DOI: 10.7860/JCDR/2026/81092.23885

Case Report

Physiotherapy Section

Multidisciplinary Rehabilitation of a Patient with Sickle Cell Disease: A Case Report

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ABSTRACT

Chronic haemoglobinopathy Sickle Cell Disease (SCD) is frequently exacerbated by recurrent infections such as aspiration pneumonitis, which may necessitate long-term airway management techniques like tracheostomy. These interventions often result in dysphagia and respiratory complications, thereby requiring a multidisciplinary approach to care. This report describes recurrent aspiration pneumonitis resulting in tracheostomy and ventilator dependence in a 19-year-old male patient with homozygous SCD. The patient underwent a structured multidisciplinary rehabilitation programme that included ENT-guided swallowing therapy and physiotherapy aimed at respiratory and musculoskeletal recovery following several unsuccessful attempts at decannulation. The patient underwent progressive mobilisation, breathing control techniques, chest physiotherapy, and staged oral-motor and swallowing exercises over a period of 4-6 weeks. Functional outcomes improved significantly following these interventions. The Volume-Viscosity Swallow Test (V-VST) showed post-treatment progression, respiratory effort improved (Modified Borg Dyspnoea Scale decreased from 7 to 2-3), and swallowing ability improved from Functional Oral Intake Scale (FOIS) Level 2 to Level 6. The Ryle's tube was removed after the patient was successfully decannulated. In patients with SCD and complex pulmonary and oropharyngeal involvement, this case emphasises the importance of early, coordinated rehabilitation-including physiotherapy and ENT collaboration- in restoring function and improving quality of life.

Keywords: Deglutition disorders, Pneumonitis, Quality of life, Respiration therapy

CASE REPORT

A 19-year-old male, known to have SCD (SS pattern), presented with a history of sickle cell anaemia for three years, and due to which he had episodes of recurrent aspiration pneumonitis. He came to the hospital with tachycardia, deranged Liver Function Test (LFT), raised C-Reactive Protein (CRP) level, and mild pleural effusion. He was admitted directly to the Intensive Care Unit (ICU) due to respiratory distress and repeated aspiration episodes. A tracheostomy was performed to secure the airway, and the patient required prolonged ventilatory support. For nutritional management, a Ryle's tube was inserted [Table/Fig-1]. Initially, antibiotics were given according to the clinical condition. Later on, platelet transfusion, stepped-up antibiotics, and ventilator support -were given to the patient. After prolonged ventilatory support of one month, an initial trial of decannulation was attempted during the hospital stay, but was unsuccessful. After a hospitalisation period of nearly one month, the patient was discharged and referred to the ENT department of the hospital for further evaluation. A second decannulation attempt was made; however, it again failed due to saliva pooling in the oropharynx. The tracheostomy tube was subsequently downsized to a cuffed tube, and the patient was placed on weekly or 10-day follow-up visits at the ENT Outpatient Department (OPD) of the same hospital.

[Table/Fig-1]: Patient is tracheostomised and ventilator dependent due to recurrent pneumonitis.

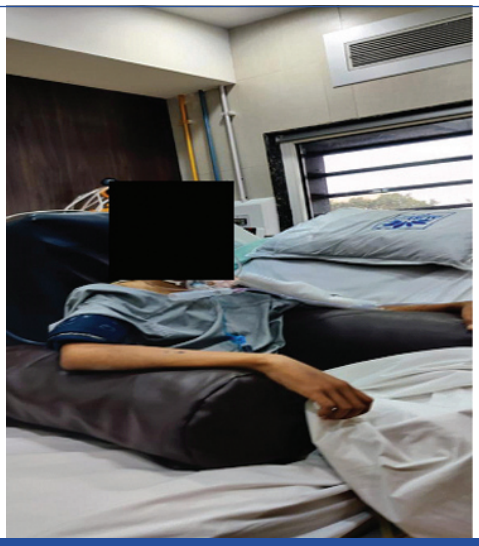
Post-discharge, the patient continued day-care physiotherapy sessions, followed by outpatient follow-up for progressive rehabilitation. The patient has been receiving treatment from the same institution since initial admission. Recurrent vaso-occlusive episodes and chronic hypoxia associated with SCD often lead to neurological complications such as stroke or cranial nerve involvement, which impair swallowing coordination. In addition, pulmonary complications, including acute chest syndrome, chronic lung injury, and pulmonary hypertension, may reduce respiratory reserve and compromise airway protection. These combined factors predispose individuals with SCD to oropharyngeal dysphagia and recurrent aspiration events. Consequently, the tracheostomy tube was downsized to a cuffed tube, and the patient was placed on weekly or 10-day follow-up visits in the same hospital. During this period, he was provided with swallowing therapy and physiotherapy exercises.

Swallowing therapy in phase 1 & 2 therapy, including targeted exercises for oral and pharyngeal musculature, is effective in reducing aspiration risk and improving functional oral intake in tracheostomised patients. Physiotherapy was incorporated to enhance respiratory capacity, maintain airway clearance, and improve overall rehabilitation outcomes. Together, these interventions addressed both the swallowing dysfunction and the reduced pulmonary reserve that predisposed this patient to repeated aspiration events. Phase 3 management home exercise programme. Patient recovery has progressed to assisted ambulation with a walker. He eventually succeeded in swallowing even with the tracheostomy and Ryle's tube in situ. After a successful trial of decuffing and tolerating sips of water, fluids, and semisolid food,

final decannulation was performed. The stoma closed naturally, and the Ryle's tube was removed, marking a significant recovery in the patient's overall condition.

As part of ongoing management, the tracheostomy tube was downsized to a cuffed version, and the patient was enrolled in a structured rehabilitation programme, including weekly or biweekly follow-up assessments.

Clinical findings: Before the evaluation, consent was obtained from the patient. The patient was observed in a seated position with forward head posture, guarded upper body movement and decreased trunk control with pillow positioning [Table/Fig-2]. On auscultation, breath sounds were reduced with crackles present in the lower lobes and decreased chest expansion. There is muscle weakness, especially in the neck flexors and extensors. Decreased core stability and postural control due to prolonged immobility. Fatigue and reduced endurance were present due to prolonged hospital stay. Initially there was a pooling of saliva in the oropharynx, difficulty in swallowing, and oral incoordination. According to the Functional Oral Intake Scale patient's score was 2, indicating tube dependency with minimal or inconsistent oral intake [1,2].



[Table/Fig-2]: Pillow positioning was done to support and stabilise the back and upper body.

Physiotherapy assessment: On postural assessment, the patient presented with a forward head posture and guarded movement due to pain and discomfort in the upper limb, and reduced trunk control due to prolonged immobility. Respiratory assessment revealed abnormal breath sounds, crackles at the base of the lungs, reduced chest expansion bilaterally, altered breathing pattern with shallow breathing, and more use of accessory muscles such as sternocleidomastoid, pectorals [3]. Weak cervical flexors and extensors due to tracheostomy, limited shoulder mobility due to inactivity, reduced trunk stability, and lower limb weakness.

Physiotherapy management: The integrated exercise plan is mentioned in [Table/Fig-3], included swallowing therapy (conducted by a physiotherapist under the guidance of an ENT surgeon), this is phase one therapy (IPD) done for one to four weeks.

Phase two: (IPD + OPD): Four to six weeks is mentioned in [Table/Fig-4].

During this period, a trial was conducted to taper the patients' dependency on the therapist for exercises. A few sessions were conducted in the IPD to observe whether the exercises were being performed correctly, followed by OPD follow-ups to ensure proper continuation of the same.

Phase three: OPD visits + Home Exercise Plan (HEP) (Four to six weeks)

Structured phase 3 management includes OPD visits and HEP is mentioned in [Table/Fig-5] [4-6].

Over the course of four to six weeks, the patient showed marked improvement in nutritional intake and physical conditioning, as well as coordination of the swallowing mechanism. Notably, he was able to tolerate oral intake even with the tracheostomy and Ryle's tube in situ. After a successful trial of decuffing and demonstrating safe swallowing of liquids and semisolids without signs of aspiration, final decannulation was performed.

The tracheostomy stoma healed spontaneously, and the Ryle's tube was removed. These interventions collectively contributed to a significant and sustained recovery in his airway protection, nutritional status, and overall quality of life. Over four to six weeks, the patient showed gradual improvement in his nutrition, physique, and both oral and oesophageal swallowing mechanisms [Table/Fig-6].

Outcome measures: Pre and post treatment outcome measures were recorded and in post-treatment it showed better outcome as mentioned in [Table/Fig-7] [1,7-9].

S. No.	Intervention	Description	Dosage
1	Oral motor exercises	Strengthening of labial, lingual, and buccal muscles to enhance bolus control and the oral preparatory phase. Procedure of treatment: lip press, lip press on tongue depressor, puff cheeks, lip retractions, lip protrusion, blowing exercises etc. It was one to one therapist to patient and either bedside or supine position	5 reps × 2 sets thrice a day
2	Controlled swallowing trials	Gradual introduction of swallowing exercises using saliva to initiate safe swallowing reflexes	5 reps × 2 sets twice a day
3	Chest physiotherapy	Techniques including percussion, vibration, and postural drainage to clear pulmonary secretions and improve lung compliance	5 reps × 2 sets thrice a day
4	Diaphragmatic and pursed-lip breathing	Breathing exercises to optimise ventilation and reduce respiratory effort	5 reps × 2 sets thrice a day
5	Neck and shoulder mobility exercises	Exercises to alleviate stiffness from prolonged bed rest and tracheostomy-related postural limitations	5 reps × 2 sets thrice a day
6	Gradual Mobilisation	Progression from bed-level activity to assisted ambulation to improve endurance and muscle tone.	5 reps × 2 sets twice a day

[Table/Fig-3]: Phase 1 management in an IPD patient with Sickle Cell Disease (SCD) and aspiration pneumonitis.

S. No.	Intervention	Description	Dosage
1	Oral motor exercises	Targeted strengthening of labial, lingual, and buccal musculature to enhance bolus control and the oral preparatory phase of swallowing	10 reps × 3 sets thrice a day
2	Controlled swallowing trials	Gradual introduction of swallowing exercises with sips of water and thickened liquids to initiate safe swallowing Controlled swallowing trials	10 reps × 3 sets twice a day as tolerated by patient
3	Chest physiotherapy	Includes percussion, vibration, and postural drainage to manage pulmonary secretions and improve lung compliance	10 reps × 3 sets thrice a day
4	Diaphragmatic and pursed-lip breathing	Increases posterior pharyngeal wall contraction to help clear pooled secretions and improve bolus propulsion	10 reps × 3 sets twice a day
5	Neck and shoulder mobility exercises	Enhances laryngeal elevation and swallowing coordination to aid airway protection during the swallow	10 reps × 3 sets twice a day
6	Gradual Mobilisation	Strengthens the posterior pharyngeal wall by restraining tongue movement during dry swallows	10 reps × 3 sets twice a day

[Table/Fig-4]: Phase 2 management in an IPD and OPD in patient with Sickle Cell Disease (SCD) and aspiration pneumonitis.

S. No.	Intervention	Description	Dosage
1.	Oral motor exercises	Targeted strengthening of labial, lingual, and buccal musculature to enhance bolus control and increase the strength of muscles.	12 reps × 3 sets twice a day
2.	Controlled swallowing trials	Gradual introduction of swallowing exercises with sips of water, thickened liquids and semi solids.	12 reps × 3 sets as tolerated by patient
3.	Chest physiotherapy	Breathing Exercises, Breath control Exercises, postural drainage to manage residual pulmonary secretions and improve lung compliance.	12 reps × 3 sets twice a day
4.	Effortful swallow manoeuvre	To increase posterior pharyngeal wall contraction and assist in clearing pooled secretions.	12 reps ×3 sets twice a day
5.	Mendelsohn manoeuvre	Used to improve laryngeal elevation and coordination during the swallow, aiding in airway protection [5].	12 reps × 3 sets twice a day
6.	Masako manoeuvre (tongue-hold)	Encouraged posterior pharyngeal wall strengthening by restraining tongue movement during dry swallows [4,6]	12 reps × 3 sets twice a day
7.	Jaw thrust	To address issues with the Upper Oesophageal Sphincter (UES) opening and hyoid elevation. It helps to open the airway and improve swallowing function by lifting the tongue away from the posterior pharynx	12 reps × 3 sets twice a day
8.	Dynamic shaker	Strengthening the muscles in the neck and upper oesophagus. The exercise helps to improve the opening of the Upper Oesophageal Sphincter (UES), a muscle at the top of the oesophagus, which is crucial for allowing food and liquids to pass into the oesophagus	12 reps × 3 sets twice a day
9.	Strengthening exercises	UL strengthening exercises with 0.5 kg of weight cuff LL strengthening exercises with 0.5kg of weight cuff and assisted ambulation with a walker	12 reps × 3 sets twice a day

[Table/Fig-5]: Phase 3 management in an OPD and home exercise programme in a patient with Sickle Cell Disease (SCD) and aspiration pneumonitis.

[Table/Fig-6]: Phase 3 management home exercise programme. Patient recovery has progressed to assisted ambulation with a walker.

To ensure continuity of therapy, maintenance of airway safety, and gradual improvement in oral motor functions during recovery, the OPD follow ups were initially scheduled for every 5th day, then weekly, and later every ten days, allowing progress to be monitored and planned. The patient was called for assessment to check for any deterioration in recovery, as the exercises were being performed independently at home. During each OPD session, all exercises were demonstrated and performed under the supervision of the therapist and ENT specialist. Exercises performed successfully were modified for progression, whereas exercises that could not be performed adequately were repeated more frequently with closer monitoring. All exercises were provided in the form of charts and instructional videos.

Outcomes	Pre treatment	Post treatment
Numerical Pain Rating Scale (NPRS) [1]	7	3
Functional Oral Intake Scale (FOIS) [7]	Level 2 (Tube dependency with minimal/inconsistent oral intake)	Level 6 (total oral diet with multiple consistency without special preparation but with specific food limitations)
Volume- Viscosity Swallow Test (V-VST) [8]	Thin liquids unsafe (cough, wet voice, desaturation). Nectar: coughing, multiple swallows. Pudding: residue present.	Thin and nectar liquids safe, no cough, no desaturation. Pudding tolerated with minimal residue.
Modified Borg Dyspnoea Scale [9]	6-7 Severe breathlessness	2-3 Mild to moderate breathlessness

[Table/Fig-7]: Pre and post-treatment outcome measures [1,7-9].

DISCUSSION

The SCD is associated with increased vulnerability to infections, which substantially elevates morbidity and mortality [10]. In India, the sickle cell gene is more prevalent among tribal populations in Maharashtra's northern Satpura ranges, the Vidarbha region, and select districts of Marathwada, where the carrier rate ranges from 0-35% [11]. Nationally, the prevalence varies from 5-34% among different tribal groups, with higher rates reported in Gujarat, Maharashtra, Chhattisgarh, Madhya Pradesh, Odisha, Andhra Pradesh, and Tamil Nadu [12].

The multisystem involvement of SCD, understanding the physiology of swallowing, is fundamental for effective clinical management. Swallowing is a complex neuromuscular process that ensures the safe propulsion of a bolus from the oral cavity to the oesophagus [13]. In addition to respiratory complications, patients with SCD are susceptible to musculoskeletal disorders, including osteomyelitis, osteonecrosis, osteopenia, and ankylosis. Avascular Necrosis (AVN) results from recurrent marrow infarctions, leading to sclerosis and alterations in bone architecture [14].

In the present case patient complains of recurrent pneumonitis and failed decannulation were attributed to persistent oropharyngeal dysfunction and poor airway clearance. A comprehensive rehabilitation approach was implemented integrating ENT- guided swallowing therapy and physiotherapy. The protocol was divided into three progressive phases. Initial therapy emphasised passive mobilisation and oral-motor retraining to restore neuromuscular control. Pulmonary complications are a major cause of morbidity in individuals with SCD and include pneumonia, acute chest syndrome, and aspiration-related lung injury [15]. Concurrently, chest physiotherapy supported airway clearance and normalisation of breathing patterns. Pulmonary complications, including aspiration pneumonitis, are common in SCD and contribute significantly to morbidity and mortality, particularly when early recognition and intervention are delayed [16].

Progression to advanced rehabilitation included Shaker and jaw-thrust exercises to facilitate UES opening and active swallowing manoeuvres, such as the Mendelsohn and Masako techniques, to enhance laryngeal elevation and coordination. Significant clinical improvement was demonstrated by the patient's transition from tube-dependent feeding to an unrestricted oral diet with minor limitations. Functional independence gains were reflected in the improvement in FOIS from level 2 to level 6, along with better Modified Borg Scale and Swallowing Quality of Life (SWAL-QOL) scores. These findings support the essential role of physiotherapy in reducing respiratory-related morbidity and improving overall swallowing function in medically complex SCD cases.

Physiotherapy goals in SCD primarily include pain management, prevention of respiratory complications, and improvement in functional capacity and quality of life [17]. In a recent study, it was

reported that both the Shaker exercise and the Masako manoeuvre improved swallowing in post-stroke dysphagia; however, the Shaker exercise produced noticeably greater improvements in Eating Assessment Tool-10 (EAT-10) scores and overall swallowing function. These findings support the use of exercises such as the Shaker manoeuvre in patients with reduced laryngeal elevation and associated impairment of UES opening and airway protection [18].

A recent case report on a child with SCD and COVID-19-induced acute chest syndrome highlights the importance of early physiotherapy in supporting respiratory recovery. To improve ventilation, reduce secretion load, and facilitate successful weaning from mechanical support, chest physical, breathing exercises, and airway-clearance techniques were crucial.

The case report by Raza M et al., demonstrated that early, targeted physical therapy plays a crucial role in minimising pulmonary complications and enhancing recovery in children with SCD and severe respiratory involvement. These findings provide external evidence supporting the role of rehabilitation strategies in medically complex SCD, independent of the observations in the present case [19].

Importantly, three key components contributed to the successful outcome: 1) early rehabilitation intervention that prevented deconditioning; 2) targeted exercises addressing both swallowing and pulmonary functions; and 3) interdisciplinary coordination that ensured individualised progression and safety monitoring. Although musculoskeletal sequelae in SCD have been well documented, pulmonary and neuromuscular complications- particularly those resulting from tracheostomy-related dysphagia- remain under recognised within rehabilitation practice. This case highlights the need for comprehensive, patient-centred treatment strategies that incorporate respiratory, nutritional, emotional, and functional considerations.

However, as a single case report, the findings have limitations and cannot be generalised to all SCD patients with tracheostomy and dysphagia. Differences in disease severity, pulmonary involvement, and access to multidisciplinary rehabilitation may affect outcomes. Objective instrumental assessments, like video fluoroscopy or fiberoptic endoscopic evaluation of swallowing, were not conducted and could have provided additional confirmation of physiological recovery.

CONCLUSION(S)

In the comprehensive management of patients with SCD complicated by tracheostomy and dysphagia, this case emphasises the vital role of multidisciplinary rehabilitation, including physiotherapy and swallowing therapy. In addition to treating respiratory insufficiency and musculoskeletal deconditioning, early intervention enhanced swallowing safety. Targeted physiotherapy techniques were incorporated to maximise airway clearance, lower the risk of aspiration, and restore functional independence. In complex SCD presentations, such comprehensive care is crucial for minimising complications, shortening hospital stays, and promoting long-term recovery.

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PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: Sep 09, 2025
- Manual Googling: Apr 10, 2026
- iThenticate Software: Apr 13, 2026 (2%)

ETYMOLOGY: Author Origin

EMENDATIONS: 8

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Jun 03, 2025**
Date of Peer Review: **Sep 27, 2025**
Date of Acceptance: **Apr 16, 2026**
Date of Publishing: **Jul 01, 2026**