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Helping Families Access Fetal Cardiac Intervention to Interrupt Evolving HLHS and HRHS

By Brita Dietzel

Ellianna Grace Foundation (EGF) provides financial, logistical and emotional support to families traveling for Fetal Cardiac Intervention and follow-up care.

The journey of a child born with a single-ventricle anatomy is uncertain. Nearly all these children face enormous challenges. These challenges begin shortly after birth, and continue throughout their lives. In recent decades, doctors learned to perform palliative surgeries that allow children to live with a single-ventricle circulation. However, the long-term future for these children remains largely unknown. As a result, a team at Children's Hospital Boston and Brigham and Women's Hospital initiated Fetal Cardiac Intervention in 2000. To date, 140 Fetal Cardiac Interventions have been performed in Boston. The goal of the Fetal Cardiac Intervention is to interrupt the progression of HLHS and HRHS and give children the chance of a two ventricle circulation. While there are varying outcomes, it is this hope that brings families from all corners of the country to Boston.

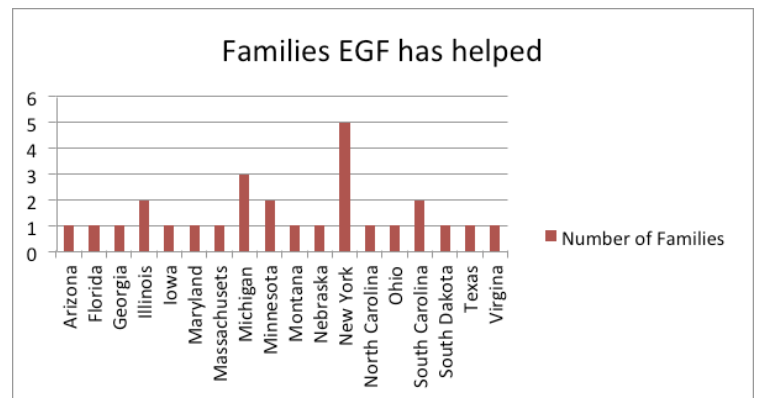
Ellianna Grace Foundation is a non-profit organization that assists families with the financial, logistical and emotional needs required to travel for the Fetal Cardiac Intervention (FCI). The organization was founded in late 2008 by two mothers, Brita Dietzel and Jessica Lindberg, who both underwent Fetal Cardiac Interventions. Their families traveled from their homes in the Midwest to Boston for their children, Ellianna and Ethan in 2004 and 2005 respectively.

For Ellianna, the Fetal Cardiac Intervention prevented Hypoplastic Left Heart Syndrome. At just under twenty weeks gestation, Ellianna was diagnosed with critical aortic stenosis with evolving Hypoplastic Left Heart Syndrome and an intact atrial septum. At this time, she presented with a dilated LV, restricted aortic valve, restricted atrial septum, and retrograde flow through the aortic arch. Within a few days of diagnosis, Ellianna and Brita underwent the FCI. Ellianna experienced bradycardia in which the doctors needed to administer atropine intramuscularly. She also developed a pericardial effusion that was drained prior to ending the procedure and monitored for the next 48 hours. Ellianna and Brita had a stable recovery and returned to Minnesota two days post-procedure. Throughout the rest of the pregnancy, Ellianna was monitored with monthly fetal echocardiograms in Minneapolis. Brita was able to return back to work after two weeks.

Ellianna's fetal echoes showed continued improvement in LV growth and function and her dilated aortic valve remained open. In January 2005, Ellianna was born with adequate LV size and function and was discharged from the hospital without any further cardiac procedures. At the time of discharge, she had mild mitral stenosis and known endocardial fibroelastosis (EFE). At four months of age, Ellianna's stiff left ventricle was causing increase in pressures in her left atrium and lungs. At this time, she underwent an EFE resection in her left ventricle and a mitral valvuloplasty.

This surgery proved to be a success as she thrived at home until she died in a car related accident at 18 months of age.

Ethan also underwent a Fetal Cardiac Intervention to treat his aortic stenosis and evolving Hypoplastic Left Heart Syndrome. His Fetal Cardiac Intervention was technically successful, and he was monitored by fetal echo bi-weekly until his birth. During the remainder of Jessica's pregnancy, Ethan's left ventricle continued to grow though it was quite stiff. Ethan was born in Boston in May of 2005 and underwent a modified Norwood with EFE resection of his left ventricle and aortic and mitral valvuloplasty. Ethan continued down the single ventricle path having his Fontan at 2 years of age. However, his left ventricle continued to grow and his surgeon continued to repair his mitral and aortic valves and resect EFE from his left ventricle. Today at 6 years of age, Ethan is in the hospital recovering from a mitral valve replacement, aortic valvuloplasty, and restriction of his atrial septum to push more blood into his LV. He has a normal LV size and function. His cardiac team has decided to proceed with a two ventricle repair in the near future.



Both children have had challenges in life, but have been greatly helped by undergoing the Fetal Cardiac Intervention. In Ethan's case, the support given to his LV in the early years to encourage growth and function has become a life-saving option today. For Ellianna, she was spared a single ventricle diagnosis after opening her aortic valve as a fetus.

After making many trips to Boston for surgeries and follow-up care, Brita and Jessica wanted to find a way to help other families access the same care their children had received. In the spirit of thanksgiving and hope, they founded Ellianna Grace Foundation to help other families access Fetal Cardiac Intervention. Once confirmed as a candidate, a family may access Fetal Cardiac Intervention in the following cases:

- 1) Critical aortic stenosis with evolving HLHS,
- 2) Pulmonary atresia with an intact ventricular septum and evolving HRHS, and
- 3) Established HLHS with an intact or highly restrictive atrial septum.

Ellianna Grace Foundation awards grants to families to assist with costs associated with traveling for a Fetal Cardiac Intervention and follow-up care. The grants help pay for airfare, lodging, meals, and/or transportation. Since December of 2008, \$22,000 in aid has been given to families across the country (below). More than 90% of the patients who were treated with the FCI lived outside Boston and the surrounding areas.¹ Thankfully, in recent years, the FCI has become more accepted therapy both nationally and internationally as more institutions are initiating Fetal Cardiac Intervention programs. In the US, the University of Michigan and University of California, San Francisco are also gaining experience with FCI. However, for majority of families, the travel requirements will remain significant financial drain. In addition to financial assistance, Ellianna Grace Foundation provides *Parent-to-Parent Connections* where families can speak directly with others who have gone through Fetal Cardiac Intervention.

Prenatal diagnosis remains the cornerstone of the Fetal Cardiac Intervention option. Early detection and diagnosis not only makes the FCI a possibility, but there is also a direct correlation to positive outcomes for the baby.² Ellianna Grace Foundation strives to educate expecting mothers to ask their obstetricians these three questions during their mid-gestation routine ultrasounds:

1. Can you see four chambers in the heart?
2. Can you see two valves entering the heart and two valves leaving the heart?
3. Are the great arteries of the heart crossing each other?

Lastly, the many families who travel to Boston for Fetal Cardiac Intervention need the support, guidance and great care of their local pediatric cardiologists. Good communication is essential. The best-case scenario happens when the local cardiologist works hand-in-hand with the team in Boston. The team in Boston is readily accessible and dedicated to each of these children. Ethan and Ellianna received superb care from doctors at Children's Hospital of Wisconsin and of Minnesota. The sharing of information between doctors has allowed for the best plan of care for these children.

If you are caring for a family who may be a candidate for Fetal Cardiac Intervention, Ellianna Grace Foundation wants to help. Please refer families to email the foundation directly at: info@elliannagrcefoundation.org or call 952.484.6196. In addition, the staff at Children's Hospital Boston and Brigham and Women's Hospital can assist in facilitating aid from Ellianna Grace Foundation.

To learn more please visit www.elliannagrace.org.

References

1. Mark Doff B. McElhinney, MD; Wayne Tworetzky, MD; James E. Lock, MD. Current Status of Fetal Cardiac Intervention. *Circulation*. 2010; 121: 1256-1263.
2. Mark Wayne Tworetzky, MD; Doff B. McElhinney, MD; V. Mohan Reddy, MD; Michael M. Brook, MD; Frank L. Hanley, MD; Norman H. Silverman, MD. Improved Surgical Outcome After Fetal Diagnosis of Hypoplastic Left Heart Syndrome. *Circulation*. 2001;103:1269.

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ELLIANNA GRACE FOUNDATION

What all pregnant families need to know about their baby's heart

Congenital Heart Defects are the most common birth defect in the United States and the world. This year **36,000 babies** will be born with a **congenital heart defect***. There are a wide range of defects. Some are so mild they don't have symptoms and resolve with time. However, there are many cases where the defect requires surgery as a newborn. Survival and outcomes are directly related to **accurate prenatal diagnosis**. This early diagnosis allows families to deliver in a hospital that has proper cardiac triage.

Early prenatal diagnosis is important! Research shows that diagnosing a heart defect before birth **improves outcomes** for the baby after birth.

There are many things you already do for your unborn baby. As part of your prenatal care, **please ask your doctor the following questions:**

1. Can you see the 4 chambers of the heart?
2. Can you see two valves entering the heart and two valves leaving the heart?
3. Are the great arteries of the heart crossing each other?

Dr. Wayne Tworetzky
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*The American Heart Association