

Respiratory physiotherapy in patients with Cystic Fibrosis and upper limb deep vein thrombosis

Tomer Israeli¹, Iris Eisenstadt¹, David Shoseyov¹, Shoshana Armoni¹, Alex Gileles-Hillel¹, Galit Cremisi¹, Malena Cohen-Cymerknoh¹, and Oded Breuer¹

¹Hadassah Medical Center

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Abstract

We report physiotherapy management of two patients with severe cystic fibrosis (CF) lung disease and upper limb deep vein thrombosis (DVT). These patients were admitted due to a pulmonary exacerbation. Following peripherally inserted central catheters they were diagnosed with an upper limb DVT. Due to their underlying lung disease, physiotherapy was mandatory for improvement. However, the DVT and anticoagulation treatment raised concerns for pulmonary emboli and hemoptysis. A framework for physiotherapy management in these patients, using a set of precautions and restrictions to maintain airway clearance while minimizing risk for pulmonary emboli and hemoptysis, was established. Using these set of instructions, the patients experienced no major adverse event while maintaining sufficient airway clearance to allow respiratory improvement. These precautions were continued until the upper limb DVTs resolved. To our knowledge there are currently no guidelines nor expert opinions available. Therefore, this framework can help guide physiotherapy management.

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Tomer Israeli BPT^{1,2}, Iris Eisenstadt MPH^{1,2}, David Shoseyov MD^{2,3}, Shoshana Armoni RN², Alex Gileles-Hillel MD^{2,3}, Galit Cremisi MSc¹, Malena Cohen-Cymerknoh MD^{2,3}, Oded Breuer MD^{2,3}

Affiliations

¹Department of Physiotherapy and ²Pediatric Pulmonology Unit and Cystic Fibrosis Center, Hadassah Hebrew University Medical Center, Jerusalem, Israel.

³The Faculty of Medicine, The Hebrew University of Jerusalem, Jerusalem, Israel.

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Corresponding author:

Tomer Israeli.

Department of Physiotherapy, Pediatric Pulmonology Unit and Cystic Fibrosis Center, Hadassah-Hebrew University Medical Center, 91120 Jerusalem, Israel, Tel: 972-2-5845026, Fax: 972-2-6435897, e-mail: sidhrta@hadassah.org.il

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Dear Editor,

Physiotherapy and intravenous antibiotics are the mainstay of treatment of pulmonary exacerbations in patients with cystic fibrosis (CF).¹ Intravenous antibiotics are commonly delivered via peripherally inserted central catheters (PICC lines)¹⁻². A relatively frequent complication of PICC lines are deep vein thrombosis (DVT) with a reported Incidence in CF of $\sim 4.5\%$ ², similar to non-CF patients.³ DVT may lead to life threatening pulmonary emboli (PE) and therefore prevention, early diagnosis and appropriate management are required.⁴

Guidelines for DVT prevention and management are available and mainly discuss the use of anticoagulation⁴ which in patients with CF may increase the risk for hemoptysis, a relative contraindication for physiotherapy¹. Clinical practice guidelines for physiotherapy management of patients with upper limb DVT are unavailable. However, guidelines for lower extremity DVT state that due to the risk for developing PE, “mobility is contraindicated until intervention is initiated to reduce the chance of emboli”.⁵

Thus, in patients with CF and an upper limb DVT it is prudent to remain alert for hemoptysis and to withhold physiotherapy until the risk for PE is reduced. However, withholding physiotherapy during a pulmonary exacerbation, may carry a risk of further respiratory deterioration.

We herein describe our experience with physiotherapy management of two patients with severe CF lung disease, admitted due to a pulmonary exacerbation, in whom a DVT developed adjacent to their PICC line catheter.

Patient A, a 16-year-old male, with DF508/W1282X *CFTR* genotype, suffering from a severe pulmonary disease with a baseline forced expiratory volume in 1 second (FEV1) of 32 %predicted and recurrent pulmonary exacerbations for which a PICC line was inserted. On March 2019, he suffered a severe pulmonary exacerbation and was admitted. On day 17th of his admission, a DVT in the left subclavian vein, adjacent to the PICC line, was identified following complaints of acute arm and chest pain.

Patient B, an 11-year-old female, homozygous for the DF508 *CFTR* mutation, suffering from a severe pulmonary disease, with a baseline FEV1 of 26 % predicted, requiring nocturnal non-invasive ventilation and listed for lung transplantation. On November 2019, she was admitted to the hospital with a life-threatening pulmonary exacerbation for physiotherapy and a prolonged course of intravenous antibiotic treatment via a newly inserted PICC line catheter. During her admission, following complaints of chest pain, she was found to have DVTs in the Rt. Jugular subclavian and brachial axial veins.

For both patients physiotherapy was considered crucial.

Standard physiotherapy care in our hospital for CF patients admitted for pulmonary exacerbations includes: A morning session consisting of exercise combined with airway clearance therapy (ACT), an afternoon exercise session at the gym, and an evening ACT session, with either a physiotherapist or by using a high frequency chest oscillator (vest). ACT, in our center, includes a combination of manual chest physiotherapy, breathing exercises (modified autogenic drainage and modified active cycle breathing techniques) and the use of positive expiratory pressure (PEP) devices.

We conducted a multidisciplinary consultation including a pediatric pulmonologist, a hematologist and senior physiotherapy staff from the CF center and the intensive care units in our hospital. We formed an approach to physiotherapy treatment which we considered both safe and effective (Table 1). The restriction were continued until DVT resolution.

Table 1. Framework for physiotherapy management of patients with CF and upper limb DVT receiving anticoagulation treatment.

Concern	Action
Hemoptysis Pulmonary emboli	Continue surveillance of sputum characteristics Avoid active upper limb movements of the affected side Withhold treatments with High-Frequency Chest Wall Oscillation device Withhold manual techniques on the affected chest
Further respiratory deterioration	Use fixation techniques on the unaffected chest Use positive expiratory pressure devices Continue cardiorespiratory exercises

Thus, management included breathing exercises, gentle fixation techniques and the use of oscillatory positive pressure devices (Aerobika, Trudell medical, Pari O-Pep); Also, patient B’s non-invasive ventilation device and soap bubble games. In addition, mild lower extremity exercises: treadmill, steps and stationary bicycle.

Patient A, following initial respiratory improvement was discharge and continued outpatient treatment. Outpatient physiotherapists were instructed regarding the required management and a senior physiotherapist from our CF center remained in constant communication.

Patient B, suffered several episodes of mild hemoptysis which resolved spontaneously, without requiring treatment modification.

The treatment framework was followed until the DVTs resolved radiologically. Neither patient suffered from PE, both maintained their respiratory condition.

To conclude, we report our experience regarding physiotherapy management in two patients with CF and severe lung disease who were diagnosed with upper limb DVTs.

To our knowledge, there are no existing guidelines nor expert opinion. This is especially relevant in patients with a more severe disease.

We present a framework established following a multidisciplinary discussion. This framework includes a set of simple instructions easily communicable. Adhering to these instructions enabled us to maintain our patients’ respiratory status without any major adverse events.

Since physiotherapy is standard treatment in all patients with CF, and especially during pulmonary exacerbation, and since DVT is not an uncommon complication and poses an increased risk for both pulmonary emboli and hemoptysis: specific guidelines are required for management of patients with CF suffering from upper and lower limb DVTs.

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