Seminars in Perinatology ▮ (2017) ▮▮▮–▮▮▮

Available online at www.sciencedirect.com

Seminars in Perinatology

www.seminperinat.com

Extracorporeal membrane oxygenation in congenital heart disease

Kiona Y. Allen, MD[a,*], *Catherine K. Allan, MD*[b], *Lillian Su, MD*[c], *and Mary E. McBride, MD, MEd*[a]

[a]*Department of Pediatrics, Northwestern University Feinberg School of Medicine, Ann & Robert H. Lurie Children's Hospital of Chicago, Box 21, 225 E Chicago Ave, Chicago, IL 60610*
[b]*Department of Cardiology, Harvard Medical School Boston Children's Hospital, Boston, MA*
[c]*Department of Pediatrics, Stanford University School of Medicine, Lucile Packard Children's Hospital, Palo Alto, CA*

ARTICLE INFO

Keywords:
ECMO
Congenital Heart Disease
E-CPR

ABSTRACT

This review article will discuss the indications for and outcomes of neonates with congenital heart disease who receive extracorporeal membrane oxygenation (ECMO) support. Most commonly, ECMO is used as a perioperative bridge to recovery or temporary support for those after cardiac arrest or near arrest in patients with congenital or acquired heart disease. What had historically been considered a contraindication to ECMO, is evolving and more of the sickest and most complicated babies are cared for on ECMO. Given that, it is imperative for aggressive surveillience for long-term morbidity in survivors, particularly neurodevelopmental outcomes.

Background

Extracorporeal support was first described in 1936 by John Gibbon, who developed a roller pump that provided flow and oxygenation via exposure of blood to ambient oxygen.[1] Around that same time, heparin became clinically available, allowing for blood to contact artificial surfaces without clotting. Artificial oxygenation and perfusion were initially created to support patients during open heart surgery, but the method of oxygenation used at the time, a direct oxygen–blood interface, resulted in organ damage and prohibitive hemolysis that kept physicians from being able to maintain patients for more than a few hours.[2] The development of the membrane oxygenator in 1956 allowed extracorporeal support to be adapted to both respiratory and cardiac indications outside of the operating room. Throughout the 1950s and 1960s, the oxygenator evolved through several different iterations, ultimately resulting in a model similar to what is used today. The first human patient was supported for, what at the time was a prolonged run, in 1971.[3]

Extracorporeal membrane oxygenation (ECMO) is well suited to support etiologies of neonatal respiratory failure, as many are self-limited, reversible, and would otherwise require the use of high ventilator settings leading to further lung injury. The first neonatal survivor of acute respiratory failure supported by ECMO was reported in the mid-1970s by Bartlett et al.[4] It was not until the 1980s that ECMO for cardiac indications began to gather momentum. Today, neonatal respiratory failure remains the most common indication for extracorporeal life support (ECLS), but the proportion of cardiac ECMO cases has increased dramatically (Fig.).[5] As newer therapies, such as surfactant, inhaled nitric oxide, and advanced ventilator technologies have become more mainstream, many children with respiratory failure are avoiding the need for ECMO support. In contrast, the use of ECMO for cardiac disease has grown with an ever shrinking number of

*Corresponding author.
E-mail address: KAllen@luriechildrens.org (K.Y. Allen).

https://doi.org/10.1053/j.semperi.2017.12.006
0146-0005/

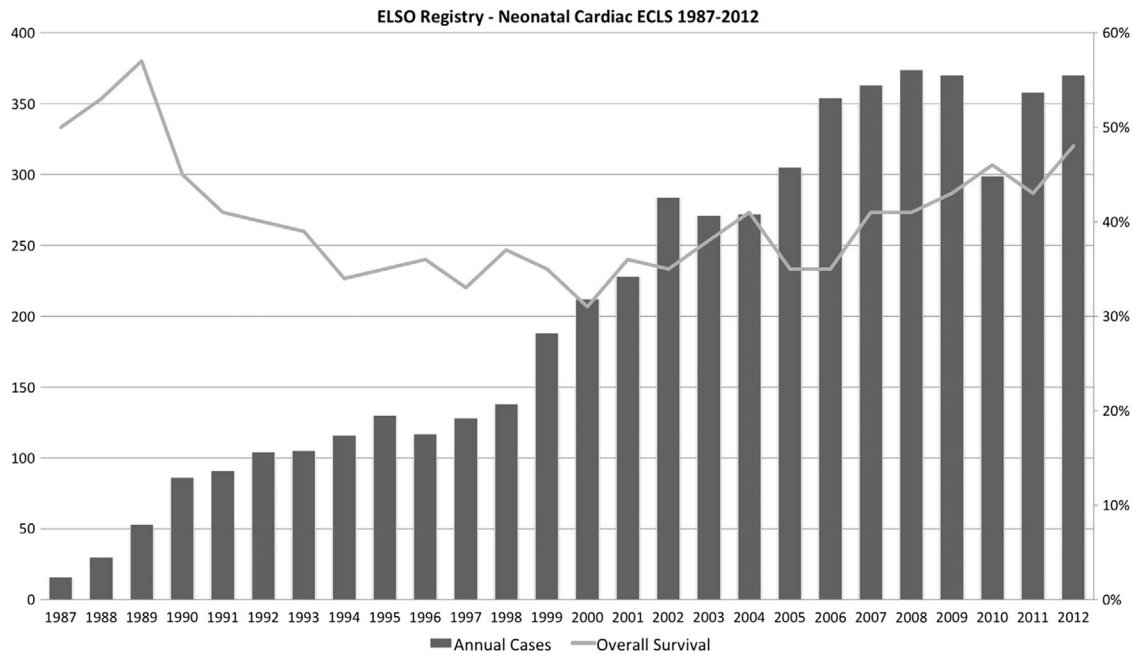

Fig. – Neonatal cardiac ECLS trends in utilization and survival. (Reprinted with permission from Andrew L. Mesher and David Michael McMullan,[5] with permission from Elsevier.)

relative contraindications. ECMO has evolved into a standard therapy for support of cardiac failure refractory to medical care alone. Between 1989 and 2015, there were nearly 40,000 neonatal ECMO cases reported to the extracorporeal life support organization (ELSO) registry. Of these, 22% were cardiac or cardiac arrest-related. Cannulation during cardiac arrest is referred to as extracorporeal cardiopulmonary resuscitation or ECPR. Since 2009, the proportion of cardiac or ECPR cases has increased to nearly 40%.[6] In this review, we will address indications and outcomes for neonatal cardiac ECMO support.

Indications

Perioperative ECMO support

The majority of neonatal cardiac ECMO occurs in the perioperative period, with 80% of the cases in the ELSO registry, excluding ECPR, occurring post-procedure.[7] Pre-operative ECMO support comprises only a small fraction of perioperative ECMO support. Gupta et al.[8] reported that 14% of perioperative ECMO cases from the pediatric health information system (PHIS) database represented patients (both neonatal and pediatric) receiving pre-operative support. Pre-operative ECMO is utilized for stabilization or recovery of myocardial end organ function prior to cardiac surgical palliation or complete repair. Data on outcomes after pre-operative neonatal ECMO support are limited, but appears to be better than other cardiac ECMO populations with survival approaching 60–70%.[8,9] Patient selection appears to be important with congenital lesions with definitive surgical outcomes [such as D-transposition of the great arteries (D-TGA) with refractory hypoxemia due to a restrictive atrial communication] or patients with acute onset of insufficient pulmonary blood flow (acute aortopulmonary shunt occlusion or refractory tetralogy of Fallot spells) demonstrating markedly better outcomes than patients with hypoplastic left heart syndrome (HLHS) with intact atrial septum, a lesion known to have a mortality approaching 50% even without requirement for ECMO.[9] In these neonates, the rising left atrial pressure immediately after birth and decrease in cardiac output related to the inability of blood to exit the left atrium can lead to worsening pulmonary venous congestion and ultimately cardiovascular collapse.[10] Some centers are employing an EXIT (ex-utero intrapartum treatment) strategy with a median sternotomy and initiation of cardiopulmonary bypass while the fetus remains on placental support.[11,12] Once the fetus is successfully cannulated onto cardiopulmonary bypass, the umbilical cord is divided and the newborn subsequently undergoes rapid atrial decompression with or without Norwood completion. Similarly, obstructed total anomalous pulmonary venous connection is a surgical emergency that may require initial stabilization with ECMO. However, as a definitive surgical repair is essentially the only therapy that will sufficiently decrease the markedly elevated pulmonary venous pressure and avoid further damage to the pulmonary vascular bed, early surgical intervention is the mainstay of therapy.

Significantly more data are available regarding indications, complications, and outcomes for post-cardiotomy ECMO support. Post-cardiac surgery, neonatal ECMO has been utilized for both failure to separate from cardiopulmonary bypass (CPB) and for clinical deterioration following initial successful separation from CPB. In one single center study of 84 neonates supported with perioperative ECMO, indications for ECMO included failure to wean from CPB (25%), post-operative cardiac arrest (46%), low cardiac output syndrome (21%), and hypoxemia (7%).[13] Post-cardiotomy ECMO in the immediate post-operative period is used to provide a period