



Conversion to central cannulation following azygous vein cannulation in right congenital diaphragmatic hernia

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ABSTRACT

Right-sided congenital diaphragmatic hernia (R-CDH) can be associated with variations in anatomy that make extracorporeal membrane oxygenation (ECMO) cannulation challenging. We report a case of azygous vein cannulation in a R-CDH patient requiring ECMO. The venous cannula passed directly from the right internal jugular vein into the azygous vein and failed to provide adequate venous return. Cannula repositioning was unsuccessful which led to an urgent planned conversion to central cannulation with a successful outcome. The risk of azygous vein cannulation is increased in patients with R-CDH due to the anatomic distortions that are unique to this subset. Azygous vein cannulation should be considered when inadequate venous return on the ECMO circuit occurs, especially in a R-CDH patient. Central cannulation should be available if the venous cannula cannot be repositioned into the superior vena cava (SVC).

1. Introduction

Congenital diaphragmatic hernia (CDH) is a congenital anomaly affecting 1 in every 2500 to 4000 live births [1]. Early stabilization with recent advances such as high-frequency ventilation, inhaled nitric oxide, and extracorporeal membrane oxygenation (ECMO) prior to surgical intervention have led to improved survival for infants with CDH [2,3]. Right-sided defects occur in approximately 20% of cases of congenital diaphragmatic hernia [3–5]. Infants with R-CDH are at higher risk of failing conventional therapy and needing extracorporeal life support [6]. The right-sided herniation in R-CDH patients can result in a leftward shift of the mediastinum, which during the course of gestation, leads to an obtuse superior vena cava (SVC)-azygous junction, and compression of the inferior vena cava (IVC) can cause dilation of the azygous vein [7]. During ECMO cannulation, the venous cannula may preferentially enter the dilated azygous vein rather than the SVC. Correction of azygous vein cannulation can be challenging in a patient requiring significant ECMO support. Infants with CDH can also have small neck veins making ECMO cannulation further challenging [8,9].

We report a case of azygous vein cannulation in a patient with R-CDH requiring ECMO support. The azygous vein cannulation was recognized in a timely fashion, followed by an attempt to reposition the cannula into the SVC. When the cannula was unable to be repositioned,

the patient was quickly converted to central venous cannulation while maintaining ECMO support via the azygous cannula.

2. Case report

A 3.08-kg baby girl with an undiagnosed right-sided congenital diaphragmatic hernia was born at 39 weeks gestation via repeat Caesarian section. Apgar scores were 6, 6, and 8 at 1, 5, and 10 min, respectively. The patient was intubated for decreased oxygen saturations (SaO₂) in the delivery room and quickly required escalation to high frequency ventilation. SaO₂ was 50–70%. Arterial blood gas showed a pH of 6.8 with a base deficit of –16. Chest x-ray to confirm placement of the endotracheal tube demonstrated a right-sided diaphragmatic hernia containing loops of bowel and a large left pneumothorax. The patient was transferred to our facility for further management. A left sided chest tube was placed for a pneumothorax with improvement in SaO₂ to 90–98%. Inhaled nitric oxide (iNO) was initiated for an increasing pre- and post-ductal SaO₂ gradient with improvement. The patient was relatively stable initially and did not meet ECMO criteria until day of life 9 when there was an acute decompensation. The patient was placed on venoarterial (VA) ECMO. An 8-French arterial catheter was placed in the right carotid artery and a 10-French venous catheter in the right internal jugular vein. ECMO support

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Fig. 1. Cross table lateral x-ray demonstrating posterior angulation of the venous catheter.

was initiated but venous return was severely limited. The ECMO circuit could not flow higher than 40–50 cc/kg/min and multiple fluid boluses were required to maintain flow. Echocardiogram could not definitively identify the cannula within the distal SVC or right atrium. A cross-table lateral x-ray showed the tip to be angled posteriorly (Fig. 1). These findings were suggestive of azygous vein cannulation.

In an attempt to reposition the cannula, the right neck was reopened at the bedside. We had direct ECHO guidance and tried several times to pull back the venous cannula but we weren't able to direct it to the SVC. The use of a guidewire was not possible as the infant was hemodynamically too fragile to be temporarily liberated from ECMO. Also, fluoroscopy was not possible given the procedure was being performed at the bedside. Patient was too unstable to be transferred to the operating room. Given the experience reported by others [7], we decided to place a central venous cannula while maintaining venous flow via the azygous cannula as a bridge to central cannulation.

Median sternotomy was performed and a 12 F right-angled venous cannula was placed directly into the right atrium and was connected to the ECMO circuit after disconnection of the azygous vein cannula. The right heart was tensely dilated upon opening the sternum consistent with suprasystemic pulmonary pressures. After switching to central venous cannulation the ECMO flows went up to 100 cc/kg/min without any difficulty, and the patient's SaO_2 increased to 100%. On day of life 13, an open transabdominal repair of the right congenital diaphragmatic hernia with Gore-Tex patch was performed. The patient was decannulated from ECMO on day of life 22 after 13 days of ECMO. The chest was closed the following day and the patient was extubated on day of life 32. At 3 months of age she underwent gastrostomy tube placement for oral aversion and fundoplication for gastroesophageal reflux. MRI of the brain showed prominent CSF spaces overlying the cerebral convexities, concerning for mild cerebral atrophy but no infarct. She was discharged subsequently, continues to do well, and is currently 9 months old.

3. Discussion

Although ECMO has a distinctive role in the management of R-CDH [10], variations in anatomy in this subset may present cannulation challenges as evidenced by our case of azygous vein cannulation. Liver herniation in R-CDH can lead to IVC compression and azygous vein dilation. Additionally, the intrathoracic liver causes a leftward mediastinal shift that alters the angle at which the azygous vein joins the SVC, making it more obtuse. Both the dilation of azygous vein and the increase in the azygous vein-SVC angle allow a venous catheter to pass directly from the right internal jugular vein to the azygous vein without difficulty during cannulation. Azygous vein cannulation should be suspected in cases of inadequate flow on ECMO in a patient with R-CDH. The diagnosis can be confirmed by absence of the catheter within the SVC or RA on echocardiography. Posterior angulation of the venous cannula on cross-table lateral chest x-ray is also an extremely helpful and a quick means to verify cannula position.

Fisher and colleagues reported a case of azygous vein cannulation with successful venous cannula advancement into the RA under echocardiographic guidance [7]. They reported a second case in which multiple attempts to direct the venous cannula into the RA failed. Support was withdrawn due to prolonged hypoxia and acidosis. Here we present a case of azygous vein cannulation that highlights a potential barrier to successful management of R-CDH and proposes an alternative management strategy. Central cannulation should be considered as an acceptable option especially when adjustment of the venous catheter is not possible or is unsuccessful. Prior knowledge of this potential complication was critical to our surgical planning. Publication of this complication is important so that other centers encountering this situation can add this to their troubleshooting list and alternatives to their armamentarium. Preventable measures such as intraoperative echocardiogram or fluoroscopy can be considered to help guide the surgeon at the time of venous cannulation.

In conclusion, the risk of azygous vein cannulation is increased in patients with R-CDH due to unique anatomic variances in this group. Physicians must recognize the potential for azygous vein cannulation and anticipate alternate strategies such as central cannulation.

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