

PROTOCOL

Title: Congenital Heart Surgeons' Society Studies in Congenital Heart Disease

Cohort: Tricuspid Atresia

Study: Tricuspid Atresia Study

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TABLE OF CONTENTS		page
	TABLE OF CONTENTS	2
	ABBREVIATIONS	3
	ABSTRACT	4
1	BACKGROUND INFORMATION AND RATIONALE	5
2	STUDY GOALS AND OBJECTIVES	6
3	DATABASE DESIGN	6
4	STUDY PROCEDURES	7
5	STUDY ENDPOINTS AND EVALUATIONS	8
6	MEASUREMENTS AND EVALUATIONS	8
7	STATISTICAL CONSIDERATIONS	9
8	SAFETY MANAGEMENT	10
9	STUDY ADMINISTRATION	10
10	PUBLICATIONS	13
11	REFERENCES	14
	APPENDIX I: CHSS DATA CENTER STUDIES	16

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ABBREVIATIONS

AV	Atrioventricular
CHSS	Congenital Heart Surgeons' Society
FHS	Functional Health Status
IRB	Institutional Review Board
LV	Left Ventricle
MUGA	Multi Gated Acquisition
PBF	Pulmonary Blood Flow
PHI	Personal Health Information
PS	Pulmonary Stenosis
QOL	Quality of Life
REB	Research Ethics Board
SPS	Systemic to Pulmonary artery Shunt
TA	Tricuspid Atresia
VA	Ventriculoarterial
VSD	Ventricular Septal Defect

ABSTRACT

Context:

Tricuspid Atresia (TA) is the third most common cyanotic congenital heart defect presenting in the neonatal period. Patients with TA & normally related great arteries are the ideal group to study surgical management along the single ventricle pathway because they have a normally developed left ventricle (LV) and mitral valve.

Goals and Objectives:

- To assemble a multi-institutional inception cohort of patients with TA and normally related great arteries
- To determine the impact of patient characteristics and management algorithms on outcomes for infants with TA and normally related great arteries
- To determine the overall Functional Health Status (FHS) and Quality of Life (QOL) of patients with TA and normally related great arteries

Study Design/Setting/Participants:

This is a prospective observational study based upon an inception cohort, age < 3 months when their index admission is to a CHSS institution. Eligible subjects will be enrolled after applicable written informed consent (and authorization, as applicable) has been obtained, and typically prior to discharge or when diagnosed with TA and normally related great arteries. Baseline demographics, diagnoses, and test results will be obtained through the subject's medical records. All subsequent clinical and surgical cardiac-related reports will be obtained as well. Annual cross-sectional follow-up will be conducted by way of a follow-up form and short questionnaires on the status of the subject's health and quality of life. The study is open to participation to all Congenital Heart Surgeons' Society (CHSS) member institutions. Those receiving Institutional Review Board (IRB)/Research Ethics Board (REB) approval for the study will be the participating sites. All study data will be stored securely and typically analyzed at the CHSS Data Center.

Study Measures:

Data will be analyzed for different patient characteristics, different treatment and management strategies, and the impact of both on subject outcome.

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1 BACKGROUND INFORMATION AND RATIONALE

1.1 Introduction

Tricuspid Atresia (TA) is the third most common cyanotic congenital heart defect presenting in the neonatal period. The true prevalence of tricuspid atresia is not known, but is estimated from post mortem and clinical studies of congenital heart disease to approach 3% in the autopsy series with a clinical prevalence rate of approximately 1.5%. Infants with TA are classified by the arrangement of the great arteries and the degree of pulmonary blood flow (PBF). The great arteries are normally related (Type I) in two-thirds of infants. The one-third with transposition of the great arteries (Type II & III) are excluded from the study cohort because they frequently have associated lesions that confound management and outcomes. In addition, TA patients are grouped according to the degree of PBF, as follows: Type A – Pulmonary Atresia, Type B – Restricted PBF Pulmonary Stenosis (PS) or restrictive Ventricular Septal Defect (VSD), and Type C – Unrestricted PBF, no PS and unrestrictive VSD.

The goal of this study is to assemble a multi-institutional inception cohort of patients with Tricuspid Atresia and normally related great arteries from which important clinical and complex questions can be answered. Parents/Guardians are being asked for permission to share their child's medical information with CHSS for the purpose of gaining knowledge for future treatment and management of children with this defect. Member CHSS institutions will participate in data collection for studies of patients with Tricuspid Atresia and normally related great arteries.

The Congenital Heart Surgeons' Society (CHSS) Data Center

The CHSS is a consortium of over 170 surgeons from more than 70 university-based hospitals/institutions in the United States, Canada, and South America. The CHSS member institutions all share an interest in the management and outcomes of surgery for congenital heart lesions. The CHSS Data Center was established in 1985 and is the coordinating site for several ongoing CHSS studies, with over 6,000 neonates, children and adults being followed (see Appendix 1: CHSS Data Center Studies). The CHSS Data Center extracts subject information and contacts families and subjects for follow-up information on a regular basis. The analyses from these data have enabled pediatric cardiovascular surgeons and cardiologists to utilize the best treatment options for a variety of congenital heart defects. They have also allowed for better counseling of patients and their families regarding prognostic outcomes. The collaboration of these institutions has led to improved treatment and management strategies in this population. Further, the CHSS has published many studies in several peer-reviewed journals.

1.2 Relevant Literature and Data

Tricuspid Atresia (TA) is the third most common cyanotic congenital heart defect presenting in the neonatal period. The true prevalence of TA is not known, but is estimated from post mortem and clinical studies of congenital heart disease to approach 3% in the autopsy series with a clinical prevalence of approximately 1.5%. (1) Infants with TA are classified by the arrangement of the great arteries and the degree of pulmonary blood flow (PBF). The great arteries are normally related (Type I) in two-thirds of infants, and one-third have transposition of the great arteries (Type II). (2-4).

The initial analysis evaluating the CHSS cohort of children with TA Type I was conducted on

150 patients (1999-2004) (5). The key findings from this study included a 5-year survival of 86% with approximately 75% of children reaching a Fontan operation by 3 years of age. More recently, the CHSS has just finalized a further analysis (accepted for publication) from this cohort evaluating 303 patients with the objective of determining the association with accessory pulmonary blood flow and outcomes during single ventricle palliation for children with TA Type I (6). The overall survival was 88% at 6 years of age with 83% of children undergoing a Fontan operation by 4 years of age. Survival for children with TA Type I undergoing an initial systemic to pulmonary artery shunt (SPS) was significantly worse than for children for whom the first operation was a pulmonary artery band or superior cavopulmonary connection. However, for children who underwent an initial SPS, survival was improved by avoiding main pulmonary artery intervention and closing the ductus arteriosus.

2 STUDY GOALS AND OBJECTIVES

A. The overall goal of this study is to assemble a multi-institutional inception cohort of patients with Tricuspid Atresia and normally related great arteries.

B. The overall objectives of this study are:

- (a) to determine the impact of patient characteristics and management algorithms on outcomes for infants with TA and normally related great arteries
- (b) to determine the overall Functional Health Status (FHS) and Quality of Life (QOL) of patients with TA and normally related great arteries

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3 DATABASE DESIGN

3.1 General Schema of Study Design

All patients evaluated and/or followed for TA Type I at CHSS participating member institutions will be considered for potential study eligibility and inclusion in the database. The database will be ongoing. Data collection will be ongoing for a subject's lifetime to assess for long-term outcomes of TA Type I.

3.2 Study Duration, Enrollment and Number of Sites

3.2.1 Duration of Study for Subject

Study duration will be for a patient's lifetime, from study initiation/subject enrollment and registration, or until the subject withdraws or is withdrawn or discontinued from the study.

3.2.2 Total Number of Study Sites/Total Number of Subjects Projected

The study will be conducted at up to approximately 70 CHSS member institutions, which may include approximately 65 investigative sites in the United States, 5 investigative sites in Canada and 2 sites in South America. Enrollment will be at a rate of approximately 25 patients per year.

3.3 Study Population

3.3.1 Inclusion Criteria

Diagnosis of Tricuspid Atresia with normally related great arteries

Protocol CHSS Studies in Congenital Heart Disease – TA Study

Version Date: 22 Sep, 2015

Page 6 of 17

Deleted: 01 Jul

Age < 3 months at time of diagnosis
Admitted to a CHSS institution on or after January 1, 1999; i.e., Date of Birth on or after October 1, 1998

3.3.2 Exclusion Criteria

Patients with Atrioventricular (AV) or Ventriculoarterial (VA) Discordance
First intervention at non-CHSS institution
Age > 3 months at time of diagnosis; i.e., Date of Birth before October 1, 1998

4 STUDY PROCEDURES

4.1 Subject Identification, Enrollment and Registration

Subjects will be identified from surgeons or cardiologists caring for these patients at the individual participating CHSS member institutions and upon new diagnosis on or after January 1, 1999. Potential cases will be identified at each participating institution in accordance with the IRB/REB approved screening methods, and enrolled in accordance with the IRB/REB approved consent and enrollment processes. The Patient Enrollment Form for the study will be used to enroll new subjects. The CHSS Data Center Registration Form for the study will be used to register new subjects.

4.2 Follow-Up

Upon consent ([and authorization, as applicable](#)) at the submitting institution and registration at the CHSS Data Center, Data Center staff will contact the parent/guardian of the subject to welcome them into the study and remind them of the annual contacts throughout their study participation. [Similarly, when the subject is eligible to and provides consent \(and authorization, as applicable\) to participate in the study him/herself, and the CHSS Data Center is provided with this information, the CHSS Data Center staff will contact the subject to welcome him/her into the study \(study continuation\) and remind him/her of the annual contacts throughout his/her study participation.](#)

Life-long annual cross-sectional follow-up will be conducted on this cohort. This phase is an essential component of establishing the cohort and is unique to this type of observational study. This is also important in understanding the progression of the patients being treated and managed with different approaches. Using this cohort of surviving subjects, the Data Center will utilize a study Follow-up Form [and Questionnaires](#) to collect information on the health status [and quality of life](#) of the subject. [The CHSS Data Center non-standardized questionnaires and the PedsQL™ standardized questionnaires \(7-14\) cover several aspects of quality of life issues for subjects such as health status, activity level, and medical care. Demographics/contact information and medical/surgical updates are collected on the CHSS Data Center Follow-up Form.](#) At yearly intervals, specially trained personnel from the CHSS Data Center will contact each subject or the parent/guardian of each subject, as applicable, for such follow-up (e.g., by telephone, mail, fax [or electronically](#), as appropriate).

Throughout the course of the study the subject's clinical and surgical cardiac-related reports will be obtained as well. Typically this information will be sent to the Data Center from the participating sites. In cases where the subject is followed at a non-CHSS institution or a non-participating CHSS institution, with the permission of the subject (parents/guardians, as applicable), the CHSS Data Center may send the subject (parents/guardians, as applicable) a

Protocol CHSS Studies in Congenital Heart Disease – TA Study
Version Date: [22 Sep, 2015](#)

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Consent for Release of Medical Information form, which the subject (parents/guardians, as applicable) can then sign and take to their health care provider requesting that the specified information be released to the CHSS Data Center. Alternatively, the subject (parents/guardians, as applicable) may themselves request through the institution that holds the information, that the information be forwarded to the CHSS Data Center.

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At certain times over the course of this study, through an IRB/REB approved amendment to the protocol, FHS testing and QOL questionnaires will be completed.

4.3 Subject Completion/Withdrawal

Subjects may withdraw (or be withdrawn by their parents/guardians, as applicable) from the study at any time without any impact on their care. Subjects may also be discontinued from the study at the discretion of the principal investigator at the participating CHSS institution if there is an inability to re-contact the subject/subject's parents/guardians and verify outcome, in which case the subject would be considered as 'lost to follow up'. The investigator may also withdraw subjects to protect the subject for reasons of safety or for administrative reasons. If a subject withdraws or is withdrawn or discontinued from the study at any stage, no further information about him/her will be collected for use in the analysis. However information already collected will continue to be used as needed to maintain the integrity of the research.

5 STUDY ENDPOINTS AND EVALUATIONS

5.1 Primary Endpoints

The primary study endpoint is survival-related, i.e., death and time to death.

5.2 Secondary Endpoints

Secondary endpoints will include risk factors that impact treatment strategies, overall health status, FHS and QOL.

6 MEASUREMENTS AND EVALUATIONS

6.1 Subject Identification and Data Collection

After obtaining applicable written informed consent/assent (and authorization, as applicable) the information to be collected will include the following:

- Completed Patient Enrollment Form
- CHSS Data Center Registration Form
- Demographic information
- Admission history and physical (to include height, weight, oxygen saturation, signs and symptoms)
- Any subsequent hospital admission (admit history and reports)
- Catheterization reports (diagnostic and/or therapeutic)
- Cardiac related operative reports
- All Echocardiography reports, including any Transesophageal Echo
- MRI reports, Holter, exercise test results
- Nuclear Medicine (Multi Gated Acquisition (MUGA) scan, lung perfusion scans)

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- Discharge summaries
- Clinic Letters
- Autopsy/death report (if applicable)
- Completed Annual Follow-up Form and Questionnaires

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The annual follow-up form and questionnaires will collect data regarding health status, quality of life, medications, activity restrictions, exercise-related symptoms, and procedures since initial diagnosis/last follow-up.

Identifying information/personal health information collected will include the following: subject and parent name; date of birth; medical record number; home address; telephone numbers; alternative contacts and contact information; referring physician information; hospital where records obtained.

Demographic, morphologic and procedural data will be abstracted from institutional medical records, echocardiograms and surgical and interventional reports. Baseline morphologic characteristics will be defined based on the initial echocardiogram. Subsequent morphologic details will be abstracted from echocardiogram reports that most immediately precede the corresponding surgical intervention.

7 STATISTICAL CONSIDERATIONS

7.1 Statistical Methods

Two expert statistical consultants (Dr. E Blackstone and Dr. B McCrindle) supervise all aspects of data analysis and summation.

The CHSS Data Center has a history of incorporating advanced statistical methodologies for generating important knowledge relating to congenital heart surgery. The methods described below have been previously utilized in our research; however, we continue to incorporate novel techniques with the help of the Quantitative Health Sciences Department Heart and Vascular Institute at the Cleveland Clinic Foundation.

Descriptive statistics will be compiled, with data described as frequencies, medians with ranges, or means with standard deviations as appropriate. In addition to traditional inferential statistics to compare groups and determine associations, advanced techniques will include multiphase parametric modeling of time-related events in the hazard domain, competing risk methodology (for competing and mutually exclusive end-states), analysis of repeated events (modulated renewal), and incorporation of time-varying covariates (usually interim procedures). These will be utilized to identify risk factors for various outcomes including survival, conversion to various end-states, and functional outcomes. Demographic, morphologic, physiologic, institutional, and procedural risk factors associated with each hazard phase of mortality, morbidity, adverse late functional or neurological outcome, or re-intervention will be sought by multivariable analysis, including hazard analysis as originally described by Blackstone and colleagues (15). The bootstrap method will be utilized to guide final variable selection for multivariable models.

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7.2 Sample Size and Power

For each subsequent study analysis to be performed the Data Center will compute sample size/power to ensure statistically meaningful results.

8 SAFETY MANAGEMENT

This is a prospective observational study and does not include any drug administration, special imaging needs or surgical intervention for research purposes. It is a minimal risk study. The only intervention is the annual follow-up form and questionnaires. The only risk is release of identifying information/personal health information (PHI) and loss of confidentiality. Every effort will be made to keep identifying information and PHI from unauthorized disclosure. Any breach will be reported.

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9 STUDY ADMINISTRATION

9.1 Data Collection and Management

1. *Privacy*: Each CHSS member institution associated with this study will have IRB/REB approval from their IRB/REB, and will utilize a Data Transfer Agreement with the CHSS Data Center to maintain the highest level of confidentiality for all study participants. After applicable written informed consent (and authorization, as applicable) is obtained, each subject's medical record will be reviewed for baseline and surgical (if applicable) data. This information, including demographic information, will be collected, and along with the consent form (and authorization, as applicable) and completed Patient Enrollment Form, will be securely transferred to the Data Center. Likewise, when the subject provides assent, and when the subject is eligible to and provides consent (and authorization, as applicable) to participate in the study him/herself, the signed forms and any updates in contact information will be securely transferred to the Data Center as well. Similarly, subsequent follow up information and questionnaires will be securely transferred to and obtained by the Data Center. Upon enrollment by the participating institutions, each subject is assigned a unique screening number. Upon registration at the Data Center, each subject is assigned a unique study number that is used for all data entry and analysis purposes, and the CHSS Data Center Registration Form is completed. Trained dedicated personnel at the Data Center will perform all data extraction and entry of specific variables, in a de-identified manner, into a secure, password protected computerized database housed at the Data Center. A master list (key) will be kept separate from the study data. Only appointed personnel at the Data Center will be able to connect the individual subject to the data. The Data Center will be responsible for maintaining a log of IRB/REB approvals and checking to ensure that participating sites are not submitting records or data without the appropriate IRB/REB approval documentation on file.

2 *Security*: Appropriate patient identity protection safeguards will be observed by the participating CHSS member institutions, as well as the CHSS Data Center, for the transmission of the patient information (e.g., baseline and follow-up patient care charts or compact discs (CDs) or echo tapes). Secure file transfer or secure courier service will be used as appropriate. The Follow-up Form and Questionnaires will be securely provided to the subjects or parents/guardians (as applicable), and returned to the Data Center by the subjects (or parents/guardians), in a secure manner acceptable to the subjects (parents/guardians, as applicable) (e.g., mail, fax, electronically). The master list (key) and the study data will all be securely stored (using a double lock system) at the Data Center, and kept separate from each

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other. Likewise, the patient information sent to the Data Center, including CDs of echocardiograms, magnetic resonance images (MRIs), CT scans and cardiac catheterizations, will be securely stored separately at the Data Center. The CDs will be sent to the Data Center securely with the subject name on the outside; once at the Data Center, the name will be removed and the study number will replace the name. When possible, the scans will only contain de-identified data prior to being sent to the Data Center. However, it may not always be possible to remove identifiers from the scans. If this is the case, all attempts at keeping the subject information confidential will be made as described above.

3. *De-identification*: Each subject is assigned a unique study number at the CHSS Data Center that is used for all data entry and analysis purposes. All data analysis, review, and published results will be performed in a de-identified manner.

9.2 Confidentiality

All data and records generated for this study will be kept confidential in accordance with applicable institutional policies, laws and regulations. The investigators and site personnel and the coordinating site will use the study data and records only for these study purposes. Safeguards are described under Data Collection and Management. The information collected as part of this study will be securely retained for 7 years after all study publications are completed. The research information will be securely destroyed according to the applicable institutional policy effective at that time.

9.3 Regulatory and Ethical Considerations

9.3.1 Compliance Statement

This study will be conducted in compliance with each participating CHSS member institutions' research policies and procedures and all applicable laws and regulations. The participating sites will perform the study in accordance with this protocol, will obtain consent [\(and authorization, as applicable\)](#) and assent (as appropriate), and will report unexpected problems in accordance with CHSS member institutions' IRB/REB policies and procedures and applicable laws and regulations. Collection, recording, and reporting of data will be accurate and will ensure the privacy, health and welfare of research subjects during and after the study.

9.3.2 Data and Safety Monitoring Plan

The study investigators will be responsible for safety monitoring. This is an observational study which involves only data collection from the hospital/institutional sources (e.g., operative notes, echocardiogram, cardiac catheterization, electrocardiogram, etc.) as well as from the annual follow-up [form and questionnaires](#) sent to the subjects or parents/guardians. This study does not dictate any specific surgical treatment or medications. All procedures performed on the patients are standard of care and the study does not involve any additional hospital visits for the subjects or parents/guardians. There is minimal safety risk with this study, mainly from the potential for breach of privacy and loss of confidentiality. The investigators and site personnel and the CHSS Data Center personnel will ensure that confidential information, including PHI/identifying personal information, will be secured as described above and will not be revealed to unauthorized parties.

9.3.3 Risk Assessment

The main risk in this study is the potential breach of privacy and loss of confidentiality. There is a minimal risk of likelihood of harm. All reasonable safeguards to secure the confidentiality of information will be taken by the investigators and their research personnel and the CHSS Data Center personnel. Safeguards are described under Data Collection and Management. We believe this study overall is minimal risk.

9.3.4 Potential Benefits of Study Participation

Information collected may contribute to the care of children in the future who have the same heart condition as those that participate in this study. The information may also improve the future management of these patients. There may be no direct benefit to the subject from participation in this study.

9.3.5 Risk-Benefit Assessment

The study as a whole represents minimal risk to the subjects. The potential benefit of identifying children with Tricuspid Atresia and normally related great arteries and enrolling them into this study outweighs the risk of participation.

9.4 Recruitment Strategy

Subjects will be identified from surgeons or cardiologists caring for these subjects at the individual CHSS member institutions. Subjects at each of the participating CHSS member institutions will be identified using their own IRB/REB approved methods for screening, consenting and enrolling subjects.

9.5 Informed Consent/Assent

Applicable written informed consent/assent ([and authorization, as applicable](#)) will be obtained at the time of the patient's routine clinical assessment at the CHSS member institution. The consent ([and authorization, as applicable](#)) will be discussed at length and ample time will be allowed for the parent/guardian and subject (as applicable) to discuss it amongst themselves and decide whether they would like to participate. The principal investigator at each participating CHSS member institution, his or her designees/study coordinator will have responsibility for subject recruitment and obtaining applicable written informed consent/assent ([and authorization, as applicable](#)) for study participation. As part of the study, written consent will include [a combined or separate authorization \(as appropriate\)](#) to have the subject's medical information securely sent to the CHSS Data Center for data abstraction and entry into the study database. Site investigators and personnel will not allow information from the subjects' charts to be sent to the CHSS Data Center unless applicable written consent and [such](#) authorization have been obtained. Once the parent/guardian or subject (as applicable) has provided applicable written informed consent, [and authorization \(as applicable\)](#), the data can be securely sent to the CHSS Data Center. The site investigator's phone number will be on the consent form and the original will be maintained in the subject's confidential study records at the respective participating site. The CHSS Data Center will not register any living patient until a copy of the signed consent form [and authorization \(as applicable\)](#) is received.

9.6 Inclusion of Deceased Patients

Inclusion in the study is requested for patients identified prospectively at the participating CHSS member institutions, but who die before written informed consent is obtained, and will be

conducted in compliance with all applicable laws and regulations, including approval for waiver/alteration of the consent process (as applicable). Families of these patients will not be contacted for research purposes. Only the information already in the medical record will be used and only aggregated data will be reported in publications, precluding families from identifying themselves in any publication. When a participating CHSS institution sends information on a deceased patient, the CHSS Data Center will follow the same procedures to include them in the study that are used for those enrolled while living patient participants.

10 PUBLICATIONS

We anticipate the results of this study will be presented at national meetings and/or published in academic journals. We will not disclose any subject PHI or identifying personal information in any presentation or publication about the study

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APPENDIX I: CHSS DATA CENTER STUDIES

Background Information

Dr. John Kirklin and Dr. Eugene Blackstone established the CHSS Data Center at the University of Alabama, Birmingham in 1985. The Data Center's role is to coordinate the CHSS cooperative studies.

The first study began in 1985, with infants with Transposition of the Great Arteries (TGA) entering participating institutions within 14 days of birth. Enrollment continued until 1989, and by this time 890 infants were entered. In subsequent years, the following studies have been initiated.

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<u>Diagnostic Group:</u>	<u>Accrual Date:</u>
Transposition of the Great Arteries [TGA]	1985-1989
Interrupted Aortic Arch [IAA]	1987-1997
Pulmonary Atresia with Intact Ventricular Septum [PAIVS]	1987-1997
Pulmonary Stenosis with Intact Ventricular Septum [PSIVS]	1987-1997
Coarctation of the aorta [CoA]	1990-1993
Aortic Valve Atresia [AVA]	1994-2000
Aortic Valve Stenosis (or hypoplasia of the LV inflow/outflow without complete atresia) [AVS]	1994-2000
Tricuspid Atresia [TA]	1999-Present
Pulmonary Conduit [PC]	2002-2014
Critical Left Ventricular Outflow Tract Obstruction [LVOTO]	2005-Present
Anomalous Aortic Origin of Coronary Arteries [AAOCA]	2009-Present
Atrioventricular Septal Defect [AVSD]	2012-Present

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Follow up for all studies initiated prior to the end of 1997 were put on hold as the CHSS Data Center was relocated at The Hospital for Sick Children in Toronto, Ontario, Canada.

How many centers participate in the studies?

At present there are over 70 centers that participate in the CHSS studies in North America, as well as representation from Argentina and Brazil.

What is the role of the CHSS Data Center?

The Data Center regularly contacts each participating institution regarding new patients being enrolled in studies. The Data Center also facilitates the follow-up of patients enrolled in studies.

Confidentiality is respected and no information that discloses patient identity is released without consent unless required by law.

The Data Center website (www.chssdc.org) is used to disseminate information to member institutions and patients.

What is the role of the participating center?

Each center has an identified contact person who is responsible for **securely** sending patient information **to the CHSS Data Center** according to identified criteria for a specific study.

Each center also participates in the regularly scheduled follow-up of patients who have been

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Protocol CHSS Studies in Congenital Heart Disease – TA Study
Version Date: **22 Sep, 2015**

enrolled in specific studies. This would include securely providing information about the patient since the last follow-up, for example: demographic information, operative notes, catheterization and echocardiogram reports.

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Follow-Up Form/Questionnaires Data Collection

The CHSS Data Center conducts annual follow-up for each of the studies. Study participants and/or their parents/guardians, as appropriate, are contacted on an annual basis and requested to complete the annual follow-up form (e.g., providing any updates in contact information and medical/surgical updates, as applicable), and also to complete the questionnaires for the study. The questionnaire data collection will be done using non-standardized CHSS Data Center questionnaires, and standardized questionnaires including the PedsQL™ quality of life and cardiac modules.

Functional Health Status Data Collection

As the participants in the prospective observational cohorts grow up from childhood to adolescence and adulthood, the Data Center may also undertake systematic data collection on their functional health status during this transition. These data collection can be specific to one or more cohorts, depending upon the patients' age in the cohort. This data collection will be done using standardized data collection instruments such as:

1. Transition Readiness Assessment Questionnaire (TRAQ)
2. The MyHeart Scale

Mechanism by which studies are conducted:

The CHSS members meet annually to review the progress of each study, decide on which lesion will be studied, determine inclusion and exclusion criteria, data sources, parts of the medical record and mechanism for follow-up. In the past, the data collection was not hypothesis driven. This was instituted when the Data Center relocated to The Hospital for Sick Children in 1997.

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