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Anthrophophysiological analysis in a patient with Progressive Ossificant Fibrodysplasia: Casuistic

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ABSTRACT

The objective was to measure anthropophysiological variables in patients with FOP, comparing them to standards. For this, ECG / ECC, tomography, pulmonary functions, manovacuometry, bioimpedance, blood analysis and goniometry were performed. The resultsAA = 27 mm; LA = 26 mm; LV = 41 mm; RV = 30 mm; FSD-LV = 20 mm; septum = 5 mm; LV-BWLV = 5 mm; LA/AA = 0.96, FE = 83%; septum/BWLV = 1.00, FDV = 74 ml; SV = 61 ml; volume/mass = 1.03 ml / g; Normalized GSF-LV; under bronchodilator, FVC, FEV1 and PEF = 54%, 64% and 81% of the predictors; FEV1% and 2575 = 118% and 116% of the predictors; MEP = 65 mmH (↓42.7%) and MIP = 55 mmHg (↓63.2%); BM = 39.0 kg; h = 1.57 m; BMI = 15.8 k/m2; AC = 64 cm; % FM = 26%; MM = 27.39 kg; BMC = 1.53 kg; FLM = 28.92 kg; RIMM = 4.5 kg / m2; OI = 63.50%; normal blood glucose, triglycerides and CRP; RS, LS, LH, RA and LA = 0º, LK = 25º, RK = 40º, LE = 55º, RH = 110º, RE = 150º. The cardiovascular differences did not cause any dysfunction. FOP caused moderate restrictive respiratory disorder, Pimax / Pmax below the predictors, MC, h, BMI, CA, bone mineral content, low MLG and IO, immobility in 05 joints and impairment in 03, with 02 preserved. It was concluded that FOP is significantly compromising the respiratory system, with moderate restrictive respiratory disorder caused by the sharp convexity thoracic scoliosis directed to the left, due to the compromised position of bipedation due to the new calcifications, in addition to compromised movement and walking.

Keywords: Physiological calcification, Body composition, Physical exercise, Heterotopic osteogenesis, Cardiovascular system, Respiratory system.

1 INTRODUCTION

Characterized as a rare genetic disorder, Fibrodysplasia Ossificans Progressiva (FOP) had its first historical record, in 1692, by the physician Guy Patin (HAIR, PEEPER, 2005); The first documented case occurred in 1740, when a London doctor described a teenager with large bone swellings in a letter to the Royal University of Physicians (LAMBERT, 2013). Most cases are caused by spontaneous mutation in gametes in the ACVR1 gene, responsible for the disease. ACVR1 encodes the *activin type-1 receptor,* BMP (bone morphogenetic protein*) receptor type-1* and mutation changes codon 206 from arginine to histadine, causing transformations of endothelial cells into mesenchymal stem cells and bone (KAPLAN, et al. 2005; HERRERA-ESPARZA, et al., 2013). The syndrome designated, in 1868, as myositis ossificans progressiva changed to fibrositis and, in 1972, to fibrodysplasia (KAPLAN, 2005).

The carriers appear normal at birth, except for monophalangism and brachydactyly in the halluces (HASAN, 2012). Subsequently, progressive heterotopic osteogenesis is manifested.Some congenital anomalies help in the diagnosis, but are not constantly observed in their entirety, given the description of a patient without characteristic congenital bone anomalies (HASAN, 2012).

According to *The International FOP Association – IFOPA* (2014), the prevalence in this rare condition is 1/2.000,000 people worldwide, totaling 800 cases (IFOPA, 2014). There are no differences related to gender, race, ethnic origin or geographic location (KAPLAN, et al., 2005). In Brazil, there are 75 confirmed cases (FOP BRASIL, 2014).

Characterized by disseminated ossification in soft tissues and congenital stigmata in the extremities, it is particularly disabling in children, presenting two fundamental characteristics, progressive heterotopic osteogenesis and congenital abnormalities of the halluces (GOSAI, et al., 2013). Other features include proximal and medial tibial osteochondromas, orthotopic fusions of the posterior elements of the cervical spine, short and wide femur neck, conductive hearing loss . The molecular mechanism of this disease is sought through the study of the FOP phenotype that maintains that the primary molecular pathology directly involves BMP signaling pathway or its interaction pathway (KAPLAN, et al., 2005).

The diagnosis of classic PFO can be made based on clinical evaluation, associated with large malformations of the feet with rapid appearance of soft tissue lesions. The clinical diagnosis can be confirmed by analysis of the DNA sequence of the ACVR1 gene, to evaluate suspected cases of atypical or variant FOP (KAPLAN, et al., 2012).

FOP has a reduced incidence in humans and much needs to be clarified about its causes and implications, especially for human physiology, since the existing literature focuses on the study of genetic causes and the molecular biology of the cells involved, those linked to mutations of the ACVR1 gene, with no cure or effective treatment (GARCIA-PINZAS, et al., 2013). It is possible to limit the development of new calcifications with Prednisone at a dose of 2mg/kg/day for 4 days in the first 24 hours of the outbreak, reducing inflammation and tissue edema seen in the early stages of ossification; when Prednisone is stopped, nonsteroidal anti-inflammatory drug (NSAID) or cyclooxygenase inhibitor in the COX-2 isoform can be used for symptomatic treatment of relapse and pain, and high doses of corticosteroids and NSAIDs, available at the primary levels of health care, can be used to mitigate the pain caused by the resurgences of the disease, improving the quality of life of patients (GARCIA-PINZAS, et al., 2013).

Chronic neurological symptoms such as prevalence of neuropathic pain, severe recurrent headaches, positive myoclonus and other sensory abnormalities have been reported, speculating that these symptoms are related to the effects of dysregulation of BMP signaling in the Central and/or Peripheral Nervous System (KITTERMAN, et al., 2012).

Patients develop thoracic insufficiency syndrome (TIS) due to costovertebral malformations with ankylosis of the costovertebral joints, ossification of the intercostal and paravertebral muscles and aponeuroses, and progressive spinal deformity, including kyphoscoliosis or thoracic lordosis. Pneumonia and congestive heart failure on the right side are the main dangers that threaten the life ofcarriers (KAPLAN, et al., 2005).

FOP causes shortening of life expectancy in decades, with death occurring on average at 45 years, although cases are reported reaching the age of 70 years exceptionally (KAPLAN, et al., 2005); at least 90% of patients die of cardiac or respiratory insufficiency, or head trauma due to falls.

After a vast review in the databases *PubMed,* Web of Science*, SciElo,* Scopus *and others, a detailed anamnesis of some anthropometric and physiological aspects of a volunteer with PFO, not mentioned in the literature, this study presents variables investigated to compare to their standard values and verify if they were influenced by the pathology.*

The objective of this study was to identify anthropometric and physiological variables of a woman with PFO and compare them to their standard values. For this, the volunteer was submitted to evaluation and verification:

a. cardiovascular, through electrocardiogram (ECG) and echocardiogram (ECC);
b. lung volumes and capacities, through spirometry;
c. of maximal respiratory pressures of inspiration and expiration, through manovacuometry;
d. glycemia, triglycerides and C-reactive protein, through blood analysis;
e. of anthropometric variables, through goniometry and bioimpedance.

2 METHODOLOGY

This is an intrinsic, unique, descriptive and holistic case study of a quantitative nature (STAKE, 2009). "Case studies [...] they are generalizable to theoretical propositions and not to populations or universes. In this sense, the case study [...] does not represent a 'sample' and, in making a case study, its objective will be to generalize theories (analytical generalization) and not to enumerate frequencies (statistical generalization)" (YIN, 2003a)).

An intrinsic case study is one in which, having previously selected the case, the researcher seeks to better understand it specifically in particular (STAKE, 2009).

It is considered as unique, since it is not intended to study it to learn about other cases, if it intends to learn about this particular case, and holistic because it envisions the voluntary as an indivisible whole that cannot be explained by its distinct anthropometric and physiological components.

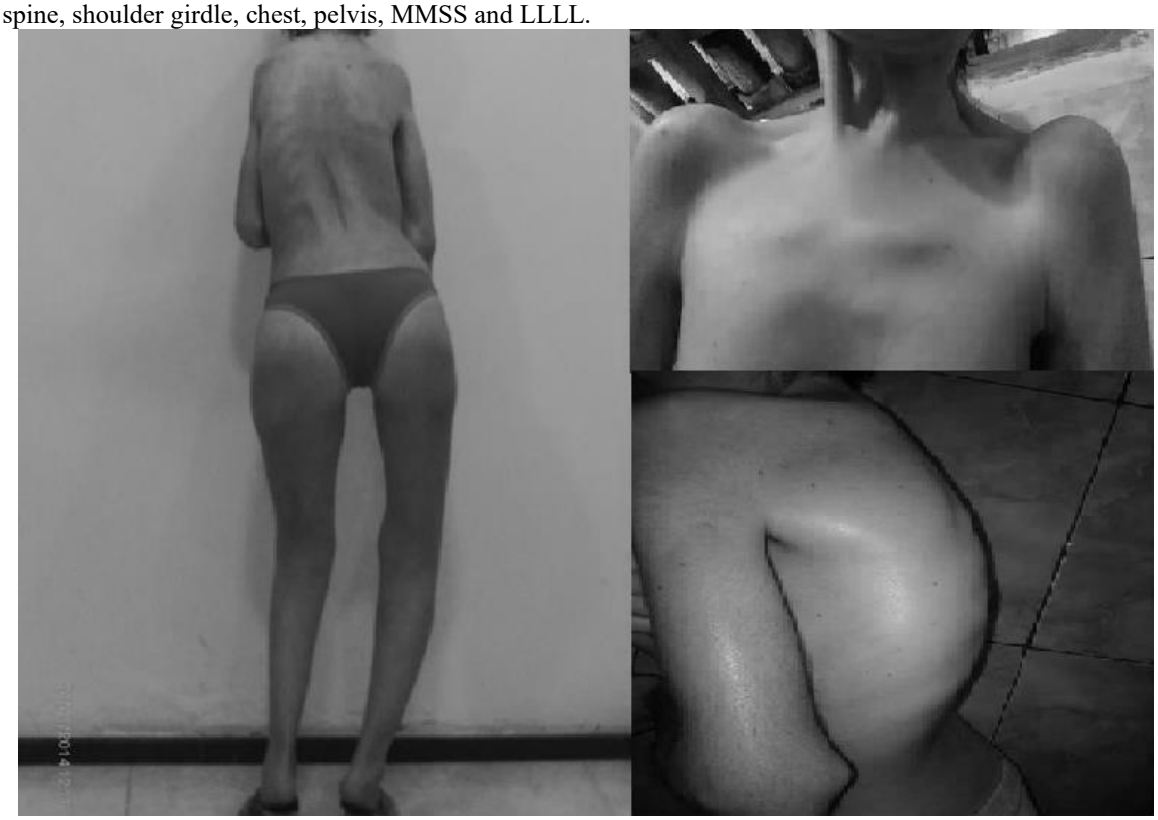
2.1 ETHICAL ASPECTS

The study was approved by the Human Research Ethics Committee of the Universidade Estadual Vale do Acaraú – UVA, Sobral-Ceará-Brazil, according to Resolution nº. 466, of 12/12/2012 of the National Health Council.

2.2 STUDY PARTICIPANT

The volunteer studied was 35 years old at the time of the study, diagnosed with PFO on 05/05/2011 (Figure 1). The pathology manifested itself in 1985, at the age of 6, with the application of smallpox vaccine. One week after vaccination, the site was shown to be fibrous, followed by fever, ear pain and reddened edema throughout the body. After regression of the edemas, the first calcifications were formed. In new outbreaks, the most common symptoms are a feeling of immobilization, pain, burning, edema, tendon shrinkage and new calcifications.

Figure 1: MLB volunteer, with FOP, 35 years old. Posterior, anterior and left lateral views showing heterotopic ossifications in the spine, shoulder girdle, chest, pelvis, MMSS and LLLL.



2.3 METHODS

- Characterization of cardiovascular conditions: ECG at rest, with a volunteer in decubitus, with records of blood pressure (BP) and heart rate (HR); hyperventilation test at rest, in decubitus, with ECG records, BP and HR measurements at the end, according to II Guidelines in Cardiogeriatrics of the Brazilian Society of Cardiology (GRAVINA, 2014).

- Helical evaluation of the chest: helical technique with 5.0 mm and 7.5 mm slices, with a volunteer in dorsal decubitus, immobile, through computed tomography, without intravenous administration of iodinated contrast.
- Pulmonary function tests (lung volumes and capacities): with volunteer comfortably in bipedal, head in neutral position, without neck flexions, with nasoclip, inspiration up to Total Lung Capacity (TLC), with post-inspiratory pause <3 sec., with tube placed immediately after inspiration on the tongue, between the teeth, with the lips clenched, performing maximal and sustained expiration (plateau 1 sec. in the VT curve) and maximum inspiration. For flow-volume curves, he performed a Forced Vital Capacity (FVC) maneuver, inhaling up to the TLC and exhaling as quickly as possible to the Residual Volume (RV), subsequently, inhaling as quickly as possible from the RV to the TLC, meeting all the requirements of the *American Thoracic Society (ATS)* (AURORA, et al., 2007). Results were recorded in liters (L) or liters per second (L/s). Forced *Vital Capacity (*FVC), Forced Expiratory Volume in the first second (FEV1), FEV1/VC*100 *(FEV1%), Peak Expiratory Flow (*PEF), mean flow between the points FEF*25 and FEF*75 ([2575]) *and* Forced Expiratory Time *(FET)* were measured.

[*]*FEF=Forced Expiratory Flow.*

- Evaluations of maximal respiratory pressures (MIP and MEP): with a volunteer comfortably in bipedal, with nose occluded by nasoclip, performed maneuvers of maximum inspiration after a normal expiration and maintained for at least 2 seconds, and maneuvers of maximum expiration performed until the RV after a normal inspiration and maintained for at least 2 seconds.
- Determination of height (h) (CZAJKA-NARINS, 2002), body mass (BM), percentage of fat mass (%FM) and category, lean mass (LM), waist circumference (WC) and category, and Relative Index of Muscle Mass (MMRI): bioimpedance was used with a volunteer in decubitus, immobile, fasting 3-4 hours before the evaluation, without physical activities for 4 hours before the evaluation and without strenuous physical activities in the 24 hours prior to the evaluation evaluation, did not drink alcohol in the 48 hours prior to the evaluation, did not take diuretic medication in the 7 days prior to the evaluation and urinated 30 minutes before the evaluation (GORDON, et al., 1988; CALLAWAY, et al., 1991); in addition to the Body Mass Index (BMI) and category (CONDE, MONTEIRO, 2006), with reference values for BMI proposed by the *Royal College of Physicians*, London, 1983, according to the equation below.

Equation 1 - Body Mass Index (BMI):

$$BMI = \frac{BM\ (in\ kg)}{H2\ (in\ m)}$$

- Determination of the Obesity Index (OI) and category: specific formulas, tables (WAITZBERG, 2001; DAMASO, 2001) and Anthropometric Measures by Gender, According to Age Classes and Brazilian Regions (IBGE, 2014).

Equation 2 - Obesity Index (IO):

$$IOA = \frac{M.C.\ (kg)}{(height \times 100)\ (m)}$$

$$IOB = \frac{mean\ M.C.\ (kg)\ according\ to\ gender,\ age,\ Brazilian\ region\ of\ origin}{mean\ height\ (cm)}$$

$$IO = \frac{IOA}{IOB} \times 100$$

- Glycemia, triglycerides and C-reactive protein: a few milliliters of blood were collected from the volunteer for analysis.
- Goniometry: with a volunteer in bipedality, amplitudes of movements were measured in the right shoulder (RE), left shoulder (LE), right elbow (DC), left elbow (CE), right wrist (PD), left wrist (PE), right knee (JD), left knee (JE), right ankle (TD) and left ankle (ET) joints, using goniometer with measurements in degrees, with volunteer performing maximum flexion movements of these joints.

3 RESULTS AND DISCUSSION

The volunteer presented structural parameters of the aorta (AA) (27 mm), left atrium (LA) (26 mm) and left ventricular (LV) end-diastolic diameter (41 mm) within the standards; right ventricular diameter-RV (30 mm) 15% above, LV end-systolic diameter (20 mm) 20% below, and diastolic septum thickness (5 mm) and LV-PPV posterior wall diastolic thickness (5 mm) 28% below these standards; These differences did not cause physiological dysfunctions in the heart. He presented mild calcification in the aortic valve, without physiological changes. AE/AA ratio (0.96), ejection fraction (83%), septum/LVPP ratio (1.00), end-diastolic volume (74 ml) and systolic volume (61 ml) were within the standard variations; there was a decrease of ± 29% in LV mass (67 g) and ± 28% in end-systolic volume (13 ml) in relation to the standard intervals. Volume/mass ratio (1.03 ml/g) was 14% higher than normal standards. These results do not denote dysfunctions, since their overall systolic function at the left ventricular level was normalized.

The helical evaluation of the chest detected a small homogeneous calcified nodule in the central portion in the transition of the anterior/posterior segments of the right upper lobe, without negative influences on the pulmonary physiology; deformity with marked left-directed convexity thoracic scoliosis, with degenerative changes in the facet joints, indicating a decrease in the left intrathoracic space, with impairment for lung expansion. Mediastinal lymph node enlargement was absent.

In pulmonary functions, FVC, FEV1 and PEF presented values of 54%, 64% and 81%, respectively, of the predictors, but FEV1% and 2575 presented values of 118% and 116%, respectively, of the predictors with measurement without bronchodilator substance effects. With bronchodilator substance, these variables continued to present values lower than the predictors, but with differences lower than 5% (statistically insignificant). Regarding the FET, pre-bronchodilator measurement showed a decrease of 8.5% in relation to the post-brochodilator. Brochodilated responses were negative. Results of CVF or forced vital capacity are compared with predicted values calculated from age, height, body mass, sex and ethnic group. FEV1 is the volume expired in the first second of the FVC. FEV1% is FEV1 divided by Vital Capacity, and FEV1/FVC*100 is also accepted as FEV1%. It was found that this volunteer has moderate restrictive respiratory disorder caused by the reduction of the intrathoracic space, due to the accentuated thoracic scoliosis of convexity directed to the left and calcification of tissues in that region.

In the evaluations of maximal respiratory pressures, after three maneuvers for MEP, a higher value was considered, 65 mmH, when in MIP the highest value considered was 55 mmHg, demonstrating that this volunteer has lower inspiratory strength than expiratory (Chart 1).

Table 1: manovacuometry results.

First. manoeuvre	PeMax 1	60	Mmhg
Second. manoeuvre	**PeMáx 2**	**65**	**mmHg**
Third. manoeuvre	PeMáx 3	60	mmHg
First. manoeuvre	PiMáx 1	55	mmHg
Second. manoeuvre	PiMáx 2	40	mmHg
Third. manoeuvre	**PiMáx 3**	**55**	**mmHg**

When compared to the predictors (BLACK, HYATT, 1969), MEP presented a value lower than the mean in 42.7% and MIP in 63.2% (Table 1).

Table 1: Normal values for maximal respiratory pressures25.

				*Pressure** (cm H2Ø)		
				Age (yr)		
Pressure	*Sex*	20 - 54	55 - 59	60 - 64	65 - 69	70 - 74
MIP	*Male*	124 ± 44	103 ± 32	103 ± 32	103 ± 32	103 ± 32
	Female	87 ± 32	77 ± 26	73 ± 26	70 ± 26	65 ± 26
MEP	*Male*	233 ± 84	218 ± 74	209 ± 74	197 ± 74	185 ± 74
	Female	152 ± 54	145 ± 40	140 ± 40	135 ± 40	128 ± 40

* Numbers represent mean ± 2SD or regression line ± 2sy·x.

Attention is drawn to studies by Lima et al. (2021), on the improvement of results for Mip. and Pemax. through the execution of a respiratory muscle training protocol (RMT) for patients with temporomandibular disorders (TMD), which could also help patients with PFO.

> This study adds to the literature a new treatment protocol for individuals with TMD and shows that the RMT resulted in increased respiratory muscle strength, lower ECOM activation, lower activation of all muscles evaluated at forced inspiration, greater serratus activation at inspiration and greater activation of the diaphragm and intercostal at forced expiration.

In body composition, there was a BM of 39.0 kg and h of 1.57 m, BMI of 15.8 kg /m2 considered low. AC resulted in 64 cm, considered low. %FM was moderate, with 26% as a result, and MM of 27.39 kg. Bone mineral content was 1.53 kg, a very low value, and fat-free mass (MLG) was 28.92 kg. The most significant result was the MRI, accusing 4.5 kg/m2, characterizing sarcopenia that, together with the low bone mineral content, is shown to compromise the volunteer, since, despite the young age, such factors are associated with the Frailty Syndrome, common in the elderly. Positively, values of BMI, WC and result of the android/gynoid ratio, found in 0.32, are opposed to the Metabolic Syndrome.

IO and its category obtained values below the standards (Chart 2), corroborating the assessment of body composition.

Table 2: IO results.

IOa	0,25 %
IOb	0,39 %
IO	**63,50 %**
IO Category	**Marasmus**

Glycemia (77.81 mg/dL) and triglycerides (36.98 mg/dL) presented desirable values; C-reactive protein (1.60 mg/L) was negative.

With goniometry, it was found that 05 joints (RE, OE, PE, TD and ET) presented total immobility as a result of the pathology. The flexibility of the CE, JD and JE joints presented impairment, but the flexibility of the CD and PD joints were well preserved (Chart 3).

Table 3: goniometry results.

Goniometry OD	0th
OE goniometry	0th
CD goniometry	150th
EC goniometry	55th
PD goniometry	110th
Goniometry PE	0th
JD Goniometry	40th
Goniometry JE	25th
TD goniometry	0th
TE goniometry	0th

According to Several researchers (KAPLAN, et al., 2005; HERRERA-ESPARZA, et al., 2013; HASAN, 2012; KAPLAN, et al., 2005; GOSAI, et al., 2013), FOP can lead to transformation of endothelial cells into mesenchymal stem cells and bone, monophalangism and brachydactyly in the halluces, thedisseminated ssification in soft tissues and congenital stigmas in the extremities, progressive heterotopic osteogenesis, thecongenital nomalias, osteochondromas Tibial proximal and medial, orthotopic fusions of the posterior elements of the cervical spine, pScotland of the short and wide femur, conductive hearing loss, TIS due to the costovertebral malformations with ankylosis of the costovertebral joints, ossification of the intercostal and paravertebral muscles, and Of aponeuroses, progressive spinal deformity, kyphoscoliosis or thoracic lordosis, pneumonia, and right-sided congestive heart failure resulting from TIS.

The classical literature does not mention alterations in other diverse anthropometric and physiological variables that may be compromised by this pathology.

Considering that in the cardiovascular conditions, the volunteer presented structural parameters of AA and LA, and LV end-diastolic diameter within the standards; RV diameter above these standards; LV end-systolic diameter, septal diastolic thickness and LVPP below, it is concluded that the differences did not cause cardiac physiological dysfunctions, not concerning PFO, since no cardiac changes or deformities caused by the gene mutation of the pathology. Although the detection of mild calcification in the aortic valve is by PFO, there is no physiological impairment. Regarding ventricular relations and functions, there was a ratio of LA/AA, ejection fraction, septum/LVPP ratio, end-diastolic volume and systolic volume within the standard variations. A decrease in LV mass and end-systolic volume was detected in relation to the standard intervals, with only the highest volume/mass ratio. These results were not supported in the literature to have, as justification, the mutation of the ACVR1 gene. As the results are not causing impairments, since the overall systolic function at the left ventricular level is normalized, the PFO is not interfering in the cardiovascular functions of this volunteer.

In the anatomo-thoracic aspect, the results of this volunteer corroborate the literature, since helical evaluation of the chest detected a small homogeneous calcified nodule in the central portion in the transition of the anteroposterior segments of the right upper lobe, in addition to a deformity with marked left-directed convexity thoracic scoliosis, with degenerative changes in the facet joints, causing reduction of the left intrathoracic space, compromising lung expansion, proving that this pathology directly affects the intrathoracic morphology of its carrier.

Regarding lung volumes and capacities, without the effects of bronchodilator substance, lower values were verified, when compared to the predictors, for FVC, FEV1, and PEF, although FEV1% and 2575 presented higher values. After bronchodilator substance, FVC, FEV1, and PEF continued to present values lower than the predictors, but with statistically insignificant differences. FET presented pre-bronchodilator measurement inferior to the post-brochodilator one, leading to the conclusion that the responses to the brochodilator substance were negative. As variable 2575 is the first parameter that decreases due to many respiratory pathologies, it is concluded that, in this volunteer, there is no interference of any disease, but analyzing the other results, which found moderate restrictive respiratory disorder, this diagnosis is attributed to PFO, which caused marked left-directed convexity thoracic scoliosis and tissue calcification in that region, decreasing the intrathoracic space and compromising the lung parenchyma in its expansiveness. Regarding maximal respiratory pressures, the results lead to the conclusion that in MIP and MEP, this volunteer presented quite significant differences lower than the predictors, especially in relation to strength inspiratory, leading to the belief that their pulmonary impairment in the face of PFO has a direct reflex in the generation of strength of the breathing muscles.

The values obtained for CM, h, BMI, WC, bone mineral content, MLG and IO were considered low for this volunteer, especially MRI, characterizing sarcopenia, leading to the conclusion that, due to these combined variables, this carrier, despite her young age, has Frailty Syndrome, caused by the consequences of the pathology. However, %MG and MM were moderate and BMI, WC and android/gynoid ratio values were low, in contrast to the possibility of involvement of Metabolic Syndrome.

Glycemia and triglycerides, which presented values within the desirable standards, and C-reactive protein that presented negative results, it is concluded that these variables were not influenced as consequences of the pathology.

In mobility and ambulation, there is impairment, since total immobility has already been manifested in 05 joints and flexibility impairment in 03 more, due to the new calcifications caused by the ACVR1 mutation, with only 02 joints preserved.

Finally, the PFO in this volunteer, in addition to what has already been reported by the classical literature, has already caused impairment in the expansion of the lung parenchyma, due to the reduction

of the intrathoracic space, causing, as a more alarming consequence, a moderate restrictive respiratory disorder, in addition to very significant decreases in respiratory pressures, especially inspiratory, with a direct negative reflex in the generation of force of the breathing muscles. In body composition, sarcopenia and Frailty Syndrome have been established, as well as their flexibility, mobility and ambulation are profoundly limited. Positively, no interference of this pathology was verified in the cardiovascular conditions, as well as in %MG, MM, BMI, CA, android/gynoid ratio, glycemia, triglycerides and C-reactive protein, ruling out an involvement of Metabolic Syndrome.

A more in-depth study is suggested that involves, if possible, the entire population with FOP in Brazil, estimated at 70 affected, for a safer confirmation of the anthropometric and physiological variables that are actually affected by this rare gene mutation.

It is concluded that, in this patient with PFO, the cardiovascular system is preserved, within the standard conditions considered acceptable, despite the manifestation of RV diameter above, LV end-systolic diameter below, and diastolic thickness of the septum and posterior wall of LV-LVPP below normal, as well as decrease in LV mass in the final systolic volume, and higher volume/mass ratio in relation to standard intervals.

In body composition, this volunteer has very low MC, BMI, CA, IO, bone mineral content and MMRI, the latter characterizing sarcopenia. Such associated factors denote a frailty syndrome. What justifies the bone mineral content to be greatly reduced when there is the incidence of new bone tissues is the fact that these new calcifications do not occur in the bone system, but in the muscular system and connective tissues, structures that are not considered in bone densitometry, as well as are not calcifications that will strengthen their support structures, protection and ambulation. This volunteer is outside the population at risk for Metabolic Syndrome, due to her BMI, WC and android/gynoid ratio being low.

Regarding blood glucose and triglycerides, verified with desirable values, and C-reactive protein with negative results, it is concluded that these physiological variables did not suffer interference from the pathology.

Goniometry showed severe immobility in 05 synovial joints, with involvement of 03 more joints, being still preserved only 02 to rticulations, in relation to flexibility, due to new calcifications as a result of the pathology. Such conditions prevent the carrier from sitting, forcing her to always be in bipedal or decubitus.

Tomography detected a calcified nodule, without damage to pulmonary physiology, indicating a decrease in the intrathoracic space, with impairment for lung expansion on the left side. More worrisome results were manifested in pulmonary functions, with FVC, FEV1 and PEF values below the predictors, although FEV1% and 2575 were above, without bronchodilator effects. Under the effect of bronchodilators, the differences of these variables were insignificant. In addition, pre-

bronchodilation FET showed a decrease in relation to post-brocodilation FET, showing that brochodilator responses were negative. Maximal respiratory pressures also presented values lower than the means. Although 2575 is the first parameter that decreases in many respiratory diseases, which was not manifested in this carrier, it is concluded that this volunteer has a moderate restrictive respiratory disorder caused by the decrease in the intrathoracic space, due to the accentuated thoracic scoliosis of convexity directed to the left and calcification of tissues in that region.

4 CONCLUSION

The PFO in this volunteer is physiologically compromising only the respiratory system, with moderate restrictive respiratory disorder caused by the accentuated left-directed convexity thoracic scoliosis, due to the impairment of the bipedal position in the face of the new calcifications.

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