CRC project protocol
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Resident: Ellis Rochelson, PGY2

Investigators:

- Ellis Rochelson, MD. Resident, Department of Pediatrics
- Brett Anderson, MD, MBA. Assistant Professor of Pediatric Cardiology
- Emile Bacha, MD. Director, Congenital and Pediatric Cardiac Surgery
- Marc Richmond, MD. Assistant Professor of Pediatric Cardiology
- Alejandro Torres, MD. Assistant Professor of Pediatric Cardiology

Title of Project:
Pre- and post-Glenn predictors of early Fontan failure

Research question:
Do pre-Glenn hemodynamics and post-Glenn course predict the likelihood of early Fontan failure?

Scientific abstract:
Children with a single functional ventricle, such as in hypoplastic left heart syndrome, typically require a series of at least three operations, culminating in a Fontan procedure. In this operation, blood from the inferior vena cava is redirected to the pulmonary circulation in order to bypass the right atrium. The goal is to establish complete cavopulmonary isolation - that is, to separate the pulmonary and systemic circuits, allowing the single functional ventricle to pump solely to the systemic circulation. It is usually preceded by several years by a Glenn or hemi-Fontan procedure, in which the superior vena cava is anastomosed to the pulmonary arteries. Though the early mortality of the Fontan procedure has greatly improved over the last several decades, early failures are still relatively common and result in Fontan takedown, cardiac transplantation, or death. Few risk factors are well established as predictors of early Fontan failure. These include poor ventricular systolic function at the time of the Fontan, elevated pulmonary artery pressure on pre-Fontan cardiac catheterization, and prolonged cardiopulmonary bypass time. Other factors, such as Fontan type, heterotaxy syndrome, and dominant left ventricle have been suggested, but their ability to independently predict outcomes is less well established. To date, no data exist regarding dynamic predictors of Fontan failure measurable in children at younger ages. Identifying early predictors of Fontan failure can help prepare families and their physicians for what can be difficult, and often devastating, outcomes. This study is a single-center retrospective analysis of patients who underwent a Fontan operation from 2005-2014, with the goal of determining if pre-Glenn hemodynamics and post-Glenn course can predict post-Fontan outcomes.

STUDY DESCRIPTION
Study purpose and rationale:
Our primary aim is to identify peri-Glenn predictors of early Fontan failure. Our secondary aim is to determine the effects of peri-Glenn predictors on post-Fontan length-of-stay. We hypothesize that patient pre-Glenn hemodynamics and post-Glenn course predict post-Fontan outcomes. In order

to achieve this aim, we will analyze pre-Glenn catheterization data and post-Glenn outcomes to determine if these measurements can predict early Fontan failure. Early Fontan failure will be measured as death, heart transplant, or re-operation prior to discharge (i.e. Fontan "takedown") within 30 days following Fontan surgery.

Study Design and Statistical Procedures:

- Subjects
    - Inclusion criteria
        - Patients ages 0-18 who underwent the Fontan operation at New York-Presbyterian/Morgan Stanley Children's Hospital from 2005-2014.
    - Exclusion criteria
        - Patients who did not have a Glenn procedure performed at New York-Presbyterian/Morgan Stanley Children's Hospital.
- Our primary predictors of interest will be measures of hemodynamics on pre-Glenn catheterization and post-Glenn complications and length-of-stay.
- Control variables will include patient age, sex, anatomic subtype, type of Fontan surgery performed (extracardiac vs. lateral tunnel).
- Outcomes will be assessed using univariable and multivariable analyses. Univariable analyses will include Chi-squared tests to assess the effects of categorical variables on categorical outcomes, and t-tests to assess the effects of continuous variables on categorical outcomes. In multivariable analyses, logistic regression will be used to assess the effects of predictors on early Fontan failure.

Potential Benefits

This study likely confers no direct benefit to the patients being studied. Analyzing our data may benefit future patients who require a Glenn or Fontan, as it may improve our understanding of risk factors and outcomes.