Annual Report
Congenital Heart Surgeons Society
Data Center

William G. Williams, M.D., FRCSC

William M. DeCampli, M.D., Ph.D

October 19, 2014

ARNOLD PALMER HOSPITAL
For Children
Supported by Arnold Palmer Medical Center Foundation

UNIVERSITY OF CENTRAL FLORIDA
College of Medicine

Personnel: Faculty

- Bill Williams, Executive Director
- Bill DeCampli, Managing Director
- Brian McCrindle
- Gene Blackstone
- Ed Hickey
- Luc Mertens

Personnel: Staff

- Sally Cai (Database Manager)
- Annette Flynn (Research Project Assistant)
- Susan McIntyre (Research Nurse Coordinator)
- Ilina Ristevska (Research Project Assistant)
- Veena Sivarajan (Research Nurse Coordinator)

Kirklin/Ashburn Fellowship

- 6th and current fellow: Travis Wilder
- Funding: salary: CHSS; tuition for advanced degree from UT: Div CT Surgery, HSC
- 7th fellow (2015-6)
  - Selection committee: DC faculty + Chairman of Research Committee
  - 11 candidates: 4 referred by CHSS members

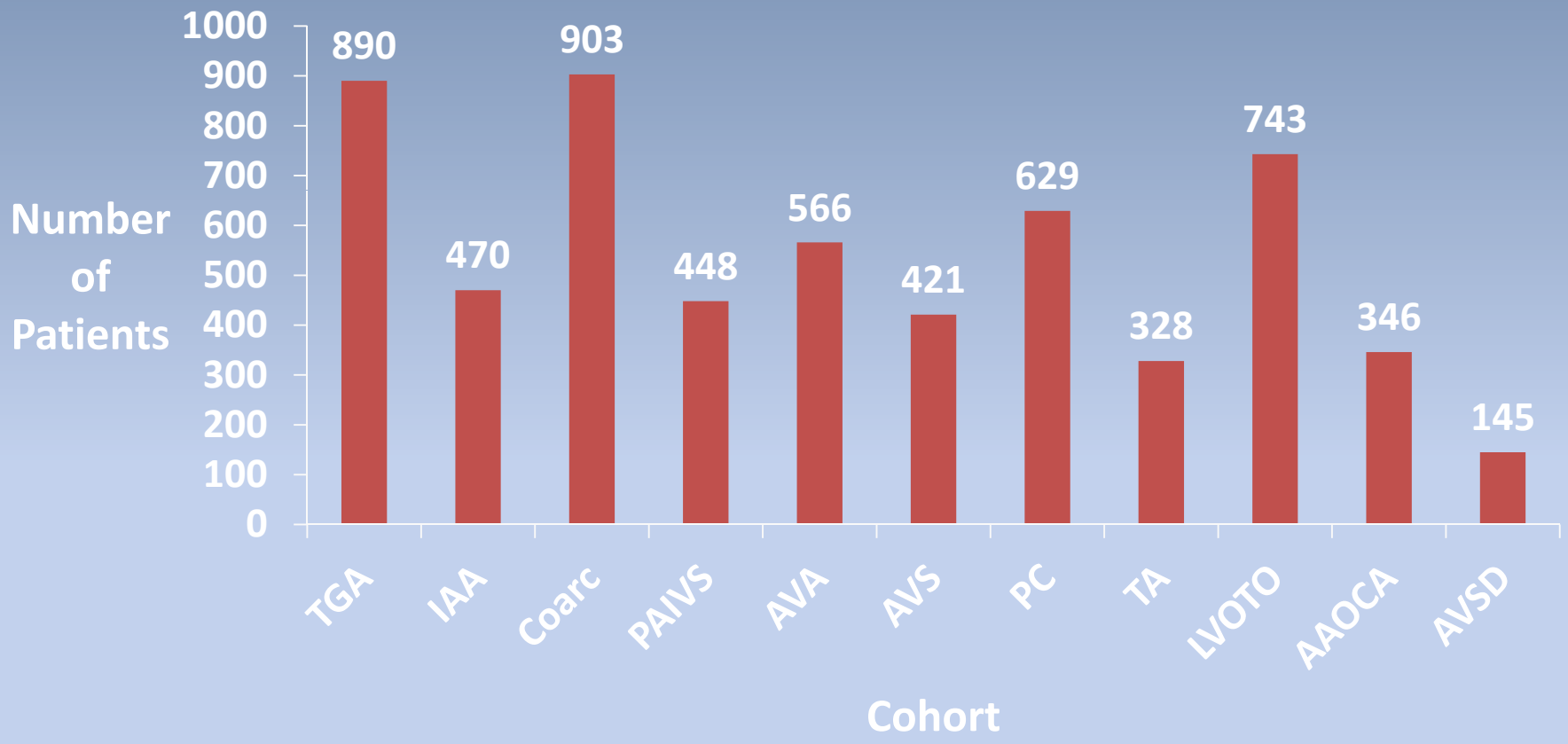
James M. Meza, M.D.

- PGY3 Surgery Resident at Duke
- Cardiothoracic Surgery track
- Also pursuing Masters of Health Science in Clinical Research at Duke
- BS Princeton, MD University of Michigan
- Referred by Jake Jaquiss

Cohorts
(as of 10/1/2014)

Diagnostic Group	Number of Institutions	Accrual Date	N
Transposition of Great Arteries	22	1985-1989	890
Pulmonary Atresia with Intact Ventricular Septum	31	1987-1997	448
Interrupted Aortic Arch	32	1987-1997	470
Coarctation of the Aorta	32	1990-1993	903
Aortic Valve Atresia	21	1994-2000	566
Critical Aortic Stenosis	21	1994-2000	421
Tricuspid Atresia	38	1999-Present	328
Pulmonary Conduit	28	2002-2014	629
Critical Left Ventricular Outflow Tract	21	2005-Present	743
Anomalous Aortic Origin of Coronary Arteries	34	2009-Present	346
Atrioventricular Septal Defect	16	2012-Present	145

Total Number of Patients in CHSS Cohorts

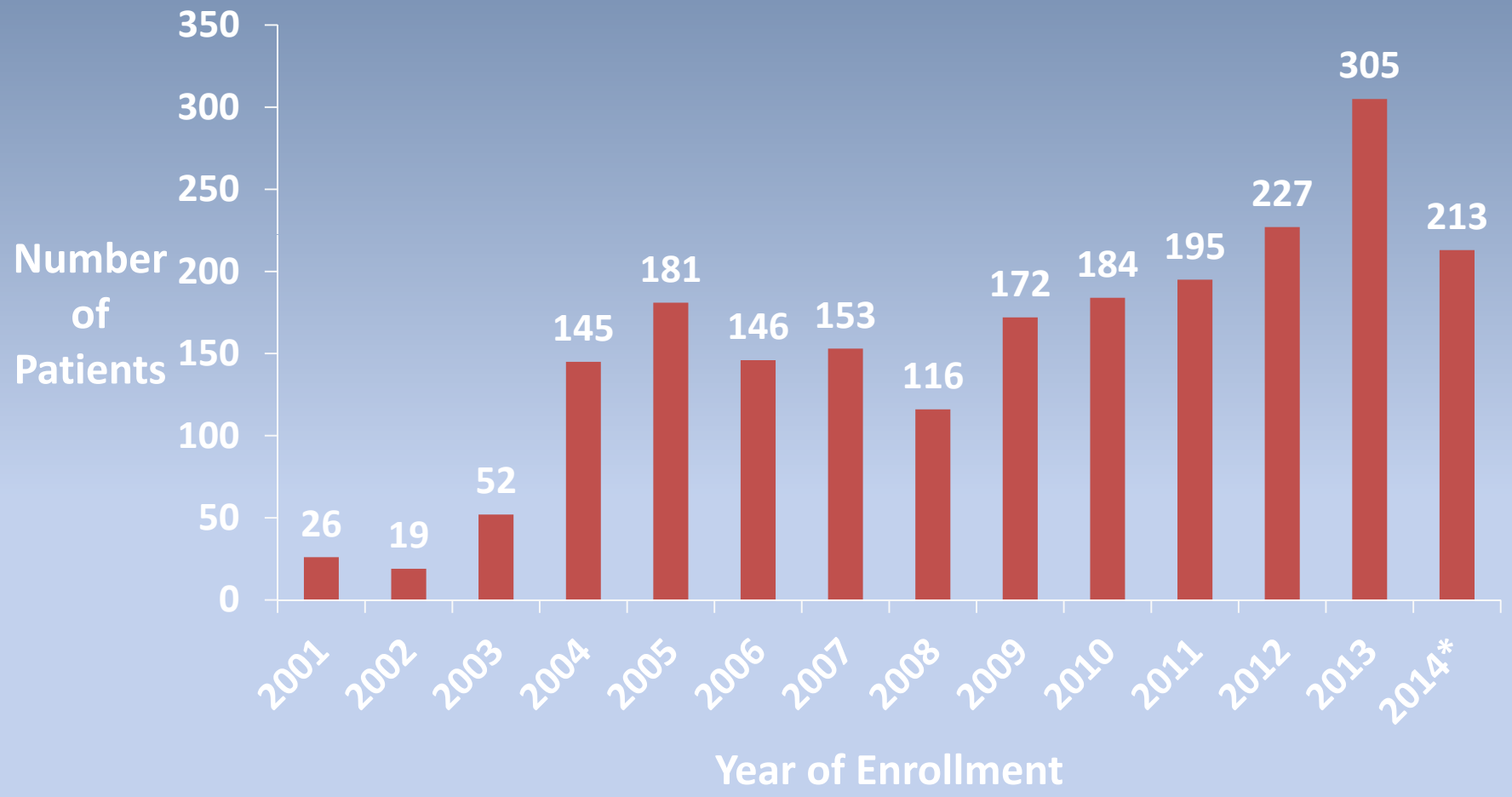


Active Enrollment Cohorts

Diagnostic Group	Number of Institutions	Accrual Date	N
Tricuspid Atresia	38	1999-Present	328
Critical Left Ventricular Outflow Tract Obstruction	21	2005-Present	743
Anomalous Aortic Origin of Coronary Arteries	34	2009-Present	346
Atrioventricular Septal Defect	16	2012-Present	145

- Proposed new cohort: Ebstein's anomaly
  - Dearani, Knott-Craig, Pizarro

Annual Enrollment



Number of Enrolling Institutions

Percentage enrollment 2013

Active Cohort	# eligible (STS)	# enrolled (DC)	Percentage enrolled
AAOCA	100	62	62%
LVOTO	709	85	12
Tricuspid atresia	208	18	8.7
Unbalanced AVSD	776	81	10.4

STS Congenital CHSS Studies
Summary

Participant 99999
January 1, 2014 - June 30, 2014

Duke Clinical Research Institute

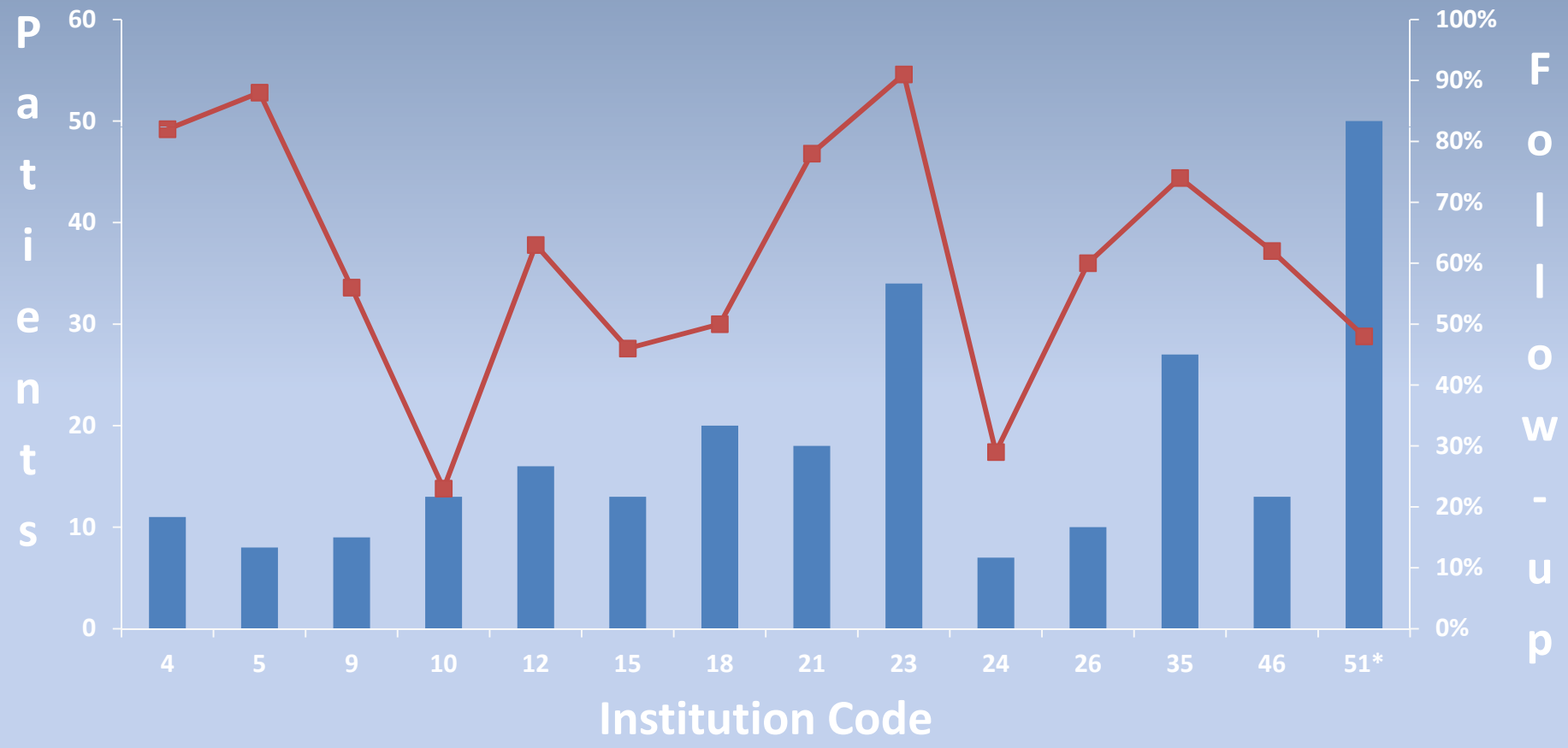
Table 1: Number of Patients Enrolled in CHSS Studies and Number Potentially Eligible for CHSS Studies

	Participant		STS	
	Enrolled[1]	Potentially Eligible[2]	Enrolled[1]	Potentially Eligible[2]
Participant's Five Most Common Procedures				
Tricuspid Atresia (TA) Study				
1590 = Shunt, Systemic to pulmonary, Modified Blalock-Taussig Shunt (MBTS)	xx	xx	xx	xx
1640 = PA banding (PAB)	xx	xx	xx	xx
1600 = Shunt, Systemic to pulmonary, Central (from aorta or to main pulmonary artery)	xx	xx	xx	xx
1670 = Bidirectional cavopulmonary anastomosis (BDCPA) (bidirectional Glenn)	xx	xx	xx	xx
1700 = HemiFontan	xx	xx	xx	xx
Pulmonary Conduit (PC) Study				
230 = Truncus arteriosus repair	xx	xx	xx	xx
380 = TOF repair, RV-PA conduit	xx	xx	xx	xx
420 = Pulmonary atresia – VSD (including TOF, PA) repair	xx	xx	xx	xx
760 = Ross-Konno procedure	xx	xx	xx	xx
1150 = Rastelli	xx	xx	xx	xx
Critical Left Ventricular Outflow Tract Study (LVOTO) Study				
870 = Norwood procedure	xx	xx	xx	xx
1640 = PA banding (PAB)	xx	xx	xx	xx
2180 = Hybrid Approach "Stage 1", Stent placement in arterial duct (PDA) + application of RPA & LPA bands	xx	xx	xx	xx
1280 = Aortic arch repair	xx	xx	xx	xx
1285 = Aortic arch repair + VSD repair	xx	xx	xx	xx
Anomalous Aortic Origin of a Coronary Artery (AAOCA) Study				
1305 = Anomalous aortic origin of coronary artery from aorta (AAOCA) repair	xx	xx	xx	xx
1310 = Coronary Artery Procedure, Other	xx	xx	xx	xx
Unbalanced atrioventricular septal defect [uAVSD] Study				
170 = AVC (AVSD) repair, Complete (CAVSD)	xx	xx	xx	xx
1640 = PA banding (PAB)	xx	xx	xx	xx
870 = Norwood procedure	xx	xx	xx	xx
1590 = Shunt, Systemic to pulmonary, Modified Blalock-Taussig Shunt (MBTS)	xx	xx	xx	xx
1670 = Bidirectional cavopulmonary anastomosis (BDCPA) (bidirectional Glenn)	xx	xx	xx	xx

[1] Stated to be enrolled, based on the v3.22 CHSS eligibility variable
[2] Stated to be not enrolled based on the v3.22 CHSS eligibility variable

Follow up

TA 2013 Follow-up
32 Institutions



*Comprised of 19 institutions contributing 5 or less patients

Follow up performance

Cohort	Cumulative Enrollment to 9/2/2014	Enrolled in 2014	Cumulative deaths through 2013 f/u	Eligible for 2014 follow up	Could not contact, or refused follow up	Percentage successful follow up
AAOCA	342	60	9	273	110	60%
LVOTO	737	39	225	473	171	64
Tricuspid atresia	313	26	36	251	79	67
Unbalanced AVSD	149	64	4	81	27	67

Grants, Studies and Publications

Active Studies

- Unbalanced AVSD
- FHS in PA/IVS
- FHS in critical AS
- LVOTO
  - Single versus 2V survival
  - Longitudinal VF, AVVF
  - Hybrid vs. Norwood vs. Sano
- AAOCA BMI

Grants

- Michael H. Ludwig Memorial Foundation
- Children's Heart Foundation
- Saving Tiny Hearts Society
- Nonin, Inc.
- Cryolife, Inc.

Grant applications

- D.O.D (Gruber)– submitted 10/17
- Le Ducq (Gruber)– multinational; genetic basis of CHD, correlation with outcomes
  - Due 12/1 pending initial approval

Papers published/submitted

- *Anomalous aortic origin of a coronary artery: A report from the CHSS Registry*. Poynter et al. WJPCHS 2014;5(1): 22-30
- *Repair of anomalous aortic origin of a coronary artery in 113 patients: A CHSS Report.* Poynter et al. WJPCHS in press
- *Surgical management of competing pulmonary blood flow affects survival before Fontan/Kreutzer completion in patients with tricuspid atresia type 1.* Wilder et al. EJCTS submitted
- *Anomalous aortic origin of coronaries arteries in the young: echocardiographic evaluation and surgical correlation; A multicenter CHSS study.* Lorber et al. JACC Imaging to be submitted.

Presentations

- *Hybrid alternatives to Norwood stage 1 are not a lower risk alternative: Norwood-RVPA offers better outcome in comparable neonates.* Wilder et al. To be presented at 2014 Scientific Sessions, AHA, November.
- *"Hybrid type" alternatives to Norwood may not be the lowest-risk solution for HLHS.* Wilder et al. Presented last night here.
- *Multiphase mixed model regression increases the utility of longitudinal data analysis in congenital heart surgery.* Wilder et al. Presented at WSPCHS meeting, Sept 2014
- *Initial management of pulmonary blood flow is associated with late outcomes in tricuspid atresia.* Wilder et al. Presented at the EACTS meeting, Oct 2014
- *Initial management of competing pulmonary blood flow affects survival in children with tricuspid atresia.* Wilder et al. To be presented at the Annual Meeting of the CCS, Oct 26-27, Vancouver

Travis Wilder, MD
Kirklin/Ashburn Fellow

- For his “hybrid alternatives to Norwood” abstract, is the winner of the AHA Council on CVDY “Outstanding Research Award in Pediatric Cardiology” to be presented at the AHA Scientific Sessions 2014, Chicago, Nov 15-19.

Abstracts submitted

- *Trends in right ventricular dysfunction and tricuspid regurgitation after single ventricle palliation of hypoplastic left heart syndrome and their differential impact on survival.* Wilder et al. Submitted for presentation at the Annual Meeting ACC, February 2015.
- *Late survival and right ventricular performance in matched children: classic Norwood-BT shunt versus Norwood-Sano modification.* Wilder et al. Submitted for presentation at the Annual Meeting AATS, May 2015.

END