Commentary: Alphabet soup: Successful management of ALCAPA in a neonate with LBW on VA ECMO

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Left ventricular dilation and dysfunction in a small neonate born preterm presents a challenging situation. In an undifferentiated patient, the diagnostic considerations are broad, including metabolic disorders, infections, anatomic lesions, and coronary anomalies. However, Skaff and colleagues present a patient in the current issue of the Journal who developed these symptoms immediately after a successful medical closure of a patent ductus arteriosus. This constellation of symptoms led to the successful diagnosis of an anomalous coronary artery from the pulmonary artery (ALCAPA).

Even with such a diagnosis in mind, it may be difficult to firmly establish ALCAPA in a neonate with low birth weight born premature. Echocardiography in this case demonstrated an abnormal flow pattern in the coronary arteries to establish the diagnosis. Although surgical management is the rule in these children, the surgical team was understandably hesitant to rush to the operating room with a small neonate and ongoing organ dysfunction. In retrospect, however, alternative surgical strategies may have avoided the dilemma posed by worsening organ failure and resultant vasoarterial extracorporeal membrane oxygenation (ECMO) that occurred in this patient.

An historic description of surgical palliation for ALCAPA involving constriction/banding of the pulmonary artery distal to the anomalous coronary origin could have been a viable option in this case. Although extreme, this approach avoids the injury of arresting an already-tenuous myocardium, can be performed via a left thoracotomy without cardiopulmonary bypass, allows a baby with low birth weight time to grow, and doesn’t preclude subsequent anatomic repair.

When the initial medical management is tolerated, as in this case, allowing the patient to grow is tempting. The benefits of a larger patient for operative repair are clear, but they must be carefully balanced with the tenuous nature of the homeostasis that has been achieved. Neonates born premature and with low birth weight are at risk for a myriad of insults over the course of their initial hospitalization, even without ongoing cardiac dysfunction. The clinical decompensation experienced by this patient was undoubtedly a result of the inability of the heart to adapt to the increased demands in a state of stress, resulting in a profound decline for this child.

The management of unrepaired ALCAPA while on vasoarterial ECMO is fraught with physiologic difficulty. The change in pulmonary vascular resistance is unpredictable, and although ECMO will undoubtedly aid in the multiorgan failure, ongoing myocardial ischemia may make postoperative recovery complicated. Nevertheless, the author’s thoughtful approach to instituting ECMO, allowing a period of convalescence, and subsequent repair proved very effective for this child. Recognizing the prolonged insult to the myocardium and allowing for a period of rest after surgical correction paved the way for

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successful recovery of myocardial function in this complicated patient.

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