National Pediatric Cardiology
Quality Improvement Collaborative

Research Explained

The Prevalence of Congenital Anomalies of the Airway or Lung in Infants with Hypoplastic Left Heart Syndrome and Differences in Midterm Outcomes: A National Pediatric Cardiology Quality Improvement Collaborative Registry Analysis

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ABOUT THIS STUDY

Why is this study important?

- The effects of congenital anomalies of their airway or lung (CAAL) may negatively impact outcomes for those with Hypoplastic Left Heart Syndrome (HLHS), but little is known at this time
    - CAAL encompass a wide variety of abnormalities, but can be summarized as a pulmonary conditions that result in respiratory difficulties.
- This study shows that infants with HLHS and CAAL are significantly more likely to die or need a heart transplant at 1 year of age.
- Infants with HLHS and CAAL are more likely to experience longer stage 1 and stage 2 combined length of hospital stay, as well as being at a higher risk of requiring additional major cardiac and non-cardiac related procedures.

What is the goal of the study?

- To determine the number of CAAL within the population of patients with HLHS
- To compare the midterm outcomes from birth through 1 year of life between patients with HLHS with and without CAAL.
- To determine if CAAL was a risk factor for death or transplantation

How was this study performed?

- The authors used data collected from a large multi-center registry of children with HLHS from the National Pediatric Cardiology Quality Improvement Collaborative (NPC-QIC).
- The authors analyzed the incidence of CAAL within the registry of HLHS patients.
- Demographic, clinical, and outcome characteristics were compared between HLHS infants with and without CAAL.

What were the results of the studies?

- 2467 infants with HLHS in the registry were evaluated
- The incidence of CAAL was low at 2.3% (57 patients) in the registry
- Infants with CAAL were more likely to be female, have lower birth weight, be older at the time of stage 1 procedure, require ventilator support before stage 1 procedure, receive a hybrid procedure as the stage 1 procedure, or not receive any type of stage 1 procedure
- Infants with CAAL had significantly longer combined stage 1 and stage 2 hospital length of stay (102 vs 65.1 days), and underwent more additional major procedures (2.04 vs 0.93 procedures).
- Infants with CAAL had significantly worse survival at 1 year of life (49.5% vs 77.2%).

What are the limitations of the study?

- The analysis is limited to infants enrolled in this registry, and may underestimate the true incidence and impact of congenital anomalies of the airway and lung in the HLHS population
- CAAL encompass a wide variety of abnormalities and the significance of each individual anomaly cannot be determined from this registry analysis
- The long term effects of anomalies of the airway and lung on Fontan circulation is still unknown

What it all means

- CAAL are rare in infants with HLHS, but are associated with significantly increased risk of death and other morbidities.
- These findings may help guide clinical decisions by providers and discussions about expectations and outcomes with families with children with HLHS and airway or lung anomalies.