



CHSS DATA CENTER NEWSLETTER

Welcome from the Managing Director

Dr. William M. DeCampi



The Newsletter is back! We have rejuvenated this medium for communicating Data Center activities, summarizing progress in the studies, and informing Members and data coordinators of important changes in our enrollment and follow up (E&F) and regulatory processes. With each issue, I will write a short summary of a pressing issue.

The success of the research mission of the CHSS depends on the commitment and collaboration of multiple groups. In particular, I cannot over-emphasize the importance of our group of institutional data coordinators (IDC). Coordinators are the “core” of our operation because they detect, enroll and provide clinical follow up of our cohort population. The task is not easy owing to the unique nature of the studies we do—long-term annual longitudinal follow up in multiple cohorts of patients. Two years ago, Linda Lambert, ARNP, of Primary Children’s organized coordinators into the “IDC Group”. Meeting periodically by conference call, this group openly discusses

the challenges of the E&F processes, including consenting and other regulatory issues. IDC members now hold a mini-conference at the CHSS Annual Meeting (and receive CE credit). There, IDC members share ideas, help each other out, and listen to study presentations that are the fruits of their labor. Highly-productive coordinators are presented awards in front of the CHSS surgeon and IDC membership.

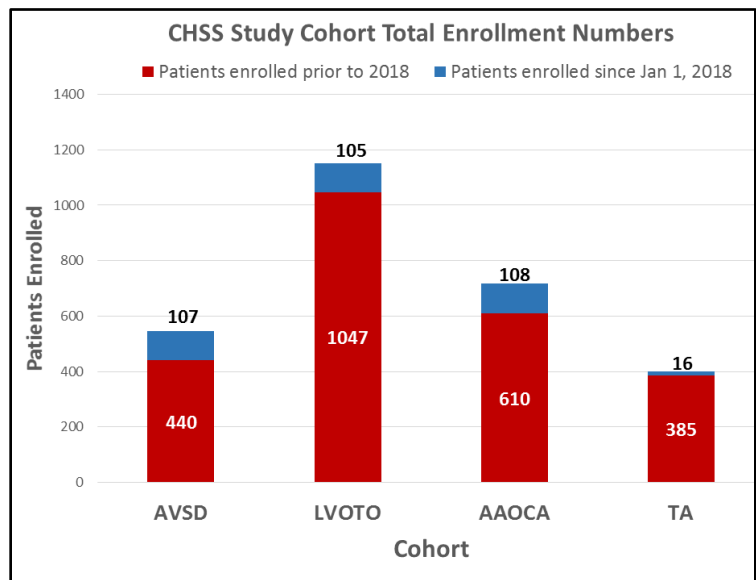
If you are a CHSS surgeon member and do not engage a coordinator in the CHSS studies, I encourage you to do so. Neither surgeons, residents nor clinical practitioners have the time or resources to capture, enroll and follow the steadily growing cohort populations. In some cases (for example, AAOCA and Ebsteins) the surgeon may not even be aware of eligible patients who are managed medically. High percentage E&F at each CHSS institution is critical to data quality in our studies. The workload can be considerable. Allot time for your coordinator to work on the cohorts. Show them appreciation for doing so. Teach them the nature, impact and uniqueness of our studies so they know they are engaged in a very special endeavor, ultimately for the benefit of children with the most complex congenital cardiac disorders.

Thank you to all the CHSS members and coordinators for your time and valuable work!

STUDY ENROLLMENT

Thank you to all site coordinators and CHSS members who helped with the these numbers. We could not do without your great work.

Figure 1. Total enrollment in actively enrolling CHSS cohorts: AVSD, LVOTO, AAOCA and TA as of January 8, 2019. The number of patients enrolled since January 1, 2018: AVSD=107; LVOTO=105; AAOCA=108; TA = 16. Total enrollment for each cohort: AVSD= 547; LVOTO=1152; AAOCA=718; TA=401.





DR. PAUL DEVLIN, KIRKLIN-ASHBURN FELLOW – PROGRESS UPDATE



As I am entering the final 6 months of the Kirklin-Ashburn Fellowship, I am appreciative of what an incredible opportunity my time in Toronto has been for my academic development and future in congenital cardiac surgery. Whether it was through weekly pathology sessions with Dr.

Williams or thoughtful discussion with many of you at Work Weekends discussing current projects, I have learned a lot regarding congenital heart surgery and the statistical analysis of longitudinal outcomes.

The current projects that I am working on include: an analysis of late survival and patient-perceived functional health status in the transposition of the great arteries cohort for presentation at the STS Meeting later this month in San Diego; analyses of the use of PA banding in the atrioventricular septal defect cohort and surgical outcomes in the AAOCA for the AATS in Toronto; completing the manuscript for the CHSS critical aortic stenosis calculator evaluation project that I presented at the AHA in November; and preparing for my thesis defense in early March, which is based on the post-Norwood aortic arch reintervention project that has been accepted for publication in JTCVS.

Thank you to all the CHSS members who have contributed to these projects through participation at the Work Weekends and on the many working group conference calls and email threads. I look forward to seeing those of you who are able to attend at the Spring Work Weekend. Finally, I'd like to extend a huge thank you to the staff at the Data Center and the Data Coordinators at each participating institution. We have made great strides in improving the tracking of patient data and streamlining the process of keeping patient records up to date in the database, while continuing to be productive in data analysis. Without the diligent and attentive work of the entire Data Center team, none of this would have been possible.

AMENDMENT ROLLOUT

In 2018 an amendment for the TGA study was implemented at participating sites.

The purpose of this amendment was to re-establish contact with subjects who have been lost to follow-up in the TGA cohort by reaching out to adult congenital heart disease clinics.

In addition, changes were also made to the CHSSDC welcome pamphlet, the annual follow-up forms, and electronic follow-up documents were added as an option for subjects who prefer electronic correspondence.

Sites participating in any of the below cohort studies can expect to receive an email from the Data Coordinating Centre with information to submit to your local IRB/REB:

- Enrolling: TA
- Non-enrolling: *IAA, PAIVS, PSIVS, AVA, AVS, PC*

EBSTEIN ANOMALY Inception Cohort

The CHSSDC is planning to launch the new Ebstein Anomaly (EA) study in early 2019. Thank you for sending your feasibility survey responses to us earlier this month. The CHSSDC is currently reviewing your responses and will reach out to the interested sites to help you with the onboarding process (IRB/REB, DTA, site specific training).

Part of this cohort will consist of a retrospective enrollment of patients diagnosed with EA at your institution between January 1, 2014 up to the date of your IRB approval date. You will then enroll patients prospectively.

This study will be looking at patients with Ebstein Anomaly who are treated:

- through monitoring
- medical management, or
- surgical intervention

Any questions, please contact Tharini Paramanathan at 1-866-477-CHSS (2477).



PATIENT TRACKING

A reminder that every participating site should have a process in place to track all patients that have been identified as potentially eligible (pre-screening), screened for inclusion, and/or enrolled in a CHSS study.

This provides important information on how many patients were eligible for enrollment vs. actual enrollment. It may also be a requirement of your IRB/REB.

Contact us at the Data Center if you need any help to develop a tracking document template. Depending on your institution, these tracking documents may require IRB/REB approval as they will contain personal health information (PHI).

RE-CONSENT AND ASSENT

One of the strengths of our research is the data obtained through annual follow-up of patients. This life-long cross-sectional follow-up is one way our studies stand out from other publications. Collecting this data annually presents many challenges. Patients get older and families move around.

Ensuring that re-consents and assents are obtained in a timely manner is essential. Sites should get in touch with their IRB/REB to make sure they are aware of their requirements.

How your institution can help:

- Know your IRB requirements related to assents, consents, and re-consents.
- Develop processes to identify patients who need re-consent and assent.
- Work with your IRB to find solutions to challenging situations i.e. can assents be obtained over the phone? Can a waiver be obtained that would allow CHSS to contact patients and obtain consent directly for those patients who will not be returning to your institution?
- Provide the Data Center with any re-consents, assents or waivers at least 2 weeks prior to follow-up start date.

SPRING WORK WEEKEND 2019

The CHSS Data Center hosts biannual Work Weekends in Toronto, Canada. This is an opportunity for the membership to learn more about the research taking place at the Data Center. We encourage all members to participate, either in person or via webinar.

Please save the date for the next Work Weekend, to be held in Toronto on **April 12-14, 2019**. Registration will open in mid-March and email invites will be sent to members via email. For updates please visit <http://www.chssdc.org/content/chss-work-weekend>

Our most recent Work Weekend was held November 16-18, 2018. We thank all participants for making the 2018 Fall Work Weekend a success! Thank you for your continued support of the CHSS research studies.



DATA CENTER CONTACTS

General phone number: 1-866-477-CHSS (2477)

General email address: chss.dc@sickkids.ca

CHSS Study	Primary Contact
Atrioventricular Septal Defect (AVSD)	Tharini Paramanathan tharini.paramanathan@sickkids.ca
Ebstein's Anomaly (EA)	Tharini Paramanathan tharini.paramanathan@sickkids.ca
Critical Left Heart Obstruction (LVOTO)	Kate Pearson kate.pearson@sickkids.ca
Pulmonary Conduit (PC) *Non-enrolling	Nabi Aghaei nabi.aghaei@sickkids.ca
Anomalous Aortic Origin of Coronary Artery (AAOCA)	Nabi Aghaei nabi.aghaei@sickkids.ca
Tricuspid Atresia (TA)	Anna Wasiak anna.wasiak@sickkids.ca



2019 ANNUAL FOLLOW-UP

In order to catch up with the annual follow ups for each cohort we have proposed a CHSS Follow-Up schedule for 2019.

We hope that the following schedule will assist you in obtaining and sending the appropriate documentation to us in time. Please contact our Clinical Research Project Assistants (Arti Singh & Julia Lo) or email chss.dc@sickkids.ca if you would like to receive a list of subjects enrolled from your site who have reached/are reaching an assenting/re-consenting age.

Cohort	Status	Last Patient Follow Up	Tentative Follow-Up
Aortic Valve Atresia [AVA]	Non-Enrolling	Sep to Nov 2014	Feb 2019
Pulmonary Conduit [PC]	Non-Enrolling	Aug to Oct 2014	Mar 2019
Tricuspid Atresia [TA]	Enrolling	May to Jul 2015	Apr 2019
Atrioventricular Septal Defect [AVSD]	Enrolling	Aug to Oct 2017	May 2019
Transposition of the Great Arteries [TGA]	Non-Enrolling	Jun to Aug 2017	Jun 2019
Critical Left Heart Obstruction [LVOTO]	Enrolling	Sep to Nov 2017	Jul 2019
Pulmonary Atresia- Intact Ventricular Septum [PAIVS/PSIVS]	Non-Enrolling	Jan to Mar 2018	Aug 2019
Interrupted Aortic Arch [IAA]	Non-Enrolling	Jun to Aug 2018	Sep 2019
Aortic Valve Stenosis [AVS]	Non-Enrolling	Nov 2018 to Jan 2019	Oct 2019
Anomalous Aortic Origin of a Coronary Artery [AAOCA]	Enrolling	Dec 2018 to Feb 2019	Nov 2019
Ebstein Anomaly [EA]	New		Dec 2019 or Jan 2020



STATUS OF CURRENT STUDIES

Study Name	P.I.	Start Date	Status	Working Group
AVSD – Outcomes in BiV Repair of Complete AVSD and Defining Unbalance	D. Overman	Aug. 2018	Data Abstraction and Study Design in Progress	P. Devlin, A. Jegatheeswaran, B. McCrindle, T. Karamlou, E. Blackstone, W. Williams, W. DeCampli, L. Mertens, C. Fackoury, P. Eghtesady, J. Jacobs, J. Baffa, C. Fleishman, A. Dodge-Khatami, C. Pizarro, K. Pourmoghadam, M. Cohen, D. Meyer, D. Overman
AAOCA – Surgical Outcomes in AAOCA	B. McCrindle	May 2018	Accepted to AATS 2019	A. Jegatheeswaran, P. Devlin, W. Williams, J. Brothers, M. Jacobs, R. Lorber, L. Mertens, C. Mery, W. DeCampli, C. Fleishman, C. Caldarone, J. Kirklin, S. Molossi, B. McCrindle
AVSD – Pulmonary Artery Banding in complete AVSD	D. Overman	Feb. 2018	Accepted to AATS 2019	P. Devlin, A. Jegatheeswaran, B. McCrindle, T. Karamlou, E. Blackstone, W. Williams, W. DeCampli, L. Mertens, C. Fackoury, P. Eghtesady, J. Jacobs, J. Baffa, C. Fleishman, A. Dodge-Khatami, C. Pizarro, K. Pourmoghadam, M. Cohen, D. Meyer, D. Overman
TGA – Late Survival and Functional Health Status	B. McCrindle	Jan. 2018	Accepted to STS 2019	P. Devlin, A. Jegatheeswaran, W. Williams, E. Blackstone, W. DeCampli, L. Lambert, K. Mussatto, C. Prospero, I. Bondarenko, B. McCrindle
LVOTO – CHSS Critical AS Calculator Evaluation	B. McCrindle	Oct. 2017	Presented at AHA 2018	P. Devlin, E. Hickey, C. Morgan, A. Jegatheeswaran, W. DeCampli, W. Williams, J. Kirklin, E. Blackstone, W. Douglas, L. Mertens, B. McCrindle
AAOCA – Features associated with ischemia in AAOCA	J. Brothers	July 2017	Presented at AATS 2018	A. Jegatheeswaran, P. Devlin, B. McCrindle, W. Williams, M. Jacobs, E. Blackstone, W. DeCampli, C. Caldarone, J.W. Gaynor, J. Kirklin, R. Lorber, C. Mery, J. St. Louis, S. Molossi, J. Brothers
LVOTO – Arch Reintervention After Norwood	T. Karamlou	Jun 2017	Presented at AATS 2018	P. Devlin, B. McCrindle, J. Kirklin, E. Blackstone, W. DeCampli, C. Caldarone, A. Dodge-Khatami, P. Eghtesady, J. Meza, P. Gruber, K. Guleserian, B. Alsoufi, L. Lambert, J. O'Brien, E. Austin III, J. Jacobs, T. Karamlou



MANUSCRIPTS

Title	P.I.	Target Journal & Submission Date	Status	Working Group
Surgery in Anomalous Aortic Origin of a Coronary Artery: A quantification of risk	J. Brothers	JTCVS 5/2019	In Development	A. Jegatheeswaran, P. Devlin, W. Williams, J. Brothers, M. Jacobs, R. Lorber, L. Mertens, C. Mery, W. DeCampli, C. Fleishman, C. Caldarone, J. Kirklin, S. Molossi, B. McCrindle
Pulmonary Artery Banding in Complete Atrioventricular Septal Defect: Association with Pre-Discharge Common Atrioventricular Valve Function and Early Outcomes	D. Overman	JTCVS 5/2019	In Development	P. Devlin, A. Jegatheeswaran, B. McCrindle, T. Karamlou, E. Blackstone, W. Williams, W. DeCampli, L. Mertens, C. Fackoury, P. Eghtesady, J. Jacobs, J. Baffa, C. Fleishman, A. Dodge-Khatami, C. Pizarro, K. Pourmoghadam, M. Cohen, D. Meyer, D. Overman
Evaluating the Congenital Heart Surgeons' Society Critical Aortic Stenosis Calculator in a New Patient Cohort	B. McCrindle	TBD 6/2019	In Development	P. Devlin, E. Hickey, C. Morgan, A. Jegatheeswaran, W. DeCampli, W. Williams, J. Kirklin, E. Blackstone, W. Douglas, L. Mertens, B. McCrindle
Late survival and patient-perceived functional health status of the CHSS TGA Cohort	B. McCrindle	Annals of Thoracic Surgery 1/2019	Submitting	P. Devlin, A. Jegatheeswaran, W. Williams, E. Blackstone, W. DeCampli, L. Lambert, K. Mussatto, C. Prospero, I. Bondarenko, B. McCrindle
Functional health status in young adults with repaired transposition of the great arteries	B. McCrindle	JACC 1/2019	Submitting	A. Jegatheeswaran, P. Devlin, K. Welke, E. Blackstone, P. Kirshbom, L. Lambert, C. Tchervenkov, R. Woods, K. Mussatto, T. Cotts, J. St. Louis, J. Jacobs, M. Jacobs, C. Caldarone, W. Williams, S. Fuller, B. McCrindle
Anomalous Aortic Origin of a Coronary Artery: Are we Closer to Risk Stratification?	J. Brothers	JTCVS 5/2018	In Revisions	A. Jegatheeswaran, P. Devlin, B. McCrindle, W. Williams, M. Jacobs, E. Blackstone, W. DeCampli, C. Caldarone, J.W. Gaynor, J. Kirklin, R. Lorber, C. Mery, J. St. Louis, S. Molossi, J. Brothers
Intervention for Arch Obstruction Following Norwood: Prevalence, Associated Factors, and Practice Variability	T. Karamlou	JTCVS 5/2018	Accepted- In Proofing	P. Devlin, B. McCrindle, J. Kirklin, E. Blackstone, W. DeCampli, C. Caldarone, A. Dodge-Khatami, P. Eghtesady, J. Meza, P. Gruber, K. Guleserian, B. Alsoufi, L. Lambert, J. O'Brien, E. Austin III, J. Jacobs, T. Karamlou



The Congenital Heart Surgeon's Society Complete Atrioventricular Septal Defect Cohort: Baseline, Preintervention Echocardiographic Characteristics	D. Overman	Seminars in Thoracic and Cardiovascular Surgery	Accepted	J. Meza, P. Devlin, D. Overman, D. Gremmels, J. Baffa, M. Cohen, M. Quartermain, C. Caldarone, K. Pourmoghadam, W. DeCampli, C. Fackoury, L. Mertens
Pre-intervention morphologic and functional echocardiographic characteristics of neonates with critical left heart obstruction - a CHSS inception cohort study	L. Mertens	European Heart Journal- Cardiovascular Imaging	Accepted	M. Slieker, J. Meza, P. Devlin, P. Burch, T. Karamlou, W. DeCampli, B. McCrindle, W. Williams, C. Morgan, C. Fleishman, L. Mertens
A novel, data-driven conceptualization for critical left heart obstruction	B. McCrindle	Computer Methods and Programs in Biomedicine	Accepted	J. Meza, M. Slieker, E. Blackstone, L. Mertens, W. DeCampli, J. Kirklin, M. Karimi, P. Eghtesady, K. Pourmoghadam, R. Kim, P. Burch, M. Jacobs, T. Karamlou, B. McCrindle
Surgical Palliation or Primary Transplantation for Aortic Valve Atresia?	G. Pettersson	JTCVS	In Revisions	K. Stackhouse, B. McCrindle, E. Blackstone, J. Rajeswaran, J. Kirklin, C. Tchervenkov, M. Jacobs, J. Jacobs, G. Pettersson