VOLUME - 10, ISSUE - 12, DECEMBER - 2021 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

Original Research Paper | General Medicine

MECHANICAL VENTILATION FOR INTERSTITIAL LUNG DISEASE BY SYSTEMIC SCLEROSIS WITH CREST SYNDROME. PRESENTATION OF A CLINICAL CASE

Katherine Esparza Maquilón*	Medical Resident ICU At "Hospital General IESS Ibarra – Ecuador. *Corresponding Author
Antonio Miguel Ornes Rodriguez	Intensivist Doctor At "Hospital General IESS Ibarra – Ecuador
Diana Mercedes Bombón Salazar	Medical Resident ICU At "Hospital General IESS Ibarra – Ecuador
Daniela Macarena Mediavilla Paredes	Medical Resident ICU At "Hospital General IESS Ibarra – Ecuador
Luis Gustavo Mediavilla Sevilla	Medical Resident "IME /Ibarra – Ecuador/
Pablo Gabriel Cevallos Morejón	Medical Resident ICU At "Hospital General IESS Ibarra – Ecuador
Zuleira Vanessa Torres Jaramillo	Medical Resident At "Hospital General IESS Ibarra – Ecuador
Ligia Elena Echeverría Serrano	Medical Resident At "Hospital Gineco Obstetrico Nueva Aurora" / Quito – Ecuador

ABSTRACT

INTRODUCTION. Interstitial lung disease (ILD) with acute respiratory failure needs ventilatory support poorly documented. One of the interstitial diseases known is the Systemic sclerosis, its advanced stages develop CREST syndrome. Faverio P, et al. (2016) suggested do not close the door to these patients and open the correct protocol, criticizing the little value that the scientific community concede to invasive mechanical ventilation (IMV).

CASE REPORT. 85-year-old male is internalized in critical care unit by pneumonia, the complementary evaluation shows a systemic sclerosis disease with CREST syndrome and it is confirmed by elevation of anti-centromere antibody and positive skin biopsy. Tomography highlights pneumonic consolidation plus interstitial lung involvement and echocardiography reveals pulmonary hypertension. The management is done with IMV, keeping the goal of driving pressure less than 15 as lung protection, recovering respiratory function in 3 weeks. Discussion. The evidence is too insufficient to establish the best decision on IMV to the management of ILD.

KEYWORDS : Interstitial lung disease (IDL), invasive mechanical ventilation (IMV), calcinosis, Raynaud phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia (CREST), pulmonary hypertension (PH).

INTRODUCTION

Interstitial lung disease (ILD) with acute respiratory failure needs an intensive management and ventilatory support poorly documented. One of the interstitial diseases known is the Systemic sclerosis, it is an infrequent disease with a global prevalence of 1 per 10,000 people and 80% involve the lung, complicating up to 25% with ILD and almost 15% with pulmonary hypertension (PH), advanced stages develop CREST syndrome, it is very infrequent situation, characterized by Calcinosis, Raynaud's phenomenon, Esophageal dysmotility, sclerodactyly, and Telangiectasia. (4-7)

Faverio P, et al. (2016) suggested do not close the door to these patients and open the correct protocol, criticizing the little value that the scientific community concede to invasive mechanical ventilation (IMV) in ILD. Non-invasive ventilation provides benefits but delays the opportunity to offer IMV at the right time. (6)

CASE REPORT.

85-year-old male with heart failure NYHA II, hypothyroidism, progressive dyspnea and immunosuppressive treatment with prednisone due to generalized pruritus, excoriations and joint stiffness without diagnosis. This patient is internalized in critical care unit by pneumonia FINE V and CURB 3, the complementary evaluation shows a systemic sclerosis disease by CREST syndrome (Figure 1) and it is confirmed by elevation of anti-centromere antibody and positive skin biopsy.

Pulmonary tomography (Figure 2) highlights right basal pneumonic consolidation and massive bilateral pleural effusion that suppress the generalized reticular pattern shown in interstitial lung involvement, also reveals the esophagomegaly characteristic of CREST syndrome.

Observation on echocardiography reveals dilation of the right atrium and hypertrophy of the right ventricle, showing furthermore pulmonary hypertension at 35mmhg.

The management is done with IMV displaying low compliance and high pulmonary resistance. Bronchodilators administration and drainage of the pleural effusion allow the tolerance of IMV with initial use of 0.8 FIO2, PEEP: 8 and tidal volume: 6cc/kg, keeping the goal of driving pressure less than 15 as protection of the lung. Several therapeutics are taken as Antibiotic and prednisone administration, also mycophenolate is suggested. Respiratory function was recovered in 3 weeks, and discharge was achieved from critical care unit, the survival was reported for more than 1 year.

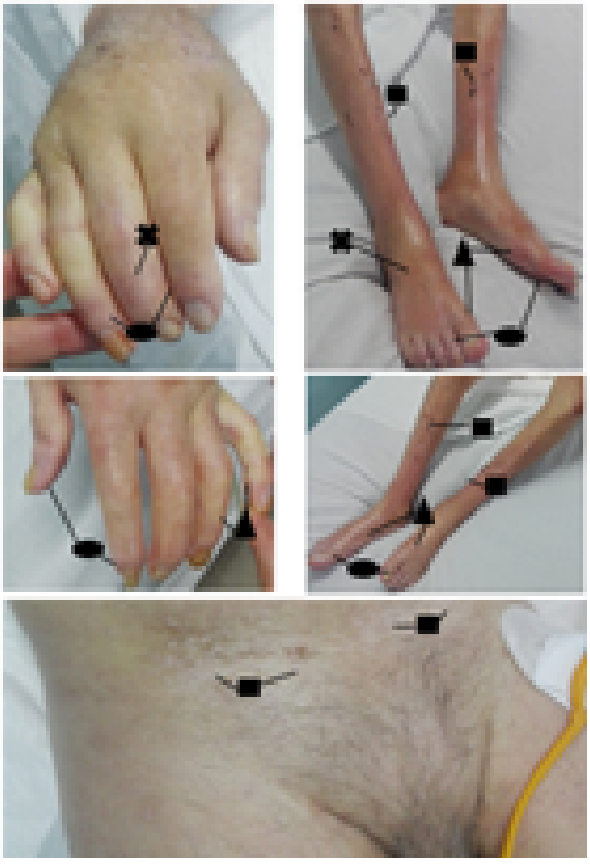

Figure 1: calcinosis and morphea skin (■), raynaud (▲), sclerodactyly (⬬) and telangiectasia (✖).

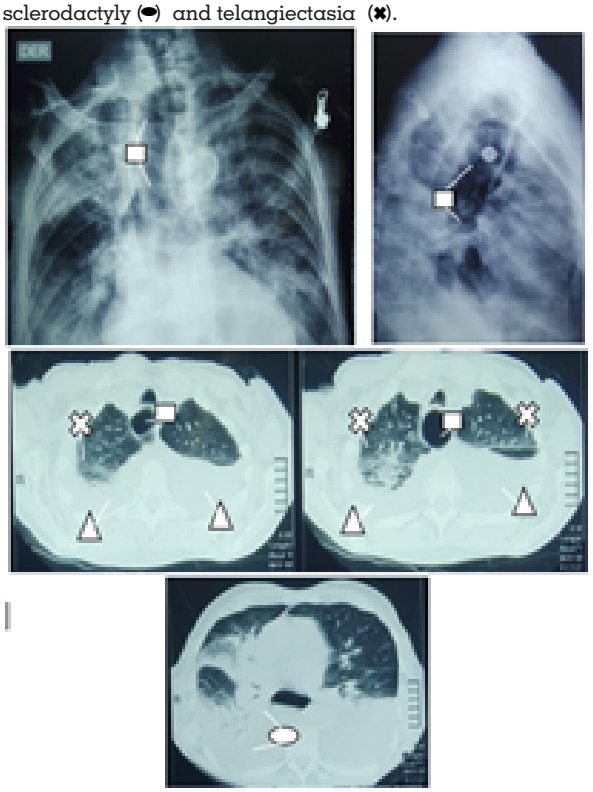

Figure 2: esophagomegaly (□), pleural effusion (△), pneumonic consolidation (⌒) and generalized reticular pattern (⊠).

DISCUSSION.

This case reports discusses the prognostic criteria described for ILD in the studies by Zafrani L, et al. and El-Abdeen A, et al.

These criteria applied in this patient estimate risk of mortality that exceeds 100% in hospitalization and higher risk of mortality per year, they mention the IMV increases the mortality when it is used in ILD. (1-2)

Zafrani L, et al. show patients with IMV having a driving pressure average of 17, which may contribute to higher mortality. (1, 8)

Currently, there is a debate on IMV for the management of acute respiratory distress syndrome (ARDS), which shows histopathological patterns as to ILD in some cases. (1, 9)

IMV techniques have been proposed to reduce pulmonary injury and offer greater diffusion of oxygen from the alveoli, one of the proposals is a driving pressure lower than 15, offering a high PEEP and low tidal volume. Patients with ILD and PH are usually excluded from these studies. (8, 10)

The evidence is too insufficient to establish the best decision on IMV to management of ILD.

In this 85-year-old patient with ILD due to systemic sclerosis with CREST syndrome, early IMV keeping pulmonary protection parameters as driving pressure lower than 15, PEEP 8 and low tidal volume of 6 cc/kg, could be related to the change in the survival prognosis.

REFERENCES:

1. Zafrani L, Lemiale V, Lapidus N, Lorillon G, Schlemmer B, Azoulay E. Acute respiratory failure in critically ill patients with interstitial lung disease. PLoS One. 2014; 9(8): e104897.
2. Ashraf Zin El-Abdeen, Lamiaa H Shaaban, Shereen Farghaly, Yara Y Omar. Prognostic factors and outcome of mechanically ventilated interstitial lung disease patients. Egypt J Bronchol. 2018; 12:226 – 232.
3. Luis Gómez Carrera, Gema Bonilla Hernan. Manifestaciones pulmonares de las enfermedades del colágeno. Arch Bronconeumol. 2013; 49(6):249–260.
4. Solomon JJ, Olson AL, Fischer A, Bull T, Brown KK, Raghu G. Scleroderma lung disease. Eur Respir Rev. 2013; 22(127):6-19.
5. Rotimi Adigun, Anis Hariz. Crest Syndrome Skinmed. 2016; 14(1):43.
6. Faverio P, Pesci A, Esquinas AM. Systemic Sclerosis and Outcome in Intensive Care Unit: Do Not Close the Door, Open the Right Protocol. J Rheumatol. 2016; 43(3): 676-7.
7. Denton CP, Khanna D. Systemic sclerosis. Lancet. 2017; 390(10103):1685-1699.
8. Amato MB, Meade MO, Slutsky AS, Brochard L, Costa EL, Schoenfeld DA, Stewart TE, Briel M, Talmor D, Mercat A, Richard JC, Carvalho CR, Brower RG. Driving pressure and survival in the acute respiratory distress syndrome. N Engl J Med. 2015; 372(8):747-55.
9. Writing Group for the Alveolar Recruitment for Acute Respiratory Distress Syndrome Trial (ART) Investigators, Cavalcanti AB, Suzumura ÉA, Laranjeira LN, Paisani DM, et al. Effect of Lung Recruitment and Titrated Positive End-Expiratory Pressure (PEEP) vs Low PEEP on Mortality in Patients With Acute Respiratory Distress Syndrome: A Randomized Clinical Trial. JAMA. 2017; 318(14):1335-1345.
10. Kacmarek RM, Villar J, Sulemanji D, Montiel R, Ferrando C, et al. Open Lung Approach for the Acute Respiratory Distress Syndrome: A Pilot, Randomized Controlled Trial. Crit Care Med. 2016; 44(1):32-42.