

Research Explained

Differences in Midterm Outcomes in Infants with Hypoplastic Left Heart Syndrome Diagnosed with Necrotizing Enterocolitis, NPCQIC Database Analysis.

Background of the Study Question

Necrotizing enterocolitis (NEC), infection/inflammation of the intestines, is a well-known complication for infants with single ventricle physiology during their initial hospitalization when they undergo their first staged surgical procedure. Immediate hospital outcomes for infants that develop NEC have been well described, but data describing outcomes after initial hospitalization are lacking. In addition, differences between those infants that developed NEC versus those that did not are unknown. The goal of this study was to compare the midterm outcomes during the interstage period for infants with hypoplastic left heart syndrome who had survived to hospital discharge that developed NEC (HLHS-NEC) versus those that did not develop NEC (HLHS-nNEC) during their initial hospitalization.

How was the study done and what did it demonstrate

This was a retrospective study using NPC-QIC database information (7/2008 – 9/2014). Infants identified with NEC were matched to patients that did not have the diagnosis of NEC based on surgical center, primary initial surgery, and gestational age +/- 1 week in a 1:3 ratio. This matching was done to take into account variations in treatment protocols between centers, differences in physiology between different surgeries, and possible major neonatal factors that would affect feeding issues. Baseline demographic, initial hospital, interstage, and data just prior second stage palliative surgery were collected.

During this time period, 1163 infants with HLHS were enrolled in NPC-QIC. Incidence of NEC was 5.8% (68/1163) (gestational age < 37 weeks 18.3% (11/60), gestational age > 37 weeks 5.2% (57/1103). Eleven infants could not be matched, so the study matched 57 HLHS-NEC with 171 HLHS-nNEC. In the HLHS-NEC group, 14 infants underwent the Norwood procedure with a BT shunt, 37 infants underwent a Norwood procedure with a RV-PA conduit, and 6 infants underwent a hybrid procedure.

HLHS-NEC infants were more likely to undergo extracorporeal membrane oxygenation, have a longer initial hospitalization, have moderate to severe valve regurgitation at time of discharge, have lower weight for Z-score at time of discharge, more likely be fed via a NG or GT, and more likely to be fed formula only compared to the HLHS-nNEC group. At the time of the second surgery, HLHS-NEC patients were still less likely be taking oral feeds, but other variables were such as weight were no longer different.

Limitations of the study

1. Retrospective study of a registry database, so could not independently verify registry data and were unable to collect any missing data points.

2. NEC diagnosis was based on individual institutions' definition and not necessarily on a uniform grading scale for diagnosing NEC.
3. Results could be affected by uncontrolled confounding issues due to variables not routinely collected by the NPCQIC
4. More quantitative data such as nutritional laboratory values, echocardiographic data, catheterization data, and specific surgical techniques were not available.
5. Registry cohort is limited to patients who were discharged home after stage I procedure and whose caregivers gave consent for them to participate in the study, so risk factors for NEC could not be fully ascertained.

What it all means

Despite similar baseline data, HLHS-NEC infants had significant differences in hospital course compared to HLHS-nNEC infants. However, infants who developed NEC were able to overcome more complicated hospital courses to have comparable nutritional outcomes to their counterparts without NEC. This is likely due to the supplemental feeding provided by the NG/GT feeds. Assuming no milk protein allergy, breast milk should be encouraged for all these patients. More studies are needed to explore if there are long-term consequences of a lack of oral feeding among those infants who developed NEC.