JACC: CASE REPORTS VOL. 2, NO. 5, 2020

MINI-FOCUS ISSUE: CONGENITAL HEART DISEASE

ADVANCED

IMAGING VIGNETTE: CLINICAL VIGNETTE

Norwood With Obstructed Total Anomalous Pulmonary Venous Connection and Tracheoesophageal Fistula Repair

Operating Room Delivery

Kyle W. Riggs, MD, Nina M. Price, Alan O'Donnell, MS, Haleh Heydarian, MD, Beth A. Rymeski, MD, David L.S. Morales, MD

ABSTRACT

A baby boy with prenatally diagnosed hypoplastic left heart syndrome variant with obstructed veins was born in the operating room (OR) and underwent emergent Norwood operation and repair of obstructed infra-diaphragmatic total anomalous pulmonary venous connection. Post-operatively, esophageal atresia with tracheoesophageal fistula was identified and repaired on day of life 11. The patient is thriving at 22 months of age. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2020;2:732-3) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

We report the successful surgical management of a patient with total anomalous pulmonary venous connection (TAPVC), double outlet right ventricle with mitral atresia, and tracheoesophageal fistula (TEF) with esophageal atresia through stage II palliation, which has never been published in the literature.

A baby boy delivered at 36 weeks gestation weighing 2.0 kg was prenatally diagnosed with double outlet right ventricle with mitral atresia and infradiaphragmatic obstructed TAPVC via fetal echocardiogram at 26 weeks gestation. Doppler echocardiography showed altered flow pattern and connection from the pulmonary venous (PV) confluence to the portal vein (Video 1). Delivery occurred in a cardiac OR, and postnatal echocardiography confirmed diagnosis. At 1 min of life, the patient developed intermittent apnea and received positive pressure ventilation within 11 min of life. Oxygen saturations were initially in the 30s to 40s, but rose to the 70s. Surgery began within the hour in a neighboring OR. A Norwood with a Blalock-Taussig shunt was performed with TAPVC repair, which he tolerated well.

Failure to pass a nasogastric tube in the intensive care unit raised the suspicion of esophageal atresia with TEF, confirmed by full body X-ray (**Figure 1**). After consulting general pediatric surgery, gastrostomy tube placement occurred on day of life 5. We held feeds and did not extubate prior to esophageal repair. Despite the posterior TAPVC repair's relationship to the TEF, the proximal and distal esophageal pouches were anastomosed without tension.

From the Cincinnati Children's Hospital Medical Center, Heart Institute, Cincinnati, Ohio. The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the *JACC: Case Reports* author instructions page.

Manuscript received March 3, 2020; accepted March 27, 2020.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2020.03.031

At 5 months of age, the patient underwent a bilateral bidirectional Glenn. We believe one should be aggressive in shunted single-ventricle patients undergoing TAPVC to ensure unobstructed PV return because they often develop fibrosis at the anastomosis secondary to their increased Qp:Qs. In this case, a dilated coronary sinus was also pushing on the confluence, which was unroofed into the confluence, and the fibrosis was resected. Oxygen saturations were in the 80s. He was discharged home on post-operative day 5 with normal function and thrives at 22 months of age.

ABBREVIATIONS AND ACRONYMS

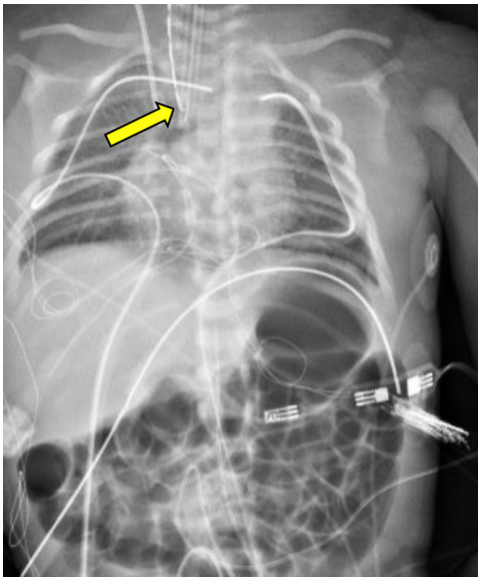
PV = pulmonary venous/vein

TAPVC = total anomalous pulmonary venous connection

TEF = tracheoesophageal fistula

Prenatal diagnosis, paramount to his successful outcome, allowed for enhanced delivery planning, parent education, and staged surgical management. We recommend OR delivery for all patients with obstructed PVs. For patients with hypoplastic left heart syndrome or transposition of the arteries with an intact atrial septum, we advise delivery in a catheterization laboratory. A possible disadvantage of OR delivery is missed diagnoses, including intracranial hemorrhaging, but term babies rarely have significant intraventricular bleeding. Cranial ultrasonography showed no bleeding, and unless massive bleeding was present, emergent cardiac surgery would likely still proceed. Additionally, relying on sump drainage of a proximal esophageal pouch in a shunted single ventricle is a precarious situation secondary to the risk of aspiration, and coexisting congenital heart disease has been found to be an independent predictor of mortality for patients undergoing TEF/EA surgical repair (1). We believe that immediate surgery allowed for pulmonary preservation and rapid recovery, making successful TEF repair possible within 2 weeks of birth.

FIGURE 1 Full-Body Roentgenogram of the 1-Day-Old Patient After Pulmonary Venous Repair and Norwood



Replogle tube **(arrow)** cannot advance past the upper intrathoracic esophagus, suggesting esophageal atresia with tracheoesophageal fistula (Video 1).

ADDRESS FOR CORRESPONDENCE: Ms. Nina M. Price, Cincinnati Children's Hospital Medical Center, Heart Institute, 3333 Burnet Avenue, Cincinnati, Ohio 45299. E-mail: nmprice22@gmail.com.

REFERENCE

1. Diaz LK, Akpek EA, Dinavahi R, Andropoulos DB. Tracheoesophageal fistula and associated congenital heart disease: implications for anesthetic management and survival. Paediatr Anaesth 2005;15:862–9.

KEY WORDS congenital heart defect, double outlet right ventricle, pediatric surgery, pulmonary circulation

APPENDIX For a supplemental video, please see the online version of this paper.