

A Review of Heart Transplantation in Children

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Reprint Request

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Received on July 29, 2017

Accepted on October 23, 2017

Abstract

A heart transplant is an operation to replace a diseased heart with a healthy one from another person. Treatment of patients suffering from end-stage heart failure (HF) leaves surgeons with limited options makes the indication of transplantation. According to the International Society for Heart and Lung Transplantation, approximately 350-500 pediatric heart transplantation procedures are performed worldwide each year, about 12% of the of the total number of heart transplants performed. An estimated 10% of congenital heart disease cases have been deemed uncorrectable. One of the most common indications for infant heart transplantation had been hypo plastic left heart syndrome (HLHS), which occurs in about one in 6000 live births. Congenital cardiomyopathy occurs in approximately one in 10,000 live births. Survival in excess of 20 years after pediatric heart transplantation has been achieved. Most programs now report that more than 70% of their recipients survive at least 5 years.

Keywords: Heart Transplantation; Congenital cardiomyopathy; congenital heart disease; Hypo plastic left heart syndrome (HLHS).

Introduction

"It is infinitely better to transplant a heart than to bury it to be devoured by worms"

-By Christian Barnard

(*Father of Heart Transplantation.*)

Cardiac transplantation in infants and children has been an accepted therapy since its successful application to infant recipients began in 1985. If a child needs a heart transplant, Parents probably feeling lots of emotions all at once-including anger, sadness, confusion, and frustration. These feelings can become overwhelming, especially when the child is waiting for a new heart. Heart transplant is an operation that replaces a dysfunctional heart with a healthy heart from another person who has had brain death but whose heart is working normally.

Adrian Kantrowitz performed the world's first pediatric heart transplant on December 6, 1967, at

Maimonides Hospital in Brooklyn, New York, barely three days after Christian Barnard's (father of heart transplantation) pioneering procedure of the world's first adult human heart transplant on Louis Washkansky on December 3, 1967 at the Groote Schuur Hospital in Cape Town, South Africa. In India Dr. Cheria performed the first coronary artery bypass surgery in 1975. He also performed the country's first heart transplant, first infant cardiac surgery and the first heart and lung transplant in 1999. Recently (2015) cardiologist successfully performed a heart transplant on a two year old boy from Russia named Baby. Gleb was diagnosed with restrictive cardio myopathy at Fortis Malar hospital, Chennai.

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Indications

Congenital malformations are still the most common indication for infant heart transplantation. An estimated 10% of congenital heart disease cases have been deemed uncorrectable. One of the most common indications for infant heart transplantation had been hypoplastic left heart syndrome (HLHS), which occurs in about one in 6000 live births. Congenital cardiomyopathy occurs in approximately one in 10,000 live births. The most common indication for heart transplantation in older children is cardiomyopathy.

The severity of heart failure in children

- Stage A
 - At risk for developing heart failure
- Stage B
 - Abnormal cardiac structure or function but no symptoms of heart failure
- Stage C
 - Abnormal cardiac structure or function
 - Past or present symptoms of heart failure
- Stage D
 - Abnormal cardiac structure or function,
 - Requiring continuous intravenous (IV) infusion of inotropes or prostaglandin E₁ (to maintain patency of the patent ductus arteriosus-PDA)
 - Requiring mechanical ventilator and circulatory support

The Indications as follows

- Cardiomyopathy
 - Dilated cardiomyopathy
 - Hypertrophic cardiomyopathy
 - Restrictive cardiomyopathy

- Cardiac tumors
- Congenital cardiac defects
 - Anatomically uncorrectable congenital heart diseases
 - Correctable conditions associated with high operative risk.
- Infections
- Toxins (either endogenous or exogenous) that causes damage to myocardium

Contraindications

Some of the absolute contraindications for pediatric heart transplantation include,

- Irreversible elevated pulmonary vascular resistance
- Malignancy
- Ectopia cordis (congenital malformation in which the heart is abnormally located either partially or totally outside of the thorax.)
- Active systemic infection
- Severe primary renal or hepatic dysfunction
- Multi-organ system failure
- Major CNS abnormality
- Infection with HIV or chronic active hepatitis B or C
- Severe dysmorphism
- Marked pre maturity (< 36 weeks)
- Small size (< 1800 g)
- Positive findings on drugs screen.
- Lack of family support systems.

Heart Transplant Team

The team members, who work together to make sure that the child has a successful transplant, probably will include

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|-----------------------------------|---------------------------|
| ➤ Cardiac surgeons | ➤ Nutritionists |
| ➤ Cardiologists | ➤ Transplant coordinators |
| ➤ <i>Advanced Practice Nurses</i> | ➤ Child Life specialists |
| ➤ Anesthesiologists | ➤ Psychologists |
| ➤ Transplant pharmacists | ➤ Social workers |
| ➤ Radiologists | ➤ Resource specialists |
| ➤ Physical therapists | |
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The transplant team will evaluate the child to determine whether a transplant will be beneficial and whether the child is a good candidate for a transplant. The evaluation will include a medical history, a physical examination, and some tests and diagnostic procedures like an Echocardiogram, an Electrocardiogram (also known as an ECG or EKG), Cardiac catheterization and biopsy.

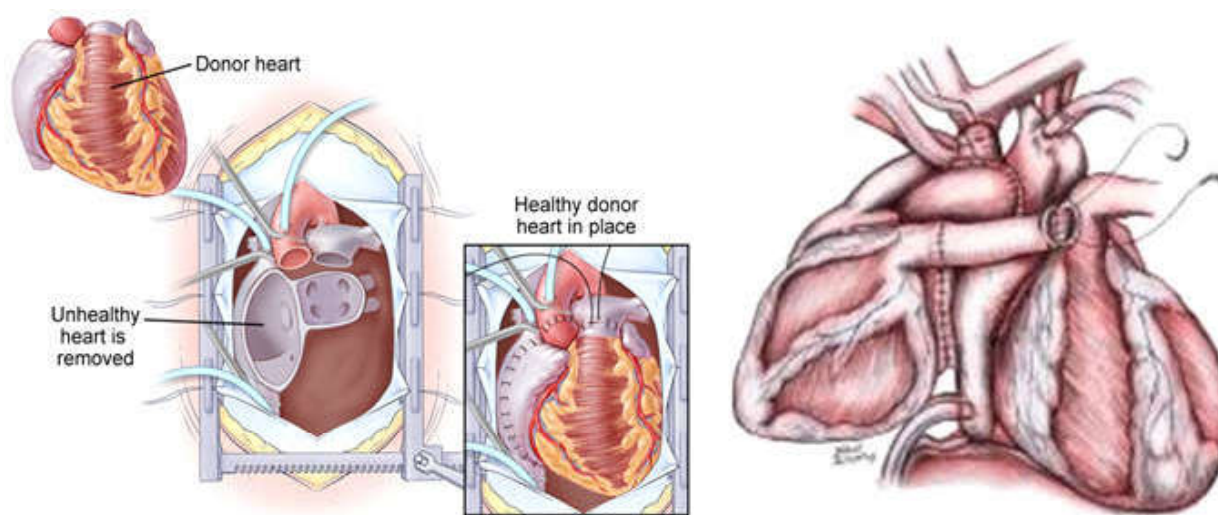
A surgical team can be dispatched from the transplantation center and can travel up to 500 miles to retrieve the needed Heart. An ischemic time up to 4 hours is considered acceptable. This allowable ischemic time permits an approximate travel time of 2.5 hours with remaining time required to implant the heart in to the patient.

Heart Transplant Procedure

Once a compatible donor organ is located, the recipient will be evaluated and started on medications in preparation for transplantation. Once the patient is under anesthesia the surgeons begin

the process by exposing the chest cavity through a cut in the rib cage. The normal functioning of the body is kept intact by a mechanical pump [ECMO, LVAD etc.] that circulates blood through the body while the diseased heart is removed after opening up the pericardium. However the back part of the left atrium is left in place. In a method called as 'Orthotopic procedure', the donor's heart is carefully trimmed and sewn to fit the remaining parts of the old heart. 'Heterotopic' heart transplantations refers leaving the recipient's heart in place and implanting a new heart to act as an additional pump or piggy back heart; this type of heart transplant rarely done in children.

The patient is given immunosuppressant both before and during the surgery in order to prevent rejection. After a successful transplantation the patient is shifted to the ICU for recovery. The duration of stay in the hospital will depend on the patient's health and how well the recipient's body is responding to the new heart.



Heart Transplant Life Expectancy and Survival Rate

There have even been rare instances where the recipients have survived for more than two decades. Generally, the heart transplant survival rate immediately after the surgery is 95% in most of the centers. If the patient survives after the first thirty days, then there are 90% chances that he/ she will cross the coveted one year mark. According to a survey conducted by the American Heart Association, the average survival rates after heart transplantation as follows

Survival rate (In yrs.)	Male	Female	Children
One year	88%	77.2%	80-90%
Five-year	73.1%	67.4%	<70-80%

Survival in excess of 20 years after pediatric heart transplantation has been achieved. Most programs now report that more than 70% of their recipients survive at least 5 years. However, although an additional 5 years of life is important for all, the goal of pediatric heart transplantation is to provide as much of a normal life span for these children as possible. The donor supply remains inadequate.

Improved public and physician awareness of donor issues is the most important factor in increasing donor supply because many potential donors are not identified as such.

Complications and Follow-Up

Even after a successful transplantation, the medical practitioner should be on the lookout for various complications such as acute rejections, bleeding from suture lines, ascending infections, cardiac allograft vasculopathy, renal failure, malignancy etc. it is essential to monitor the child for signs of infection and rejection, including fever, tiredness, difficulty breathing, vomiting, weight gain, and poor appetite. In the long term, regular checkups are needed to monitor for complications. At first, these checkups will occur often (perhaps weekly). They gradually become less frequent, though, and eventually may be necessary only once or twice a year.

Conclusion

Many kids who have heart transplants live normal, healthy lives once they recover from surgery. Some feel better than they ever have before. Even though it is a costly line of treatment it has been a miraculous breakthrough in the medical field in the care of children with abnormal congenital cardiac defects. Heart transplant's prognosis is uncertain and places a person on a lifelong financial burden and drug therapy; it is still opted as the most sought out

therapy for end stage congestive heart failure. General public awareness and media attention focused on the need for donors have also contributed to an increase in the available donor pool and transplantation activity.

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