Congenital Heart Surgeons’ Society Data Center

October 20-21, 2013
October 21, 2013

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- Congenital Heart Surgeons’ Society members and member institutions

**Institutional Support**

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- Children’s Heart Clinic (Dr David Overman)
- Primary Children’s Hospital (Dr John Hawkins)
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- The Children’s Heart Foundation
- Saving tiny Hearts Society
- Cryolife®, Inc.
- Michael H. Ludwig Memorial Foundation
- Nonin, Inc.

Sincerely,

W.G. Williams, MD, FRCSC
Executive Director

C. Caldarone, MD, FRCSC
Managing Director
Introduction

Update of Current Cohorts

Anomalous Aortic Origin of a Coronary Artery  Travis Wilder
Late Functional Health Status in Critical Aortic Stenosis  Brian McCrindle
Aortic Valve Atresia  Katie Stackhouse
Left Ventricular Outflow Tract Obstruction  Travis Wilder

New Projects

Technical Performance  Chris Caldarone
Registry of Registries  Peter Gruber
Echo Core Labs  Bill Williams
Ebstein Anomaly  Christian Pizarro
               Chris Knott-Craig
               Joe Dearani

Transformation  Chris Caldarone
### Congenital Heart Surgeons’ Society Cohorts

<table>
<thead>
<tr>
<th>Diagnostic Group</th>
<th>Number of Institutions</th>
<th>Accrual Date</th>
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<tr>
<td>Transposition of Great Arteries</td>
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<td>Coarctation of the Aorta</td>
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<td>Aortic Valve Atresia</td>
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<td>Critical Aortic Stenosis</td>
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<td>Tricuspid Atresia</td>
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<td>1999-Present</td>
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<tr>
<td>Pulmonary Conduit</td>
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<td>Critical Left Ventricular Outflow Tract Obstruction</td>
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<td>2005-Present</td>
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<td>Anomalous Aortic Origin of Coronary Arteries</td>
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<td>2009-Present</td>
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<tr>
<td>Atrioventricular Septal Defect</td>
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### Congenital Heart Surgeons’ Society Studies

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<td>Late Functional Status in Cohort of Survivors with PAIVS</td>
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Total Number of Patients in CHSS Studies

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<th>Cohort</th>
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<td>AAOCA</td>
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<td>AVSD</td>
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Annual Enrollment

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<td>2012</td>
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<td>2013</td>
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</table>
Number of Enrolling Institutions

Patients transitioning to adulthood in CHSS studies

Increased Electronic Data Management
Anomalous Aortic Origin of a Coronary Artery

Summary/Description:

The natural and unnatural history of patients with anomalous aortic origin of a coronary artery (AAOCA) is unknown. Evidence-based criteria to guide decisions between observation and intervention strategies are not well developed. To identify evidence based criteria for management of these patients, the CHSS began multi-center enrollment in 2009.

Status:

To date, 272 patients < age 30 years (men age at diagnosis 9.7 years) are enrolled from 31 CHSS institutions. 139 underwent surgical intervention, 104 have been medically managed.

Data:

Our current analysis is based on 243 patients with updated data entry. It is directed at comparing preoperative imaging with the "true" surgical findings. Our objective is aimed at identifying which morphologic variables can be reliably identified within and between imaging techniques. Initially, the comparison was made between echocardiogram reviews as reported by a core group of expert echocardiologists with surgery and institutional echocardiogram reports with surgery. This analysis will eventually be carried forward to include preoperative CT and MRI studies.

Plan:

- An abstract based on correlations between demographics, symptoms, coronary anatomy and management of the initial 200 patients has been accepted by the American Heart Association and the AAOCA working group is in the process of finalizing the manuscript.
- In addition, a profile of what was observed in surgery and surgical techniques on the first 113 patients was accepted as an STS abstract and will soon have finalized manuscript.
- The core group expert echocardiologists are in the process of producing a "best practice" technical paper to detail optimal techniques for diagnosing AAOCA.
- Analysis moving forward will require completion of cross-sectional follow up to determine factors influencing long term outcomes.

Publications:

Late Functional Health Status of a Cohort of Survivors with Critical Aortic Stenosis

Summary/Description:

Management of the neonate with critical aortic stenosis (AS) represents a complex problem, in part due to the wide spectrum of aortic valvular and annular pathology as well as variable hypoplasia of left heart structures. The most difficult of these patients have critical AS and intermediate hypoplasia of left heart structures, making the decision to pursue biventricular repair (BVR) or univentricular repair (UVR) in these "borderline" patients particularly difficult. Previous CHSS analyses of our extant AS cohort (with study enrollment from 1994-2000) include comparison of surgical and balloon aortic valvotomy for AS, and identification of factors influencing survival after BVR or UVR which led us to devise a calculator which may be used to assist in triage to BVR vs. UVR. Importantly, we determined that a bias toward BVR may exist and that many patients are inappropriately triaged into the wrong pathway according to the calculator. However, there is a paucity of other studies examining the implications of a bias toward BVR in the AS population, if one exists. Meanwhile, survival after management of critical AS has risen in the contemporary era, and the research necessarily must now include late functional outcomes. Therefore, we submitted a grant to the Children’s Heart Foundation to fund a study that would address the following question: "In patients with critical AS and "borderline" left heart morphology, is selection of BVR associated with improved functional health status or exercise performance?"

Status:

- Dr. Brian McCrindle has received a grant from the Children’s Heart Foundation for this study.
- Pending the ethics approval.

Plan:

This study will recruit from 264 surviving and tracked patients in the CHSS AS cohort from 25 member institutions. These patients are between 13 to 19 years of age. Based on participation in other cohorts within this population, we expect 184 patients to participate. To assess functional health status, they will be asked to complete these standardized questionnaires:

- Behavior Assessment System for Children-2
- Behavior Rating Inventory of Executive Function
- Habitual Activity Estimation Scale
- Pediatric Quality of Life Inventory
- Pediatric Cardiac Quality of Life Inventory
- Hollingshead Four-Factor Scale

Simultaneously, standardized exercise testing will be performed to obtain an objective assessment of their capacity for exercise. Serial available echocardiographic reports will also be collected. The final dataset for analysis will therefore consist of clinical and neonatal anatomic data acquired before any intervention, available serial echocardiographic studies, and current measures of functional health status and exercise capacity. Correlation between neonatal echocardiographic findings, current echocardiographic data, choice of management strategy and prospectively gathered indices of functional health status and exercise capacity will be evaluated. Serial echocardiograms will permit us to track longitudinal changes in left heart structures as these patients enter adulthood and will also be accounted for in our examination of factors influencing late functional health status and exercise capacity.
Aortic Valve Atresia

Summary/Description:

As the vast majority of neonates with severe congenital heart defects such as aortic atresia survive operative treatment and live into adulthood, their functional health status (FHS) becomes of paramount importance as does their success in transitioning from pediatric to adult cardiac care for sequelae of their palliative repairs or transplantation. We will use the aortic atresia cohort to investigate current FHS of adolescents living after intervention for aortic atresia, characterize behaviors and barriers related to transition of care, and how FHS impacts transition behaviors.

Status:

613 neonates from 27 institutions were prospectively enrolled in the aortic atresia cohort from 1994-2000. At last annual follow-up, 257 patients were alive. The response rate for the last completed annual follow-up was 46%.

Data:

Several FHS and transition of care questionnaires were added to the annual follow-up packet, and to date, 75 patients have responded. Data collection is ongoing.

Plan:

Follow-up is ongoing with this cohort currently. Data analysis to characterize this cohort, describe long-term outcomes, and investigate FHS and transition of care will begin in Jan 2014.

Publications:


Critical Left Ventricular Outflow Tract Obstruction

**Summary/Description:**

The CHSS has made fundamental contributions to our understanding of left sided obstructive lesions based on the aortic valve atresia (AVA) and aortic valve stenosis (AVS) cohorts. Despite advances in surgical and interventional management, outcomes in patients with critical left ventricular outflow tract obstruction (LVOTO) remain poor relative to other congenital heart diseases. Anticipating the need for evaluation of emerging strategies (e.g. Sano shunts, hybrid procedure) the CHSS initiated the development of a contemporary cohort of neonates with LVOTO in 2005. Surgical detail has been tracked extensively (~200 variables) and will permit detailed analysis in these patients.

**Status:**

Since January 1, 2005, 672 eligible neonates have been enrolled from 21 CHSS institutions. To date, data entry is complete on nearly all index surgical procedures and baseline echo reports. A large amount of date pertaining to subsequent procedures, follow up echocardiograms, and catheter interventions is still in need of entry.

**Data:**

Of the 672 eligible patients, 664 have had an index procedure (7 died prior to intervention, 3 alive w/out intervention). Early analysis shows an initial procedure consistent with single ventricle repair strategy in 83% of patients and bi-ventricle repair in 15%. Overall there were 206 deaths after initial procedure, representing an early survival of approximately 70%.

Initial management procedures include:

- Norwood procedure (N=439; 66%),
- hybrid stage I (N=111; 17%) or balloon aortic valvotomy (N=97; 15%),
- Transplant (N=5; 1%),
- Yasui (N=4; 1%)

**Plan:**

We propose an analysis to determine how demographics, morphology, and choice of initial management influence early survival and transition to subsequent procedure. This analysis would incorporate a detailed account of the techniques of Stage I palliation, including a large population of patients managed with an initial hybrid procedure.

**Publications**

Surgical Technical Performance Score (TPS) in Congenital Heart Surgery

Summary/Description:
Emile Bacha and his colleagues have done pioneering work in demonstrating usefulness of measuring surgical Technical Performance Scores (TPS) in congenital heart surgery. They have demonstrated that the TPS can be measured successfully in a single institution - and can be used to predict morbidity and mortality. The concept of breakdown a complex procedure into discrete components which may be individually evaluated in terms of technical success – or failure – allows a surgeon to identify areas to improve performance with far greater precision than is possible when evaluating general outcomes (e.g., hospital death). The TPS concept was developed in single institutions, but there is currently no mechanism for surgeons outside these institutions to compare their technical performance against a large dataset.

A CHSS Working Group was established to evaluate the potential for creating a TPS tool which is freely available to all members. The Working Group’s plan developed the structure to create a web-based TPS entry platform which can allow participating members to calibrate their technical performance against the entire CHSS as a quality assurance tool. The Data Center has created and maintains the backbone infrastructure for the TPS project. This data will be used to generate confidential reports allowing a surgeon to evaluate his/her technical performance against his/her institution and the entire CHSS. Although designed as a quality improvement project, subsequent research projects are expected to develop using this infrastructure.

Status:
The Data Center has created and internally tested the software tool for the Technical Performance Project based on the Working Group's concept for five operations (VSD, TOF, AVSD, ASO with VSD, ASO without VSD). The members of the Working Group have been invited to beta test the system before it is rolled out to the Society.

Plan:
The surgical performance data is uploaded in de-identified format into a cloud based Research Electronic Data Capture (REDCap) system. It has built-in controls to help ensure accuracy and data quality. The REDCap data entry system takes about a minute per patient to enter the data. Technical performance score for a completed case is available instantly.

Working Group Members:
Emile Bacha, Chris Caldarone, Bill Williams, John Karamichalis, Jeff Jacobs, Marshall Jacobs, Erle Austin, Bill DeCampli, Eugene Blackstone, Peter Gruber, David Overman

References (Bacha et al):
Summary/Description:

A new cohort of patients with a diagnosis of Atrioventricular Septal Defect was established to achieve the following objectives:

- Define the anatomic limits of balanced Atrioventricular Septal Defect (AVSD) using a data-driven approach
- Determine demographic/morphologic/physiologic factors that are associated with selection of surgical strategy
- Identify relationships between patient characteristics, morphology, selection of surgical strategy, and outcomes (survival, freedom from re-intervention)
- Develop and evaluate a clinically relevant prediction model to assist in selection of surgical strategy with specific focus on the evaluation of balanced and unbalanced AVSD.

Inclusion Criteria:

1. Diagnosis of or referral with complete AVSD at a CHSS member institution after January 1, 2012 and within first year of life (Entry criteria include: Tetralogy of Fallot, Double Outlet Right Ventricle with complete AVSD)
2. Atrioventricular and Ventriculoarterial concordance

Exclusion Criteria:

1. Partial or Transitional AVSD
2. Aortic Atresia
3. Heterotaxy
4. Total or Partial Anomalous Venous Connection

Status:

The Data Center has invited participation through direct email and announcements, newsletters and research coordinators. Twelve institutions are currently able enroll patients in the cohort. In total 84 eligible patients have been enrolled. A group of echocardiographers have set up a virtual core lab and are prepared to train research teams at participating institutions on types of images to be collected during patient visits. The virtual echo core lab will allow participating institutions to upload de-identified echo images for independent review.

Plan:

The Data Center invites your participation. Instructions to initiate participation in the cohort have been uploaded on our website at [http://www.chssdc.org/studies](http://www.chssdc.org/studies). Please let us know of your interest in participating in the cohort by writing to chss.dc@sickkids.ca.