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CASE REPORT

Impact of pulmonary rehabilitation pre- and post-lung transplantation in a child with cystic fibrosis: a case report

Impacto da reabilitação pulmonar pré e pós–transplante de pulmão em paciente pediátrico com fibrose cística: relato de caso

Impacto de la rehabilitación pulmonar en el pretrasplante y en el postrasplante pulmonar en paciente pediátrico con fibrosis quística: un reporte de caso

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ABSTRACT | Cystic Fibrosis (CF) is a genetic disease that reduces quality of life. Lung transplantation (LTx) is a strategy for end-stage lung disease treatment in CF. Pulmonary rehabilitation (PR) in LTx is effective, however, only one study has determined its effectiveness in children, and most studies have not included CF exclusively. Thus, reports showing components for PR protocols and outcomes not considered in previous studies of PR in LTx due to CF in children are still needed. To report this case, written informed assent and consent of patient and parent were obtained. Ethical Requirement was formally waived by the institution. A 12-year-old patient with CF was referred to PR due to LTx. A general and respiratory training was conducted daily for six months (pre) and two years (post) the transplantation, with the parents' full support. General training included treadmill and cycle ergometer use and upper limbs exercises. Respiratory protocol included inspiratory training and respiratory physical therapy. We observed improvements in pulmonary function, exercise capacity, inspiratory muscle strength, and quality of life, including school functioning, with progress maintenance after 2.5 years of continuous intervention. This case presents a PR protocol pre- and post-LTx with good long-term results. These components for treatment protocols and outcomes may be useful to consider in clinical interventions or future investigations.

Keywords | Lung Transplantation; Rehabilitation; Cystic Fibrosis; Child; Case Reports.

RESUMO | Fibrose cística (FC) é uma doença genética que reduz a qualidade de vida. O transplante pulmonar (LTx) é uma estratégia para o tratamento de doenças pulmonares em fase terminal na FC. A reabilitação pulmonar (PR) no LTx é eficaz, porém apenas um estudo determinou sua eficácia em crianças, e outros estudos não incluíram a FC exclusivamente. Portanto, relatórios que mostram componentes para protocolos e resultados de RP, não considerados em estudos anteriores de RP em LTx devido à FC em crianças, ainda são necessários. Assim, descreve-se o caso de um paciente de 12 anos com FC encaminhado para RP devido à LTx. Foi obtido o consentimento informado e por escrito do paciente e dos pais. O requisito ético foi formalmente renunciado pela instituição. Um treinamento geral e respiratório foi realizado por seis meses antes e dois anos após a LTx, diariamente, com total apoio dos pais. O treinamento geral incluiu esteira, cicloergômetro e exercícios para

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os membros superiores. O protocolo respiratório incorporou treinamento inspiratório e fisioterapia respiratória. Foram observadas melhorias na função pulmonar, capacidade de exercício, força muscular inspiratória e qualidade de vida, incluindo o funcionamento escolar, com manutenção dos avanços após 2,5 anos de intervenção contínua. O caso apresenta um protocolo de RP pré e pós-LTx com bons resultados a longo prazo. Os componentes para protocolos de tratamento e resultados encontrados podem ser úteis para intervenções clínicas ou investigações futuras.

Descritores | Transplante de Pulmão; Reabilitação; Fibrose Cística; Criança; Relatos de Casos.

RESUMEN | La fibrosis quística (FQ) es una enfermedad genética que reduce la calidad de vida de los afectados. El trasplante de pulmón (LTx) es una estrategia para el tratamiento de enfermedades pulmonares en fase terminal en FQ. La rehabilitación pulmonar (RP) en el LTx es eficaz, aunque solo un estudio evaluó su eficacia en niños, y otros estudios no han tratado exclusivamente de FQ. Por lo tanto, se necesitan informes que contengan elementos de los protocolos y de los resultados de RP y que no habían sido considerados en los estudios anteriores sobre RP en LTx por FQ pediátrica. Así se describe el caso del paciente de 12 años de edad con FQ que fue remitido a RP debido a LTx. Para ello, se obtuvieron el consentimiento informado y el consentimiento por escrito del paciente y sus padres. La institución renunció formalmente a los requisitos éticos. Se realizó entrenamiento general y respiratorio durante seis meses antes de LTx y dos años después de LTx, diariamente, con apoyo de los padres. El entrenamiento general incluyó cinta de correr, cicloergómetro y ejercicios para extremidades superiores. El protocolo respiratorio incluía entrenamiento inspiratorio y fisioterapia respiratoria. Se observaron mejorías en la función pulmonar, capacidad de ejercicio, fuerza muscular inspiratoria y calidad de vida del participante, incluida en la limitación de actividades, con mantenimiento de los logros tras 2,5 años de la intervención continua. El caso del estudio presentó un protocolo de RP pre- y post-LTx con buenos resultados a largo plazo. Estos elementos de los protocolos de tratamiento y los resultados encontrados pueden ser útiles en las intervenciones clínicas o en futuras investigaciones.

Palabras clave | Trasplante de Pulmón; Rehabilitación; Fibrosis Quística; Niño; Informes de Casos.

INTRODUCTION

Cystic Fibrosis (CF) is a genetic disease that affects several organs and reduces quality of life[1]. The most frequent cause of death is respiratory disease[2], and lung transplantation (LTx) is a strategy for end-stage lung disease[1].

Pulmonary rehabilitation (PR) in LTx increases survival rates, muscle strength, exercise capacity, and quality of life in adults[3,4]. However, only one study has determined its effectiveness in children, and other studies have not included CF exclusively[5]. Furthermore, studies show pre- or post-LTx protocols[4,6], and school functioning and inspiratory training have not been considered as outcome or PR program component, respectively[5]. Thus, case reports that show components for pulmonary rehabilitation protocols and outcomes not considered in previous studies of PR in LTx due to CF in children are still needed. The only case of PR pre- and post-LTx in a CF child with good long-term outcomes is from the Dr. Luis Calvo Mackenna children hospital.

CASE DESCRIPTION

Written informed assent and consent of patient and parent were obtained following the Ethics Committee's recommendation. Ethical Requirement of Research Ethics Committee approval for this project was formally waived by the institution.

A 12-year-old girl with a severe CF phenotype was referred to pre- and post-PR due to LTx. The patient lived with her parents and grandmother. Before LTx, she was absent from school for one year due to respiratory exacerbations. Physical activities were dropped, and socialization was reduced due to symptoms and exacerbations. She presented progressive and rapid impairment with infection by resistant *S. aureus* in 2009 and *A. xylosoxidans* in 2012, and an onset of supplementary oxygen and non-invasive ventilation in 2014. In 2016, she underwent several hospitalizations with ICU admission. In 2017, she was registered on the national waiting list for LTx, and six months before LTx, the PR program was started.

In pre-LTx and pre-PR evaluation, underweight and severe impairment in pulmonary function, exercise capacity, inspiratory muscle strength, and quality of life were observed, with the highest impairments in physical and school functioning (Table 1).

A pre-LTx PR protocol was designed, which included general and respiratory training. The patient performed outpatient training twice per week, and home-based training daily, once per day. General training was performed on a treadmill for 20 minutes (three minutes at 50% of the slope and speed obtained in ICT and two minutes at 80%, to complete four series (interval training)), and upper limbs exercises without weights. Perceived exertion was used to set the workload (initial Borg scale 3-4 and final Borg scale 5-6). Respiratory training consisted of inspiratory exercises using the Threshold® device for 15 minutes at 30% MIP, regular respiratory physical therapy using CoughAssist®, Flutter®, or Acapella®, and respiratory physical therapy administered by a physical therapist at the hospital or by the parents at home. After the first three months of rehabilitation, the protocol was modified due to reduced exercise capacity and fatigue. Outpatient training was changed to a daily home-based training, once per day, and monitored by a physical therapist twice per week. The 20-minute exercise on the treadmill was replaced by a 10-minute exercise on a cycle ergometer at 50% of theoretical maximum HR (constant load). The rest of the protocol was not modified. Pre-LTx PR was performed for six months. During treatment, the patient had the support of her close relatives, who adhered to treatment and changed habits and their home structure to improve disease management.

Table 1. Evaluation pre- and post-lung transplantation of a pediatric patient with cystic fibrosis from Chile

Evaluation			
Item	**Pre-Tx/Pre-PR**	**Three months Post-Tx**	**Two years Post-Tx/Post-PR**
BMI	17.9	20.8	21.4
FEV_1	19%	75%	79%
6MWT	210m 30% predicted	600m 94% predicted	605.4m 95% predicted
Basal/final HR (6MWT)	113bpm/150bpm	71bpm/124bpm	103bpm/150bpm
Oximetry (SpO_2) (6MWT)	95% with 1.5L O_2	>95% without supplemental oxygen	>95% without supplemental oxygen
ICT in treadmill	Slope: 10 Speed: 1.8Kph	Slope: 14 Speed: 5.6Kph	-
CLT in cycle ergometer	4min with 3L O_2	30min no O_2	-
MIP	31cmH_2O (25% predicted)	150cmH_2O (122% predicted)	179cmH_2O (146% predicted)
PedsQL			
Physical functioning	32/32	3/32	2/32
Emotional functioning	12/20	5/20	5/20
Social functioning	1/20	0/20	0/20
School functioning	16/20	3/20	5/20

BMI: body mass index; CLT: constant load test; FEV_1: forced expiratory volume in the first second; HR: heat rate; ICT: incremental cardiopulmonary test; MIP: maximal inspiratory pressure; 6MWT: 6-minute walk test; PedsQL: Pediatric Quality of Life Inventory.

In 2018, bilateral lobar transplantation was performed, with good clinical results. Two weeks after LTx, post-LTx PR was started. Training was home-based and performed daily. General training was performed twice per day and consisted of 10-minute exercises on a cycle ergometer at 80% of theoretical maximum HR (Figure 1). In addition, general training included upper limbs exercises without weights for 10 minutes, in which elbow flexion and extension were performed in three sets of 20 repetitions each. Respiratory training was performed once per day and consisted of inspiratory exercises using the Threshold® device for 15 minutes at 30% MIP, with three sets of three minutes of exercise and two minutes of rest. Perceived exertion until level 6 of the Borg scale was used to set the workload of each component. Regular respiratory physical therapy was performed three times per day, administered by parents at home, and included diaphragmatic re-education, controlled inspiratory debit exercise, and lung volume increase therapy with devices. This protocol was performed for one year, after which the frequency of training was reduced to once per day during the second year. After three months of post-LTx PR, re-evaluation showed recovery in BMI, FEV1, 6MWT, ICT, CLT, MIP, and PedsQL, in which the highest improvements were physical and school functioning (Table 1).

Figure 1. Patient training at home on a cycle ergometer

Post-LTx PR lasted two years, after which the evaluation showed maintenance of progress, with an increase from 75% to 79% in FEV1 and from 150 to 179cmH_2O in MIP, and no hospitalizations (Table 1). The training sessions were performed daily, following the post-LTx protocol, using telerehabilitation by videoconference supervised by a physical therapist twice per week.

DISCUSSION

One study has reported the effects of post-LTx PR in children[5], in which training occurred three times per week for three months, whereas training post-LTx in our case was daily, twice per day. Training 3–6 times per week for 1–3 months has been reported in adults[7,8]. The study included aerobic and resistance training using weights, gross motor activities, and stretching[5]. Intensity of aerobic conditioning was set using the Borg scale[5], however, our case added percentage of theoretical maximum HR. The aforementioned study did not consider respiratory training[5], whereas other studies have included respiratory exercises with no devices[9].

PR has been studied before or after LTx, with similar results[4,6]. However, due to the severe impairment throughout the patient's life with CF, incorporation of a pre- and post-LTx protocol could be a useful treatment strategy, whose effectiveness should be determined.

Inspiratory muscle training pre-LTx has been reported in one study, which observed increased respiratory muscle strength and exercise capacity[10]. Our case included inspiratory muscle training pre- and post-LTx, using the same protocol, with similar results.

This case showed school functioning as one of the most affected dimensions and most susceptible to treatment. Moreover, adherence of caregivers was considered a facilitator to accomplish the goals.

Most studies have determined outcomes immediately after PR completion (three months)[5]. This case reported a two-year PR program with evaluation after two years, which showed improvements in pulmonary function and inspiratory muscle strength. This suggests that long-term protocols must be further investigated.

Limitations are related to the type of study. Besides the consequent bias of a case report, which cannot show a cause-and-effect relationship, the current PR protocol combines general and respiratory training with respiratory physical therapy, which makes it difficult to attribute the effect of intervention to any of these components. Furthermore, modifications had to be included in the protocol due to clinical impairment. However, this report could be useful when considered as a comprehensive treatment strategy with good long-term results.

CONCLUSION

This case showed a PR protocol pre- and post-LTx, which included inspiratory muscle training, respiratory physical therapy, and school functioning evaluation, with good long-term outcomes. Considering that the determination of effectiveness of PR in LTx due to CF needs more investigation, with a lack of clinical guidelines for PR in LTx, this case presents components for treatment protocols and outcomes, which have not been included in previous studies related to the effectiveness of PR in LTx due to CF in children, which may be useful to consider in clinical interventions or future investigations.

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