Romanian Journal of Medical Rehabilitation,
Physical Medicine and Balneoclimatology
RJMRPMB; ISSN 3044 – 8786; ISSN-L 3044 – 8778

Volume I, Issue 2
PP 101–108
August 2024

ORIGINAL PAPER

PULMONARY REHABILITATION IN PECTUS EXCAVATUM: FROM THEORY TO MEDICAL PRACTICE

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Received 28th July 2024, Revisions received 19th August 2024, Accepted 20th August 2024
https://www.doi.org/10.59277/RJMRPMB.2024.2.05

Abstract. Background: Pectus excavatum (PE) is the most common developmental abnormality of the chest, which is determined by the abnormal development of the sternum and ribs with the appearance of a concavity in the anterior chest wall. **Case presentation:** We present the case of a 43-year-old man, diagnosed at the age of 5 with Pectus excavatum, for which surgical intervention was performed, and who later, at the age of 8, underwent right lower partial lobectomy for pulmonary cysts. The patient included in the pulmonary rehabilitation program at the Rehabilitation Clinical Hospital, Iasi, Romania, returns for exertional dyspnea, chest constriction, moderate physical asthenia. The patient goes through all the complex multidisciplinary assessments and continues the pulmonary rehabilitaton (PR) program, and at the end of the two weeks of hospitalization the benefits are quantified, he continues the PR program at home. **Conclusions:** Pulmonary rehabilitation is one of the most powerful evidence-based therapies, constituting a cornerstone alongside surgical management of congenital chest malformation.

Keywords: *pectus excavatum, dyspnea, exercise capacity, training, quality of life*.

Background

Pectus excavatum (also known as sunken sternum or funnel chest) is a congenital malformation of the chest caused by the abnormal development of the sternum and ribs with the appearance of a concavity in the anterior chest wall. The incidence rate of the condition is 1:1000 cases, being the most common developmental abnormality of the chest (95%), followed by pectus carinatum, Cantrell's syndrome, asphyxiating thoracic dystrophy and spondylo-thoracic dysplasia [1–3].

Therapeutic management includes surgical correction based on resection and reconstruction of the chest wall, followed by a customized rehabilitation program to improve the fitness of the cardiorespiratory system, to correct the vicious position of the spine, improve muscle contracture, increase the laxity of the joints that participate in breathing, toning the respiratory muscles, increasing the effort capacity, etc [4–6].

Case presentation

We present the case of a 43-year-old male patient, non-smoker, with no history of occupational exposure, who at the age of 5 (1981) was diagnosed with Pectus excavatum for which he underwent surgery (sternochondroplasty).

Later, at the age of 8 (1984), he underwent right lower partial lobectomy for pulmonary cysts. The patient has associated respiratory (pulmonary emphysema), cardiovascular (essential arterial hypertension grade 1) and rheumatological (dextroconvex dorsal scoliosis, operated L_5–S_1 disc herniation) pathology. He presents himself to the Pulmonary Rehabilitation Clinic of the Rehabilitation Clinical Hospital Iasi for grade III dyspnea on the modified Medical Research Council (mMRC) scale, cough with mucous expectoration, predominantly in the morning, anterior chest constriction and moderate physical asthenia.

The clinical examination at admission reveals a relatively good general condition, BMI=19.6 kg/m^2, pale skin, hyperemic thenar and hypothenar eminences, venous ectasias in the lower limbs bilaterally, asymmetric chest, sunken sternum (sternal excavation depth = 3 cm) (Fig. 1), excursions asymmetric ribs of reduced amplitude, post-operative scars located at the level of the right hemithorax and anterior thorax, pulmonary auscultation diminished vesicular murmur bilaterally, crackling rales disseminated throughout the lung area, SaO_2 = 98% aa, modified precordial area, apex shock in the IV intercostal space on the left midclavicular line, rhythmic heart sounds, HR = 82/min, BP = 130/80 mmHg.

The biological balance reveals discrete non-specific inflammatory syndrome, normochromic normocytic anemia.

Assessment of dyspnea at rest using the mMRC scale and the BORG scale during exertion in our patient showed grade III and 7, respectively.

[*Photo Source:* Personal Archive]

Figure 1 – Appearance of the thorax in Pectus excavatum.

When assessing current daily activities using the Lawton Instrumental Activities of Daily Living Scale, the patient scored 7, which places him at assisted independence.

The electrocardiogram shows a deviated axis to the right, negative P waves in V1, incomplete right bundle branch block (rSR' in the right precordial leads – V1) and biatrial hypertrophy. Echocardiography shows a small left atrium (25/37/40 mm), absence of intracavitary formations, normal pericardium, no signs of PH, mild mitral insufficiency and left ventricular ejection fraction = 69%.

Respiratory function, spirometry reveals severe restrictive ventilatory dysfunction – FVC=44.6% (2.44L), FEV1= 34% (1.51L), FEV1/FVC=71.8 and DLCO = 78% (value at the lower limit of normal).

Imaging, postero-anterior and profile chest X-ray reveals right apical pachypleuritis, with small areas of fibrosis, bilateral basal accentuated lung hypertransparency, with well-defined appearance on profile X-ray (obs. large emphysema bubbles) and right costo-diaphragmatic sinus symphysis. The chest CT examination with contrast material highlights:

1) deformed and asymmetric thorax, with the displacement of the cardiomediastinal elements to the left side,
2) right apical fibromicronodular lesions,
3) right lung – hyperdense suture material at the level of the lower lobar bronchus (partial lower right lobectomy),
4) at the base of the right lung – extensive pulmonary hypertransparency with a poor lung pattern, with fine pleuro-septal adhesions and a few (2 mm) subpleural micronodules,
5) tracheal diverticulum with a diameter of 5mm,
6) the left adrenal gland with a hyperplastic appearance,
7) the dorsal spine in dextroconvex scoliosis, with degenerative arthrosic dorsal vertebral changes, small dorsal intraspongy hernias, anterior subsidence of the T_{12} and L1 vertebral bodies (Fig. 2).

A) B) C)

[*Photo Source: Personal Archive*]

Figure 2 – A) and B) – CT axial and coronal section – Deformed thorax (with reduction of antero-posterior diameter) and asymmetric (with dimensional reduction of the right hemithorax), with displacement of the mediastinum and the heart to the left side. C) Coronal CT Section – Right Lower Partial Lobectomy.

Exercise capacity was assessed by the 6 minutes walk test (6MWT), cardiopulmonary exercise test (CPET) and respiratory muscle testing.

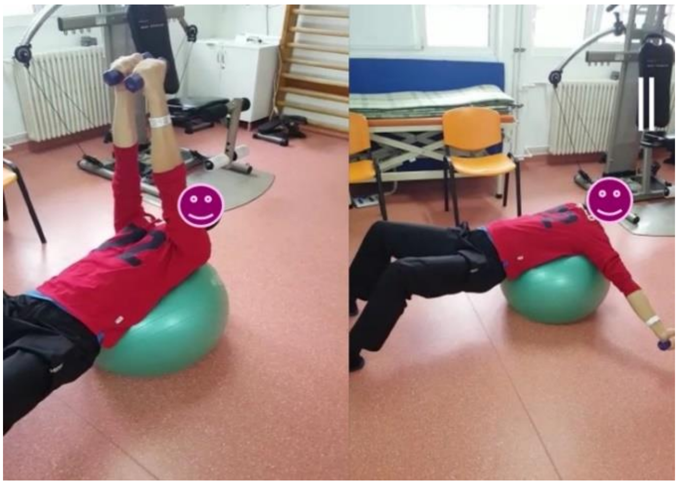
- 6MWT – Patient walks 70% of predicted distance in 6 min walk test (570 m out of 816 predicted meters). Initially, SpO_2 = 98% aa, HR = 78/min, BP =130/80mmHg, dyspnea (BORG scale) = 2. Final, SpO_2 = 95% aa, HR = 98/min, BP =145/95 mmHg, dyspnea (BORG scale) =7.
- CPET – Maximum level of effort = 44% of the maximum predicted value, on the Borg scale =15. Oxygen consumption – maximum value of 1421 ml/minute (53%). Anaerobic threshold – not reached. Respiratory exchange rate (RER = $VCO_2/V0_2$) = 0.94. Ventilation per minute = 34.3 liters/minute → 49% of the ventilation reserve, being reduced. SaO_2 = = maintained at 95%. FC-114 bpm → 166 bpm (94% of maximum predicted value). Oxygen pulse = 8.9 ml/min/beat (57% – important reduction). BP = 130/80 mmHg → 190/100 mmHg. Conclusions: Slight reduction of functional capacity (Weber class A); a heart rate target of 125 beats/minute is recommended for aerobic exercise.
- Evaluation of the respiratory muscles – the maximum inspiratory pressure at the level of the oral cavity is 40 cmH_2O (the highest value among the three determinations). Initially, the patient will train with a pressure of 30% of the initial value, which is 12 cmH_2O.

During hospitalization, the patient receives cortisone treatment in sulfur water aerosols, expectorant, parenteral hydration, cardiorespiratory medication (Bisoprolol and Theophylline) and the pulmonary rehabilitation program.

Exercises performed by the patient within the personalized PR program (Figs. 3–8).

[*Photo Source:* Personal Archive]

Figure 3 – In orthostatism, with the lower limbs apart, with the cane in front: Time 1: Inhale with the cane above the head; Time 2: Expires with the rear stick; Time 3: Inhale with the cane above the head; Time 4: Exhale with flexion of the trunk and bringing the stick to the level of the knees.



[*Photo Source:* Personal Archive]

Figure 4 – The patient sitting supine on the physioball, holding 1 kg dumbbells in his hands, performs abduction movements of the arms on inhalation and adduction on exhalation.

[*Photo Source:* Personal Archive]

Figure 5 – The patient in orthostatism is seated with his right side to the trellis, with his right hand at the level of the hip and the left hand attached to the trellis at the level of the head. Perform the inclination of the trunk to the left with the opening of the left hemithorax on inhalation and return on exhalation.

[*Photo Source:* Personal Archive]

Figure 6 – Training with the multifunctional device, to which threshold weight can be added to also train the muscles of the upper limbs in inhalation and exhalation. The patient performs exercise training, the degree of difficulty of which can be progressively increased.

[*Photo Source:* Personal Archive]

Figure 7 – Dosed exercise training with the cycle ergometer for the muscles of the lower and upper limbs. On the cycle ergometer, continuous or interval training can be performed.

[*Photo Source:* Personal Archive]

Figure 8 – Respiratory muscle training with specialized medical devices – PowerBreathe device.

At the end of the two weeks of hospitalization, the patient's effort capacity increased as quantified by increasing the distance in the 66 MWT (78.7% of the predicted distance = 635 meters, compared to 570 meters from the hospitalization) and the respiratory muscles quantified by the increase of the maximum inspiratory pressure from the level of the oral cavity to 52 cmH_2O, and when evaluating dyspnea on the mMRC scale – the patient obtained a score of 1 (compared to 3 on admission).

The patient's prognosis is relatively good as long as he continues the PR program at home and avoids intense physical exertion. The presence of right apical pulmonary micronodules requires a periodic imaging evaluation to detect their possible dimensional increase.

Discussions

Pectus excavatum occurs as a result of the abnormal development of the sternum and ribs with the appearance of a concavity in the anterior chest wall. The depth of the excavation can affect the functions of the thoracic organs, causing the appearance of cardiorespiratory symptoms such as: intolerance to effort, dyspnea, wheezing, previous chest pains, heart rhythm changes, repeated respiratory infections, fatigue and osteoarticular changes. The etiopathogenesis of PE is based on a series of factors such as intrauterine trauma, malformations of the diaphragm, rickets or the dyschondroplastic theory [1, 3].

Therapeutic management includes surgical correction based on chest wall resection and reconstruction, followed by a personalized rehabilitation program [7].

Our patient benefited from the Nuss procedure as a technique for surgical correction of the malformation, which involves the retrosternal placement, under thoracoscopic control, of a precurved special blade (made of steel, titanium or nickel) – initially, which is inserted with the concavity up and then through rotation at 180º, the concavity of the blade is directed downwards, correcting the deformity by raising the sternum. Surgical techniques have evolved and today there are multiple options for intervention [8, 9].

The American Thoracic Society / European Respiratory Society defines pulmonary rehabilitation as “an evidence-based, multidisciplinary and complex intervention for patients with chronic respiratory disease who are symptomatic and often have limited daily activities” and “is designed to reduce symptoms, to optimizing functional status, increasing adherence to treatment and reducing care costs, by stabilizing or ameliorating the systemic manifestations of the disease.” The current definition of PR, according to the American Thoracic Society Practice Guideline published in August 2023, is based on the definitions previously developed, based on scientific evidence, by specialists in respiratory recovery, which were published starting in 1974 and updated in 2006, 2007, 2013, 2015 and 2021 [10, 11].

Dosed physical training is one of the most important components of physical therapy and PR programs. The physical exercise program is individualized for each patient depending on: the degree of severity of the disease, age, sex, associated diseases, training methods, duration, rhythm, intensity of effort, motivation and the patient's choice [7].

Respiratory muscle training is a valuable method that provides additional PR benefits by improving both muscle strength and endurance. Its purpose is to improve the function of the respiratory muscles and relieve dyspnea. Respiratory muscle testing measures the maximum inspiratory or expiratory pressure a subject can generate in the oral cavity. The test consists of performing three maneuvers, which must have a maximum variability of 20%, from which the best result is chosen, and during the hospitalization the patient will train at a percentage of 30–60% of the initial value. At the end of the PR program, they are tested again to note the patient's progress, which is usually favorable [12, 13].

Breathing exercises can be varied by: the frequency of breaths per minute, the manner of movement of the chest and abdomen, the duration of the respiratory phases, the combination of the respiratory phases, the execution posture, directing the air at the level of the airways [12, 14].

The techniques and objectives of the pulmonary rehabilitation program in PE patients are represented in Figs. 9 and 10.

In the case of the patient with PE, who was compliant and followed all medical indications, who was included in the respiratory recovery program in 2008, significant progress was observed regarding exercise tolerance, posture correction, respiratory muscle toning, control and breathing coordination. All these benefits allowed him a high quality of life and a favorable socio-professional reintegration. In the specialized literature, there are few publications on the effects of respiratory recovery in patients with pectus excavatum. The rehabilitation process as such is often referred to as conservative care before surgery. There is no unequivocal evidence that rehabilitation has an impact on chest defect reduction [5, 7].

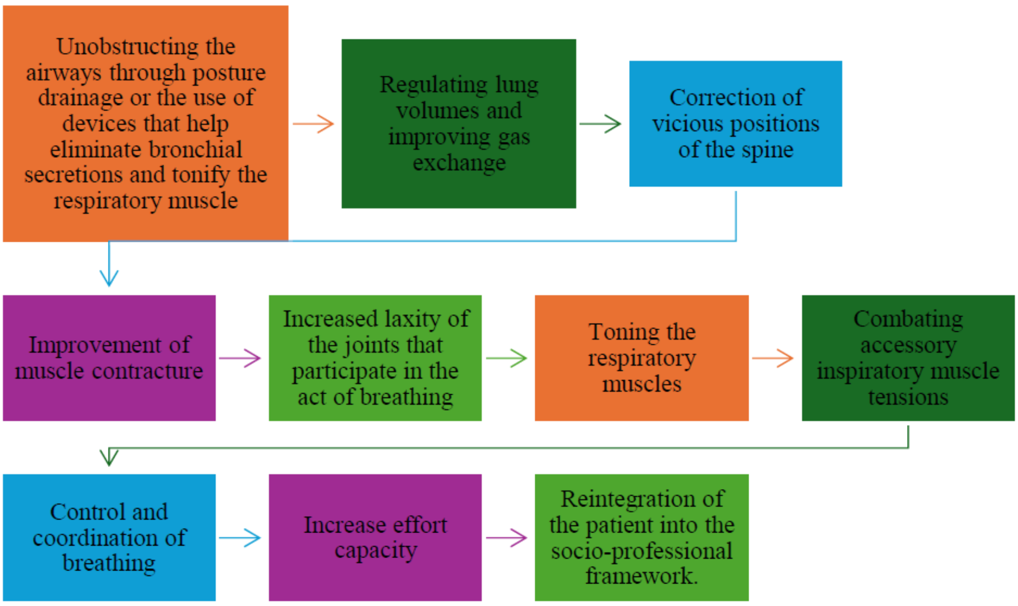


Figure 9 – Objectives of the PR Program [12].

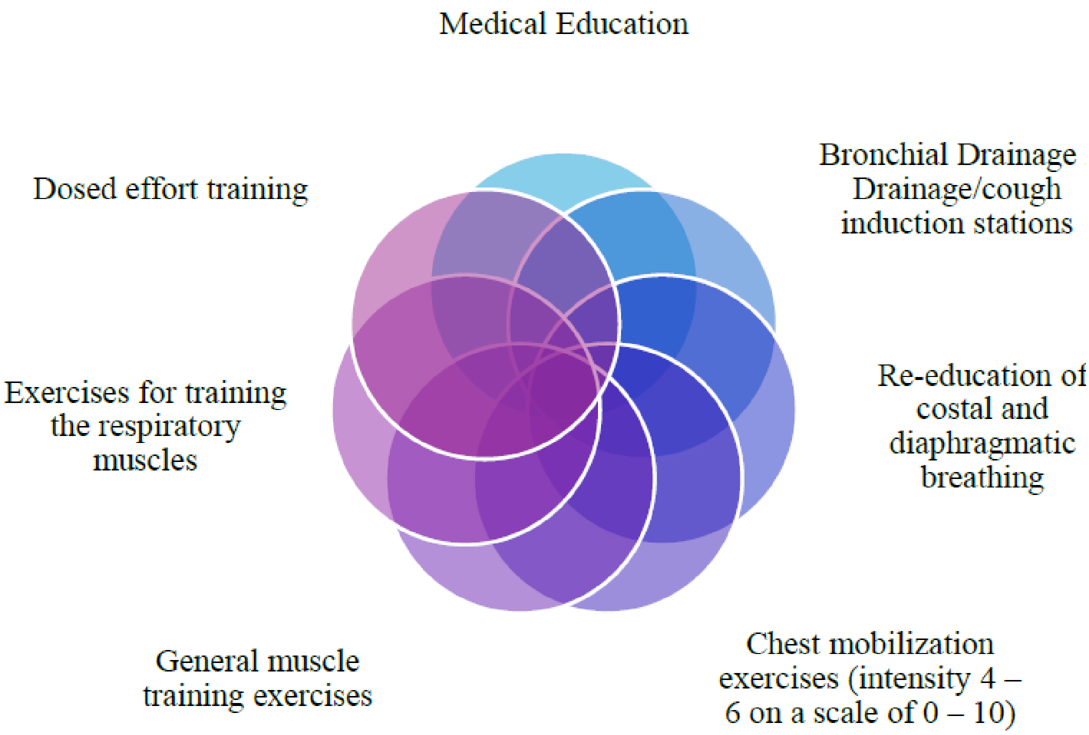


Figure 10 – Techniques of the PR Program [12].

Conclusions

Pulmonary rehabilitation is one of the most powerful evidence-based therapies, forming the cornerstone, along with surgical management, of congenital chest malformation. The particularities of this case are represented by:

1) the presence of a congenital malformation that is less noticeable at birth and that accentuates and becomes evident with advancing age,
2) the compression exerted by the blocked sternum on the intrathoracic and mediastinal organs, with the displacement from their normal position,
3) although surgical intervention was performed to correct the malformation in childhood, it recurred with the recurrence of the excavation,
4) inadequate aesthetic appearance and
5) alterations of cardiac and pulmonary functions (right bundle branch block, mitral insufficiency, severe restrictive ventilation dysfunction).

Compliance with Ethics Requirements:

"The authors declare no conflict of interest regarding this article."

"The authors declare that all the procedures and experiments of this study respect the ethical standards in the Helsinki Declaration of 1975, as revised in 2008(5), as well as the national law. Informed consent was obtained from all the patients included in the study."

"No funding for this study."

Acknowledgements: *None*

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