

Clinical considerations and outcomes of a patient with Hermansky-Pudlak syndrome pulmonary fibrosis status post lung transplantation in inpatient pulmonary rehabilitation

Sharon Matos

New York University Langone Health, United States

Abstract

Hermansky-Pudlak syndrome (HPS) is a rare disorder characterized by oculocutaneous albinism, bleeding diathesis, granulomatous colitis, and pulmonary fibrosis (PF). Lung transplantation (LT) is the only curative therapy, yet thoracic surgery has been a contraindication due to bleeding risk. The purpose of this case study is to highlight the PT considerations/outcomes of a 48 year old with HPS-PF s/p bilateral LT in Inpatient Rehabilitation (IR). Post LT, pt presented to IR with poor visual acuity and nystagmus, dyspnea on exertion, decreased balance/strength/endurance, dominant chest/apical breathing pattern. Interventions included balance/endurance training, breathing retraining, strengthening, stretching. 10 meter walk test improved from .61 to .73 m/s. Timed up and go improved from 19.81 to 10.7 seconds. There was no change in Dyspnea-12 questionnaire score. University of California, San Diego Shortness of Breath Questionnaire improved from 41 to 42/120. 6MWT improved from not safe to perform to 542ft with a rollator. At evaluation, pt walked 32 ft. At discharge, pt walked 90 ft ind or 542 ft (rollator and supervision). Pt did not have active bleeding post LT. PT considerations include impact of vision on mobility, screening for bleeding, balance screen, and fatigue as a primary limiting factor. Wickerson et al's guidelines for outpatients for LT and American College of Sports Medicine guidelines for COPD were referenced due to the lack of guidelines. Resistance training at 60% 1 repetition max (RM) was trialed and was too difficult for pt. A low-moderate intensity, using Borg scale and 10-12 RM were feasible for exercise prescription leading to improved functional mobility.

Biography:

Sharon Matos completed her doctoral of physical therapy degree from Stony Brook University. She is a senior physical therapist at NYU Langone Health. She is part of the cardiopulmonary strategic planning group, has performed performance improvement projects in diabetes and COPD at NYU Langone Health.

3rd World Physical Medicine and Rehabilitation

Conference; Webinar- June 18-19, 2020

Abstract Citation:

Sharon Matos, Clinical considerations and outcomes of a patient with Hermansky-Pudlak syndrome pulmonary fibrosis status post lung transplantation in inpatient pulmonary rehabilitation, Rehabilitation Health 2020, 3rd World Physical Medicine and Rehabilitation Conference; Webinar- June 18-19, 2020.

(<https://rehabilitation.healthconferences.org/2020>)

