CASE REPORT

# Results of a physical fitness program for a patient with severe hemophilia A

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## ABSTRACT

Hemophilia is a blood clotting disorder that causes the decrease or absence of blood coagulation factors VIII or IX. This disease causes a person to bleed longer than a normal person if it is not treated. A 31-year-old male with hemophilic arthropathy in the left elbow and right ankle was evaluated. The program lasted 20 months, with aerobic and muscle strength training. The muscles involved were the pectoralis major, latissimus dorsi, biceps brachii, triceps brachii, deltoideus, quadriceps femoris, and hamstring. The patient performed two series of 10 repetitions with 45 seconds between series. The intensity was based on the initial test of 10 maximum repetitions in accordance with the rate of perceived exertion on the Borg Scale. Aerobic exercise was performed on a horizontal stationary bike for 20 minutes. Pulse rate was registered at rest, after 10 and then 20 minutes of workout, and again after three minutes of recovery. Six months before the initiation of the fitness program the patient suffered three hemorrhages: two spontaneous in the left elbow and left ankle and one in the right leg due to a small unspecified trauma. All three hemorrhages were treated by means of clotting factors (CF's). During the program period, the first hemorrhage treated with CF's occurred in the left elbow by trauma after increasing the exercise load. Twelve months later, the patient had spontaneous bleeding in the same joint. The least improvement in strength was found for the triceps brachii with 33%, whereas the knee extensors improved the most with 257%. The average muscle strength increase was 121%. Monitored physical exercise is an important vehicle in treating of people with hemophilia, presenting the need for resistance training specific for hemophilic patients to prevent injuries and avoiding early wear of the musculoskeletal system. The objective of this study was to present the results of a physical activity program to prevent bleeding episodes in a person with severe hemophilia A and the prophylaxis without administering any clotting factors.

**Keywords:** Hemophilia A, Physical Conditioning, Human, Muscle Strength, Exercise

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## INTRODUCTION

According to the *Federação Brasileira de Hemofilia* (Brazilian Federation of Hemophilia) (2011), this pathology is caused by a genetic and/or hereditary blood disorder, characterized by a defect in coagulation.[1] This disorder is due to a deficiency in proteins called coagulation factors, specifically the factors VIII (deficiency responsible for Hemophilia A) and IX (deficiency responsible for Hemophilia B) that aid in the staunching of hemorrhages that occur especially in the muscles and joints. Hemorrhages, which are frequent in the joints, may generate significant musculoskeletal problems if they are not treated correctly with the administration of a coagulation or anti-hemophilic factor (AHF).[2]

The incidence of hemophilia A and B is approximately 1:10.000 (A) and 1:40.000 to 1:50.000 (B) male births,[3] with type A being the most common with 80% of the cases. According to the Brazilian Ministry of Health (2011), there are currently about 11,000 hemophiliacs in the country, with approximately three thousand being severe cases.[3] Depending on the quantity of AHF, the hemophilia is classified as severe (< 1% AHF), moderate (1% < AHF < 5%), and light (5% < AHF < 40%). Half of all the patients in the world are classified as having severe hemophilia.[4]

Hemophiliacs do not bleed faster than those without the disease, but they may bleed for a longer period, compromising the structures and tissues of the body.[5] The bleedings can occur due to traumas or can be spontaneous, with spontaneous hemorrhages rarely classified as light.[6]

Hemophilia is not transmissible and it is mostly hereditary, but people without a family history may have hemophilia, due to genetic mutations that afflict 30% of the cases. Although most patients are males, there are rare occurrences in females. For that to occur, a hemophiliac man needs to have a child with a woman who has the hemophilic gene - a combination to be avoided, although rare.[7]

Some people develop what are called TARGET joints, especially in childhood, that are characterized by multiple bleedings in the same joint and which can cause deterioration in the joint cartilages causing hemophilic arthropathy, with the progressive destruction of cartilage in these regions at every new hemarthrosis - or bleedings in the joints.[4] The repetition of various hemorrhagic episodes in the same joint may result in deformities and reduction in the range of motion of the joints.

Physical activities at school as well as resistance exercises and low impact sports are important for improving the musculoskeletal and psychosocial conditioning, preventing future hemorrhages, and increasing self-esteem, social integration, balance, flexibility, and muscle strength.[3] The low level of physical conditioning of hemophiliacs is associated with a greater frequency of joint bleedings in comparison with healthy individuals. To interrupt this vicious circle, physical treatments and sports are recommended.[8]

Many studies corroborate the importance of physical exercises in the treatment of people with hemophilia.[2,9,10] Falk et al.[11] described a high level karate athlete, an Israeli champion, with severe hemophilia A, who suffered bleedings only after trauma, and even so, rarely, without the occurrence of spontaneous hemorrhages.[11]

Herbsleb et al.[12] showed that people with hemophilia have a lower performance in exercising than healthy individuals due to their anatomical limitation - arthropathies and muscle shortenings - and to the fear of bleeding. Regardless of being a hemarthrosis or a hematoma, hemorrhages vary in intensity and in frequency due to various factors such as age, prophylaxis, physical exercises, and phenotype.[12]

The purpose of the present study was to present the results obtained from a patient with severe hemophilia A submitted to a physical conditioning program as a prophylaxis without the administration of an AHF.

## CLINICAL CASE

R.S.C., a male, lawyer, 31 years old, with severe hemophilia A and hemophilic arthropathy in the left elbow and right ankle.

Before starting the program, the patient did not practice any type of exercise and, due to his work, remained seated all day in a chair at the office. He received physiotherapy for two years before starting the current training program.

R.S.C. agreed to participate in this study and signed the free and informed consent form. All the trainings and evaluations were carried out at the Physical Medicine and Rehabilitation Institute at the Hospital das Clínicas of the University of São Paulo School of Medicine, IMREA HC FMUSP - Lucy Montoro Network)/Lapa Unit.

### Clinical evaluation

The participant was clinically examined by a physiatrist from the IMREA HCFMUSP - Rede Lucy Montoro to initiate the Physical Conditioning and was approved to perform the physical exercises.

### Physical evaluation

The patient was submitted to physical evaluation: flexibility of the lower limbs (sit and reach using the Wells bench), sitting rising test (SRT) (sitting and rising from a chair five times, timing the repetitions), body weight, and height (Chart 1).

### Strength test

The strength of the lower and upper limbs was evaluated in machines specific for weight lifting (Chart 2). To prevent an overload on the muscles and joints, the evaluation measured the strength of each segment according to the effort rate perceived by the Borg scale from 11 to 13. The load used for the resistance training was the value obtained in the performance of 10 complete and efficient repetitions, that is, without postural compensation to facilitate the effort necessary to the movement.

### Training

The program lasted for 20 months, with two one-hour weekly sessions, supervised by a physical education teacher. The aerobic training consisted of pedaling for 20 minutes on a horizontal ergometric bicycle (Movement, Perform H3 model); the resistance training lasted 30 minutes and was done on specific weight-lifting machines (Sportin); the session was then concluded with 10 minutes of stretching.

The patient initiated the aerobic training on a horizontal ergometric bicycle at the average speed of 30 rpm. His pulse rate (PR) was recorded at rest, after 10 and 20 minutes of training, and three minutes after the end of the session. The resistance training was applied to 10 muscle groups (pectoralis major, latissimus dorsi, deltoideus, biceps brachii, triceps brachii, upper and lower abdomen, quadriceps, ischiotibialis, and triceps suralis). The patient performed two series with 10 repetitions, with 45 seconds between each series. The intensity was based on the initial

**Chart 1.** Values obtained in the final evaluation

| Test | Initial Values | Final Values |
|---|---|---|
| SRT | 11"94s | 06"10s |
| Weight | 75kg | 83 kg |

**Chart 2.** Final results of the exercises

| Exercise | Repetitions | Initial Load | Final Load |
|---|---|---|---|
| Chest Fly | 10 | 8kg | 14kg |
| Shoulder Adduction with High Pulley | 10 | 20kg | 40kg |
| Shoulder abduction | 10 | 3kg | 7kg |
| Curls | 10 | 10kg | 20kg |
| Triceps Pulley | 10 | 15kg | 20kg |
| Knee Extensions | 10 | 7kg | 25kg |
| Knee Flexion | 10 | 10kg | 25kg |

test, between 11 and 13 on the Borg scale. The training load was increased 20% as the patient reported that it was becoming easier.

## RESULTS

After 20 months, improvements in global (Figure 1 and Chart 2) muscle strength and in the SRT (Chart 1) could be observed. The evaluations were made by the same teacher and with the same measuring equipment to maintain the reliability of the data. The smallest gain in strength was from the triceps brachii with 33%, while the largest gain was for the knee extensors with 257%; the average gain in muscle strength was 121%. As for the SRT functional test, there was improvement of 95.74% and an increase of 10.67% in body weight.

## DISCUSSION

Six months before the physical conditioning program, the patient suffered three hemorrhages: two were spontaneous in the left elbow and ankle and one in the right thigh due to a small-unidentified trauma. The three hemorrhages were treated with an AHF.

Eight months into the program, a hemorrhage occurred and was treated with an AHF: the bleeding occurred in the left elbow caused by trauma during the exercises, after an increase in the load. Twelve months later, the patient had a spontaneous hemorrhage in the same joint.

There were other bleedings that were not treated with an AHF; two spontaneous in the left elbow and three small-unidentified traumas in the right thigh and arm. These last episodes, where no AHF was used, would have been treated as important bleedings two years earlier, making immediate use of an AHF.

This is not necessarily that the quantity of blood factor had increased, but that the strengthening of muscle structures and the development of proprioception probably controlled joint instabilities and the hemarthroses that occurred had been less severe because of that. However, Groen et al.[13] and Koch et al.[14] showed that during aerobic training in patients with light to moderate hemophilia A, there was an increase in the activity of the endogenous Factor VIII 10 minutes and 30 minutes after the end of sessions, consecutively.[13,14]

In hemophilia, self-esteem, anxiety, and body image are important characteristics that improve with exercising, because it benefits posture, tones the muscles, and creates possibilities of performing physical activities without fear of injury.[2,6]

Many times, people have a light bleeding and immediately apply the medication, not knowing how to evaluate the severity of the event, and whether it will be controlled naturally or only with cryotherapy applications (local treatment with ice). With exercising, physical improvements such as increases in muscle strength, proprioception, physical and cardiorespiratory fitness, joint stability, and self-knowledge, making the person with hemophilia perceive when the hemorrhage can be controlled or not - that is, when the coagulation factor will be necessary or when the body will naturally control and reabsorb the bleeding by itself.

It is possible, with exercises and prophylaxis associated with the phenotype, to perform individual treatments and increase the quality of life of people with hemophilia, as it already happens in many countries in which the life expectancy for hemophiliacs is equal to the general population and makes hemophilia the genetic disease with the most success in its treatment.[15]

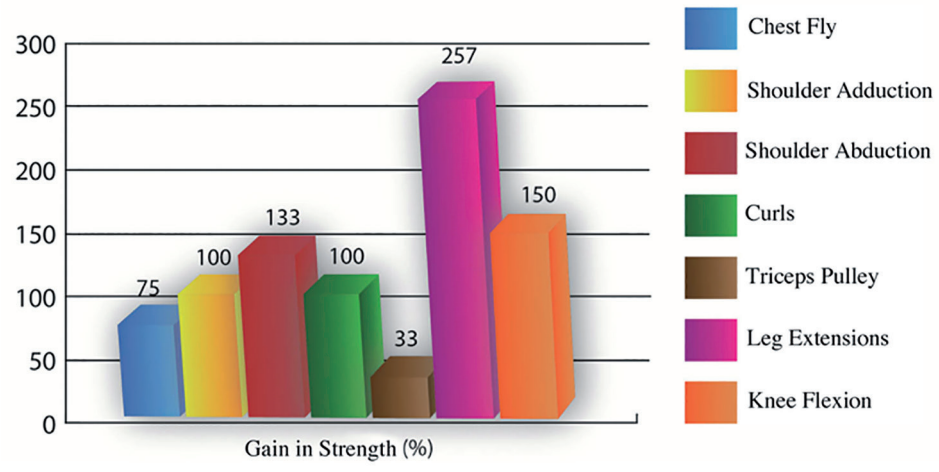


**Figure 1.** Percentage of strength gain at the end of the program

## CONCLUSION

The supervised practice of physical exercises is an important instrument to help the treatment of people with hemophilia and it demonstrates the need for strength and muscle fitness training for this group of people to prevent injuries, avoiding the wear and compromising of the musculoskeletal system. Studies on the same theme with a larger number of individuals are necessary to verify the efficiency of this physical exercise program model.

## REFERENCES

1. Federação Brasileira de Hemofilia. O que é hemofilia? [texto na Internet]. Caxias do Sul: FBH [citado 2013 Jun 14]. Disponível em: http://www.hemofiliabrasil.org.br/hemofilia.php
2. Buzzard BM. Physiotherapy, rehabilitation and sports in countries with limited replacement coagulation factor supply. Haemophilia. 2007;13 Suppl 2:44-6. DOI: http://dx.doi.org/10.1111/j.1365-2516.2007.01506.x
3. Brasil. Ministério da Saúde. Manual de reabilitação na hemofilia. Brasília: Ministério da Saúde; 2011.
4. Broderick CR, Herbert RD, Latimer J, Curtin JA, Selvadurai HC. The effect of an exercise intervention on aerobic fitness, strength and quality of life in children with haemophilia (ACTRN012605000224628). BMC Blood Disord. 2006;6:2. DOI: http://dx.doi.org/10.1186/1471-2326-6-2
5. World Federation of Hemophilia. What is hemophilia? [text on the Internet]. Quebec: WFH; 2012. [cited 2013 Jun 14]. Available from: http://www.wfh.org/en/page.aspx?pid=646
6. Srivastava A, Brewer AK, Mauser-Bunschoten EP, Key NS, Kitchen S, Llinas A, et al. Guidelines for the management of hemophilia. Haemophilia. 2013;19(1):e1-47. DOI: http://dx.doi.org/10.1111/j.1365-2516.2012.02909.x
7. World Federation of Hemophilia. How do you get hemophilia. [text on the Internet]. Quebec: WFH; 2012. Quebec; Available from: http://www.wfh.org/en/page.aspx?pid=644.
8. 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(1):170-8.
9. Czepa D, von Mackensen S, Hilberg T. Haemophilia & Exercise Project (HEP): the impact of 1-year sports therapy programme on physical performance in adult haemophilia patients. Haemophilia. 2013;19(2):194-9. DOI: http://dx.doi.org/10.1111/hae.12031
10. Gomis M, Querol F, Gallach JE, González LM, Aznar JA. Exercise and sport in the treatment of haemophilic patients: a systematic review. Haemophilia. 2009;15(1):43-54. DOI: http://dx.doi.org/10.1111/j.1365-2516.2008.01867.x
11. Falk B, Portal S, Tiktinsky R, Weinstein Y, Constantini N, Martinowitz U. Anaerobic power and muscle strength in young hemophilia patients. Med Sci Sports Exerc. 2000;32(1):52-7. DOI: http://dx.doi.org/10.1097/00005768-200001000-00009
12. Herbsleb M, Hilberg T. Maximal and submaximal endurance performance in adults with severe haemophilia. Haemophilia. 2009;15(1):114-21. DOI: http://dx.doi.org/10.1111/j.1365-2516.2008.01860.x
13. Groen WG, den Uijl IE, van der Net J, Grobbee DE, de Groot PG, Fischer K. Protected by nature? Effects of strenuous physical exercise on FVIII activity in moderate and mild haemophilia A patients: a pilot study. Haemophilia. 2013;19(4):519-23. DOI: http://dx.doi.org/10.1111/hae.12111
14. Koch B, Luban NL, Galioto FM Jr, Rick ME, Goldstein D, Kelleher JF Jr. Changes in coagulation parameters with exercise in patients with classic hemophilia. Am J Hematol. 1984;16(3):227-33. DOI: http://dx.doi.org/10.1002/ajh.2830160304
15. Mannucci PM. Treatment of haemophilia: building on strength in the third millennium. Haemophilia. 2011;17 Suppl 3:1-24.