Severe Acute Respiratory Distress Syndrome (ARDS) or Severely Increased Chest Wall Elastance?

Simon Lindner 1, 2, Daniel Dürschmied 1, 2, Ibrahim Akin 1, 2, Simone Britsch 1, 2

1. Department of Cardiology, Angiology, Haemostaseology and Medical Intensive Care, University Medical Center Mannheim, Mannheim, DEU
2. European Center for Angioscience (ECAS) and German Center for Cardiovascular Research (DZHK), Partner Site Heidelberg/Mannheim, Mannheim, DEU

Corresponding author: Simon Lindner, simon.w.lindner@gmx.de

Abstract

Esophageal manometry can be used to calculate transpulmonary pressures and optimize ventilator settings accordingly. We present the case of a 31-year-old male patient with ataxia-telangiectasia (Louis-Bar syndrome) and a BMI of 20 kg/m², admitted to our intensive care unit for coronavirus disease 2019 (COVID-19) pneumonia. The patient soon required mechanical ventilation; however, there was very poor respiratory system compliance. Cholecystitis complicated the clinical course, and veno-venous extracorporeal membrane oxygenation (ECMO) was initiated as gas exchange deteriorated. Esophageal manometry was introduced and revealed severely increased intrathoracic pressure and chest wall elastance.

Introduction

All parts of the respiratory system must be considered when mechanical ventilation is initiated in patients with acute respiratory distress syndrome (ARDS). The overall mechanical characteristics are influenced by both the lung and the chest wall [1]. Increased stiffness of the chest wall that surrounds the lung requires higher pressures to ventilate the lung even when the lung itself shows normal mechanical behavior. The pressure that is required for inflation to 1 liter above resting position is defined as elastance. To assess the individual contribution of lung and chest wall to the overall respiratory system elastance, intrathoracic pressures can be obtained by esophageal manometry. The calculated transpulmonary pressures reveal lung and chest wall compliance and can then be used to individualize ventilator settings [2]. Especially in morbidly obese patients, where pleural pressures are elevated due to more weight on the thorax and abdomen, this method has been useful to guide ventilation [3]. However, high chest wall elastance due to stiffness of the thoracic cage or increased intraabdominal pressures can similarly lead to increased pleural pressures [4], without patients showing obvious clinical characteristics.

Case Presentation

A 31-year-old male patient was admitted to our hospital for coronavirus disease 2019 (COVID-19) pneumonia to receive oxygen support. After three days, he was transferred to our intensive care unit due to ARDS of increasing severity. The patient was suffering from ataxia-telangiectasia (Louis-Bar syndrome), a rare neurodegenerative, autosomal recessive disease, which, in this patient, had resulted in paresis of the torso and arms, as well as plegia of the lower extremities. He had, however, been mobile in an electric wheelchair. He received no medication prior to hospital admission and had a BMI of 20 kg/m². Louis-Bar syndrome has a poor prognosis with a median life expectancy of 19-25 years, although there are cases reaching an age of 40 years [5]. In a case series comprising critically ill patients, three of seven patients survived till intensive care unit discharge. One of seven patients had survived for three years post discharge [6]. However, this patient had exceeded the median life expectancy. Thus, a less severe disease progression was assumed. As no other negative prognostic factors were present, neither mechanical ventilation nor extracorporeal membrane oxygenation (ECMO) was contraindicated.

Non-invasive respiratory support was started for our patient with alternation between high-flow nasal cannula and non-invasive full-face mask ventilation. However, tolerance of non-invasive mask ventilation was poor and could not be improved sufficiently by the administration of sedatives. Thus, mechanical ventilation was initiated. Initial empiric positive end-expiratory pressure (PEEP) was set to 10 cmH₂O, following the ARDS network table [7]. A driving pressure (ΔPaw) of 25 cmH₂O was required to achieve sufficient ventilation, resulting in a tidal volume of 230 ml (4.8 ml/kg predicted body weight (PBW)) and a plateau pressure (ΔPplat) of 55 cmH₂O. To maintain safer Pplat limits, the PEEP was reduced to 7 cmH₂O. Prone positioning was performed two hours after intubation. The patient then developed acute abdomen due to acute abdomen due...
to acute cholecystitis. To safeguard gas exchange during surgery, veno-venous extracorporeal membrane oxygenation (ECMO) was initiated. With a blood flow of 2.5 l/min and a sweep gas flow of 3 l/min, sufficient gas exchange was achieved. PEEP was set to 10 cmH\textsubscript{2}O in an attempt to prevent de-recruitment, and ΔP\textsubscript{aw} was reduced to 20 cmH\textsubscript{2}O to achieve a P\textsubscript{Plat} of 30 cmH\textsubscript{2}O. However, these settings resulted in tidal volumes < 100 ml (< 2.1 ml/kg PBW).

Esophageal pressure (P\textsubscript{es}) monitoring was started (Table 1). End-expiratory P\textsubscript{es} was 22 cmH\textsubscript{2}O, and end-inspiratory P\textsubscript{es} was 26 cmH\textsubscript{2}O. The PEEP was adjusted to 22 cmH\textsubscript{2}O to achieve a transpulmonary end-expiratory pressure (PEEP minus end-expiratory P\textsubscript{es}) of 0 cmH\textsubscript{2}O. In contrast, with a PEEP of 10 cmH\textsubscript{2}O, the transpulmonary end-expiratory pressure had been -12 cmH\textsubscript{2}O, probably resulting in end-expiratory alveolar collapse. Inspiratory pressure was then raised to increase tidal volume to approximately 6 ml/kg PBW. A tidal volume of 300 ml (6.3 ml/kg PBW) was accomplished with a P\textsubscript{Plat} of 41 cmH\textsubscript{2}O and a resulting transpulmonary driving pressure (P\textsubscript{Plat} minus end-inspiratory P\textsubscript{es}) of 15 cmH\textsubscript{2}O.

The measurements were repeated five hours later (Table 2). End-expiratory P\textsubscript{es} was now 19 cmH\textsubscript{2}O, resulting in a transpulmonary end-expiratory pressure of 3 cmH\textsubscript{2}O. End-inspiratory P\textsubscript{es} was 25 cmH\textsubscript{2}O. The PEEP was reduced to 19 cmH\textsubscript{2}O to restore a transpulmonary end-expiratory pressure of 0 cmH\textsubscript{2}O. To maintain an identical tidal volume, ΔP\textsubscript{aw} could also be reduced, resulting in a transpulmonary driving pressure of 9 cmH\textsubscript{2}O.

Gas exchange improved consistently over the following days. After open cholecystectomy, a complicated postoperative course developed, with acute renal failure and the need for continuous renal replacement therapy. These complications resulted in a delay in ECMO discontinuation. The ECMO was eventually discontinued after 13 days.

**Discussion**

The depicted case demonstrates challenges that can arise from special anatomic features in rare diseases, which can be difficult to assess at the bedside. Esophageal manometry can be used to assess chest wall...
elastance [4]. Measurements in this case clearly indicate high chest wall elastance, probably due to higher chest wall stiffness and increased intraabdominal pressure due to cholecystitis. Adjustment of respirator settings according to calculated transpulmonary pressures resulted in improved gas exchange. Our case highlights the potential of transpulmonary pressure assessment to maintain lung-protective ventilator settings during ECMO therapy. In ARDS, higher airway pressures may be required when high chest wall stiffness, obesity, or increased abdominal pressures are present [1]. When conventional airway pressure safety limits are used, initial PEEP and ΔPaw can be inadequately low. The resulting de-recruitment poses a direct threat of further lung injury, and hypoventilation can delay the discontinuation of ECMO. Esophageal manometry and transpulmonary pressure calculations can identify optimal airway pressures and thus justify transgression of conventional limits while maintaining, or even improving, lung-protective ventilation.

Conclusions
Transpulmonary pressure calculations were useful to maintain and even improve lung-protective ventilation under ECMO therapy in this case. Not only morbidly obese patients can show severely elevated esophageal pressures. This patient with a normal BMI showed high esophageal pressure due to a severely increased chest wall elastance. Adjustment of ventilator settings to account for chest wall elastance rapidly improved gas exchange and led to the discontinuation of ECMO.

Additional Information

Disclosures
Human subjects: Consent was obtained or waived by all participants in this study. Conflicts of interest: In compliance with the ICMJE uniform disclosure form, all authors declare the following: Payment/services info: All authors have declared that no financial support was received from any organization for the submitted work. Financial relationships: All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. Other relationships: DD is a member of SFB1425, funded by the Deutsche Forschungsgemeinschaft (DFG, German Research Foundation) – Project #422681845.

References
1. Pelosi P, Cereda M, Foti G, Giacomini M, Pesenti A: Alterations of lung and chest wall mechanics in patients with acute lung injury: effects of positive end-expiratory pressure. Am J Respir Crit Care Med. 1995, 152:531-7. 10.1164/ajrccm.152.2.7635705
2. Talmor D, Sarge T, Malhotra A, et al.: Mechanical ventilation guided by esophageal pressure in acute lung injury. N Engl J Med. 2008, 359:2095-104. 10.1056/NEJMoa0708638
3. Eichler L, Truskowska K, Dupree A, Busch P, Goetz AE, Zölfer C: Intraoperative ventilation of morbidly obese patients guided by transpulmonary pressure. Obes Surg. 2018, 28:122-9. 10.1007/s11695-017-2794-3
4. Gattinoni L, Chiumello D, Carlesso E, Valenza F: Bench-to-bedside review: chest wall elastance in acute lung injury/acute respiratory distress syndrome patients. Crit Care. 2004, 8:350-5. 10.1186/cc2854
5. Crawford TO, Skolasky RL, Fernandez R, Rosquist KJ, Lederer HM: Survival probability in ataxia telangiectasia. Arch Dis Child. 2006, 91:610-11. 10.1136/adc.2006.094268
6. Lockman JL, Iskander AJ, Benbea M, Crawford TO, Lederer HM, McGrath–Morrow S, Easley RR: The critically ill patient with ataxia–telangiectasia: a case series. Pediatr Crit Care Med. 2012, 13:e84-90. 10.1097/PCC.0b013e318219281c
7. Brower RG, Matthay MA, Morris A, Schoenfeld D, Thompson BT, Wheeler A: Ventilation with lower tidal volumes as compared with traditional tidal volumes for acute lung injury and the acute respiratory distress syndrome. N Engl J Med. 2000, 342:1301-8. 10.1056/NEJM200005043421801