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Extracorporeal membrane oxygenation support in individuals with thoracic insufficiency

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Abstract

Introduction: Respiratory failure is the leading cause of mortality in individuals with congenital spine and rib deformities. We present a case report of a child with Jeune syndrome surviving respiratory failure using extracorporeal membrane oxygenation (ECMO). We also summarize thoracic insufficiency syndrome cases reported in the Extracorporeal Life Support Organization (ELSO) registry.

Case Report: A two-year-old male with a chest circumference less than a third percentile for age was admitted with influenza pneumonia developing a peak oxygenation index of 103.5. The child survived to baseline pulmonary function after nine days of venous-arterial ECMO support.

Discussion: The ELSO registry contained 27 individuals with a surrogate diagnosis of thoracic insufficiency (0.05%). There was no significant difference in survival to discharge for thoracic insufficiency patients (52%) compared to a previously healthy population supported with ECMO.

Conclusion: ECMO is safe and may be effective in supporting individuals with thoracic insufficiency.

Keywords

thoracic insufficiency; extracorporeal membrane oxygenation; Jeune syndrome; pediatrics; thoracic surgery

Introduction

The definition of thoracic insufficiency syndrome (TI) is the inability of the thorax to support normal respiration and growth.¹ Without surgical intervention, these chest wall deformities decrease lung function, leading to an early death.² The standard of care is

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support with prolonged mechanical ventilation, which has significant complications, including muscle deconditioning, ventilator-associated pneumonia and lung injury.³ In other diseases with acute on chronic exacerbations, physicians are increasingly using extracorporeal membrane oxygenation (ECMO) to improve survival and pulmonary rehabilitation.⁴ The Extracorporeal Life Support Organization (ELSO) has only previously reported on two individuals with TI who survived with ECMO support. The purpose of this report is to determine if ECMO support is safe to offer to individuals with severe TI.

Materials and Methods

The University of Texas Health Science Center at San Antonio (UTHSCSA) Institutional Review Board and ELSO approved a 1990 to 2013 retrospective case review and case report. The case review ICD9 diagnostic codes⁵ used to identify individuals in the ELSO Registry with TI were kyphoscoliosis and scoliosis (737.3, n=8), congenital musculoskeletal deformities of the spine (754.2, n=5), fusion of the spine, congenital (756.15, n=2), other anomalies of the ribs and sternum (756.3, n=5), chondrodystrophy (Jeune syndrome (756.4), n=6) and spondylocostal dysostosis (Jarcho Levin syndrome (756.9), n=1). As a comparison, we collected data from individuals, excluding infants with meconium aspiration (skewed survival rate), who received ECMO support for respiratory failure. For statistical analysis of outcomes from the case review, we used Fischer's exact test due to the small sample sizes, except in rare instances where the sample size was too large, in which case, we used the Chi-square analysis with Yates' correction. Data analysis was performed using Prism 6 for MAC OS X, v 6.0 (GraphPad Software, Inc., San Diego, CA).

Thoracic insufficiency (TI) ECMO case

A two-year-old male with Jeune syndrome and a chest circumference less than a third percentile for age was admitted with acute on chronic respiratory distress due to influenza pneumonia. The surgical history included two vertical expandable prosthetic titanium ribs (VEPTR), placed at eighteen months and two years of age, with one post-operative complication of a partial lobe decortication. The patient presented with a one-week history of congestion, fever and cyanosis. He rapidly progressed to high frequency oscillatory ventilation (HFOV), hypercapnia ($p\text{CO}_2$ 116.9) and an oxygenation index of 103.5. The family consented to ECMO support, consisting of a 14 Fr right carotid arterial and a 16 Fr left internal jugular venous catheter, a Stockert S3 roller pump, an AVECOR I-3500 silicone oxygenator and an arterial filter. Blood flow was titrated to 110-130 ml/kg/hr to provide lung and cardiac rest. The anticoagulation protocol used unfractionated heparin, targeting an activated clotting time (ACT) between 180 and 220. The physician team chose veno-arterial (VA-) over venovenous (VV-) ECMO because the patient had a history of increased pulmonary vascular resistance due to a very small thoracic cage. Throughout his 9-day ECMO course, the only ECMO-related complication was cannulation site bleeding that required bedside surgical exploration. After resolution of pneumonia and tolerating a one-hour trial off ECMO, he was decannulated to conventional support. Forty-one days after admission, he was discharged to home with baseline pulmonary function.

Registry review results

The ELSO registry from 1990 to 2013 contained complete records for 27 individuals supported with ECMO and a surrogate diagnosis of TI. This was 0.05% of all ECMO patients, with an average age of four months and 7.96 days of support. The TI patients received ECMO support for respiratory failure (69.0%), cardiac failure (24.1%) and cardiopulmonary arrest and resuscitation (ECPR, 6.9%). Survival to discharge for respiratory failure with ECMO support with and without TI were 52% and 62% ($p=0.28$), respectively. Table 1 documents the percentage change in survival and hours of ECMO runs. For TI patients, there was no difference in survival between veno-arterial and veno-venous mode of ECMO support; 46.6% and 50.0%, respectively. The pediatric population ($n=14$) in the TI cohort had increased complications. Pulmonary hemorrhage in patients with respiratory failure and TI ($n=10$) were 20% vs. 8%, respectively ($p<0.04$). Patients with TI and cardiac failure ($n=4$) had complications of seizure activity 50% vs. 8.9% ($p<0.04$), hemofiltration requirement 75% vs. 21.2% ($p<0.03$) and myocardial stun 50% vs. 5.1%, ($p<0.01$). Only two pediatric patients with TI received CPR with ECMO (ECPR) and both survived to discharge.

Discussion

Advances in expandable rib and posterior spinal fusion technology allows TI patients to live well into early adulthood.^{6,7} ECMO has been successful in improving outcomes in a wide variety of conditions, including polymicrobial pneumonia, sepsis and multi-organ failure.^{8,9} Based on this success, we performed a case review to determine if our patient with TI would be a good candidate for ECMO support.¹⁰ Although lifesaving, ECMO support is invasive, costly and has a high rate of complications. The only significant complication in TI patients with respiratory failure and ECMO support was pulmonary hemorrhage, probably caused by a combination of the need for systemic anticoagulation and previous thoracic surgeries.¹¹ Complications from ECMO support in TI patients with cardiac failure may arise from the pathophysiology of TI syndrome. Jeune syndrome develops a restrictive thorax, causing pulmonary hypertension leading to chronic cor pulmonale and heart and renal failure.¹² The limitations of our study include low sample size, selection bias in the case examples and an incomplete data set.

Conclusion

ECMO support for patients with TI syndrome may be an effective therapy to improve survival. Future studies should address if earlier use or type of (veno-venous and veno-arterial) ECMO support would improve survival.

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Table 1.

Survival Based on ECLS Support Mode.

Support Mode	Entire Registry		Individuals with Thoracic Insufficiency		p-value*
	# Runs	% Survived	# Runs	% Survived	
NEONATAL (Birth to 30 days)					
Respiratory VA	19372	71.3	5	40	0.29
Respiratory VV	6667	83.5	3	66.7	0.42
Cardiac	26	34.6	1	0	1
PEDIATRIC (30 days to 18 years old)					
Respiratory VA	3367	50.8	6	50	1
Respiratory VV	2153	66.6	4	50	0.61
Cardiac	6392	49.5	4	25	0.62
ADULT (18 years old and up)					
Respiratory VA	710	41.7	1	0	1
Respiratory VV	3118	60.6	1	0	0.39
Cardiac	3362	39.9	2	100	0.16

VA: veno-arterial; VV: venovenous.