely manifest the disorder1-3. It is estimated that in Brazil there are about 12,000 hemophilia patients, of whom 3,000 present the severe form of the disease, which leads, in some cases, to the individual's disability due to frequent hemorrhages[2,4].

Hemarthrosis represent one of the most frequent and disturbing clinical complications since it is predictive of functional limitations[2,3,5]. The degenerative process is usually polyarticular affecting mainly the articulation of the knees and ankles[2,6,7]. Conversely, hematomas occur mainly in the muscular system, more frequently in the iliopsoas muscle[2,8,9]. Thus, postural changes, particularly abnormal spinal deviations due to structural deformities, muscle imbalance and/or contractures, and poor postural habits[10,11], are one of the complications that these individuals are subject to[7].

Posture is characterized as the position that the body adopts in space, with the aim of establishing a balance between support structures, muscles and bones[12,13]. A good posture involves causing minimal stress on joints and a low energy expenditure in muscular activity[13]. Poor body alignment can alter the distribution of loads and pressures on joint surfaces, contributing to joint degeneration and muscular tensions[7,12,13]. Postural evaluation is used as a method for the diagnosis of posture changes and parameter for monitoring the effectiveness of a treatment and the adoption of preventive measures[14].

In view of the severity of joint complications due to hemophilia, its high frequency and lack of studies, in Brazil and particularly in the city of Salvador, on the incidence of postural changes in this population, it is necessary to carry out research on the issue. Therefore, the present study aims to estimate the frequency of postural changes in hemophilia patients attending the Hematology and Hemotherapy Center of Bahia, and identify possible associated factors.

## Material and methods

This is a descriptive cross-sectional study conducted with patients older than 18 years, diagnosed with hemophilia, followed at the Fundação de Hematologia e Hemoterapia da Bahia (Hematology and Hemotherapy Center of Bahia, HEMOBA), located in the city of Salvador, Bahia, Brazil. Data collection was conducted from August 2016 to March 2017. Everybody who was followed at the HEMOBA in the period of collection and accepted to participate in the study was included. Patients with postural changes from other diseases and those who reported previously diagnosed musculoskeletal and neurological diseases or episodes of hemarthrosis/ hematoma less than 15 days before data collection were excluded due to pain and edema.

Postural assessment and collection of the data were performed by previously trained researchers. A pilot study was carried out, in which a semi-structured form prepared by the authors and was applied and we performed the postural assessment through visual analysis. The pilot study was carried out with five hemophilia patients, followed at the Hemoba, during one week, aiming to calibrate and improve the collection instrument and standardize the postural evaluation. The semi-structured form presented sociodemographic, clinical and anthropometric variables.

With regard to socio-demographic variables, we asked about age in full years, skin color according to the IBGE, collapsed in white or not-white, schooling, marital status and work activity[15]. Regarding the anthropometric variables, we investigated the body weight in kg, height in meters, waist circumference and body mass index (BMI). From the BMI values it was possible to obtain the percentile classification of each participant, and their categorization of weight in leanness/eutrophic and overweight/obesity.

The type of hemophilia, whether A or B, classified according to factor VIII or IX deficiency, respectively, was included in the evaluation. The degree of haemophilia was inserted according to its severity

correlated with the presence of the activity of the coagulation factor: below 1%, severe, between 1 and 5%, moderate or between 6 and 30%, mild. In the present study we looked for the presence or not of the inhibitor of the coagulation factor and the type of factor replacement, whether by demand or prophylaxis[1]. We also evaluated, the presence of hemophilic arthropathy and how many joints were affected: only one or more. The presence of the target joint, deformity in flexion or extension, and physical therapy follow-up were also investigated. Laterality was defined as right-handed or left-handed. Physical activity was considered to be present for whom claimed to practice physical activity at least three times a week.

For postural evaluation, the patients were instructed to wear appropriate clothing, such as shorts, for men, and shorts and top for women. The postural evaluation of the subjects in the orthostatic position was subjectively previously evaluated, laterally and posteriorly, through visual analysis[14]. A semi-structured form was also used, in which, the information observed was marked in the standardized observation options. Subsequently, photographs of the participants were taken, in the same positions of the postural evaluation, in order to confirm the findings related to the postural evaluation, by the same examiner who performed the visual analysis. Digital photography was performed with a cellphone camera (Samsung® J5, 13 megapixels, resolution of 4128 x 3096 pixels) with use of flash and without zoom. The cellphone was positioned two and a half meters away from the volunteers and one meter high from the floor, the cell was perpendicular to the individual[16].

In the previous view, the presence or absence of asymmetry of the scapular and pelvic girdles was assessed, if there was leveling between the medial malleoli and the positioning of the ankles, whether neutral, pronated or supinated. We observed the angle of Thales, whether it was symmetrical, larger to the right or larger to the left. In a lateral view the position of the head was evaluated, whether centralized, anteriorized or extended; the positioning of the shoulders, if aligned in antepulsion or retrogression; the alignment of the knees, if aligned, curved or flexed; and whether there was an increase or decrease in thoracic kyphosis and lumbar lordosis. In the posterior view, the symmetry of the knees through the articular interline was verified, if leveled, larger to the right or larger to the left.

The Adams Forward Bend Test was selected to verify the presence of gibbosities. To perform the test, the participants were asked to stand in the orthostatic position, barefoot, with their feet together, their knees extended, and to lean their torso forward trying to touch their toes. The Adams test was considered positive when there was spinal deformity or alteration in the spine due to the projection of spinal apophyses[14].

The length of the lower limbs was verified according to the Magee method using a Universal Medical Products INC® measuring tape, with the volunteers positioned in a supine position on a stretcher, with a neutral hip rotation and medial malleoli the closest to the body's sagittal plane. Subsequently, the examiner, positioned next to the limb to be measured, located, through palpation, the anatomical points of the antero-superior iliac spine and the most distal and medial point of the medial malleolus of the same side. Using a tape measure, we measured from one point to another, recording the result in centimeters[14]. The type of foot, whether neutral, flat or cavus, was evaluated with the aid of a Cris Medical® podoscope, Model ref.552.9[15].

The database was created in the Microsoft Office Excel 2007 program, and Epi Info® (version 3.5.2) was used for data analysis – for calculation of means, standard deviation, absolute and relative frequencies to describe the variables of interest. The association between the variables of the study was verified by Fisher's exact Chi-square test (two-sided) and Chi-Square test with the Yates correction. Associations with $p < 0.05$ (95% CI) were considered statistically significant.

The research project was approved by the Research Ethics Committee of the Bahia State University (CEP-registration 055417/2016 UNEB on 06.13.2016) in accordance with the standards of the National Ethics Council Resolution 466/12 on Research (CONEP).

## Results

There were 37 hemophiliac individuals, followed-up by the Hemotherapy and Hematology Foundation of Bahia (HEMOBA). Three of them (8.1%) did not meet the inclusion criteria and five (10.8%) refused to participate in the study, totaling 29 individuals.

Table 1 shows the sociodemographic characteristics of the hemophiliacs. The sample was exclusively male, 29 (100%) and 22 (75.9%) were in the age group of 18 to 40 with a mean age of 34.9±12.6 years, 22 (75.9 %) considered themselves to be non-whites. As to schooling, 23 (79.3%) had complete or incomplete high school or college, 15 (51.7%) worked and 21 (72.5%) lived without a partner.

**Table 1.** Sociodemographic and anthropometric characteristics of the hemophiliacs followed-up at the HEMOBA. Salvador, Bahia, Brazil 2017.

| Variables | n=29 | % | Mean | SD |
|---|---|---|---|---|
| **Sex** | | | | |
| Male | 29 | 100 | - | - |
| Female | - | - | | |
| **Age** | | | | |
| 18-40 | 22 | 75,9 | 34,9 | 12,6 |
| >40 | 7 | 24,1 | | |
| **Skin color** | | | | |
| White | 7 | 24,1 | | |
| Non-white | 22 | 75,9 | - | - |
| **Schooling** | | | | |
| Illiterate /primary education | 6 | 20,7 | | |
| High school/complete or incomplete university | 23 | 79,3 | - | - |
| **Labor activity** | | | | |
| Work | 15 | 51,7 | - | - |
| Do not work | 6 | 20,7 | | |
| Retired/Receiving benefits | 8 | 27,6 | | |
| **Marital status** | | | | |
| Living with a partner | 8 | 27,5 | | |
| Living without a partner | 21 | 72,5 | - | - |

Source: Data from the research. SD=Standard Deviation.

Regarding the classification of the type of hemophilia, 22 (75.9%) were type A. With respect to severity of the disease, 21 (72.4%) were classified with the severe form, 21 (72.4%) reported that they had no coagulation factor inhibitor and 15 (51.7%) conducted a prophylactic replacement of the factor. Hemophilia arthropathy in one or more joints was reported in 25 (86.2%) individuals, 15 (51.7%) underwent physiotherapeutic treatment and 23 (79.3%) did not practice regular physical activity. Regarding body mass index (BMI), 15 (51.7%) were lean or eutrophic. (Table 2).

**Table 2.** Clinical characteristics of the hemophiliacs followed-up at the HEMOBA. Salvador, Bahia, Brazil, 2017

| Variables | n=29 | % |
|---|---|---|
| **Hemophilia Type** | | |
| A | 22 | 75,9 |
| B | 7 | 24,2 |
| **Level** | | |
| Mild | 2 | 6,9 |
| Moderated | 6 | 20,7 |
| Severe | 21 | 72,4 |
| **Presence of inhibitor** | | |
| Yes | 8 | 27,6 |
| No | 21 | 72,4 |
| **Type of replacement factor** | | |
| Demand | 14 | 48,3 |
| Prophylaxis | 15 | 51,7 |
| **Presence of arthropathy** | | |
| Yes | 25 | 86,2 |
| No | 4 | 13,8 |
| **Physiotherapy treatment** | | |
| Yes | 15 | 51,7 |
| No | 14 | 48,3 |
| **Physical activity** | | |
| Yes | 6 | 20,7 |
| No | 23 | 79,3 |
| **BMI** | | |
| Lean/eutrophic | 15 | 51,7 |
| Overweight/obese | 14 | 48,3 |

Source: Data from the research. BMI = Body Mass Index.

Table 3 shows the frequencies of postural changes in the hemophiliacs. The flexion deformity of one or both knees corresponded to 20 (69%), 20 (69%) presented a decrease in the change in plantar arch height, 20 (69%) had a difference in leg length greater than one centimeter, and 11 (37.9%) had ankle pronation changes. Increased lumbar lordosis was verified in 20 (69%) individuals, and the increase of thoracic kyphosis in 15 (51.7% ). The Adams test was negative in 15 (51.7%) hemophiliacs and 21 (72.4%) had asymmetry in the Thales angle.

**Table 3.** Frequency of postural changes in hemophiliacs followed-up at the HEMOBA. Salvador, Bahia, Brazil, 2017.

| Variables | n=29 | % |
|---|---|---|
| **Flexed knees** | | |
| No | 9 | 31 |
| Yes | 20 | 69 |
| **Type of foot** | | |
| Aligned | 3 | 10,3 |
| Flat | 20 | 69 |
| Cavus | 6 | 20,7 |
| **Lower limbs length** | | |
| ≤1 cm | 9 | 31 |
| >1cm | 20 | 69 |
| **Ankle** | | |
| Aligned | 10 | 34,5 |
| Pronated | 11 | 37,9 |
| Supinated | 8 | 27,6 |
| **Lumbar lordosis** | | |
| Physiological | 9 | 31 |
| Increased | 20 | 69 |
| Diminished | 0 | 0 |
| **Thoracic kyphosis** | | |
| Physiological | 14 | 48,3 |
| Increased | 15 | 51,7 |
| Diminished | 0 | 0 |
| **Adams' test** | | |
| Positive | 14 | 48,3 |
| Negative | 15 | 51,7 |
| **Thales' angle** | | |
| Symmetrical | 8 | 27,6 |
| Asymmetrical | 21 | 74,4 |

Source: Data from the research.

Table 4 shows the results of the association between the Adams test and the sociodemographic, clinical and posture variables of hemophiliacs. The presence of scoliotic posture, detected by the Adams test, was more frequent in the age group > 40 years, with a statistically significant value (p=0.031). Another variable that also revealed a statistically significant association with the Adams test was the number of joints with hemophilia arthropathy, with a statistically significant value (p=0.039). There was no other statistically significant relationship between the variables.

**Table 4.** Association between sociodemographic, clinical and postural variables with the Adams test in hemophiliacs followed-up at the HEMOBA. Salvador, Bahia, Brazil, 2017

| Variables | Positive Adams' test | | Negative Adams' test | | P-value |
|---|---|---|---|---|---|
| | n | % | n | % | |
| **Age** | | | | | |
| 18-40 | 8 | 36,4 | 14 | 63,6 | 0,031* |
| >40 | 6 | 85,7 | 1 | 14,3 | |
| **BMI** | | | | | |
| Lean/eutrophic | 6 | 40 | 9 | 60 | 0,364 |
| Overweight/Obesity | 8 | 57,1 | 6 | 42,9 | |
| **Laterality** | | | | | |
| Righthanded | 12 | 46,2 | 14 | 53,8 | 0,473 |
| Lefthanded | 2 | 66,7 | 1 | 33,3 | |
| **Physical Activity** | | | | | |
| Yes | 2 | 33,3 | 4 | 66,7 | 0,360 |
| No | 12 | 52,2 | 11 | 47,8 | |
| **Thales angle** | | | | | |
| Symmetrical | 2 | 25 | 6 | 75 | 0,128 |
| Asymmetrical | 12 | 57,1 | 9 | 42,9 | |
| **Flexed knees** | | | | | |
| No | 2 | 22,2 | 7 | 77,8 | 0,067 |
| Yes | 12 | 60,0 | 8 | 40 | |
| **Lower limbs length** | | | | | |
| ≤1 cm | 2 | 25 | 7 | 75 | 0,104 |
| >1cm | 12 | 57,1 | 8 | 42,9 | |
| **Number joints by haemophiliac arthropathy** | | | | | |
| Only 1 | 1 | 16,7 | 5 | 83,3 | 0,039* |
| + 1 | 13 | 68,4 | 6 | 31,6 | |

Source: Data from the research. Fisher's exact test. *P-value < 0,05.

**Table 5.** Frequency of postural changes in individuals with hemophilia in follow-up (HEMOBA), according to their hemophilia grade. Salvador, Bahia, Brazil, 2017.

| Variables | Mild Hemophilia | | Severe/moderated hemophilia | | P-value |
|---|---|---|---|---|---|
| | n | % | n | % | |
| **Thales Angles** | | | | | |
| Symmetrical | 1 | 12,5 | 7 | 87,5 | 0,482** |
| Asymmetrical | 1 | 4,8 | 20 | 95,2 | |
| **flexed knees** | | | | | |
| No | 0 | 0 | 9 | 100 | 0,946¥ |
| Yes | 2 | 10 | 18 | 90 | |
| **Ankle** | | | | | |
| Aligned | 1 | 10 | 9 | 90 | 0,578** |
| Non aligned | 1 | 5,3 | 18 | 94,7 | |
| **Lower limbs length** | | | | | |
| ≤1 cm | 1 | 11,1 | 8 | 88,9 | 0,532** |
| >1cm | 1 | 5 | 19 | 95 | |
| **Lumbar lordosis** | | | | | |
| Physiological | 0 | 0 | 9 | 100 | 0,084¥ |
| Increased | 2 | 10 | 18 | 90 | |
| **Thoracic Kyphosis** | | | | | |
| Physiological | 1 | 7,1 | 13 | 92,9 | 0,701** |
| Increased | 1 | 6,7 | 14 | 93,3 | |
| **Adams' test** | | | | | |
| Positive | 0 | 0 | 14 | 100 | 0,494¥ |
| Negative | 2 | 13,3 | 13 | 86,7 | |

Source: Data from the research. **Fisher's exact test. ¥Chi-square test with Yates correction.

Table 5 shows the frequencies of postural changes in hemophiliacs in follow-up at the HEMOBA according to the degree of haemophilia. It was verified that there was no statistically significant deference when analyzing Thales angles (p=0.482), flexed knees (p=0.946), ankle alignment (p=0.578), difference in the length of the lower limbs (p=0.532), (p = 0.084), alteration of thoracic kyphosis (p=0.701) as well as the variables alteration in lumbar lordosis (p=0,084), thoracic kyphosis (p=0,701) and Adams' test (p = 0.494). There was also no statistical difference in the postural variables, in relation to the type of replacement factor (Table 6).

**Table 6.** Postural changes of hemophiliac individuals followed-up at the HEMOBA, according to the type of replacement factor. Salvador, Bahia, Brazil, 2017.

| Variables | Demand | | Prophylaxis | | P-value |
|---|---|---|---|---|---|
| | n | % | n | % | |
| **Thales Angles** | | | | | |
| Symmetrical | 4 | 50 | 4 | 50 | 0,457** |
| Asymmetrical | 10 | 47,6 | 11 | 52,4 | |
| **flexed knees** | | | | | |
| No | 6 | 66,7 | 3 | 33,3 | 0,495** |
| Yes | 8 | 40 | 12 | 60 | |
| **Ankle** | | | | | |
| Aligned | 5 | 50 | 5 | 50 | 0,894† |
| Non aligned | 9 | 47,4 | 10 | 52,6 | |
| **Lower limbs length** | | | | | |
| ≤1 cm | 4 | 44,4 | 5 | 55,6 | 0,550† |
| >1cm | 10 | 50 | 10 | 50 | |
| **Lumbar lordosis** | | | | | |
| Physiological | 3 | 33,3 | 6 | 66,7 | 0,249** |
| Increased | 11 | 55 | 9 | 45 | |
| **Thoracic Kyphosis** | | | | | |
| Physiological | 8 | 57,1 | 6 | 42,9 | 0,364† |
| Increased | 6 | 40 | 9 | 60 | |
| **Adams' test** | | | | | |
| Positive | 6 | 42,9 | 8 | 57,1 | 0,579† |
| Negative | 8 | 53,3 | 7 | 46,7 | |

Source: Data from the research. ** Fisher's exact test. †Chi-square test.

## Discussion

In this study, the frequency of postural changes in individuals with hemophilia was high. The main alterations were deformity in knee flexion, decrease in the height of the plantar arch, increase in lumbar lordosis, increase in the angle of Thales and scoliotic posture. These findings corroborate those reported by Boccalandro et al.[17], in a study conducted in Milan, Italy, involving children with and without hemophilia, in which the frequency of postural changes was higher in those with the disease. However, the authors did not describe which postural changes were found.

In our study, the sex of the sample was exclusively male, and our findings resembles the results presented by Garbin et al.[18] in a study performed at a hospital school in the State of São Paulo. A possible explanation for these findings may be due to hemophilia being rare in females, since both X chromosomes should be affected[1]. As for age, a higher frequency was observed in young adults; similar results were found by Garbin et al.[18] in the aforementioned study. This low frequency of older individuals may be a consequence of the low life expectancy of these individuals, since the offered treatment was limited.

Regarding labor activity, it was observed that the majority of the individuals worked, however, it is worth noting that because it is a population composed predominantly of young adults, a significant portion of this population received social benefits or was retired due to the disease. Similar results were reported by Caio et al.[19] in a study performed with hemophiliacs, regularly followed at

the Hemocenter of the State University of Campinas (UNICAMP), Campinas, São Paulo. The authors compared hemophiliac individuals with their siblings without the disease, respecting the age group. It was possible to verify that the major obstacles of hemophilia patients are socioeconomic in nature – in addition to the reduced chances of access to regular employment and higher retirement rate, probably due to disability.

The practice of physical exercises can contribute to better quality of life and health, increasing muscle strength, joint function, balance and flexibility. Despite this, most participants in this study did not practice physical activities regularly. This finding corroborated the results of the study by Nunes et al.[20], in which the authors categorized the quality of life of hemophiliacs who were being followed-up at a regional hemocenter of São Paulo. These results differ from those found by Von Mackensene et al.[21] in a study conducted at four haemophilia centers across the UK that demonstrated a higher frequency of individuals performing physical activity, including contact sports, who had a lower rate of joint changes. One possible explanation for this divergence may be the culture of developed countries to promote primary prophylaxis in childhood, thus avoiding further joint damage.

This study also demonstrated a high frequency of deformities in knee flexion. These findings may be associated with the high frequency of haemophiliac arthropathies in the individuals from our study. Galante et al.[10] in a study with hemophiliac children with or without synovitis, in the age group of 3-7 years, reinforced this association between joint impairment and postural alteration. They showed that children with synovitis present a decrease of the knee angle (semiflexion) in relation to children without synovitis. In addition, they stated that the more global and precocious the articular alteration in the hemophiliac patient, the greater the chance of damages in joint alignment. This indicates the importance of early postural assessment, because when orthopedic problems are not corrected in childhood they can lead to limitations in adult life.

Santos et al.[11] also showed a high frequency of deformities in knee flexion, in a study performed with hemophiliacs accompanied at the Haemotherapy and Hematology Center of Espírito Santo. In this study, it was found that the most affected joints, due to deformities, were the knees. Similar data were demonstrated in the study by Bastos et al.[22] carried out in Vale do Paraíba Paulista, through a retrospective, descriptive, documentary research. Data collection was carried out in the medical records of hemophilia patients from the Hematology Outpatient Clinic. The authors reported a high incidence of hemarthrosis in these joints. This may be suggestive of a knee flexion contracture, since the frequencies of hemarthroses favored hemophilic arthropathy, with knee flexion contracture being one of its complications.

The increase of lumbar lordosis was verified in the majority of the individuals participating in this study. Although they did not find studies that specifically described this lumbar spine condition in hemophiliacs, Galatro et al.[9] reported that hematomas favor muscular shortening and/or fibrosis. When they occur in the iliopsoas muscle associated with the frequent hemarthroses in the knees and pain, they usually influence the individual to adopt an antalgic resting posture, with knees flexed, pelvic anteversion and consequent hyperlordosis. Kim et al.[23] investigated the relationship between trunk muscle strength and lumbar lordosis in 31 individuals without hemophilia, mean age 35 years. The authors observed that imbalance in muscle strength of the trunk may significantly influence the lordotic curve of the lumbar spine. These findings may justify the high frequency of changes in the lumbar spine, since for the maintenance of the orthostatic posture there is a need for strong, flexible muscles, easily adaptable to changes in the environment.

The presence of scoliotic posture, identified through the positive Adams test, was significantly associated with the number of joints affected by hemophilic arthropathy. This result is consistent with the literature[11]. Hoepers[24] reiterates this with his study conducted with hemophiliacs living in the State of Santa Catarina, in which it was observed that

hemophilic arthropathy mainly affects the joints of the knees. Hemophilic arthropathy generates a set of fixed contractures in the flexion of these joints, favoring a compensatory response of hip flexion and a position in ankle flexion, which can lead to a scoliotic posture[11]. No other studies were found that defined this correlation.

We observed a high presence of scoliotic posture detected by the Adams test in hemophiliac individuals over than 40 years old. This finding may be justified by the existence of a greater difficulty in accessing the treatment of this disease in the past, which favors a greater occurrence of alterations in the musculoskeletal system. No other studies have found that found this correlation with hemophiliac individuals.

It was not possible to establish a statistically significant relationship between the variables of postural changes and the degree of haemophilia. However, it was possible to observe that in the hemophiliacs with severe/moderate haemophilia the postural alterations were more frequent, mainly the changes related to lumbar lordosis. These findings may be associated with the increased predisposition to joint and muscle damage that these individuals are subject to[5]. We also did not find other quantitative studies performed with the same population, of the present study, that observed this relationship.

Despite factor replacement, by primary or secondary prophylaxis, preventing joint complications, there were no statistical associations between the type of factor replacement, if demand or prophylaxis, with the frequencies of postural changes. This result can be explained by the late implementation in Brazil of primary prophylaxis by the Ministry of Health in December 2011 and secondary prophylaxis in 2012[25]. It is also important to point out that the time in years that the hemophiliacs of this study reported prophylaxis was not sufficient to predict a differentiated postural pattern or to reverse postural changes that have occurred over the years, given that this is not the purpose of prophylaxis. No studies were found to contribute with similar data.

The use of cross-sectional design precludes causal inferences, even though such methodology has relevance to characterize the population. The low cost and easy access for data collection were advantages of this study, however, the non-accomplishment of a sample calculation and the reduced sample number can be considered as a fragility since it does not allow an extrapolation of results for individuals with hemophilia. Another issue to be considered is the fact that the postural evaluations were carried out by different evaluators and through visual analysis, a fact that may have generated a bias. To minimize this bias, previous training was conducted with the team of evaluators, in addition to conducting a pilot study.

## Conclusion

The results obtained in this study demonstrated a high frequency of postural alterations, mainly in the lower limbs and in the curvatures of the spine, with a significant association for the elderly over 40 years old and with more than one joint affected by hemophilic arthropathy. These findings can help to justify the need for the implementation of programs for prevention and early screening for health professionals and managers working directly with this population.

These actions may help to reduce the incidence of these disorders, as well as promote the early detection of the alterations, which is essential for the prevention of future complications that may contribute to the quality of life of these individuals and their functionality. However, further research on postural changes in this population, with a control group with healthy individuals and the association of postural assessment instruments, should be encouraged. It is thus advisable to reinforce the implementation of these programs as to investigate other associated variables.

**Authors' contributions**

Santos RS participated in the conception, design, search and statistical analysis of the research data, interpretation of the results, writing of the scientific article and sending the scientific article. Figueirôa GR participated in the design, search and statistical analysis of the research data, interpretation of the results and writing of the article. Machado BA participated in the search and statistical analysis of the research data, interpretation of the results and writing of the article. Mamede CA participated in the design, search and statistical analysis of the research data, and interpretation of the results.

**Competing interests**

No financial, legal or political competing interests with third parties (government, commercial, private foundation, etc.) were disclosed for any aspect of the submitted work (including but not limited to grants, data monitoring board, study design, manuscript preparation, statistical analysis, etc.).

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