

Extracorporeal membrane oxygenation for pulmonary hypoplasia and prematurity: any chance of survival?

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Extracorporeal membrane oxygenation (ECMO) is a valuable and lifesaving treatment, but unfortunately is also labour-intensive. A Hong Kong paediatric intensive care unit was consulted about a premature infant born at 34 weeks' gestation in cardiopulmonary shock with antenatal history of pulmonary hypoplasia, anhydramnios, and renal dysgenesis for whom treatment with ECMO was being considered. The patient had severe lactic acidosis and required cardiopulmonary resuscitation. Unfortunately, he succumbed before transfer to an ECMO centre.

In medical settings with limited resources, acute care physicians often need to make critical decisions about challenging cases. Considerations for ECMO include prematurity, birth weight, severity of shock, supraphysiologic ventilatory support, inhaled nitric oxide, and multiple inotropes.^{1,2} There are also other roles for ECMO in borderline cases, such as maintaining stability whilst awaiting a diagnosis or treatment decision. The need for central (right atrial and aortic) or cervical (carotid and right internal jugular vein) cannulation is technically challenging but achievable in premature infants. The risks of haemorrhage or ischaemia in the brain and other tissues are significant. When considering such an invasive procedure in a critically ill neonate, it is crucial to carefully evaluate the ultimate objective and the specific function it serves. It is hoped that ECMO allows time for the hypoplastic lungs to grow and mature; however, treatment with venoarterial ECMO is usually only possible for 7 to 10 days in premature infants.

Mortality trends in neonatal ECMO for pulmonary hypoplasia have been recently reviewed.¹ Despite new technologies and treatments, such as ECMO and nitric oxide, there has been little improvement in the outcomes for infants with significant pulmonary hypoplasia.³ Cuestas et al¹ reported that 34% of patients survived to discharge. The mortality rate was significantly higher among

patients with congenital diaphragmatic hernia (CDH) than those with pulmonary hypoplasia secondary to renal dysplasia. Mortality decreased significantly over time for all groups, but the proportion who survived to discharge increased for patients with CDH but decreased for patients with pulmonary hypoplasia secondary to renal dysplasia. Based on a 2017 retrospective review of the Extracorporeal Life Support Organization Registry, no patient with a diagnosis of renal agenesis/dysgenesis or cystic kidney disease survived.⁴ Further, no patients with a primary diagnosis of pulmonary hypoplasia and renal agenesis treated with ECMO survived.⁴

Prematurity and low birth weight have been relative contraindications for ECMO; however, these criteria may not be evidence-based. In an often-cited report of three premature infants with birth weights <2 kg and CDH who underwent cervical venoarterial cannulation for ECMO and repair of their CDH, all received long courses of respiratory support attributable to lung hypoplasia but there were no short- or long-term complications directly attributable to the ECMO treatment. All three were alive at 2 years of age and were reaching normal developmental milestones.⁵ The authors argued that infants with low birth weights may benefit from ECMO and should not be denied such therapy based on arbitrary guidelines regarding gestational age or weight alone.

However, the outcomes of significant pulmonary hypoplasia in premature infants without CDH, as in the case we described, have not been reported. This may be due to a reporting bias related to the poor prognosis of such cases. There was a case report of an infant born borderline prematurely (36 + 2/7 weeks' gestation) with lower urinary tract obstruction and pulmonary hypoplasia who was supported by ECMO for 10 days and survived, though he required long-term oxygen therapy and peritoneal dialysis.⁶ Another retrospective study reported three infants (>36 weeks' gestation) with

congenital renal disease and pulmonary hypoplasia who received ECMO and survived; however, a fourth (35 weeks' gestation; birth weight, 2.24 kg) did not survive.⁷

There is an urgent need for comprehensive data from large multicentre registries to facilitate informed parental counselling. Until more information regarding the outcomes of ECMO becomes available, offering this invasive treatment to premature infants with severe pulmonary hypoplasia but without CDH may be of little benefit and potentially futile.

Author contributions

All authors contributed to the concept or design of the study, acquisition of data, analysis or interpretation of data, drafting of the manuscript, and critical revision of the manuscript for important intellectual content. All authors had full access to the data, contributed to the study, approved the final version for publication, and take responsibility for its accuracy and integrity.

Conflicts of interest

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