Management of Complete Tracheal Rings in a Patient With Tetralogy of Fallot

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INTRODUCTION

Congenital tracheal anomalies usually present as respiratory distress, stridor, or respiratory tract infections in the first few days of life, but can be asymptomatic in some patients. Long-segment tracheal stenosis can be life-threatening so emergent airway management with mechanical ventilation is necessary. If mechanical ventilation is not possible prior to surgical repair, extracorporeal membrane oxygenation (ECMO) or stenting may be temporizing measures to oxygenate and ventilate.

CASE DESCRIPTION

A 3 day-old full term female was postnatally found to have tetralogy of Fallot with a unicuspid pulmonary valve, severe subpulmonary and pulmonary valve obstruction, ventricular septal defect with right to left shunt, and patent ductus arteriosus (PDA). She was stable on room air on prostaglandins while awaiting surgical repair. On day 3 of life, she was found unresponsive with significant desaturation to 30%. Multiple attempts to intubate the patient yielded a grade I view but 3.0, 2.5, or 2.0 endotracheal tubes were unable to be passed due to an obstruction immediately distal to the vocal cords. The patient remained an easy mask ventilation between each attempt. Transthoracic echo was performed at bedside which showed adequate pulmonary blood flow. The patient was transported to the operating room with mask ventilation for emergent direct laryngoscopy and bronchoscopy.

Otolaryngology was unable to insert a rigid 2.5 mm bronchoscope into the trachea, though a 1.9 mm bronchoscope was able to be passed. Complete tracheal rings were seen immediately distal to the vocal cords down to carina. The surgeon was also unable to intubate with 2.0 endotracheal tube. Tracheostomy was not an option due to complete tracheal rings throughout the entire trachea, therefore the airway was temporized with a laryngeal mask airway. The cardiac surgeon cannulated the patient's right internal jugular vein and carotid artery for veno-arterial ECMO. To prevent fluid overload to the lungs, the arterial cannula was inserted with the tip in the descending aorta to minimize flow through the PDA, and the prostaglandin drip was discontinued.

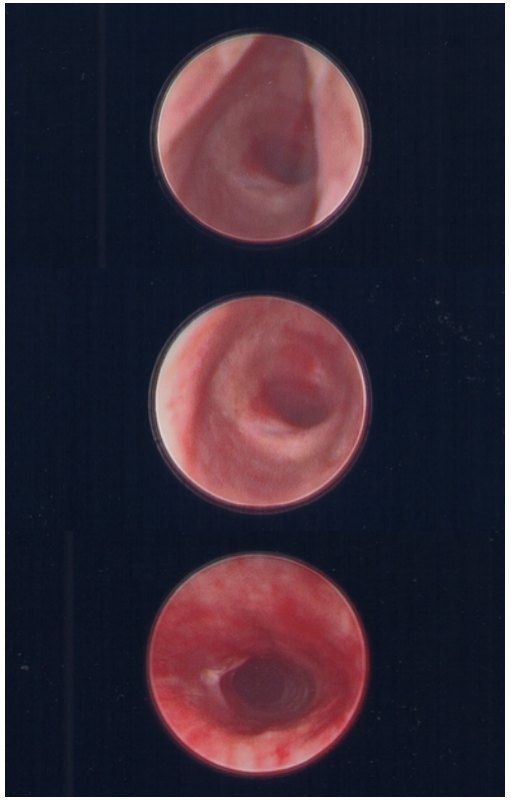

Figure 1: Complete tracheal rings seen on direct laryngoscopy, bronchoscopy

DISCUSSION

Oxygenation and ventilation were particularly concerning in this patient due to the risk of tet spells with hypoxia, hypercarbia, and acidosis. In addition to the risks of ECMO cannulation including infection, bleeding, and neurologic injury, there were other factors to consider for this patient. First, ECMO would require closure of the PDA to decrease flow through the PDA to her lungs. The arterial cannula was inserted into the right common carotid artery and advanced to the descending aorta to decrease blood flow through the PDA. The prostaglandin infusion was turned off after cannulation. Secondly, the patient would require cardiopulmonary bypass for the tetralogy of Fallot repair and slide tracheoplasty. Surgeons note improved surgical exposure and more precise surgical repair without an endotracheal tube obstructing the field.

CONCLUSION

ECMO can be used successfully as a bridge to slide tracheoplasty or other tracheal reconstruction in patients who cannot be conventionally ventilated due to severe or long-segment stenosis.

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