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.[1] 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 norepinephrine, 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 decompensation, vasopressor requirement increased to a maximum of 0.03 units of vasopressin, 40 mcg of norepinephrine, 45 mcg of phenylephrine, and 50 ng of veletri. 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 P_{high} 30, P_{low} 10, T_{high} 3, and T_{low} 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 veletri. Stabilizing her hemodynamics allowed for tertiary transfer that same day. Upon tertiary center arrival, patient was placed on V-V ECMO and successfully decannulated after five days.

Figures

A

B

Figure A demonstrates patient's CXR upon ED presentation. Here, opacification is noted within the right lower lung fields, suggestive of possible infiltrate. This was prior to patient acutely decompensating requiring transfer, intubation, and vasopressor therapy. Note also the lack of left lung fields due to pulmonary hypoplasia.
Figure B demonstrates patient's CXR after acute decompensation into septic shock, around 12 hours after ED presentation. Note patient's patchy infiltrate throughout the entire right lung fields. At this point is when providers suspected patient was in severe ARDS.

C

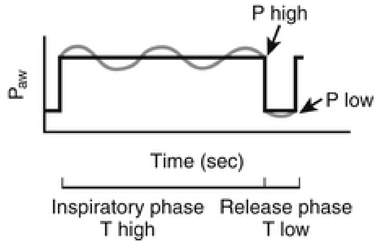

Figure C. This is an example schematic of airway pressure release ventilation (APRV). You set 4 specific parameters: P_{high} (typically set at the P_{plat} or around 30-35cm H_2O), P_{low} (set at 0), T_{high}, and T_{low}. APRV is classified as pressure controlled intermittent mandatory ventilation, where mandatory breaths are given but patient can spontaneously breath during and between these cycles. Mandatory breaths are given at the P_{high} for a duration of T_{high}. There is then a reprieve down to P_{low} for T_{low} duration.[5] The reprieve is generally <1.5 seconds.

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.[2-4]

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.

Conclusion

The goal of this case report is to bring awareness to the management of VACTERL patients and the usefulness of APRV in medically complex individuals with ARDS, especially those that have failed conventional therapy.

References

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