Rapid Development of ARDS in VACTERL Patient with Lung Hypoplasia and Pulmonary Hypertension Treated with APRV

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Introduction

VACTERL Syndrome is a rare, genetic disorder accompanied with numerous birth defects involving the vertebrae, anus, heart, trachea, esophagus, kidneys, and limbs. These defects can vary greatly within organ system and the combination of defects differ between individuals. Little to no research is reported on management of these patients with ARDS. Here, we describe a case of a VACTERL patient with pulmonary hypertension and pulmonary hypoplasia who rapidly progressed into ARDS.

Case Presentation

A 32-year-old VACTERL female with past medical history of pulmonary hypertension, mitral regurgitation with mitral clips, atrial fibrillation, left side pulmonary hypoplasia, horseshoe kidney, hemivertebrae, and a BMI of 16 presented to an outlying ED with suspected aspiration pneumonia. Patient’s poor hemodynamics required transfer to our facility’s ICU, where she was started on noradrenaline, vasopressin, steroids, and broad-spectrum antibiotics. Patient progressed to septic shock, severe ARDS, and was intubated within a 12-hour window from ED presentation. During decompression, vasopressor requirement increased to a maximum of 0.03 units of vasopressin, 40 mcg of norepinephrine, 45 mcg of phenylephrine, and 50 ng of velteli. While on VC-AC mode, patients oxygenation deteriorated with high inspiratory pressures over 40 and a P/F ratio of 64.6 on 100% FiO2. P/F ratio after one hour post proning was 51.6. Making ventilatory adjustments while on VC-AC was difficult due to her severe ARDS and her underlying left side pulmonary hypoplasia. Because of her poor oxygenation and high inspiratory pressures, she was switched to PC-APRV with Phigh set at 30, Plow set at 10, Thigh set at 3, and Tlow set at 0.8. Oxygenation improved with a P/F ratio of 102.7 one hour after. Her pressor requirements severely decreased and were eventually titrated down to 10 mcg of norepinephrine, 45 mcg of phenylephrine, and 50 ng of velteli. Stabilizing her hemodynamics allowed for tertiary transfer that same day. Upon tertiary center arrival, patient was presented to an outlying ED with suspected aspiration pneumonia. Patient's primary diagnosis was VACTERL with severe ARDS, let alone VACTERL patients with pulmonary hypertension and left side pulmonary hypoplasia developing ARDS. Some of the literature suggests APRV as a useful tool in the treatment of severe ARDS. The theory is that APRV provides prolonged exposure at high pressures to maximize functioning of available alveoli, decrease the stress associated with alveoli being collapsed and opened continuously (like what you see in standard VC-AC mode), and improve overall oxygenation.

Discussion

Little information has been reported in the literature on patients with VACTERL with severe ARDS, let alone VACTERL patients with pulmonary hypertension and left side pulmonary hypoplasia developing ARDS. Some of the literature suggests APRV as a useful tool in the treatment of severe ARDS. The theory is that APRV provides prolonged exposure at high pressures to maximize functioning of available alveoli, decrease the stress associated with alveoli being collapsed and opened continuously (like what you see in standard VC-AC mode), and improve overall oxygenation.

Our case highlights the complexity of ARDS management, especially in those with congenital malformations leading to a worsened disease state. Our patient was stuck in a never-ending cycle of poor oxygenation and worsening hemodynamics necessitating high dosages of vasopressors leading to worsening oxygenation. APRV became useful in this scenario since we were able to set the pressure parameters and times spent at these specific values. This allowed her oxygenation status to significantly improve and her vasopressor requirement to decrease significantly. This became crucial because she was then deemed stable for tertiary center transport and was later placed on V-V ECMO.

References