Success story of a case of arthrogryposis multiplex congenita with restrictive lung disease who underwent pulmonary rehabilitation using a robotic patient-guided suspension system

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Abstract
We present a case report of a 31-year-old female with arthrogryposis multiplex congenita who suffered from restrictive lung disease and who underwent pulmonary rehabilitation successfully with a robotic device. This device falls into the category of a patient-guided suspension system. The patient was admitted to the intensive care unit after an episode of pneumonia. In the general ward, she could not be weaned off oxygen and was not able to ambulate more than 3 m with assistance. During the first session of training with the robotic device, she ambulated 162 m. After six sessions of robotic therapy, she was weaned off oxygen and could walk independently. The use of patient-guided suspension systems should be explored in pulmonary rehabilitation.

Keywords
Arthrogryposis, restrictive lung disease, pulmonary rehabilitation, robotic device

Introduction
Patients who have had a bout of severe respiratory infection leading to severe deconditioning and requirement of oxygen support are frequently referred for inpatient rehabilitation. In many instances, the level of participation of the patient depends on how well the patient is able to oxygenate. Rehabilitation may be very limited in these patients with early and profound exercise-induced oxygen desaturation in addition to their dyspnoea and peripheral muscle weakness. This is the case with this patient. In addition, the effects of pulmonary rehabilitation in patients with restrictive lung diseases are not known as well compared with pulmonary rehabilitation in chronic obstructive pulmonary disease (COPD) patients.

Case report
Written consent was obtained from this patient. IRB was not applied as this is a case report and written consent would suffice. The patient is a 31-year-old Indian woman who was born with arthrogryposis multiplex congenita and suffered from contractures of the knees, hypoplastic muscles, severe scoliosis and restrictive lung disease. She underwent release of the knee contractures at the age of 6 years old. When she was in her late teenage years, she underwent spinal instrumentation for severe scoliosis. In her early adulthood, she was community ambulant without aids although her exercise tolerance was low. She needed to take breaks when ambulating in the community. She was working in a travel agency doing reception and office work. After her retrenchment, she seldom went out and became homebound. She was admitted to hospital for dyspnoea as well as infections on several occasions.

In the most recent admission, she was admitted for pneumonia complicated with hypercapnic respiratory
failure requiring intubation and ventilator support at the Intensive Care Unit. When she was in the general ward, she could not be weaned off oxygen and was advised to be discharged with home oxygen. She was only able to ambulate 3 m with oxygen with minimal one-person assist, limited by fatigue, desaturation and tachycardia. She also required external high-frequency chest wall oscillation using an inflatable vest that wraps around the chest also known as VEST (high frequency chest wall oscillation) therapy to help to clear her secretions. She was then referred to inpatient rehabilitation.

In the physical examination, range of motion is full in all limbs and manual muscle testing (MMT) were 4+ on all extremities, except for hip flexors which were 4 bilaterally. MMT remained the same on discharge. Hand grip strength was measured to be 22.6 pounds (normal range: 74.1) on the right hand (dominant) and 24.6 pounds (normal range 68) on the left hand.

On the day after her transfer for inpatient rehabilitation, she was tried on the Andago™, which is a robotic device in the category of patient-guided suspension system (Figure 1). This system allows for partial weight support and over-ground walking at the same time. She was able to ambulate with 7.5 kg partial weight support on both sides as well as oxygen supplementation of 2 L/min. She ambulated a total of 162 m on the first session with multiple breaks. No further session of VEST therapy was needed as she was clearing her secretions better. On session 6, she was able to cover a distance of 325 m with 2 L/min oxygen and 2.5 kg partial weight support on each side. After six sessions of robotic therapy, the patient was able to cover 80 m with a walking stick for normal over-ground walking. Patient however continues further robotic therapy, for about three more sessions, to improve endurance while waiting for oxygen concentrator to be available. The oxygen concentrator was needed for standby use. She was successfully weaned off oxygen 5 days before discharge and was able to walk short distances to the bathroom without a gait aid. She was discharged home after a total of 14 days on inpatient rehabilitation.

**Discussion**

Arthrogryposis multiplex congenita is a rare congenital syndrome that presents at birth. The specific symptoms and findings can differ, with a wide range in severity. Patients can present with non-progressive contractures of various joints, hypoplastic muscles, slender and fragile long bones, central and peripheral nervous system disorders. This patient also presented with restrictive lung disease secondary to severe scoliosis.

The effects of pulmonary rehabilitation in patients with restrictive lung disease are less known compared with patients with COPD. According to Salhi et al., patients with restrictive lung disease responded better after 24 weeks of pulmonary rehabilitation as compared with 12 weeks. There were clinically significant improvements in exercise capacity, muscle force and quality of life. It appeared that patients on oxygen at baseline and those with severe disease should not disqualify from a pulmonary rehabilitation program as they may benefit more. A longer duration of rehabilitation may be beneficial, especially for those with worse symptoms and require a lower exercise intensity at the start. If this patient was discharged in that functional state, it was very likely that she would become bedbound. She was therefore given a trial of inpatient rehabilitation.

Conventional therapy would be at very low intensity at the start, due to early and severe desaturation and dyspnoea. This may lead to further inactivity and deconditioning. As the rehabilitation process could be limited by the poor effort tolerance, a device with partial weight support was chosen to help the patient reduce her load and start her walking. Prior to trying the patient on Andago™, history was taken to exclude past fractures, which may suggest fragile bones. The patient weighed 55 kg. She was offloaded 27% of her body weight by having total partial weight support of 15 kg. Even on the first session, the patient was able to walk a distance of 162 m. That was 54 times more than what she could do with normal walking.

This patient has a height of 1.33 m. The lower height limit for this robotic device is 1.35 m. However, the patient was able to use this system, as the suspension system was able to come down to accommodate her height. This system is also preferred to using the treadmill with the ceiling hoist which is available, but not tried on this patient. This is preferred as the patient is able to select her gait speed, and it allows her to take breaks whenever she needs to catch her breath.

Pulmonary rehabilitation has been shown to be useful in restrictive lung diseases secondary to scoliosis. In a study of pulmonary rehabilitation in children with scoliosis, it was found that it improved the oxygen saturation and forced vital capacity. Moreover, sarcopenia and frailty in chronic respiratory disease is increasingly recognised. This patient even at the age of 31 suffered from frailty even before this hospitalisation episode. She became housebound, suffered from low physical activity, weakness and exhaustion. Frailty is associated with poor outcomes in chronic respiratory disease including falls, hospitalisations and greater levels of disability, and a predictor of mortality. Pulmonary rehabilitation has been shown
Conclusion
In conclusion, patients with restrictive lung disease should undergo pulmonary rehabilitation. The patient-guided suspension system should be explored in selected patients undergoing pulmonary rehabilitation. Its novel use in a young patient with restrictive lung disease had an effect on her recovery. Usually pulmonary rehabilitation in patients with severe and early desaturation and fatigue may be a slow process. However, this patient was successfully weaned off oxygen and achieved independent ambulation after six sessions of robotic therapy. This system is usually used in conditions such as stroke, spinal cord injury, brain injury and general deconditioning. It can also be useful in pulmonary rehabilitation when the patient has severely reduced effort tolerance.

Authors’ contributions
TSS conceived the study. TSS, CAV, EMZ researched literature. TSS wrote the first draft of the manuscript. All authors reviewed and edited the manuscript and approved the final version of manuscript.

Availability of data and materials
Not applicable

Conflict of interest
The authors declare that there are no conflicts of interest.

Ethical approval
None.

Informed consent
Written informed consent was obtained from the patient for their anonymised information to be published in this article.

Funding
The author(s) received no financial support for the research, authorship, and/or publication of this article.

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