LUMEN ET VIRTUS

Study of the influence of regular practice of water aerobics on joint flexibility of a Fibrodysplasia Ossificans Progressiva (FOP) patient: A single case study

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Rômulo Carlos de Aguiar[1]

ABSTRACT

Fibrodysplasia Ossificans Progressiva (FOP), with 700 cases worldwide, is a chromosomal genetic disorder (2q23-24), caused by a spontaneous mutation in the ACVR1 gene, transforming endothelial cells into bone, restricting the movements and vascularization of the muscles affected by bone neoformation. As there are no specific therapies, the objective of this study was to identify the effects of a water aerobics exercise program on some indicators of body functionality and structure, with the aim of providing a better quality of life to patients. The research was an intrinsic, descriptive, quantitative, investigation-action case study with a volunteer with FOP. A multidisciplinary group was organized to characterize the physiological and anthropometric conditions and to develop a program of water aerobics exercises. From January to July 2014, a total of 50 hydrogymnastics sessions of 50 minutes each were carried out for 3 days a week to verify the effects on anthropometric variables and joint flexibility. Considering the evaluation of the effects of the water aerobics exercise program on flexibility, the results revealed that the amplitudes of the right shoulder, left shoulder, left wrist, left knee, right ankle and left ankle joints did not change; The right elbow, left elbow, right wrist, and right knee joints increased the flexion angle. Regarding the range of joints, it was possible to observe gains in the range of joints not fully calcified by the disease, which suggests that some joint mobility exercises may be beneficial in patients with FOP. The study is unique, without a control group and, being a case study of a volunteer with a disease of faster progression at her age, it makes it difficult to interpret the results. The results obtained are unexpected and opposite to those often obtained with healthy individuals or with other pathologies. Thus, this study suggests that this program of physical exercises of water aerobics in patients with FOP may be indicated, and further studies are needed to confirm these results or to identify the most recommended type of exercise for these patients.

Keywords: Fibrodysplasia Ossificans Progressiva (FOP), Aerobics, Goniometry, Joint, Flexibility.

INTRODUCTION

In addition to pathologies, the population also faces the consequences of the natural aging process that often decreases quality of life, reducing the ability to independently perform activities of daily living and instrumental activities of daily living. Some pathologies can restrict or totally

[1] Postdoctoral
Vale do Acaraú State University – UVA
Inta University Center – Uninta

compromise this ability and the adoption of methods that involve strengthening sedentary individuals is an indispensable condition for their life, health and general well-being. This process, by itself, already leads to a decline in several functions, especially with regard to movement and ambulation. If this is associated with the involvement of some pathology considered a rare disease, whose specific drugs and treatments practically do not exist, the situation becomes more critical.

Fibrodysplasia Ossificans Progressiva (PFO), RD caused by gene mutation, mainly compromises the locomotor system, with excessive formation of progressive heterotopic ossification, conversion of soft tissues (muscles, tendons, ligaments, aponeuroses and other connective tissues) into bone tissue and immobilization of the joints, manifesting itself throughout the body, being characteristically more common in the tibiae, femurs and cervical spine, being disabling to its sufferers (Gosai et al., 2013).

FOP shortens the life expectancy of its patients to 45 (forty-five) years and one of the most common causes of death is head trauma due to falls, due to the limitation for ambulation (Kaplan et al., 2005).

Figure 1: Symmetrical malformations of the great toes with hallux valgus of the feet and heterotopic ossifications in the right hand of the volunteer.



The syndrome had several designations until McKusick (1972), in 1970, began to call it fibrodysplasia ossificans progressiva, which indicates the progressive transformation of soft tissues of the body into bones and affects the muscles secondarily. Due to the stony appearance of the carriers, caused by the immobility of the joints, characteristic of the advanced form of the disease, it is also called *Stone Man Syndrome*. The PFO phenotype includes, as a defining feature, congenital malformation of the halluxes (see Figure 1) and progressive heterotopic calcifications with certain anatomical patterns.

According to Kaplan and Kantanie (2004), joint immobilizations occur following the development of the PFO that occurs from the axial skeleton to the appendicular skeleton, affecting the cervical region, the spine and the shoulder girdle anterior to the elbows, pelvic region and knees. Also, according to the authors, it also occurs in the craniocaudal direction, affecting the shoulder

girdle and elbows anterior to the pelvic region and knees, and from the proximal area to the distal area, affecting the shoulder girdle anterior to the elbows and hands, and the pelvic region anterior to the knees and feet, with progression of calcifications in childhood, Initially, in the cervical region, in the shoulder girdle and in the thorax, and later the pelvic region and knees are affected, in adolescence or early adulthood. According to Delai et al. (2004), this progression can be spontaneous or be accelerated by traumas such as falls from one's own body.

Connor (2004) states that physical therapy is not recommended for FOP, since passive hyperextension of soft tissues of a compromised joint can result in pain. Active movement should be considered with caution and with comfort for the patient, to avoid new outbreaks.

As FOP presents symptoms such as joint involvement, it can be assumed that they may undergo changes to the point of providing a better quality of life to patients, if the regular practice of physical exercises is combined with pharmacological and therapeutic treatments. However, initially, the affected joints and the conditions of these patients for physical exercise should be identified, which makes each case a particular study, since FOP causes partial or total physical disability.

In this study, a program of regular practice of water aerobics sessions in a pool with heated water was developed and applied in a woman with FOP, when it was intended to verify whether it is capable of inducing some type of joint functional improvement.

Thus, in general, the objective was to identify the effects of a physical exercise program consisting of hydrogymnastics sessions on the flexibility of some joints not yet fully affected by the pathology.

Specifically, for this, the objective was to characterize the effects of the hydrogymnastics exercise program on the conditions of movement, ambulation and amplitude related to the joint flexibility of a PFO patient.

METHODOLOGY

CHARACTERIZATION OF THE PARTICIPANT

The participant in this study is Caucasian, born at term without intercurrences, in 1979. His early childhood apparently became normal when in 1985, at the age of six (6), during the application of an intramuscular smallpox vaccine, with a technique using an automatic syringe injector gun, the pathology manifested itself. After a few days, fever, ear pain, scalp lesions, as well as reddened and swollen areas in various parts of the body appeared. After regression of the edema, the first calcifications were formed. There was also a progressive atrophy and a significant loss of mobility in the left arm, where the bulge had previously manifested. Over time, the atrophy of the arm worsened, causing a tilt of the cervical region to the left side.

Figure 2: Volunteer with Fibrodysplasia Ossificans Progressiva. Posterior, anterior and left lateral views showing heterotopic ossifications in the spine, shoulder girdle, thorax, pelvis, upper limbs and lower limbs.



In 1995, at the age of 16 (sixteen), the pathology began to affect his spine and, later, his lower limbs (LL). The joints gradually became immobilized and the lower limbs stiffened and no longer flexed. The limitations worsened and difficulties in sitting were manifested. The tendons of the feet atrophied and she began to need the use of sandals with unevenness of 12.5 cm on both sides to correct her posture, since her feet were in constant hyperextension. In 1996, at the age of 17 (seventeen), she no longer raised her upper limbs (MMSS), tendons in her feet atrophied even more, always remaining on the tips of her feet, in addition to having her spine calcified, preventing her from flexing her trunk and sitting. At the beginning of May 2011, his pathology was diagnosed as Fibrofisplasia Ossificans Progressiva – FOP, irreversible and, until then, without therapies or prognosis of cure (see Figure 2).

PROCEDURES

In an internet search, looking for rare diseases, this carrier was found. A study on the pathology was then carried out, before a previous contact with the patient, a face-to-face meeting and the approach to her interest and availability to participate in an innovative and unprecedented research for her pathology, with interest on her part.

The research project was submitted to the Human Research Ethics Committee of the Vale do Acaraú State University – UVA in Sobral-Ceará-Brazil, in accordance with Resolution No. 466, of December 12, 2012 of the National Health Council, obtaining an approval opinion. A new contact was made with the patient to clarify the intention and objectives of the research, to resolve any doubts and answer any questions, followed by the signing of the Informed Consent Form (ICF) for participation in the research by the now volunteer, after all clarifications. Next, a multidisciplinary group was organized to monitor the participant, consisting of a physical education professional, a physical therapist, a cardiologist, an occupational therapist, and a pulmonologist.

JOINT FLEXIBILITY ASSESSMENT

Flexibility consists of the ability of the joint to reach its maximum points of flexion and extension, adduction and abduction, according to the joint, having been measured by active goniometry in January 2014 and July 2014. A WCS stainless steel mechanical goniometer was used, with a reading range of 0º-360º, reading resolution of 1º (one degree), with degree checks of the range of motion in the right shoulder (RE), left shoulder (LE), right elbow (CD), left elbow (CE), right hand (MD), left hand (ME), right knee (JD), left knee (JE), right foot (DP) and left foot (PE), and the volunteer was asked only for the maximum flexion movement of these joints, performing 03 measurements and considering the highest result. In the measurements, the right hip joint (DFC) and the left hip joint (LVC) were disregarded, because the volunteer already had these joints totally calcified by the heterotopic ossification characteristic of the pathology. The starting point or 0º (zero degree) in the volunteer was considered the relaxation position with comfort of the measured body segments.

WATER AEROBICS PROGRAM

The water aerobics physical exercise program was implemented with the participation of a physical therapist and developed with the monitoring of a Physical Education professional, whose number of sessions and duration of each session was determined after analysis of the results of the initial anamnesis, in addition to the administration of the overload principle that depended on the evolution/involution of the volunteer's conditions.

The program was composed as follows:

- ✓ Program period: February 10, 2014 to August 27, 2014.
- ✓ Weekdays: Mondays, Wednesdays and Fridays.
- ✓ Total sessions: 50 (fifty) sessions.
- ✓ Duration of each session: 60 (sixty) minutes, on average.
- ✓ Division of the session:
  - 10 (ten) minutes for warm-up with displacements;
  - 30 (thirty) minutes for localized exercises:
    - (a) dynamic exercises for upper limbs: shoulder adduction and abduction, elbow flexion and extension, shoulder adduction and abduction associated with elbow flexion and extension;
    - (b) dynamic exercises for lower limbs: hip adduction and abduction, knee flexion and extension, stationary gait;
    - (c) breathing exercises;
  - 10 (ten) minutes to return to calm.

- ✓ Number of sets per session: 02 (two) sets;
- ✓ Number of repetitions/interval per set: from 05 (five) to 20 (twenty) for each exercise, with 01 (one) minute interval between each set.
- ✓ Accessories used: neoprene suit, ear protectors, cap, cervical vest, boards, floats, long poles (spaghetti), ethyl vinyl acetate shin guards and cannulas of varying lengths.

In the dynamic exercises, in the supine position, movements of the right arm were performed, consisting of abduction and adduction of the RE, flexion and extension of the CD, and flexion and extension of the MD with the arm submerged, and flexion and extension of the fingers of the MD, also with the arm submerged; Movements with the left upper limb were not possible due to the immobility caused by the pathology (see Figure 3). With the lower limbs, without the anklets, abduction and adduction of the legs, circular movements with both legs simulating pedaling in the water, flexions and extensions of the JD and JE, as well as kicks with both legs in extension (see Figure 3); All movements lasted approximately 01 (one) minute.

Figure 3: Volunteer performing movements with her right arm and lower limbs.

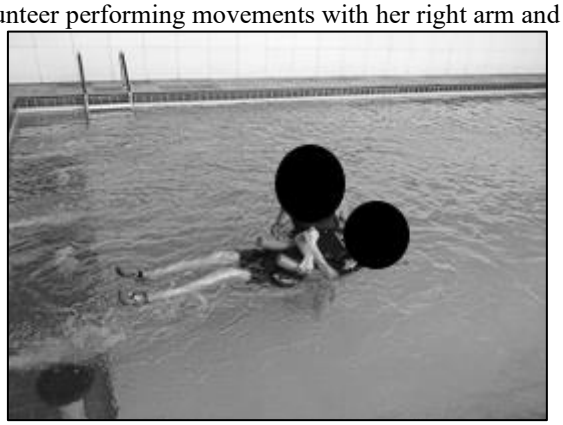
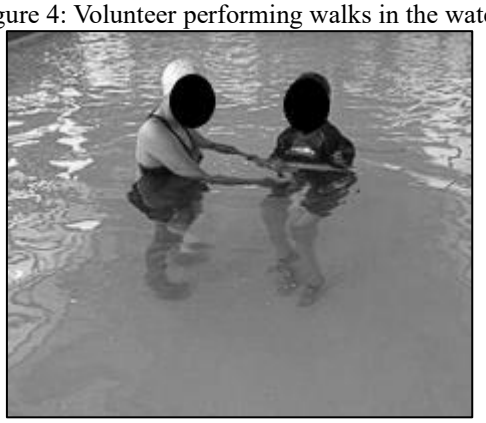


Figure 4: Volunteer performing walks in the water.

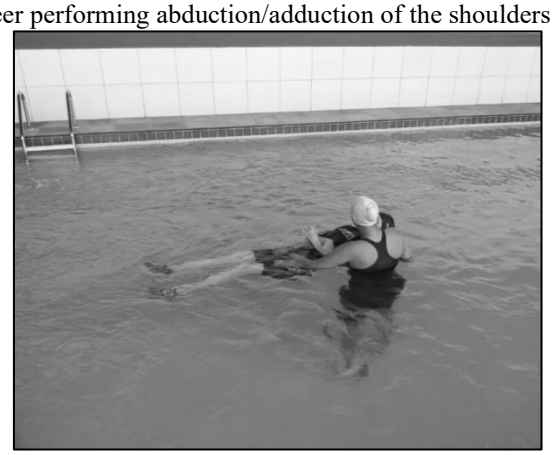


In the bipedal position, with the body immersed up to the height of the chest, walks were made in different directions (front, side and back), performing 04 (four) laps in the pool, always

breathing during the walk. After each one-month period, the volunteer was asked to walk with gradually longer steps (see Figure 4).

In the dorsal position, the patient performed abduction/adduction of shoulders and legs, simultaneously, in combined movements (see Figure 5).

Figure 5: Volunteer performing abduction/adduction of the shoulders and lower limbs.



RESULTS

Considering the evaluation of the effects of the water aerobics exercise program on flexibility, the results revealed that the joint amplitudes of the RE, EE, PE, JE, TT and ET did not change; the CV, CE, PD, and JD joints increased the flexion angle (see Table 2).

With the measurements, in January 2014, it was found that in 07 (five) joints this volunteer already presented total immobility as a result of the pathology: OD, OE, ME, CFD, CFE, PD and PE. The flexibility of the EC, JD and JE joints was already compromised due to the pathology, but the flexibility of the DC and MD joints was still quite preserved. In July 2014, goniometry showed that flexibility in the DC, CE, PD and JD joints increased in relation to January 2014. The flexibility of the other joints remained unchanged, as shown in Table 1.

Table 1: Comparative pre-test/post-test results of goniometry in January 2014 and July 2014.

Joint	January/2014	July/2014	Difference Jan.14/Jul.14
CD	1400	1500	↑7.1%
EC	400	550	↑37.5%
MD	950	1100	↑15.8%
JD	300	400	↑33.3%
JE	250	250	=

Legend: CD (right elbow); FB (left elbow); MD (right hand); JD (right knee); JE (left knee).

DISCUSSION

In the present study, it was possible to verify that a water aerobics program that includes joint mobility exercises, applied to a PFO patient, was able to induce an increase in the flexion angle of all joints that were not yet calcified.

Although, according to the classic study by Silver (1923), there are standard values for maximal joint flexions (see Table 2), considering the anatomical position of the individual as the starting point or 0º (zero degree), in this study, the starting point or 0º (zero degree) in the volunteer was established as the position of relaxation with comfort, since the heterotopic calcifications caused by FOP, in many of the joints, it prevented him from positioning himself in the anatomical position according to the specific literature.

Table 2: Standard values for elbow, hand and knee joint flexions evaluated in the volunteer, in degrees.

Joint	Default Value for Maximum Flexion (*)
Elbow	1400
Hand	700
Knee	1300

Adapted from Silver (1923).

Regarding flexibility, mobility and ambulation, there is impairment in all these variables, since total immobility was manifested in 07 (seven) joints and impairment of flexibility in another 03 (three), due to the calcifications caused by the pathology, with only 02 (two) joints still fully preserved. However, comparing pre-test goniometry with post-test, changes were found in the DC, CE, MD and JD joints, denoting an alteration in the amplitude of these joints that can be attributed to the practice of water aerobics in the EC and JD joints. These results were also obtained by Zambon et al. (2015) in a study with elderly women who practice water aerobics, and the authors found that flexibility in hip flexion and extension movements in elderly women who practice water aerobics and combined training increased, suggesting that these activities can provide improved hip mobility, at the level of the sagittal plane, thus being able to be applied to other joints.

CONCLUSION

Regarding flexibility, in the joints with total impairment, due to immobility due to calcifications (OD, OE, CFD, CFE, TD, TE and PE), there were no changes, but it was concluded that in other joints DC, CE, PD and JD there were increases in their amplitudes, suggesting the fact that they were regularly requested for 06 (six) months during the movements triggered in the hydrogymnastics sessions, caused changes in the volunteer's movement. Therefore, the regular practice of water aerobics or other physical activity may enable a gain in the joint range of some joints already partially compromised by immobilization, but caution is necessary, as it may also cause osteoarticular injuries.

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