

**Key Findings:** Prenatal Diagnosis, Birth Location, Surgical Center, and Neonatal Mortality in Infants with Hypoplastic Left Heart Syndrome

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The journal *Circulation* published a study in October 2013: “Prenatal Diagnosis, Birth Location, Surgical Center, and Neonatal Mortality in Infants with Hypoplastic Left Heart Syndrome.”

<http://circ.ahajournals.org/content/early/2013/10/17/CIRCULATIONAHA.113.003711.abstract>

The NPC-QIC Research and Publication Committee has reviewed this article and a summary of the findings can be found below.

**Main Finding from this Study:**

The authors of this study concluded that infants with HLHS born closer to a surgical center that performs cardiac surgery on infants have a higher survival rate than infants born far from a surgical center.

**About this study:**

- **Why is this study important?**

Over the past several decades there has been dramatic improvement in surgical survival in infants born with HLHS. However, ~20% of infants born with HLHS still die within the first months of life. We are constantly looking for ways that we can improve the survival of infants with HLHS, trying to identify improvements we can make in the way we care for these infants. This study attempts to identify risk factors that we might be able to address to improve this survival.

- **How was this study performed?**

The researchers in this study used information from the Texas Birth Defects Registry and identified 463 infants with HLHS born in Texas from 1999-2007. The researchers then looked at where these infants were born, and what the distance and driving travel time was between their birth hospital and the closest surgical hospital that performed stage 1 surgical palliation (Norwood procedure). The researchers then looked for a relationship between this distance and the likelihood of survival of these infants.

- **What were the results of the research?**

- Data for a total of 588 infants with HLHS born in Texas between 1999-2007 was available in this Registry.

- Several factors were found to be associated with better or worse survival for infants born with HLHS:
  - The researchers concluded that infants born far from a surgical hospital (more than 90 minutes driving distance away) have **worse** survival.
  - Infants with a birth weight <2.5 kg had **worse** survival.
  - Infants who had surgery at a surgical hospital who performed more Norwood surgeries had slightly **better** survival after that surgery.
- While prenatal diagnosis was not by itself associated with better or worse survival, prenatal diagnosis is a very important factor related to distance from the surgical hospital; 66% of infants who were born less than 10 minutes from the surgical hospital were diagnosed prenatally as compared to only 13% of the babies born more than 90 minutes from the surgical hospital.

- **What are the limitations of this study?**

Studies that use databases like the Texas Birth Defects Registry are nice because they allow researchers to look at a large number of patients. This is especially helpful when trying to learn about patients with rare problems like HLHS. However, one of the problems with research using databases is that there often is incomplete information about the patients. The following are some of the other things that limit the interpretation of findings from this study.

- Survival was better in infants born in the more recent time period (2003-2007) than in the older (1999-2002) period. More infants were diagnosed prenatally in the more recent time period (49% versus 25.5%) and that number has likely continued to increase in recent years.
- More infants living more than 90 minutes from the surgical center were Hispanic (30% of mothers born in Mexico) and lived in poverty, suggesting they may have had more limited access to care, including lack of prenatal diagnosis. These socio-demographic factors were not included in the study of factors that may affect survival.

- **What are the takeaway messages considering the results and limitations of this study?**

There are many factors that affect survival of infants born with HLHS. There are some findings from this study that may allow us to improve survival of these infants moving forward.

- This study did note worse survival in infants with HLHS born far from surgical hospital. However, there are likely other factors that influence that finding, including the changes in surgical practices over time and socio-economic factors that may influence survival.
- Prenatal diagnosis is important because it may reduce mortality if mothers living far away can deliver close to a cardiac surgery center. Prenatal care is also important because we know that infants with HLHS who are born prematurely or with a low birth weight (<2.5kg) have worse survival. We should be doing everything we can to make sure that pregnancies receive adequate prenatal care and appropriate referrals.

- We need to continue to understand the relationship between surgical volume (the number of specific procedures that are performed at a surgical hospital) and the survival before and after surgery in infants with complex heart problems like HLHS. Some studies, including this one, have suggested that the more surgeries that are done, the better the survival. Collaborative work, like that going on in NPC-QIC will be required to understand the answer to this question.