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Correlation between hemophilic severity and ventilatory function

Shahinaz Abdella[1], Bassant H. Elrefaay[2], Awny F. Rahmy[3] and Ayman A. Sadek[4]

[1]*Department of Physical Therapy, Damanhour Medical National Institute, Damanhour, Egypt.*
[2]*Department of Physical Therapy for Cardiovascular/Respiratory Disorders and Geriatrics, Faculty of Physical Therapy, Cairo University, Cairo, Egypt.*
[3]*Dean of the Faculty of Physical Therapy, Pharos University, Alexandria, Egypt.*
[4]*Director of Department of Tuberculosis, Chest hospital, Damanhour, Egypt.*

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ABSTRACT

Hemophilic patients are inactive persons and they have sedentary life style because of fear of bleeding and decrease strength of skeletal muscle and this include diaphragm and inspiratory muscles. The purpose of this study was to investigate the correlation between severity of hemophilia and ventilatory function. *Materials and Methods:* Thirty male patients with hemophilia type (A). Twenty hemophilic patients with mild severity received their medical treatment and on their normal activities and 10 hemophilic patients with moderate severity received their medical treatment and on their normal activities. The ventilatory function was assessed. *Results*: The Spearman's rank-order correlation between (Severity scale and FVC) revealed that there was no correlation ($p>0.05$). This means that change in the severity scale grades is not consistent with change in FVC. As well as, there was no correlation ($p>0.05$) between (Severity scale and MVV). This means that change in the severity scale grades is not consistent with change in MVV. *Conclusion:* from these findings we can conclude that there are no correlation between (severity and FVC), and between (severity and MVV).

Keywords: Hemophilia , ventilatory function test, MVV, FVC

Introduction

Hemophilia is a congenital disorder resulting from a deficiency of clotting factors. Affected individuals lack factor V111 or 1X procoagulant activity and consequently experience repeated, often spontaneous, hemorrhagic episodes in organs, including the locomotor tract, muscles, and may gradually encounter serious problems in their activities of daily living (Van Meeteren, 2000).

Haemophilia is an inherited bleeding disorder which is characterized by recurrent spontaneous bleeding in joints and muscles. Without treatment most people with haemophilia die in childhood or‘early adult life (WFH, 2003).

Hemophilia A is four times more common than hemophilia B. The incidance of factor VIII deficiency is thought to be approximately 1 per l0 000 birth; for factor IX deficiency, the incidance is approximately l per 30000- 50000 births (Kasper, 2000 and Soucie *et al.,* 2005).

The severity of hemophilia is determined by the level of clotting activity of factor VIII in the blood. There are three levels of severity: mild, moderate, and severe. In mild hemophilia the percentage of factor activity in blood from 5% to40% of normal, in moderate hemophilia froml% to 5% of normal, while in sever hemophilia the percentage of factor activity in blood is less than 1% of normal (Srivastava, 2005).

Haemarthrosis, sinovitis and bruising are the most frequent problems for the haemophilic patient, and are frequently accompanied by marked muscle atrophy around the joint, possibly because of a poor use of this muscle (Tiktinsky *et al.,* 2002).

A fundamental requirement for many of the activities of daily living is the ability to perform predominantly aerobic, oxygen-using, work. Such activities require the integrated efforts of the heart, lung, and circulation to deliver oxygen to the metabolically active muscle mass (Fleg *et al.,* 2000).

Physical activity during childhood and adolescence is essential for health as well as social reasons. Children with hemophilia are less physically active compared with healthy peers. This low

Corresponding Author: Shahinaz Abdella, Department of Physical Therapy, Damanhour Medical National Institute, Damanhour, Egypt.

activity level may influence their physical capacity, muscular ability and bone strength (Falk *et al.,* 2000).

Also, children with hemophilia who are less active than their healthy peers generally have decreased muscle strength and flexibility, and have lower aerobic working capacity (Lane *et al.,* 2004).

Pulmonary function test are valuable tool for respiratory system evaluation, representing an important adjacent to (patient history, various lung imaging studies and invasive testing such as bronchoscope and open lung biopsy). Pulmonary function test is normally repeated at least three times to ensure reproducibility, so, it can only be used only on patients who are able to understand and follow instructions, thus, this test is not suitable for patients who are unconscious, heavily sedated, have limitations that would interfere with vigorous respiratory efforts or patient with recent heart problem (Steckbeck *et al.,* 2002).

The normal range of FVC is approximately 80% or more of predicted. A reduced FVC may be indicative of restrictive lung disease. But the FVC can also be reduced from significant airflow limitation, muscle weakness, or poor effort or technique. Also for asthmatics the bronchioles are constricted during forced expiration and the air trapped in the lung that leads to decreased FVC. A similar problem exists with individuals who smoke or have bronchitis (Berger and Halastala, 2001; Rundell *et al.,* 2001).

The MVV is a test of the overall function of therespiratory system. It is influenced by the status of the respiratory muscles, the compliance of the lung-thorax system, the condition of the ventilatory control mechanisms, and the resistance offered by the airways and tissues. The MVV is typically decreased in subjects with moderate or severe obstructive disease. This may be the result of the increased airway resistance caused by bronchospasm or secretion of mucus (Susan and Cairo, 2004; Ruppel, 2003). MVV may be normal in patients who have restrictive lung disease. Diseases that limit lung or chest wall expansion may not interfere significantly with airflow. Patients who have restrictive lung disease can compensate by performing the MVV maneuver with low tidal ventilation (Vt) and high breathing rates (Merk, 2001).

Unfortunately, there are few studies available that report the pulmonary function test in hemophilic patients. Therefore, the current study is developed to investigate the correlation between severity and FVC, and between severity and MVV.

Subjects, Materials, and Methods

1**-Subjects:**

The study was carried out on thirty men patients suffering from hemophilia A (mild and moderate), the study was conducted from March 2016 to April 2018 at physical department of Damanhour Medical Institute.

2- Inclusion criteria

All the patients:
1- Had hemophilia A (mild or moderate).
2- Had not any deformity that interfere with the procedures.
3- BMI< 29.9
4- The ages of the patients ranged from20-40 years.

3-Exclusion criteria:

1- Patients were suffering from any evidence of internal organs bleeding.
2- Patients with unstable cardiovascular problems.
3- Patients with uncontrolled hypertension.
4- Patients with obstructive lung diseases.
5- Obesity.

Ethical consideration:

The procedure of the study was explained for every participant. All participants signed on consent form before the study procedures. The protocol of the study was approved by ethical commeitte, Faculty of physical therapy, Cairo University.

Instrumentation & Materials:

A - Evaluation equipments:

1-spirometric assessment:
Spirometry was used for ventilatory function test which quantitatively determined the following ventilatory parameters:
-Forced vital capacity (FVC).
-Maximum voluntary ventilation (MVV).
It was carried out in the sitting position using the spirometer.

Methods

A-Evaluation procedures:

Weight and height:
The evaluation procedure had been done for all patients. Body mass index (BMI) was calculated according to the following

$$BMI = Weight\ (kg) / Height^2\ (m^2).$$

Ventilatory function test:
The subject asked to remain sitting on chair and throughout the test with a nose clip that was tightly fitted to the nostrils so no air could escape during the test.

A mouthpiece was inserted into the turbine sensor by at least 0.5cm and then placed at least two centimeters into the subject's mouth.

Subject preparation:
The subject was required to:

1- Not eat a heavy meal just before the test because a full stomach may prevent lungs from fully expanding.
2- Not perform any excess effort for 6 hours before the test.
3- Wear loose clothes that don't restrict breathing in any way.

Before starting the test a thorough explanation of the various command such as (take a deep breath) (blow all air out) (hold your breath) (breath naturally) was done for every subject individually in simple terms and demonstrated to him before doing its steps.

Instrument Preparation:
The following steps were followed by the same therapist:

1- Connect the apparatus to the power and switch it on.
2- Calibaration of unit (only to be carried out with the first test of the day, after every significant temperature changes, or after changing the sensor).
3- Press new key to enter the patient data including age, sex, weight and height.
4- Place the disposable mouth piece in the sensor and press the function test key to select the forced vital capacity or the maximum voluntary ventilation according to the desired test.

FVC maneuver:
The subject was asked to fill his lung as much as possible, the close his lips tightly around disposable mouth piece and then exhale as forcelly and completely as possible and blow all the air out.

Forced vital capacity (FVC) was obtained and recorded by the apparatus from such maneuver. Forced vital capacity (FVC) is the maximum amount of air that can be forcefully and rapidly exhaled after a deep breath (maximal inspiration) and measures approximately 5 liters for an adult male (Jones *et al.,* 2003).

MVV maneuver:

It is the maximum air, which can be expired in a minute by deepest and fastest breathing. Normally equals 60:120 L/min for females and 80:160 L/min for males. It can never be normal in presence of lung disease. It requires the subject to breathe as maximally and rapidly as possible for 15 seconds (Chupp, 2001; Hughes & Pride, 2002).

The subject was asked to put a disposable mouth piece in his mouth tightly **,** inhale and exhale fully as completely ,as forced as possible for 12 seconds.

- Each maneuver was repeated for three successive times and the greatest reading was obtained and recorded.
- All the previous measures were recorded and stored then repeated again at the end of the study period.

Statistical analysis

Statistical analysis was conducted using SPSS for windows, version 23 (SPSS, Inc., Chicago, IL). The Spearman product-moment correlation was used to determine the strength and direction of a linear relationship between Severity scale, FVC and MVV. The alpha level was set at 0.05.

Results

The Spearman's rank-order correlation between (Severity scale and FVC) revealed that there was no correlation ($p>0.05$). This means that change in the severity scale grades is not consistent with change in FVC. As well as, there was no correlation ($p>0.05$) between (Severity scale and MVV). This means that change in the severity scale grades is not consistent with change in MVV. (table 1 and fig1and 2).

Table 1: Correlations between Severity scale, FVC and MVV.

		FVC	MVV
Severity scale	**Correlation Coefficient Spearman's) (ρ)**	0.008	0.157
	p-value	0.996	0.407

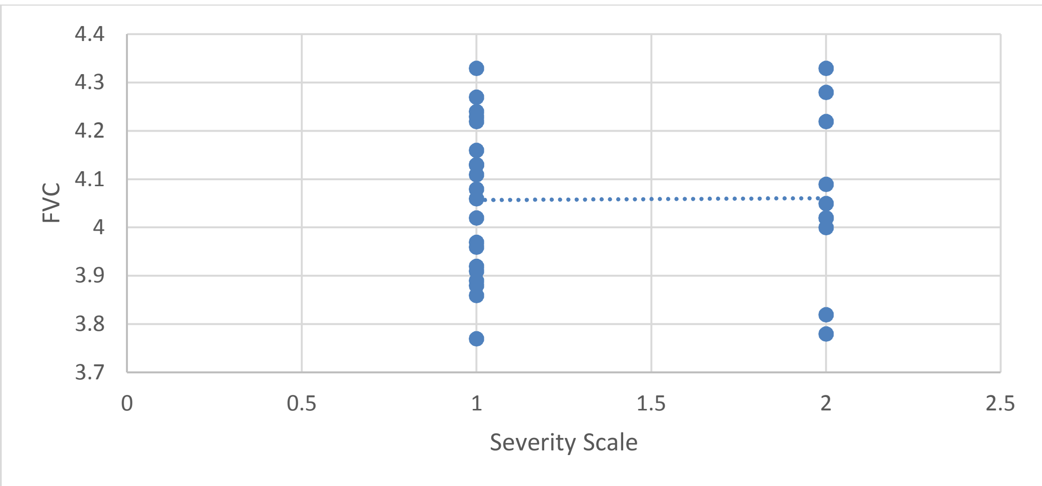

Fig. 1: Scatter plot for the bivariate correlation between severity scale and FVC.

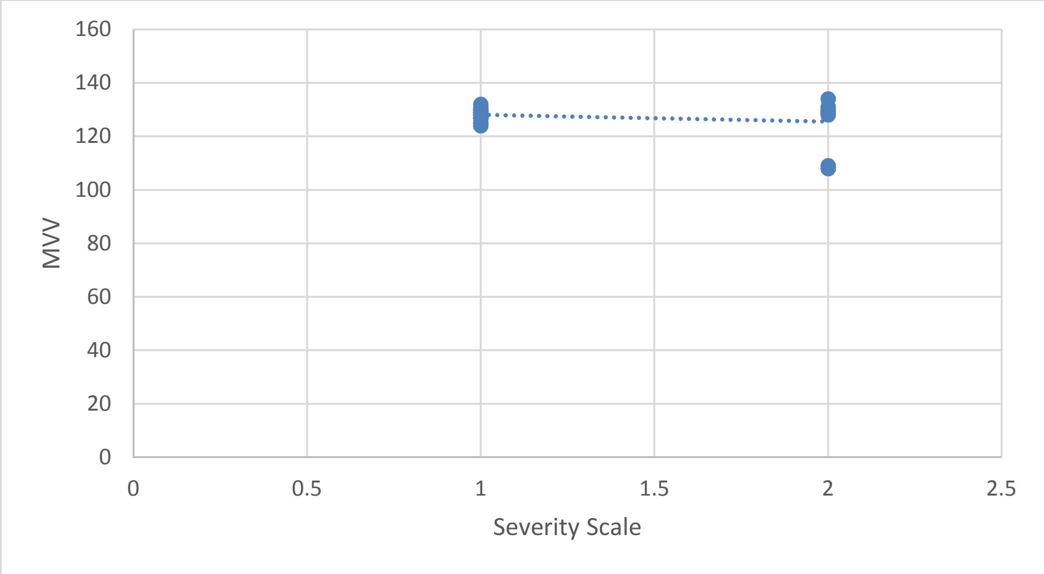

Fig. 2: Scatter plot for the bivariate correlation between severity scale and MVV.

Discussion

People with haemophilia still have decreased fitness and strength compared with age-matched individuals without haemophilia (Falk *et al., 2005).*

Physical fitness is a dynamic physical state comprising cardiovascular /pulmonary endurance; muscle strength, power, endurance, and flexibility; relaxation; and body composition that allows optimal and efficient performance of daily and leisure activities (APTA, 2009).

The inspiratory muscles, including the diaphragm, are morphologically and functionally skeletal muscles and therefore should respond to training in the same way as would any locomotor muscle if an appropriate physiological load is applied (Kraemer *et al.,* 2002).

A comparison of assessed movement behaviour of hemophilic adults on prophylactic treatment with healthy adults was done by Timmer *et al.* (2018) and he found that In absolute hours, adults with severe haemophilia spent more time sitting and standing and were less physically active (walking and running) than healthy adults.

The pulmonary capacity of the individual is related to the body structure of the people as well as the oxygen need of the sports branch performed. The respiratory systems of the people who have trained adequately adapt to the increasing oxygen need during the exercise rapidly (Taşgın and Dönmez, 2009).

The current study revealed no correlation between severity of hemophilia and ventilatory function. This means that the 2 grades of hemophilia don't affect ventilatory function. The current study has some limitations a larger sample size is warranted. The inclusion of sever grade is also recommended in future researches.

Conclusion:

We can conclude that there are no correlation between (severity and FVC) ,and between (severity and MVV).

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Nil.

Conflicts of interest:

There are no conflicts of interest.

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