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An unusual way to improve lung function in congenital myopathies: the power of singing

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Congenital myopathies (CMs) are a clinically and genetically heterogeneous group of disorders characterized by early onset weakness, hypotonia and characteristic structural abnormalities in muscle fibres. Hypotonia and weakness can be present at birth or appear in infancy, and a static or slowly progressive clinical course may present with muscle weakness, loss of spontaneous movement, involuntary muscle activity, and muscle atrophy. Often patients develop a restrictive syndrome and respiratory failure and require respiratory support
In our case, we described lung improvement and respiratory muscle training due to singing in a young patient, affected by CMs with a poor adherence to non-invasive mechanical ventilation.

Key words: congenital myopathies, FVC, respiratory muscle training, singing

Introduction

Respiratory insufficiency is one of the main causes of death in CMs [1,2]. These patients have a restrictive ventilatory pattern, hypoventilation, chronic hypercapnia, and sleep disturbances. Respiratory failure is associated with increased morbidity and poor quality of life perception [3]. We describe the case of a young woman, affected by congenital miopathy, she had not compliance and tolerance with non-invasive mechanical ventilation or alternative respiratory support devices. The young patient started to take singing lessons for recreational purposes, meanwhile we were noting down the variation of her respiratory function during the months.

Case description

Caterina, 30 years old woman with spastic tetraparesis and drug-treated epilepsy was monitored for evaluation of daytime dyspnea in our Department. We performed pulmonary function tests, and resulted a restrictive pattern, as attended. The patient was in treatment with cycle of diurnal non invasive mechanical ventilation with a mouthpiece mask, because of poor adherence with nasal mask due to claustrophobia and anxiety of not being able to call a family member, if necessary. However, the patient complained of episodes of panic attack, and reduced control of epilepsy. We tried to adapt the patient to alternative respiratory support modalities such as Intermittent Abdominal Pressur Ventilation (IAPV) [4]. The patient reported tightness, claustrophobia, panic attacks and worsening epilepsy. Therefore, after a few adaptation sessions, she interrupted the respiratory support.
Forced vital capacity (FVC) and forced expiratory volume in one second (FEV1) were measured with spirometer. The pulmonary function test revealed a severe reduction of respiratory capacity: FVC was 0.93 L (28%) and FEV1 0.91 (31%). MIP and MEP were measured, with a value of 24 and 36. PCEF was 60 L/sec. Dyspnea was assessed with the dyspnea VAS scale. The index has 10 numerical values, rated on a scale from 0-10, each value corresponding to an intensity of breathlessness.

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Meanwhile, the patient decided to take private singing lessons just for fun, as a hobby and in an attempt to relax. The program included exercises aimed at controlling emotion and anxiety, exercises to coordinate breathing, with deep inhalations and exhalations, and finally diaphragmatic breathing exercises. The duration of the lessons was about 1 hour.

After three months of singing lessons, the patient was reevaluated as routinely happened at the referral centre. Spirometry showed an improvement in diurnal respiratory function with values of FVC equal to 1.08 L (32%) *vs* 0.93 L (28%) and FEV1 equal to 1.04 L (35%) *vs* 0.91 L (31%), accompanied by an improvement in occurence of symptoms as tiredness and dyspnea.

Six months after the beginning of singing program, there was a further improvement in pulmonary function as can be seen in the Table I (FVC = 1.21 L (86%); FEV1 = 1.15 (39%). MIP was 36, MEP was 48, PCEF was 120 L/sec (Tab. I)

We evaluated how much singing positively affected the patient's quality of life with a VAS scale, the score obtained on the question "How much does singing improve your quality of life?" The value was 10, the maximum on the VAS scale.

Table I. Respiratory parameters observed pre and post Singing lessons.

	FVC (lt)	FEV1 (lt)	Dyspnea (VAS scale)	MIP	MEP	PCEF
Pre Singing Lesson	0.93	0.91	8	24	36	60
3 months post	1.08	1.04	5	32	40	100
6 months post	1.21	1.15	4	38	48	120

Discussion

Respiratory function in CMs patients is characterized by progressive inspiratory and expiratory muscle weakness, which causes a restrictive respiratory syndrome [2,3]. Singing for people with respiratory disease is perceived to be beneficial in managing dyspnoea, increasing well-being and Quality of Life, and reducing social isolation [5]. In our case we observed an unexpected improvement in respiratory function. One hypothesis to explain the improvements in lung volume is that the application of the singing lesson could help maintain lung and chest wall compliance, prolongs the patient's inspiratory and expiratory time with an improvement in alveolar ventilation and resolution of atelectasis. In addition, singing improves muscle strength, therefore it could, in some cases, improve restrictive breathing patterns.

It has been developed in recent years "Singing for Lung Health" (SLH), a rehabilitation programme borned in the UK and has become increasingly popular, although heterogeneous singing approaches are applied in other countries [5]. SLH included physical, vocal and breathing exercises, with a focus on improving strength, endurance and flexibility of the respiratory muscles [6,7]. The British Lung Foundation initiative SLH includes systematic training of singing teachers in lung physiology/pathophysiology and a methodological approach to singing as a physical activity providing respiratory control [5].

With regard to physical outcomes, singing may improve respiratory muscle strength, coordination and performance; reduce hyperinflation; improve lung function; and enhance functional exercise capacity [8].

The focus is on the learning of breathing control and postural techniques that enable effective singing, where song is used to improve and reduce breathlessness. Therefore it may offer an innovative and practical means to provide insight into patient perspectives and perceptions of breathlessness [9,10].

In a randomised controlled trial, the SHL programme was compared with conventional exercises training as part of a 10-week community-based pulmonary rehabilitation programme. Based on their findings, the authors concluded that a singing programme was not inferior to an exercise training programme in improving functional exercise capacity [10].

Multiple psychological well-being benefits from singing are reported across studies, such as increases in positive mood. Patients with lung diseases taking a singing lesson reported increased energy, amateur singers sometimes report an increased sense of self-expression. Choral singers also had significantly higher scores on quantitative measure of positive mood, personal growth, vitality. Temporary increases in immune response (e.g. salivary immunoglobulin A) may occur after choral singing. Biological stress responses such as cortisol or heart rate may vary in these patients [11].

Currently, there is no international consensus guideline on singing as a training intervention for lung disease [12]. While respiratory protocols for respiratory muscle tests are well defined as respiratory management protocols for patients with neuromuscular conditions in general [13], the specific approach to respiratory treatment in CMs is not standardized yet [3]. In fact, there are limited data on the natural history of changes in pulmonary function over time in patients with CMs; furthermore the clinical presentation of respiratory involvement varies between patients with different pathologies. Therefore the management protocols for respiratory care in these patients vary based on the patient's muscle weakness [1,3].

Pulmonary rehabilitation is a cornerstone in pulmonary diseases



Figure 1. Caterina and her teacher during a singing lesson.

management. However, pulmonary rehabilitation adherence is generally low, and barriers include availability, economic issues, motivation and an inability to attend or perform physical training [14].
Therefore, alternative, evidence-based pulmonary rehabilitation activities are required. Singing may have benefits for quality of life (QoL), respiratory control and well-being, but the impact on the pulmonary rehabilitation key outcome, physical exercise capacity, is still uncertain. Singing is also proposed to improve diaphragmatic control, and further study are necessary.

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Conflict of interest statement

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Authors' contributions

AA and GF: conceptualization; MRV and LA: methodology; MRV, AA, GF: writing - original draft preparation; MRV, AA and LA: writing - review and editing. All authors have read and agreed to the published version of the manuscript

Ethical consideration

the authors obtained consensus to publish data and photo

Refererences

1 Fujimura-Kiyono C, Racz GZ, Nishino I. Myotubular/centronuclear myopathy and central core disease. Neurol India 2008;56:325-32. https://doi.org/10.4103/0028-3886.43451

2 Thaha F, Gayathri N, Nalini A. Congenital myopathies: clinical and immunohistochemical study. Neurol India 2011;59:879-83. https://doi.org/10.4103/0028-3886.91369

3 McEntagart M, Parsons G, Buj-Bello A, et al. Genotype-phenotype correlations in X-linked myotubular myopathy. Neuromuscul Disord 2002;12:939-46. https://doi.org/10.1016/s0960-8966(02)00153-0

4 Annunziata A, Coppola A, Polistina GE, et al. Daytime alternatives for non-invasive mechanical ventilation in neuromuscular disorders. Acta Myol 2021;40:51-60. https://doi.org/10.36185/2532-1900-042

5 Lewis A, Cave P, Stern M, et al. Singing for Lung Health - a systematic review of the literature and consensus statement. NPJ Prim Care Resp Med 2016;26:16080. https://doi.org/10.1038/npjpcrm.2016.80

6 Lewis A, Cave P, Hopkinson N. Singing for Lung Health: service evaluation of the British Lung Foundation programme. Perspect Public Health 2018;138:215-222. https://doi.org/10.1177/1757913918774079

7 Goldenberg RB. Singing lessons for respiratory health: a literature review. J Voice 2017;32:85-94. https://doi.org/10.1016/j.jvoice.2017.03.021

8 Gick ML, Nicol JJ. Singing for respiratory health: theory, evidence and challenges. Health Promot Int 2016;31:725-734. https://doi.org/10.1093/heapro/dav013

9 Oxley R, Harrison SL, Rose A, et al. The meaning of the name of 'pulmonary rehabilitation' and its influence on engagement with individuals with chronic lung disease. Chron Resp Dis 2019;16:1479973119847659. https://doi.org/10.1177/1479973119847659

10 Vaes AW, Spruit MA, Franssen FME, et al. The efficacy of singing versus exercise training: do the data really support the authors' conclusions? Eur Respir J 2022;59:2102857. https://doi.org/10.1183/13993003.02857-2021

11 Gick ML, Nicol JJ. Singing for respiratory health: theory, evidence and challenges. Health Promot Int 2016;31:725-734. https://doi.org/10.1093/heapro/dav013

12 Lord VM, Cave P, Hume VJ, et al. Singing teaching as a therapy for chronic respiratory disease - a randomised controlled trial and qualitative evaluation. BMC Pulm Med 2010;10:41. https://doi.org/10.1186/1471-2466-10-41

13 McNamara RJ, Epsley C, Coren E, et al. Singing for adults with chronic obstructive pulmonary disease (COPD). Cochrane Database Syst Rev 2017;12:CD012296. https://doi.org/10.1002/14651858.CD012296.pub2

14 Kjærgaard J, Juhl CB, Lange P, et al. Adherence to early pulmonary rehabilitation after COPD exacerbation and risk of hospital readmission: a secondary analysis of the COPD-EXA-REHAB study. BMJ Open Respir Res 2020;7:e000582. https://doi.org/10.1183/23120541.00173-201