ABSTRACT BOOK

28th International Symposium on

Adult Congenital Heart Disease

'Tackling & Transforming Outcome of Complex CHD'

June 5-9, 2018
Toronto, Ontario

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ISACHD AWARD WINNERS:

Winner of the ISACHD Young Investigator Research Award

Oral Presentation: Friday, June 8 @ 3:55pm

FEASIBILITY AND EFFICACY OF NEGATIVE PRESSURE VENTILATION IN THE AMBULATORY FONTAN POPULATION-(FONTAN-CMR) - A PILOT STUDY

Pradeepkumar Charla; Shi Joon Yoo; Gauri Rani Kaur; Kenichiro Yamamura; John T Granton; Erwin Oechslin; Ashish Shah; Eric Horlick; Lee Benson; Osami Honjo; Rachel M Wald; *Toronto Congenital Cardiac Center for Adults, Toronto General Hospital, Peter Munk Cardiac Center, University of Toronto, Toronto, Canada*

Background: The Fontan operation is the procedure of choice for children born with single ventricle physiology. Although cardiovascular complications are relatively uncommon in childhood following successful Fontan palliation, rates of morbidity and mortality are strikingly high in adulthood and can be attributed to a chronically low cardiac output (CO) state. Treatment options for adults with failing Fontan physiology are extremely limited. While short-term augmentation of CO with negative pressure ventilation (NPV) immediately following Fontan surgery has been documented, the effects of NPV in the ambulatory setting have never been studied. Using a novel, non-invasive, portable ventilator (United Hayek Medical) never before applied to the ambulatory congenital heart disease population, our aim was to explore the safety, feasibility and efficacy of NPV in the adult Fontan population. Hypothesis: We hypothesized that NPV will result in an improvement in CO as measured by cardiovascular magnetic resonance imaging (CMR).

Methods: Primary: To explore the impact of NPV on CO in adults with a Fontan circulation as compared with healthy controls. Secondary: To evaluate safety and tolerability of the device by questionnaire. Material and methods: Inclusion criteria: Adult patients (> 18 years) with lateral tunnel or extracardiac Fontan connections and a dominant left ventricle were screened for inclusion from an existing database. Healthy controls were matched 1:1 based on age and gender. Exclusion criteria: Patients were excluded if they had evidence of failing Fontan physiology (as determined by his/her primary cardiologist), significant valve regurgitation/stenosis, obstruction within the Fontan circuit, resting oxygen saturation 35 kg/m2), chest wall deformities and/or contraindications to CMR imaging. Phase contrast (PC) flow measurements using CMR were completed at multiple locations (ascending aorta, descending aorta, pulmonary arteries, vena cavae and Fontan circuit) at baseline and at various timepoints and pressure settings (as shown in Figure 1). Flow measurements were expressed as L/min/m2. Continuous data within and between groups were analyzed using Wilcoxon Mann-Whitney test.

Results: We enrolled 20 subjects: 10 adult Fontan patients and 10 age-matched healthy controls. We completed CMR studies in 8 Fontan patients and 10 controls (2 CMR studies pending at the time of abstract submission). Patient demographics are presented (Table 1). At baseline, Fontan patients had decreased cardiac index (CI) compared to healthy controls (Figure 2). A brief period of intermittent NPV resulted in significant increase in CI from 2.65 to 3.26 L/min/m2 (23% mean increase) and pulmonary blood flow from 2.15 to 2.72 L/min/m2 (26% mean increase) in the Fontan group as compared to controls (Figures 3 and 4). Patients reported better tolerability of the device in the intermittent as compared to the continuous NPV setting; no significant adverse events were observed during the study.

Conclusions: A significant improvement in CO can be achieved in adult Fontan patients using intermittent NPV in the ambulatory setting. This pilot data require further validation in larger populations of Fontan survivors and the potential impact of enhanced CO on clinical outcomes should be the focus of future studies.

Winner of the ISACHD Cardiac Care Associate Investigator Award :

Oral Presentation: Friday, June 8 @ 4:10pm

HEALTH LITERACY IN ADULTS WITH CONGENITAL HEART DISEASE

Susan Fernandes; Christy Sillman; Tara Dade; Emily Dong; Christiane Haeffele; George Lui; Ian Rogers; Anitra Romfh; Caroline Scribner; Lee Sanders; *Stanford University School of Medicine, Stanford, California, USA*

Background: Cognitive impairment, neurodevelopmental issues, and dementia are frequently noted in patients with congenital heart disease (CHD) and likely place patients at increased risk for health literacy issues. Health literacy, defined as the understanding and application of words (prose), numbers (numeracy), and forms (documents) has not been specifically examined in adult CHD (ACHD) patients. Given this, we sought to describe the health literacy in ACHD patients utilizing a validated health literacy assessment tool.

Methods: In this single center study, we administered the "The Newest Vital Sign" survey to assess health literacy in subjects > 18 years of age with CHD. Subjects were asked to review a nutritional label and then answer 6 related questions. Per the tool scoring guidelines, answering 4-6 questions correct indicates adequate literacy, 2-3 correct answers indicates the possibility of limited literacy and 0-1 correct answers suggests high likelihood of limited literacy. Additional demographic questions were asked of the patient including questions regarding known learning disabilities. A retrospective chart review was also completed to obtain cardiac history.

Results: A total of 166 subjects agreed to participation. Median age was 38.7±13.4 years, 50% were male, 17% were Hispanic and 50% were White. The CHD complexity was simple in 33%, moderate in 34% and complex in 33%. A college degree was held by 55% of participants; 16% noted a previously diagnosed learning disability. Health literacy was noted as adequate in 78% of participants, possibly limited in 13% and likelihood of limited literacy in 9%. Multivariable logistic regression identified Hispanic ethnicity, non-White race, limited English proficiency and history of a learning disability as independent predictors of inadequate health literacy. Age, gender, educational level, CHD complexity, number of cardiac surgeries and oxygen saturation were not predictive.

Discussion: Our study indicates that health literacy is limited in more than 20% of our subjects. This may make educating patients about their disease and clinical and or surgical management plans more difficult. It may also contribute to gaps in care. Health literacy screening should be considered for all ACHD patients and educational interventions targeting those with inadequate health literacy is warranted.

ORAL PRESENTATIONS:

Wednesday June 6 @ 9:12am

CONTEMPORARY OUTCOMES AND MORTALITY RISKS OF EBSTEIN'S ANOMALY

Varisara Pornprasertchai; Chodchanok Vijarnsorn; Supaluck Kanjanauthai; Paweena Chungsomprasong; Prakul Chanthong; Kritvikrom Durongpisitkul; Jarupim Soongswang; *Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok, Thailand*

Background/Objectives: Ebstein's anomaly (EA) is a complex congenital heart disease with wide anatomical variations and vary age-dependent clinical symptoms. Even though this disease has been long described, long term data is still sparse due to remarkable anatomic severity of the disease and relatively small sample population. This present study is aimed to assess survival outcome and predictors of mortality in patients with EA

Methods/Overview: A retrospective reviewed was performed on all patients with EA who diagnosed by echocardiogram in our institution between year 1994-2016. Patients who had EA with complex transposition of great arteries were excluded. Baseline characteristics, treatments and outcomes were reviewed. Survival analysis was performed at the end of 2017. Multivariate analysis was utilized to assess mortality risks.

Results: A total of 153 patients (age 18 ± 19.5 years, 60% female) were analyzed. 64 patients underwent at least one surgical intervention with median age of 17 years (1 day - 64.4 years). During follow up period with median of 5.2 years (3 days - 23.5 years), death occurred in 32 patients (20.9%) due to major cardiac adverse events. Overall survival rates at 1, 5, and 10 years follow up period were 89%, 82.2% and 79%, respectively. Among 64 EA patients who underwent surgeries, survival rate at 1, 5 and 10 years were 87.5, 82.4%, and 77.7% respectively which had no statistically difference when compared to 89 non-operated EA patients, survival rate were 89.9%, 87.5%, and 81.8%, at 1, 5, and 10 years follow up period. Significant overall mortality risks were age at diagnosis ≤ 2 years, TV z-score > 3.80, TV

Conclusions: Patients with EA overall had a moderately good survival in contemporary era. We report, five important predictors of death in this patient population.

Wednesday June 6 @ 9:22am

MID-TERM OUTCOMES IN ADULT PATIENTS WITH SURGICALLY OPERATED EBSTEIN ANOMALY

Norihisa Toh; Yasuhiro Kotani; Teiji Akagi; Yosuke Kuroko; Kenji Baba; Shin-ichi Otsuki; Shingo Kasahara; Hiroshi It; *Department of Cardiovascular Medicine, Japan*

Background: Ebstein anomaly (EA) is a quite rare form of congenital heart disease and various operative techniques have been described due to its heterogeneous anatomy. However, limited data is available on the long-term outcomes in adult patients with surgically operated EA. We sought to review our experience of adult survivors of operated EA.

Methods: Thirty-five adult patients with operated EA (1994 to 2017) were identified from our database. Surgical history, clinical, ECG, echocardiography, and outcome data were analyzed. Unfavorable cardiovascular events included death, heart failure admission, ventricular arrhythmia, and re-intervention.

Results: Median age at the first clinical visit was 17 years (range 16 to 58, 37% male). On the initial visit, 24 patients had tricuspid valve (TV) repair (cone repair [n=10], Carpentier technique [n=9], and others [n=5]), 2 patients had TV replacement, 4 patients had one-and-a-half ventricular repair (1.5VR), and 5 patients had Fontan palliation. Their demographic, surgical characteristics, and laboratory data are shown in Table. Median follow-up of duration was 7.1 years. One patient died after the re-operation. Six patients experienced heart failure admission and 3 of 6 required reoperations (Fontan revision [n=1], Fontan palliation [n=1], and cone repair [n=1]). One patient experienced sustained ventricular tachycardia followed by reoperation (1.5VR and implantable cardioverter-defibrillator implantation). One patient underwent re-TV replacement. Event-free survival was 94%, 90%, 77%, and 64% at 1, 5, 10, and 15 years (Figure). In multivariable Cox proportional hazards analysis, serum BNP at the initial visit > 85 pg/ml was independently predictive for unfavorable cardiovascular events.

Conclusions: Adult patients with surgically operated EA continue to have high rates of morbidity and mortality, with need for re-operations. While this subset of patients might not be representative of all adult operated EA survivors, continued follow-up at centers specialized for adult congenital heart disease is recommended for adults with operated EA.

Baseline variable	All patients (N=35)	Patients with unfavorable events (N=9)	Event-free survivors (N=26)	P-value
Age, yrs	23.4 ± 12.7	32.8 ± 15.5	20.2 ± 10.0	0.008
Status at the initial visit, n				0.035
TV plasty	24	5	19	
TV replacement	2	2	0	
1.5VR	4	0	4	
Fontan palliation	5	2	3	
BNP, pg/ml	83.2 ± 34.7	215.9 ± 60.0	31.2 ± 37.6	0.014
ECG parameters				
QRS width, msec	133.4 ± 42.7	124.7 ± 58.7	137.1 ± 34.9	0.473
WPW syndrome, n	5	2	3	0.600
Echo parameters				
Left ventricular EF, %	(N=28)	67.4 ± 8.5 (N=7)	68.6 ± 8.1 (N=21)	0.711
Tricuspid regurgitation grade				0.082
Mild	11	2	9	
Moderate	10	1	9	
Severe	7	4	3	

Friday, June 8 @ 9:30am

REPEATED MEASUREMENTS OF CARDIAC BIOMARKERS IMPROVE RISK PREDICTION IN PULMONARY ARTERIAL HYPERTENSION ASSOCIATED WITH CONGENITAL HEART DISEASE

Alexandra C. van Dissel; Ilja M. Blok; Aeilko H. Zwinderman; Arie P.J. van Dijk; Anthonie L. Duinhouwer; Barbara J.M. Mulder; Berto J. Bouma, *Academic Medical Center, Amsterdam, The Netherlands*

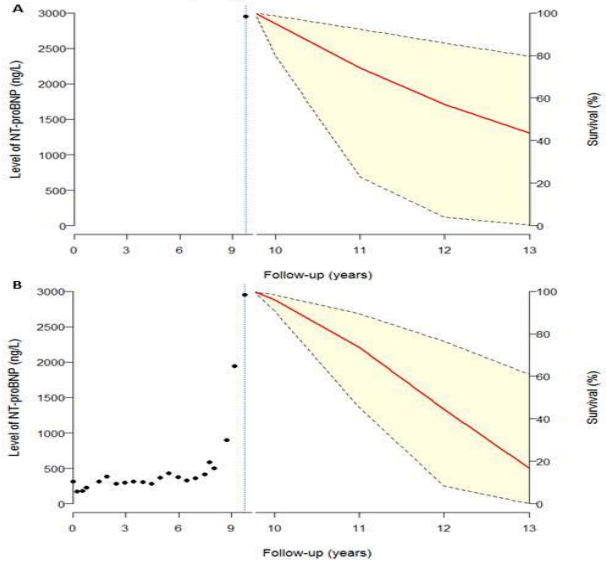
Background: Accurate risk stratification in pulmonary arterial hypertension associated with congenital heart disease (PAH-CHD) remains challenging. The use of cardiac biomarkers is promising to identify patients at high risk, who could be targeted for preventive measures. We evaluated the value of repeated and combined measurements of four biomarkers, representing myocardial stress (N-terminal pro-brain natriuretic peptide [NT-proBNP]), myocyte injury (troponin-T), cardiorenal dysfunction (cystatin-C) and fibrosis (galectin-3) in 98 PAH-CHD patients.

Methods: Between January 2004 and January 2016, patients were prospectively followed at two Dutch university hospitals and underwent regular measurements of serum cardiac biomarkers. Biomarkers were log-transformed. The association with mortality was assessed using fitted mixed-effect, Cox regression and joint models, adjusted for patient age and gender.

Results: Mean age of patients was 43±16 years, 34% was male and 69% had Eisenmenger syndrome. During a median follow-up of 6.9 years, 40 patients (41%) died. The average number of repeated biomarkers per patient during follow-up was 9 for NT-proBNP, 5 for troponin-T, 4 for cystatin-C and galectin-3. Median levels of repeated biomarkers were: NT-proBNP 518 ng/L (IQR 223-1433), troponin-T 0.011 µg/L (IQR 0.005-0.024), cystatin-C 0.97 mg/L (IQR 0.81-1.23), and galectin-3 15 µg/L (IQR 12-18). The correlations among the repeated biomarkers were weak to moderate (all r<0.4). Biomarker levels in patients who died progressively increased before time of death compared to their counterparts who remained alive. During follow-up, one SD increase in biomarker level was 250% for NT-proBNP, 150% for troponin-T, and 50% for cystatin-C and galectin-3. Each SD increase in biomarker level was associated with a doubled risk of death at any particular time (all biomarkers p<0.001), HR as follows: NT-proBNP HR 2.2 (95% CI 1.7 to 2.9); troponin-T HR 2.2 (95% CI 1.6 to 2.3); cystatin-C HR 1.8 (95% CI 1.4 to 2.4); galectin-3 HR 1.7 (95% CI 1.2 to 2.3). Risk prediction with repeated measurements was more accurate than with single-moment measurements (Figure). Of the four repeated biomarkers, NT-proBNP, troponin-T and cystatin-C achieved high discriminatory abilities for 10-year mortality (c-index 0.81-0.92), whereas galectin-3 had a c-index of 0.60. Using a model combining the former three biomarkers yielded a similar c-statistic of 0.85, indicating that a combination of several repeated biomarkers does not provide additional information.

Conclusion: Repeated biomarker measurements had an approximately 2-fold higher mortality risk per one SD increase and were better predictors of mortality than single measurements. Combining several biomarkers integrating diverse biological pathways, however, had no additional value. Therefore, regular assessment of one biomarker, e.g. NT-proBNP, is recommended for assessing risk in individual persons.

Figure. Dynamic survival predictions for an individual patient based on NT-proBNP measurements.
(**A**) The probability that this patient will survive for the next 3 years after the last observed time (dotted vertical line) for which a longitudinal measurement (dot) was available is 45%, with wide 95% confidence interval (shading). (**B**) The survival probability of the same individual patient is only 18% based on the complete biomarker history (dots), with much narrower 95% confidence interval (shading).



Friday, June 8 @ 9:40am

PULMONARY HYPERTENSION IN ADULTS WITH SYSTEMIC RIGHT VENTRICLES REFERRED FOR CARDIAC TRANSPLANTATION

Yuli Kim; Katherine Awh; Michael Acker; Pavan Atluri; Christian Bermudez; Maria Crespo; Joshua Diamond; Stephanie Fuller; Jonathan Menachem; Emily Ruckdeschel; David Drajpuch; Rhondalyn Forde-McLean; Lee Goldberg; Jeremy Mazurek; Chrsitopher Mascio; Eduardo Rame; Lynda Tobin; Joyce Wald; *Division of Cardiovascular Medicine, Hospital of the University of Pennsylvania, Division of Cardiology, The Children's Hospital of Philadelphia, Philadelphia, USA*

Background/objectives: Adults with systemic right ventricle (RV) physiology are at risk for RV dysfunction and end-stage heart failure necessitating referral for orthotopic heart transplantation (OHT). Pulmonary hypertension (PH) frequently coexists in adult congenital heart disease and can complicate the assessment for OHT.

Methods: A single-center, retrospective case review of systemic RV patients ≥ 18 years referred for heart transplant evaluation from 2008-2017.

Results: Eighteen patients with systemic RV physiology (mean age 40.8 ± 7.8 years, 56% female) were referred and evaluated for OHT, of whom one-third (n=6) had PH defined as mean pulmonary artery pressure (mPAP) > 25 mmHg. Clinical characteristics are summarized in Table 1. At time of referral for OHT consideration, all 6 patients had pulmonary vascular resistance (PVR) > 3 Wood Units. Two of the 6 patients were considered OHT-ineligible due to PH among other comorbidities including medication nonadherence in one and multiorgan failure in the other. Of the remaining 4, three were listed for heart-lung transplant (HLTx) and one for OHT alone. Clinical details are summarized in Table 2. Patient 1 had double outlet right ventricle with ventricular septal defect (VSD) status post VSD closure and left ventricle to pulmonary artery conduit who presented in cardiogenic shock and new diagnosis of post-capillary PH. Initially listed for HLTx, his listing was changed to OHT alone after aggressive PH-directed medical management, including milrinone and nitroprusside. He underwent OHT with extubation to inhaled flolan and was discharged on sildenafil for 5 months post-transplant. Patient 2 had L-transposition of the great arteries (TGA) status post atrial septal defect repair and mechanical tricuspid valve replacement with known pre-capillary PH on treprostinil and sildenafil. She presented with decompensated heart failure and underwent HLTx during that admission. Patient 3 has D-TGA status post Mustard with pre-capillary PH on epoprostenol and is actively listed 1AE for HLTx. Patient 4 had D-TGA status post Mustard and combined pre- and post-capillary PH with mPAP 34, pulmonary capillary wedge 20, and PVR 3. He was started on sildenafil. Three years later, he presented with syncope from ventricular tachycardia in the setting of decompensated heart failure and referred for OHT. After diuresis and institution of milrinone, catheterization demonstrated improved hemodynamics with nitroprusside challenge and he was listed for OHT alone. Post-transplant day 1 he had a pulmonary hypertensive crisis with anoxic brain injury and care was withdrawn shortly thereafter

Conclusion: Pulmonary hypertension is common in systemic RV patients referred for OHT. Systemic RV dysfunction places these patients at risk for post-capillary PH but pre-capillary PH can coexist. Despite management with selective pulmonary vasodilators and afterload reduction, the criteria for listing such patients for HLTx versus OHT are not known and need further elucidation.

Friday, June 8 @ 9:50am

PULMONARY ARTERIAL HYPERTENSION AND PREGNANCY - A SYSTEMATIC REVIEW OF PREGNANCY OUTCOMES IN A CONTEMPORARY COHORT

Ting Ting Low; Nita Guron; Robin Ducas; Kenichiro Yamamura; Rohan D'souza; John Granton; Candice Silversides; *National University Heart Centre, Singapore, Toronto General Hospital, Toronto, Canada*

Background: Women with pulmonary arterial hypertension (PAH) have significant risks during pregnancy. However, with advances in PAH therapies and more organized care models in the past 10 years, contemporary outcomes may be better than historical reports and need to be systematically examined. Objective: To perform a systematic review of adverse pregnancy outcomes in pregnant women with PAH in the last decade. Adverse outcomes included: 1) maternal mortality; 2) maternal cardiac morbidity; and 3) adverse fetal and neonatal outcomes.

Methods: We searched the Medline, Embase and Cochrane database for articles describing outcomes in pregnancy cohorts published between 2008-2018. We excluded case reports, case series with less than 8 cases, and studies that did not report PAH specific treatment. Two reviewers independently reviewed 3663 titles and performed a preliminary extraction of data from 10 publications. Additional publications are expected to be included in the final analysis. Pooled incidences and percentages of maternal and perinatal outcomes were calculated.

Results: Altogether, there were 224 pregnancies reported in 213 women with PAH. In the 8 studies (n=186 pregnancies) that reported early pregnancy outcomes, there were 46 (25%) therapeutic abortions and 7 (4%) spontaneous miscarriages. In the 46 terminations; 4 women died early postpartum, of which 1 died during the intervention. There were 174 pregnancies that continued beyond 20 weeks of gestation. In those studies that reported functional class, 79% (111/141) were in functional class I or II pre-pregnancy and the mean systolic pulmonary artery pressure (n=116 pregnancies) was 75±30 mmHg. The most common diagnosis was PAH associated with congenital heart disease (69%), followed by idiopathic PAH (18%) and other (12%). With regards to targeted PAH therapy during pregnancy; 21% of women were treated with a prostacyclin analogue, 23% with phosphodiesterase-5 inhibitors and 8% received calcium channel blockers. Combination therapy was used in 12% (17/137) of pregnancies. The mean gestation age at delivery was 33 ± 5 weeks (n=116). Most women had Caesarean deliveries (77%); and, where reported, 36% (52/144) of pregnancies had general anesthesia. The overall maternal mortality rate for pregnancies that continued beyond 20 weeks gestation was 11% (19/174); 6 maternal deaths occurred in 33 pregnancies with idiopathic PAH (18%) and 13 deaths in 124 pregnancies with congenital heart disease (11%). There were no deaths in 26 mothers with PAH from other etiologies. The causes of death included pulmonary hypertension crisis, heart failure and shock. Premature births occurred in more than half of the pregnancies (54%; 81/150). Stillbirth occurred in 3% (5/174) of pregnancies and neonatal mortality rate was 1% (2/174).

Conclusions: In this systematic review of a contemporary cohort of pregnant women with PAH, maternal mortality continues to be a significant risk. Continued prospective studies are needed to improve outcomes and to better understand the role of PAH therapy.

POSTER PRESENTATIONS:

1

PERCUTANEOUS BAFFLE INTERVENTIONS IN ADULT SURVIVORS OF THE ATRIAL SWITCH OPERATION: A SINGLE CENTER EXPERIENCE

Heiko Schneider; Andrew Leventhal; Ashish H Shah; Mark Osten; Lee Benson; Eric Horlick; *Toronto General Hospital, PMCC, TCCCA, Toronto, Canada*

Background: Baffle-related complications are known to occur in adults after the atrial switch operation. Given the high risk of repeat surgery, there is considerable interest in their percutaneous treatment. Despite the significance, limited data exist on the indications for and technical aspects of these interventions. We report our institution's experience with catheter-based treatments for baffle stenoses and leaks in an exclusively adult population.

Methods: This is a single center retrospective study. We interrogated the Toronto Congenital Cardiac Centre for Adults database to identify patients with atrial switch procedure, who have undergone trans-catheter interventions for baffle related complicated. Patient characteristics, procedural aspects, and clinical findings were recorded and analyzed. Each intervention was individually reviewed and data was collected describing indication, procedural steps, immediate success and outcomes. The local research and ethics board approved the study.

Results: Key patient and procedural characteristics are summarized in Table 1. We identified 292 adults who were treated with the atrial switch procedure. Of these, 20 patients had 26 percutaneous interventions (mean age: 33.6±8.1 years; n=12 male; n=8 female). The indication for interventions was: superior systemic venous baffle stenosis (SVC-S; n=13) or total occlusion (SVC-O; n=4), superior systemic venous baffle leak (SVC-L; n=4), Inferior systemic venous baffle stenosis (IVC-S; n=3), Inferior systemic venous baffle leak (IVC-L; n=1) and pulmonary venous baffle stenosis (PVB-S; n=1). Trans-venous pacing / implantable cardioverter defibrillator implantation associated with baffle stenosis was the most common indication for intervention (n=12). There were no major peri-procedural complications. Four procedures were not successful (15% failure rate). A total of n=4 stenotic or occluded baffles were balloon dilated only (SVC-O=1, SVC-S=2, IVC-S=1). A wide variety of stents was used to treat obstructive lesions (see Table 1). Leaks were closed with AmplatzerTM septal occluders (n=4, Abbott Vascular, St Paul, MN, USA) or CardioSEALTM devices (n=1, Nitinol Medical technologies, Boston, MA, USA). One patient suffered from pulmonary venous baffle stenosis which was stented successfully. The most important risk factor for procedural failure was total occlusion of the superior systemic venous baffle. Follow-up from a limited number of cases, suggests that baffle interventions can potentially provide a durable treatment.

Conclusion: Baffle related complications are uncommon, however, trans-catheter intervention offer viable outcomes. Interventions to treat a completely occluded baffle were associated with higher failure rates. There are no standard criteria for interventions and the decision to intervene must be made on a "case-by-case" basis in a multidisciplinary setting. The interventional equipment utilized in all cases varied widely and requires in-depth knowledge of, at times, non-standard use of the interventional armamentarium.

Procedure #	Patient #	Sex	Age	Lesion	Success	Device	Indication
1	1	M	37	SVC-O	Yes	Covered CP Stent® & P5014 Palmaz® stent	TED
2	2	M	24	SVC-S	No	None	SVC syndrome
3	2	M	24	SVC-S	No	None	SVC syndrome
4	3	F	47	SVC-S	Yes	Palmaz® 5014 stent	TED
5	4	M	34	SVC-S	No	None	Exercise intolerance
6	5	M	27	SVC-O	Yes	4 cm Palmaz® stent	Persistent pleural effusion
7	6	M	31	SVC-O	No	None	Reduce tricuspid regurgitation
8	7	F	44	SVC-O	Yes	16 mm Z-Med II-X® balloon	SVC syndrome
9	8	M	33	SVC-S	Yes	Palmaz® Genesis® 5014 stent	TED
10	9	F	39	SVC-S	Yes	Palmaz® 5014 stent	TED
11	10	F	27	SVC-S	Yes	18 mm Owen's® LP balloon	SVC syndrome
12	11	M	34	SVC-S	Yes	Palmaz® Genesis® 5910 stent	TED
13	12	F	37	SVC-S	Yes	18 mm Z-Med II-X® balloon	Varicose veins
14	13	M	32	SVC-S	Yes	Palmaz® 5014 stent	Concomitant IVC narrowing
15	14	M	22	SVC-S	Yes	36 mm Max® LD stent	TED
16	15	M	26	SVC-S	Yes	5 cm Johnson & Johnson stent	TED
17	16	F	27	SVC-S	Yes	Palmaz® Genesis® 5910 stent	TED
18	4	M	34	IVC-S	Yes	20 mm Z-Med II-X® balloon	Exercise intolerance
19	12	F	37	IVC-S	Yes	Palmaz® 5014 stent	Varicose veins
20	17	M	18	IVC-S	Yes	5 cm Johnson & Johnson stent	Hemodynamic gradient
21	4	M	34	PVB-S	Yes	Palmaz® XL 3110 stent	LV dilation and dysfunction
22	18	F	26	SVC-L	Yes	18 mm ASO	TED
23	19	M	41	SVC-L	Yes	10 mm ASO	TED
24	20	F	50	SVC-L	Yes	28, 28, 23 mm CardioSEAL®	Exertional desaturation
25	3	F	47	SVC-L	Yes	8 mm ASO	TED
26	8	M	33	IVC-L	Yes	22 mm ASO	TED

Table 1.: Summary of patient characteristics, demographics and key procedural data. SVC-O: Superior Systemic Venous Baffle Obstruction, SVC-S: Superior Systemic Venous Baffle Stenosis, IVC-S: Inferior Systemic Venous Baffle Stenosis, SVC-L: Superior Systemic Venous Baffle Leak, IVC-L: Inferior Systemic Venous Baffle Leak, PVB-S: Pulmonary Venous Baffle Stenois; TED: Transvenous Electric Device; F=Female, M=Male.

2

DEDICATED TEAM IMPROVES EFFICIENCY IN ASD/PFO CLOSURE IN AN ADULT CATHETERIZATION LABORATORY

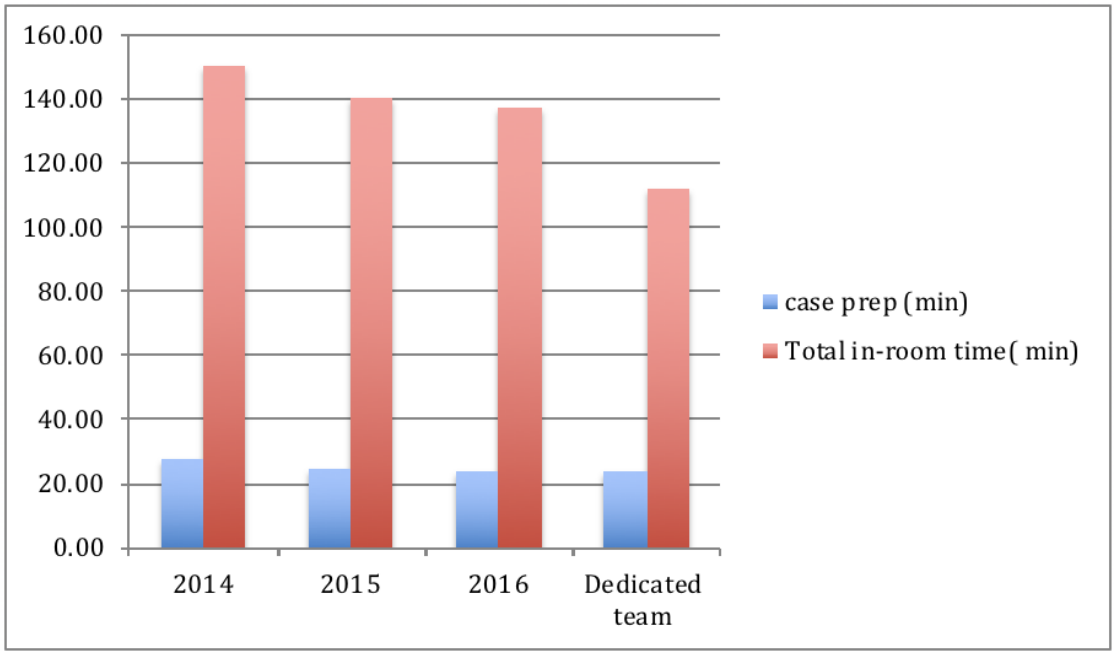
Stephanie Fuentes Rojas; Akanksha Thakkar; Ponraj Chinnadurai; Eunice Karanja; Danish Bawa; C. Huie Lin; *Houston Methodist Hospital, Houston, Texas, USA*

Background: The rising prevalence of adult congenital heart disease (ACHD) poses important challenges in the cardiac catheterization lab of adult-only hospitals. The resources needed to render an adult catherization lab ready for ACHD cases are not well understood. We propose that a dedicated 'core team' approach may improve the efficiency of ACHD interventions.

Methods: We reviewed all consecutive atrial septal defect (ASD) and patent foramen ovale (PFO) closures performed by an ACHD/Interventional-trained operator working in an adult hospital cath lab with staff inexperienced in ACHD and ASD/PFO closures from January 2014 to April 2018. In May 2016, we implemented a dedicated biplane cath lab room, day, and a consistent "core team" comprised of two radiation technologists and two nurses. Time spent in the catheterization lab prior to procedure start (case preparation time) and the total time spent in the procedure room (case time) were measured. Cases involving additional procedures such as atrial fibrillation ablation were excluded from analysis.

Results: Before the institution of the 'core team', median time to case start (in minutes) in 2014 (n=13) was 28 (IQR 22-33), 2015 (n=7) was 25 (IQR 25-28) and 2016 (n=12) was 24 (IQR 19-28). Median total in-room time (in minutes) was 150 (IQR 125-161), 140 (IQR 125-156), and 137 (IQR 121-165) respectively. Since 'core team' implementation (n=13), the median time to case start has been reduced to 24 minutes (IQR 22-29) and median total time has been reduced to 112 minutes (IQR 97-118).

Conclusions: While case time was modestly reduced over three years of PFO/ASD closures, the introduction of consistent and dedicated staff reduced case time by nearly 20%. These data may be a model for establishing a successful learning curve for ACHD care in adult hospitals and further justify dedication of specialized resources when developing an ACHD program.



3

Going Proximal to Distal: A Novel Strategy for Prosthetic Valve Fracture to Enable Transcatheter Pulmonary Valve Implantation and Complex Right Ventricular Outflow Tract Intervention

Stephanie Fuentes Rojas; Akanksha Thakkar; Ponraj Chinnadurai; Danish Bawa; Eunice Karanja; Gary Monteiro; Thomas MacGillivray; John Breinholt; *Houston Methodist Hospital, Houston, Texas, USA*

Background/Objectives: Pulmonary valve replacement is increasingly being used in patients with childhood repair of Tetralogy of Fallot for late complications. Over time, patient-prosthesis mismatch may develop, necessitating re-intervention. Percutaneous valve-in-valve (ViV) therapy has been used as a solution to early device failure to avoid repeated sternotomies. We report a novel proximal to distal approach to pulmonary ViV implantation to treat patient prosthesis mismatch followed by interventions on bilateral stenotic pulmonary arteries.

Case: A 30 yo F with ToF presented with decreased exercise tolerance. Initial repair was at age 4. Bioprosthetic pulmonary valve (PV) replacement was at age 24 (Carpentier-Edwards Perimount 21 mm). Non-invasive imaging showed normal LV, large RV with signs of volume and pressure overload, severe pulmonic regurgitation (PR), moderate right pulmonary artery (RPA) stenosis. Internal diameter of the PV on CTA was 18 mm. On cath, there was severe stenosis of the RPA and moderate stenosis of the left pulmonary artery (LPA); pressures in the RV were 99/15 mm Hg, main pulmonary artery 55/12 mm Hg (mean, 23 mm Hg), LPA 33/14 mm Hg (mean, 19 mm Hg) and RPA 26/13 mm Hg (mean, 17 mm Hg). Interventions on the stenotic PV and pulmonary arteries were planned. The valve presented a 'Russian doll' problem as deploying a valve within an already stenotic valve would further reduce the valve area. Therefore, we planned to fracture the Perimount frame prior to performing ViV implantation. On reviewing available devices, a 23 mm Edwards SAPIEN transcatheter pulmonary valve was found to be most appropriate. However, the length of the device delivery system for the transcatheter pulmonary valve necessitated deviation from convention by using a proximal to distal approach where we performed ViV implantation first followed by interventions on the pulmonary arteries. A 22mm x 4cm balloon was advanced into the Perimount frame through a 14 Fr Mullins sheath. It was inflated to burst pressure which fractured the valve ring. Using a 24 mm x 4 cm balloon, the fractured ring expanded to 22 x 23.5mm. A 23 mm Edwards SAPIEN transcatheter pulmonary valve was deployed in the Perimount valve frame using rapid RV pacing to stabilize deployment and CTA-fluoroscopy fusion to optimize C-arm angles. Balloon angioplasty of the LPA using a 24mm x 4cm balloon was performed. RPA stenosis had a more significant gradient; CTA-fluoroscopy image fusion was used to deploy a balloon expandable P3110 XL stent. Initial attempt to deploy the stent using a 20mm x 4cm balloon was unsuccessful as the frame of the newly placed valve ruptured the balloon. Deployment was then performed using a 20mm x 2cm balloon. RV pressure decreased to 55/10 mm Hg with a residual gradient of 14 mm Hg from RV to RPA and 18 mm Hg from RV to LPA.

Conclusions: In contemporary ACHD care, complex RVOT interventions will require modifying existing prosthetic valve apparatus as well as counter intuitive approaches. Pre-procedural planning and CTA-fluoroscopy image fusion in our case enabled safe transcatheter valve-in-valve PV implantation and bilateral PA intervention.

4

OUR EXPERIENCE OF COMBINED PERCUTANEOUS OCCLUSION OF THE LEFT ATRIAL APPENDAGE (LAA) AND AN INTERATRIAL COMMUNICATION.

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Background: Patients may have indications for both LAA and interatrial communication closure. Closure of the interatrial communication will impede access to the left atrium at a later date. It is therefore prudent to close the LAA prior to closing an interatrial communication.

Methods: A retrospective study of combined LAA and ASD/PFO closure in our center from 2009 to 2016.

Results: We performed 8 cases of LAA closure combined with ASD/PFO closure in same sitting. Three patient underwent the procedure for multiple strokes and Atrial fibrillation (AFIB) and five patients for right heart volume overload due to ASD complicated by AFIB. They ranged in age from 53-78 years of (average 66 years). Half of the patients were female. Only one patient had impaired renal function with an eGFR of 30ml/min/m2. The mean CHA2DS2VASc was 5 in the the stroke group and only 2 in the patients with right heart volume overload. For the LAA closure, we used the Amplatzer Cardiac Plug in 4 patients , Amulet Device in 2 patients and LAmbre LAA occluder in 2 patients. The Amplatzer septal occluder and Amplatzer PFO occluder were used for the interatrial communications. There were no documented complications either during the procedures or post procedure.

Conclusions: Combined LAA and ASD devices implantation is a useful procedure for patients with atrial fibrillation and interatrial communications.

5

CLOSURE OF A VENTRICULAR SEPTAL DEFECT COMPLICATED BY HEMOLYSIS REQUIRING TRANSCATHETER RETRIEVAL OF AMPLATZER OCCLUDER

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Background: Transcatheter closure of residual ventricular septal defects (VSDs) after prior surgical closure is considered if Qp : Qs is >1.5. Hemolysis due to transcatheter ventricular septal defect (VSD) closure is infrequently reported and ideal management is unclear.

Case: 78 yo M with prior surgical VSD repair/RV infundibular resection presented with orthopnea. Exam revealed a systolic murmur at the left upper sternal border and bilateral LE edema. CTA showed a 16 mm membranous VSD with L-R shunt. Qp:Qs was 2 by CMR. Three defects in the ventricular septum (2 membranous at prior patch repair site, 1 high muscular) were seen on LV gram; the largest was closed with an 8 mm Amplatzer™ Septal Occluder (St. Jude Medical, Inc., MN) by TEE and 2D-3D CTA fusion. Residual shunt was noted, but expected to resolve with endothelialization of the device. The patient however, developed tea colored urine on post-operative day 0 with a drop in hemoglobin from 14.5 to 11 g/dL, platelets from 145 to 86 k/µL. LDH was 1218 U/L and haptoglobin was <10 mg/dL. Due to the risk of hemoglobin nephropathy induced acute renal failure, we chose to attempt transcatheter retrieval of the device as open surgical risk was unacceptable. The patient was urgently returned to the cath lab and the device snared and re-sheathed. Post retrieval, his urine color was normal in 24 hours and CBC returned to baseline at 2 weeks. Six months later, the patient was taken back to the cath lab and VSD closure was once again attempted. A 10mm/8mm AmplatzerTM Duct Occluder was used to close the lower of the defects which was about 5 mm on balloon sizing. A repeat ventriculogram revealed a larger superior defect (about 7 mm on diameter on balloon sizing) which was closed using a 12mm/10mm AmplatzerTM Duct Occluder. A small amount of residual shunt was noted through the devices and inferior to the disc of the 10mm/8mm device. The patient had an uncomplicated post-procedure course without recurrence of hemolysis. There was also a marked improvement in his functional status with normalization of RV size and function on 1 month follow up.

Conclusion: There is no consensus on the management of hemolysis after device closure of VSDs. Watchful waiting may lead to hemoglobin associated nephropathy, coils may embolize and do not necessarily eliminate residual shunt, and there is a high risk of morbidity with emergency thoracic surgery. This case highlights an infrequently reported complication of trans-catheter VSD closure. Conservative or surgical management may be appropriate in some cases; however, until further data becomes available, it may be prudent to retrieve the device to prevent further complications. Additionally, optimal device selection to completely occlude all relevant defects and minimize residual shunt may reduce risk of post-op hemolysis. Choice of such a device may require more advanced pre-op planning such as 3D printing.

6

PERSPECTIVES ON ADVANCE CARE PLANNING AND PALLIATIVE CARE AMONG ADULTS WITH CONGENITAL HEART DISEASE

Jill Steiner; James Kirkpatrick; James Kirkpatrick; Karen Stout; J. Randall Curtis, *University of Washington, Seattle, USA*

Background: Symptom management, advance care planning (ACP), and palliative care are gaining recognition as critical components of comprehensive care for adults with congenital heart disease (ACHD). Previous research suggests patients want information on life expectancy and to have discussions about future care. Patients want their doctors to initiate conversations, preferably when they are healthy. The purpose of this study was to explore, among patients with ACHD, their (1) willingness to participate in advance care planning and (2) preferences for with whom to have these discussions.

Methods: This was a cross-sectional survey of consecutive outpatients seen in the ACHD Clinic at a large academic medical center. Survey items consisted of questions regarding prior discussions about care preferences or participation in ACP, willingness to participate in future discussions, and questions about perceived health. We also asked about potential barriers and facilitators for participation in discussions. Demographic data were obtained from the electronic health record and by survey. We examined descriptive statistics and conducted multivariate logistic regression to identify correlates of willingness to participate in ACP.

Results: Surveys were completed by 150 of 206 eligible patients (72.8% response rate). Only 6.7% had previously discussed their care preferences with their ACHD clinician, 4.0% with another cardiologist, and 3.4% with their primary care physician. A minority of participants (28.7%) had done some prior ACP, accomplished by naming a durable power of attorney for healthcare (4%), completing a living will or advance directive (7.3%), or both (17.3%). The majority of participants (78.9%) were at least moderately willing to have a meeting to discuss care preferences, 69.1% to participate in ACP, and 91.3% to speak to a palliative care specialist. We identified correlates of being "very willing" to participate in ACP and found that being married and believing "you have a shorter lifespan than your peers" were both significantly associated with willingness (Table), while age, gender, education, and severity and perceived severity of ACHD were not. When asked with which provider patients would prefer to have future discussions, 48.6% chose their ACHD doctor, 6.9% indicated another cardiologist, 12.5% chose their primary care physician, and 17.4% chose multiple providers. The most commonly identified barrier to ACP was "I have not felt sick enough to talk with my doctor about end-of-life care." The most commonly identified facilitator was "I worry that I could be a burden on my friends and family if I were to become very sick."

Conclusion: Patients with ACHD are willing to have advance care planning discussions. They prefer to have these conversations with their ACHD doctor, but are willing to meet with a palliative care specialist if recommended. Patients with shorter self-perceived lifespan and those who were married were particularly interested in ACP. Identification of common barriers and facilitators, such as symptom status and concern for burden on family and friends, may help guide ACP interventions.

Table: Predictors for Being Very Willing to Participate in Advance Care Planning

Predictor	Odds Ratio	95% CI	P-value
Older Age	0.98	0.95-1.01	0.19
Male Sex	1.45	0.71-2.94	0.31
Education Beyond High School	1.43	0.68-3.03	0.35
Being Married	2.16	0.68-3.03	**0.047**
CHD Lesion Severity	0.95	0.55-1.63	0.84
Perceived CHD Lesion Severity	1.02	0.62-1.67	0.95
Anticipating Shorter Lifespan	0.52	0.30-0.89	**0.017**

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PERCEPTIONS OF ADVANCE CARE PLANNING AND PALLIATIVE CARE AMONG ACHD CLINICIANS

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Background: Moderate and complex congenital heart disease (CHD) remains a life-shortening illness. Symptom management, advance care planning (ACP), and palliative care (PC) are gaining recognition as critical components of comprehensive care. Previous research has indicated that although most adults with CHD would like providers to initiate ACP discussions earlier in the disease course, these often do not occur, and the optimal setting and timing for introduction of PC remain debated. The objectives of this study were to evaluate ACHD clinicians' (1) comfort level with managing physical symptoms and psychosocial needs and (2) perspectives on the decision/timing of initiation of ACP and referral to PC.

Methods: This was a cross-sectional study of adult CHD clinicians attending an ACHD symposium. The survey, developed for this study, included descriptions of six hypothetical patients, most of whom had been recently hospitalized for heart failure. Each case was followed by a series of questions regarding clinician comfort managing specified domains of care (response options ranged from 0 = "not at all comfortable" to 10 = "extremely comfortable"). Other case questions included initiation of ACP and PC referral. This was followed by questions about participant background, including ACP/PC training, knowledge and experience. Descriptive results are presented here**.**

Results: Fifty clinicians completed surveys; physicians comprised 72% of the sample. Altogether, respondents self-rated their PC knowledge as 5.3 ± 2.1 on a 0-10 scale; 55% reported no previous training in PC. Average comfort level in managing domains of care are shown in the Table. Across the six cases, the average percentage of clinicians who would initiate ACP ranged from 25-67%, and probable referral to PC ranged from 14-32%. In general, when asked about the best time to initiate discussions of ACP, 22% of respondents said they would wait until the development of a life-threatening complication or hospitalization and 8% preferred to wait until the occurrence of multiple complications or hospitalizations.

Conclusions: Clinicians appeared more comfortable managing physical symptoms and discussing prognosis than psychosocial needs. Clinicians noted high value in advance directives yet were themselves often unlikely to initiate ACP and refer to PC. This raises the critical questions of how and by whom discussion of these important matters should be initiated, and how to best support ACHD clinicians in these endeavors.

	Comfort managing the following domains				**Importance of advance directive**	**Provider would probably:**	
Case	Fluid retention	Dyspnea	Psycho-social	Discussing prognosis		Initiate ACP	Refer to PC
1	6.6 ±2.6	6.5 ±2.4	5.9 ±2.0	7.1 ±2.0	8.7 ±1.6	47%	30%
2	7.3 ±2.2	7.2 ±2.1	6.2 ±1.9	7.3 ±1.7	8.7 ±1.5	35%	14%
3	7.5 ±2.2	7.2 ±2.1	6.2 ±2.0	7.4 ±2.1	8.3 ±1.9	34%	16%
4	8.0 ±2.2	7.6 ±2.3	6.6 ±1.9	7.2 ±2.0	7.9 ±1.9	30%	30%
5	7.6 ±2.3	7.4 ±2.1	6.4 ±2.0	7.6 ±1.9	9.4 ±1.3	67%	32%
6	n/a	n/a	6.3 ±2.2	6.8 ±2.2	8.3 ±1.9	25%	29%

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DETECTION OF ARRHYTHMIAS IN ADULT CONGENITAL HEART DISEASE PATIENTS WITH IMPLANTABLE LOOP RECORDERS

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Background: Rhythm disorders are the leading cause of morbidity and mortality in adults with congenital heart disease (ACHD). Non-invasive arrhythmia monitoring may not detect infrequent or asymptomatic arrhythmias. Implantable loop recorders (ILRs), such as the Reveal LINQTM (Medtronic), have been shown to be useful in long-term arrhythmia monitoring in adult patients to document occult atrial fibrillation. However, to date there have been no reports of the use of ILRs in the ACHD population. We proposed that the ILRs will detect arrhythmias, missed by other monitoring modalities, which result in change in management in ACHD patients.

Methods: This is a retrospective review of the use of Reveal LINQTM ILRs in ACHD patients followed at a single center from December 2014 to December 2017. Medical records were reviewed to determine cardiac diagnosis, indication for implant, ILR findings, and changes in management.

Results: Twenty-three ACHD patients, mean age 30 years (range 21 - 50 years), underwent ILR implantation were followed for an average of 23±13 months. Among the 23 patients 8 patients had Fontan palliation (35%), 7 had Tetralogy of Fallot (TOF) (30%), 2 had pulmonary stenosis (10%) and 6 patients had other CHD (26%). Reasons for implant were palpitations (70%), documented arrhythmia (69%), dizziness (43%), and syncope (39%); several patients had multiple indications for implant. Arrhythmias resulting in change in therapy were documented in 8 (35%) patients; 4 (17%) had medication change, 1 (4%) underwent implantable cardioverter-defibrillator (ICD) implantation, 1(4%) underwent pacemaker implantation, 1 (4%) had an electrophysiology study (EPS), and 1 (4%) had a cardioversion. Underlying diagnoses among patients with positive findings were: Fontan (4/8, 50%), TOF (1/8,12%), Ebstein's Anomaly (1/8,12%), Interrupted Aortic Arch (1/8,12%), and Congenital Aortic Stenosis (1/8,12%). Seven of the 8 patients with positive findings (88%) had routine Holter or event monitor which demonstrated no arrhythmia. The average time from implant to positive finding was 361+ 316 days. Among the 8 patients with Fontan who had an ILR implanted, 4 patients (50%) had positive findings which resulted in cardioversion, EPS, ICD placement, or initiation of anti-arrhythmic medication. Only 1 patient with TOF had a positive finding of sinus node disease with sinus pause and syncope, resulting in pacemaker placement. Pertinent negative events occurred in 1 patient (4%) who had Fontan and syncope corresponding to sinus rhythm on ILR. ILR reprogramming was needed in 1 patient due to asymptomatic tachycardia events corresponding to sinus tachycardia with exercise. Over-sensing resulting in false positive tachycardia events occurred in 3 patients, all of whom had TOF; two had double counting of QRS due to right bundle branch block.

Conclusion: This is the first study to utilize ILRs as an adjunct for arrhythmia monitoring in the ACHD population with pertinent positive and negative findings affecting management in 39% of patients. Patients with Fontan palliation had the highest rate of positive findings (50%) affecting management. ILR is rate-based detection without morphology discrimination; therefore, programming consideration should focus on primary indication for implant and patients' underlying conduction system.

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INCIDENCE, MANAGEMENT AND MECHANISMS OF ARRHYTHMIAS IN ADULT PATIENTS POST RASTELLI PROCEDURE: TWENTY-FIVE YEARS SINGLE CENTRE EXPERIENCE

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Background: Arrhythmias are observed in patients who underwent the Rastelli procedure however little is known about the clinical course and mechanisms of these arrhythmias in long term follow up. This study aims to evaluate the incidence, mechanism and management of arrhythmias post-Rastelli procedure.

Methods: In this retrospective study, we analyze 37 adults post Rastelli procedure, referred to an adult congenital cardiac clinic.

Results: N=37 patients. Male=60%. During median follow-up period of 25 years, tachy-arrhythmias occurred in 43% patients . The median time of arrhythmia onset post-operation was 23 (IQR-21) years. Supraventricular tachycardia (SVT) was the commonest type of tachyarrhythmia observed in 14(37%) patients, ventricular tachycardia (VT) in 5(13%) and both SVT/VT in 3(8%). Twelve ablation procedures were performed in 6(16%) patients. SVTs were intra-atrial reentrant tachycardias (IARTs) including cavo-tricuspid (CTI) isthmus and right atrial incisional scar related tachycardia. VTs were macro-reentrant or micro-reentrant mechanisms occurring around the base of the right ventricle to pulmonary artery conduit. Ablation procedures (median 2/patient) were successful in achieving arrhythmia free survival in all patients (IQR-2years) without any complications. 32% patients underwent device implantation of which 21% were pacemakers for complete atrioventricular block and 13% were AICD implantations. Post-index device implantation, complication rate of 10% was observed during the follow-up.

Conclusion: The incidence of late arrhythmias is high (43%) after Rastelli procedure. The tachyarrhythmias are primarily scar-related with both macro-reentry and micro-reentry mechanisms seen. Ablation in this patient population is safe but more than one procedure may be needed to successfully treat the arrhythmias.

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HIGH DENSITY MAPPING FOR ABLATION OF ATRIAL TACHYARRHYTHMIAS IN ADULTS WITH CONGENITAL HEART DISEASE

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Background: Ablation of atrial tachyarrhythmias in adults with congenital heart disease (ACHD) is complex. The use of 3-dimensional electroanatomic mapping systems (EAM) is recommended and acute success rates using EAM are estimated to be greater than 80%. High density mapping with the PentaRay® (Biosense Webster) catheter has been used safely for ablation of ventricular tachycardia in ischemic and non-ischemic cardiomyopathy patients; however, high density mapping has not been studied in ablation of atrial tachyarrhythmias in ACHD patients. We proposed the addition of high density mapping with PentaRay® mapping catheter, in addition to 3D EAM, is safe and effective for ablation of atrial tachyarrhythmias in adults with CHD

Methods: This is a single center retrospective cohort study of adults with CHD who underwent ablation procedures for atrial tachyarrhythmias from 2013 - 2017. Patients were divided into two cohorts: those who underwent ablation with EAM and those who underwent ablation with EAM with addition of PentaRay® mapping catheter (EAM+P).

Results: Fifteen ablations were performed in 13 patients using standard EAM, and 11 ablations were performed in 10 patients using EAM+P. There was no difference in mean age (38 vs 33 years, p=0.08) or complexity of CHD (66% vs 64% complex CHD, p=1). The proportion of combined catheterization and electrophysiology (EP) study vs EP study only was higher, though not significant, in EAM group compared to EAM+P (53% vs 18%, p=0.1). Median fluoroscopy time was longer in EAM group (26.2 mins, IQR 12-54min) compared to EAM+P group (5min IQR 1-7.5min) p=0.0017. A higher number of sheaths were used for EAM cases compared to EAM+P (p=.00142). Although not statistically significant, the EAM group had longer EP procedure time compared to EAM+P group: median (IQR) of 207 min (148-381min) vs. 187 min (94-201min). Acute success rates of ablation were similar in the two groups with 93% success in EAM and 100% success in EAM+P. There were no procedural related complications in either group.

Conclusion: This is the first study to demonstrate the safety and efficacy of high density PentaRay® mapping catheter in addition to 3D mapping system for ablation of atrial tachyarrhythmias in ACHD patients. Use of PentaRay®mapping catheter demonstrated shorter fluoroscopy time and decrease number of access sites needed with a trend toward shorter procedure time. There was no difference in acute success rate with the addition of PentaRay® mapping catheter in the ablation of atrial tachyarrhythmias.

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HIS BUNDLE PACING IN EBSTEIN'S ANOMALY

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Purpose: Ebstein's anomaly is a malformation of the tricuspid valve (TV) and right ventricle (RV). In the presence of cyanosis and heart failure, surgery, which often includes repair of the TV, is indicated. Development of postoperative AV nodal conduction block warrants permanent pacemaker implantation. Transvenous lead mplantation in the RV proper carries the risk of injury to the surgically repaired TV. Epicardial lead implantation is cumbersome and fraught with long-term sensing and capture issues. Placement of the ventricular lead in a His position, without crossing the valve, eliminates risk of both acute TV damage and progressive insufficiency.

Methods: Due to cyanosis and symptoms of heart failure, a 27 year old male underwent tricuspid valve repair with posterior annular plication, annuloplasty ring placement, ASD closure, right atrial reductionplasty, and biatrial CryoMaze procedure. The patient was extubated on postoperative day 1 and weaned off inotropic and pressor support on postoperative day 2. Due to persistent complete AV block, the patient underwent dual-chamber transvenous pacemaker implantation. A Medtronic 3830 SelectSecure MRI Surescan lead was placed proximal to the TV in the His bundle position, where it captured atrialized RV myocardium.

Results: His pacing with lead placement proximal to the TV eliminated the risk of lead induced TV insufficiency. The resultant nonselective His capture engaged the proximal His-Purkinje conduction system and resulted in a relatively narrow paced QRS morphology without the typical left bundle morphology of RV apical pacing. Transthoracic echocardiogram prior to discharge revealed adequate mobility of the TV with trivial insufficiency and no atrial level shunt.

Conclusions: Injury to the compact AV node is an inherent risk of TV repair in Ebstein's anomaly. As demonstrated in this case, His pacing can be successfully achieved in Ebstein's patients following TV repair. It avoids TV injury and eliminates risk of lead-induced TV insufficiency. Furthermore this strategy may avoid the development of LV and RV dysfunction, which can be associated with long term RV apex pacing. His bundle pacing should be considered as the mode of choice in Ebstein's anomaly patients.

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MOBILE HEALTH IN ADULT PATIENTS WITH CONGENITAL HEART DISEASE REDUCES EMERGENCY HOSPITALIZATIONS

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Background: Adult patients with congenital heart disease (CHD) have residual sequelae and may experience emergency presentations because of heart failure (HF) and/or arrhythmias. As they are relatively young of age and under lifelong surveillance, they seem an appropriate population for mobile health (mHealth), as timely intervention might prevent clinical deterioration. The aim of this study is to evaluate whether telemonitoring through mHealth in adult patients with CHD reduces emergency hospitalizations.

Methods: Symptomatic adult patients with CHD were enrolled in an mHealth telemonitoring program that evaluates blood pressure (BP) and weight twice a week and single lead EKG once a week. Data was matched with individualized thresholds and EKGs were analyzed by trained nurses. If data exceeded patient specific thresholds consecutively twice or more, or if arrhythmias were found, patients were contacted by their treating cardiologist for treatment adjustments or reassurance. If data was missing, patients received an app notification. Adherence was registered. Emergency hospitalizations were defined as cardiac hospitalizations and emergency care presentations. Patient reported outcome measures were determined using three different questionnaires; CaReQoL, EQ-5D-5L for quality of life and PAM-13 for self-management. Questionnaires were performed at baseline and repeated every three months.

Results: So far, 55 symptomatic adult patients with CHD (median age 45 years (range 19-70), 34,5% male and CHD severity of mild (6), moderate (29) and severe (20)) were included; 22 patients experienced HF NYHA class ≥II, 43 had palpitations or arrhythmias and eight had hypertension. Mean follow-up was 3.0 months, adherence was 97%. During follow-up there were three emergency hospitalizations, in contrast to the 19 emergency hospitalizations during a similar time period in historical data (P=0,044). So far, PROM questionnaire results showed a trend towards improvement in quality of life (CaReQoL and EQ-5D) with an increase of 17% in questionnaire scores and towards decrease in self-management (PAM-13) of 7,3%.

Conclusion: This mHealth telemonitoring program in adult patients with CHD showed an excellent adherence compared to previous telemonitoring studies and a reduction of emergency hospitalizations compared to historical data. A tendency towards a better quality of life was observed.

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TRENDS IN HOSPITALIZATION FOR HEART FAILURE PATIENTS WITH GROWN-UP CONGENITAL HEART

Mike Saji, Morimasa Takayama; Tomomi Ueda; Satoshi Yazaki; Tadahiro Yoshikawa; Katsuhiko Mori; Naoki Wada; Makoto Ando; Shuichiro Takanashi; yukihiro Takahashi; Mitsuaki Isobe; *Sakakibara Heart Institute, Japan*

Introduction: Living well is as important as living longer. Quality of life (QOL) has gained much attention in biomedical science over the past few decades. Reducing rehospitalization has been targeted as a top strategic priority in an effort to improve patient care and decrease costs. For patients' side, as the heart failure stage progresses, episodes of acute heart failure are likely to happen more often, and patients become less likely to make a good recovery. Therefore, multiple rehospitalizations will result in decline in QOL. For medical side, additional costs associated with high rehospitalization rate may lead some to question the cost-effectiveness. Although QOL in Japanese population has been reported recently, relationship between heart failure and rehospitalization in this population remain unknown. This study aims to demonstrate the real-world data regarding rehospitalization in heart failure patients with grown-up congenital heart in high-volume center in Japan.

Methods: We reviewed 1610 patients who were diagnosed with congenital heart disease, and were older than 20 years in our out-patient clinic. Complexity of congenital heart disease was determined according to administrative data and patient cohort was divided into three groups; simple, moderate, and great complexity. Brain natriuretic peptide (BNP) and N-terminal pro-brain natriuretic peptide (NT-pro BNP) were examined at out-patient clinic. Our institutional review board approved the study. All information was retrospectively obtained from patients' medical records or telephone interview. Therefore, there was follow-up and accountability of all patients.

Results: Patients at out-patient clinic in our hospital has increased from 624 to 761 patients/year (121%) for last three years. Out of 1610, 1009 (63%) had blood sample of BNP or NT-pro BNP. Patients with BNP or NT-pro BNP level greater than normal limit had more frequent hospitalizations than those within normal limit (6.0 vs. 3.1, $p < 0.001$). Patients with great complexity disease had greater level of NT-pro BNP than those with moderate complexity or simple disease (1140 vs. 806 vs. 390 pg/ml respectively, $p < 0.001$). Furthermore, Patients with great complexity disease had larger number of hospitalizations than those with moderate complexity or simple disease (6.6 vs. 3.8 vs. 1.7 times/after starting follow-up respectively, $p < 0.001$). In Kaplan-Meier analysis, estimated cumulative survivals were 73.4 ± 4.5% in patients with BNP or NT-proBNP level greater than normal limit, and 97.1 ± 1.6% in those within the limit with mean follow-up of 36 ± 0.4 years, respectively (log rank $p < 0.001$).

Conclusion: Follow-up patients in adults with grown-up congenital heart are growing in high-volume center in Japan. Rehospitalization was more frequent in patients with heart failure and in patients with congenital heart disease with greater complexity. Those patients should be managed appropriately by multidisciplinary heart team consisting of pediatric cardiologists, pediatric surgeons, interventional cardiologists, cardiovascular surgeons and heart failure specialists.

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COMPREHENSIVE THREE-DIMENSIONAL RECONSTRUCTION OF ADULT CONGENITAL HEART DISEASE

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Background: The complex 3-dimensional anatomy often present in unrepaired and repaired adults with congenital heart disease (ACHD) is difficult to appreciate using conventional 2-D images. Volume-rendered 3-D reconstructions permit more precise anatomical understanding.

Methods/Results: We present an illustrative guide to our techniques for 2-D computed tomographic (CT) acquisition and subsequent 3-D reconstruction utilizing endocast and various forms of virtual imaging, including virtual dissection, highlighting its potential utility in ACHD (slides 1-5).

Conclusion: 3-D CT reconstruction utilizing both endocast and virtual dissection imaging may be useful in improving anatomical understanding in.

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ATRIAL SEPTAL DEFECT: COMPREHENSIVE THREE-DIMENSIONAL RECONSTRUCTION USING DUAL-SOURCE COMPUTED TOMOGRAPHY

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Background: The atrial septal defect (ASD) is a common adult congenital heart lesion. Transesophageal echocardiography (TEE) is currently accepted as the gold standard for pre-procedural assessment of the 3D anatomy of the lesion and its surrounds. TEE commonly provides optimal results. In some cases, however, it has limitations in visualizing the correct anatomy due to its narrow field of view, suboptimal quality of the 3-D images, or wide anatomical variation in the individual structural anatomy.

Methods/Results: We present an illustrative guide to our techniques for 2-D computed tomography (CT) acquisition and subsequent 3-D reconstruction utilizing virtual dissection imaging, comparing this to 3D TEE and autopsy dissection (slides 1-5).

Conclusions: This technique of virtual dissection may be an optimal alternative where TEE shows suboptimal findings. It can demonstrate in comprehensive fashion the 3-D anatomy of the communication between the atrial chambers in relation to the surrounding structures.

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PLANNING AND IMPLEMENTATION OF A TRANSITION PROGRAM

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Background: Due to advances in surgical and medical management there is a rapidly growing population of adolescent and young adult survivors of congenital heart disease (CHD). Recent data reveals there are more adults than children with CHD. These CHD survivors are at risk of both cardiac and non-cardiac morbidity and premature mortality. The majority do not undergo proper transition and experience lapses in care (Mackie et al., 2016; Reid et al., 2004; Wray et al., 2013). These lapses in care place them at a higher risk of late cardiac complications (Yeung et al., 2008). At our institution in 2017 there were 3,291 unique CHD patient visits of teens between the ages of 12-18. Unfortunately, in 2017 only 43 patients between 18-22 years old transferred care to an adult congenital cardiologist. We hypothesized that many young adults are being lost to follow up due to lack of a formal transition program and education. To address this issue, we developed a transition of care program to empower our adolescent patients with education, life skills, and to ensure proper follow-up.

Planning and Implementation: The transition program is based upon national guidelines and upon the gottranstion.org framework. A needs assessment was performed among the pediatric cardiology department which showed an overwhelmingly positive response to the need for a program. Departmental approval was sought for and obtained to establish a program and to hire a transition clinician. The program was subsequently developed by a doctoral nurse practitioner student and the director of the adult congenital cardiology program. The transition program is designed to begin at age 12 with an introduction to the transition team and education on the transition policy. We created an online database for tracking of transition activities using REDCap software. Periodic transition assessments will be performed to assess transition readiness and to tailor education to patient's needs. A medical health passport will be used to summarize pertinent medical information. Finally, a transfer of care checklist was created to ensure proper hand-off from the pediatric to adult provider.
For assessment of efficacy of the program, a health care transition process measurement tool developed by gottransition.org was used. It is an objective tool that utilizes the three main categories of implementation in practice, youth/family feedback and leadership, and dissemination in practice. The highest possible score is 100. Our score prior to the planning process was 0, and the current score just after planning and development is 38. We anticipate this score to reach near 100 within the first year of implementation.

Conclusions/Future Decisions: As the survival of children born with CHD continues to improve, formal transition programs become an integral part of the congenital cardiology practice. We hope to collect data on an ongoing basis and improve the efficacy of our transition program on an ongoing basis. Our goal is to provide seamless care through our transition practice in order to improve health outcomes and also ensure that patients are not lost to follow up.

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FACTORS ASSOCIATED WITH HAVING A PRIMARY CARE PROVIDER AMONG PATIENTS WITH ADULT CONGENITAL HEART DISEASE

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Background: Similar to all patients with a chronic health care issues, it is recommended that patients with adult congenital heart disease (ACHD) have a primary care provider (PCP) for management of non-cardiac conditions, prevention and promotion of health including mental health. However, many ACHD patients do not have an established PCP. The aim of this study is to examine factors associated with having an established primary care provider among ACHD patients.

Methods: From July 2017 to December 2017, 427 patients (47% male) completed clinical questionnaires including whether the patients had an established PCP. Patient demographics including age, gender, education level, disease complexity, and presence (or absence) of an established PCP were analyzed. Patients without an ACHD diagnosis were excluded.

Results: Mean age was 32 + 10 years (range: 17-74 years). Disease complexity was classified as simple (n=78, 18%), moderate (n=209, 49%), and complex (n=140, 33%). Of 427 patients, 222 answered the question on presence or absence of established PCP. Overall, 83% (184 patients) indicated having an established PCP. Of these patients, rates of having an established PCP by disease complexity was 78.3% (n=36) for those with simple defects, 85.1% (n=86) for moderate complexity, and 82.7% (n=62) for those with complex disease. Those patients with an established PCP were older than those without (32.3 years versus 27.9 years, $p < 0.01$). There was no significant difference in gender, education level, or disease complexity among these groups. When controlled for age, there was no significant association between disease complexity and having an established PCP (OR=0.96, 95% CI=0.49-1.87).

Conclusions: In a cohort of ACHD patients followed by a single ACHD center, higher age was the only factor associated with having an established PCP, and gender, education level, or disease complexity were not found to be associated factors; however, the data is limited as the PCP status of those not completing the questionnaire is unknown. Further study is needed to better understand the limitation to obtaining an established PCP in the ACHD population.

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RISK-BENEFIT RATIO MAY NOT JUSTIFY A FURTHER DECREASE IN THRESHOLD FOR PULMONARY VALVE REPLACEMENT LATE AFTER TETRALOGY OF FALLOT REPAIR: AN EXPERIENCE WITH 2579 PATIENTS

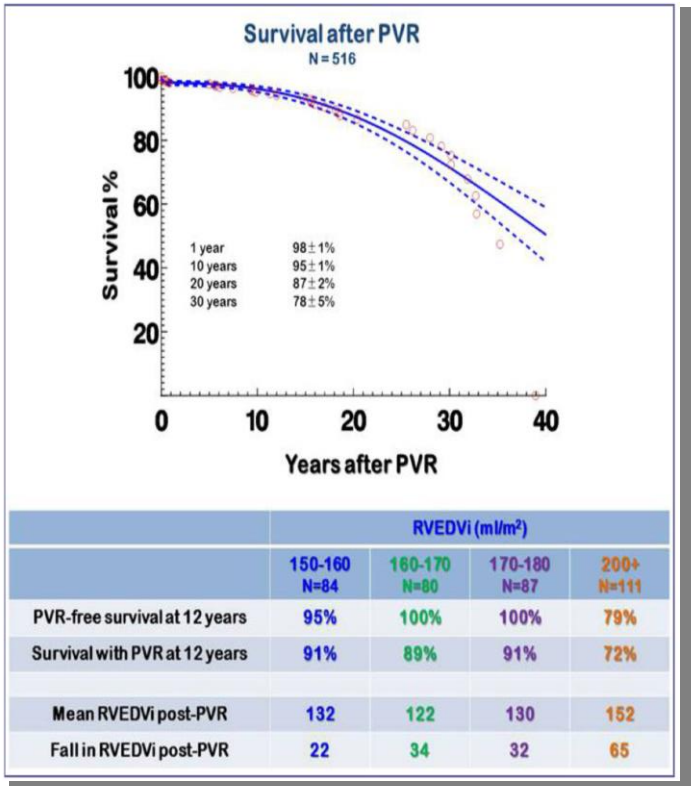
Edward J Hickey, Ting Ting Low; Susan L Roche; Candice Silversides; Erwin Oechslin; Jack Colman; Andrew Crean; Lee Benson; Eric Horlick; Mark Osten; Rachel Wald; *The Hospital for Sick Children, Toronto General Hospital, Toronto, Canada*

Background/Objective: Recommended thresholds for pulmonary valve replacement (PVR) in asymptomatic patients after tetralogy of Fallot (TOF) repair are continually decreasing. We studied the natural history versus outcomes after PVR in various categories of indexed right ventricular end-diastolic volume (RVEDVi).

Methods: Acquisition of all repeated measure datapoints and cross-sectional review in 2579 patients (born 1924 - 2011), including 7553 echos, 2579 MRI scans and all interventional data. Analysis was via parametric competing risks techniques and time-related regressions adjusted for repeated measures.

Results: Survival was 95%, 81% and 73% at age 20, 40 and 60 years respectively. Freedom from PVR (N=516, mean age 25 years) was 87%, 66% and 45% at 20, 40 and 60 years. Overall, survival after PVR was 98%, 95% and 87% at 1, 10 and 20 years. PVR-free survival in all 345 patients with MRI RVEDVi > 150 was 95% at 15 years. Comparative survival showed no survival advantage with PVR versus natural history for RVEDVi 150-160, 160-170, 170-180 (table); these categories had excellent PVR-free survival approaching 100% at 10 years. Patients with RVEDVi > 200 had late survival decrements with PVR or without. PVR resulted in a large and significant reduction in RVEDVi (mean 40 ml/m2, P<.0001), after which RVEDVi remained stable (P=.10). Patients with RVEDVi 150-160, 160-170 or 170-180 had similar reductions in RVEDVi after PVR to comparable levels (table). 35% of children transitioning to adult care had RVEDVi > 150 ml/m2

Conclusions: Lowering the RVEDVi threshold for PVR does not appear to offer a clear survival advantage and offers small differences (if any) to RV geometry, potential procedure-related morbidity and endocarditis risk and would mean intervening on many teenagers with repaired TOF who could otherwise anticipate intervention in later life.



	RVEDVi (ml/m²)			
	150-160 N=84	160-170 N=80	170-180 N=87	200+ N=111
PVR-free survival at 12 years	95%	100%	100%	79%
Survival with PVR at 12 years	91%	89%	91%	72%
Mean RVEDVi post-PVR	132	122	130	152
Fall in RVEDVi post-PVR	22	34	32	65

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SAFETY AND EFFICACY OF EXERCISE TRAINING IN SYMPTOMATIC PATIENTS WITH MODERATE AND SEVERE CHD: A RANDOMIZED CONTROLLED TRIAL

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Background: The large majority of studies on the effect of exercise training in adults with congenital heart disease (CHD) have been performed in asymptomatic patients (New York Heart Association (NYHA) class 1). Moreover, most studies are limited by short duration and standardized nature of the exercise program with low compliance. Purpose: To assess whether long-term, home-based, self-selected, individualized exercise training is safe, improves peak exercise capacity (peakVO2), and quality of life and lowers serum N-terminal prohormone brain natriuretic peptide (NT-proBNP) levels in adults with CHD and heart failure symptoms (NYHA ≥2). In addition, we expected high compliance because patients could self-select their exercise program.

Methods: Forty CHD adults in NYHA class II or III were randomized to an intervention group (n=20) with three home-based, self-selected exercise training sessions of 40 minutes three times per week for 6 consecutive months, or to a control group (n=20) with usual care alone. Exercise training sessions were individualized based on heart rate reserve (80%) determined at baseline cardiopulmonary exercise test. Randomization was stratified by CHD complexity. At baseline and 6 months follow-up, we determined peakVO2, serum NT-proBNP levels, and quality of life through SF-36 and TAAQOL-CHD questionnaires. The study is registered at http://clinicaltrials.gov, identifier: NCT02825472.

Results: Forty patients (mean age 39.9±12 years, NYHA II 90%, male 55%, severe CHD complexity 63%) were included, of whom 34 patients completed the protocol and were analyzed. Reasons for drop-out were lack of interest (n=4) and lost-to-follow-up (n=2). None of the patients in the intervention group had to discontinue the training program due to adverse events related to exercise. The majority of patients chose to perform interval training with cardio-fitness (73% of patients), swimming or running (both 20% of patients). At baseline, peakVO2 was 23.9±5.9 ml/kg/min in the exercise group and 26.9±6.3 ml/kg/min in the control group (p=0.153). PeakVO2 increased significantly in the exercise group (mean Δ 1.85 mL/kg/min, 95%CI: 0.52 to 3.17; p=0.010), mainly in patients with severe CHD complexity, but not in the control group (mean Δ 0.75 mL/kg/min, 95%CI: -0.40 to 1.90; p=0.184) (Figure 1). No significant changes were found in serum NT-proBNP levels or quality of life.

Conclusion: Home-based, self-selected exercise training is safe and beneficial in symptomatic CHD adults, especially with severe CHD complexity, with reasonable compliance. Physicians should encourage CHD adults with heart failure symptoms, after individual work-up, to perform exercise training of their preference. Figure 1. Change in peakVO2 between the two randomization groups. Boxplot with 25th, 50th and 75th percentiles, error bars represent 95% confidence interval of the median, red dots represent mean change in peakVO2.

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SELF-CARE IN ADULT CONGENITAL HEART DISEASE: DO YOUNG ADULTS EXHIBIT PREFERENCES FOR HEALTH CARE AUTONOMY?

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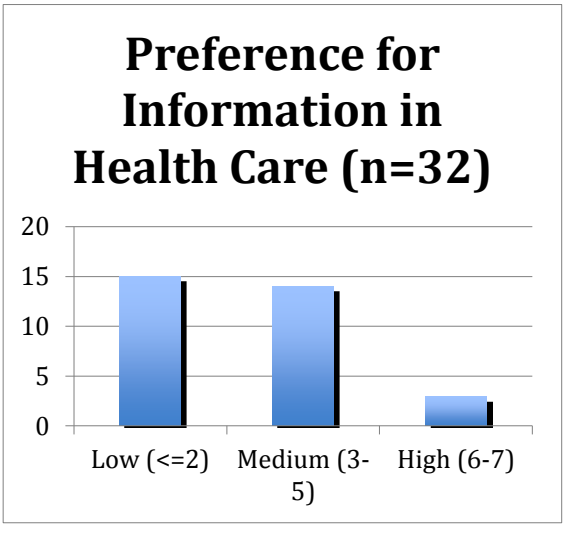
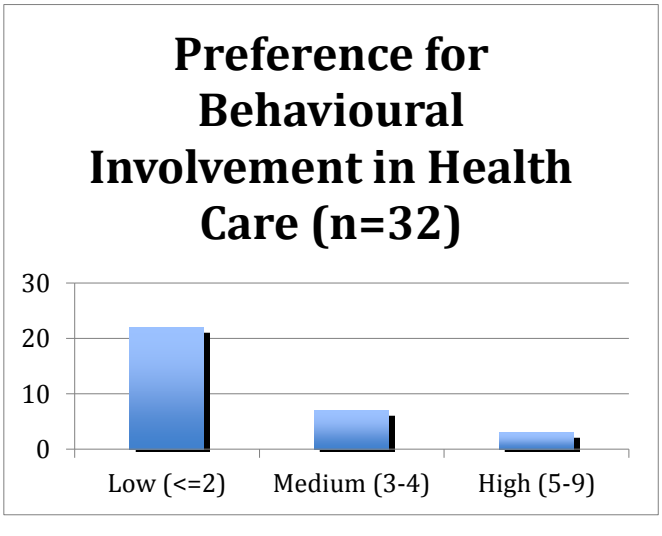
Background: The majority of patients born with congenital heart disease (CHD) now live to adulthood and facilitating successful transition to adult care has been deemed a priority. Health care autonomy has been identified as a predictor of successful transition. The Krantz Health Opinion Survey (HOS) is designed to assess individual attitudes toward health care and is a validated measure to assess preferences for receiving medical information and involvement in health care decision-making. We performed the Krantz HOS on adults with CHD in Saskatchewan to evaluate their desire for health care autonomy.

Methods: This project was one component of a wider study assessing transition success among ACHD patients in Saskatchewan, Canada. We contacted patients with CHD who were deemed to require lifelong cardiac care and should have transitioned to adult care between 2007 and 2014. We report the Krantz Health HOS component of this study which was one component of the telephone survey conducted.

Results: Of the 106 patients contacted, 32 consented to participate in the telephone survey portion of the study (30% response rate). The majority of respondents (n=19, 59%) were male with an average age of 22 +/- 2 years. All but one of the participants, who immigrated from the Phillipines at age 14, were born in Canada and their first language was English. Twenty-two respondents (69%) had CHD of moderate to severe complexity while the remainder (n=10, 31%) had simple CHD. The majority of participants (n=24, 75%) received a low overall score on the Krantz HOS survey (6 or less). The remainder (n=8, 25%) received a medium score. None of the patients surveyed received a high score suggestive of health care autonomy. When stratified based on preferences for receipt of medical information and self-care behaviours (Figures 1 & 2), more participants (n=17, 53%) indicated a preference for receiving information about their health than those who wanted to be actively involved in decision-making about their health (n=10, 31%).

Conclusion: Young adults with CHD express passive preferences toward receiving information about their health and decision-making. This presents a challenge as most current transition strategies rely upon patient engagement in educational efforts. These preferences reflect an external locus of control regarding health. They could represent innate personality traits of young adults with CHD but, because they were almost universal among those surveyed, they more likely result from decades of engagement with the health care system and reflect a learned coping strategy. Further research should focus on behavioural interventions to improve self-care practices while investigating and acknowledging the psychological effects of chronic illness on young adults.





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GENETIC PREDICTORS OF ADVERSE RIGHT VENTRICULAR REMODELLING IN TETRALOGY OF FALLOT POST-REPAIR

Marie-A Chaix, Oyediran Akinrinade; Myriam Lafreniere-Roula; Rachel Van der Laan; Luis Altamirano Diaz; Lynn Bergin; Erwin Oechslin; Glen van Arsdell; Cedric Manlhiot; Connie Bezzina; Seema Mital; *Labatt Family Heart Centre, The Hospital for Sick Children, University of Toronto, Toronto, Canada*

Background: Tetralogy of Fallot (TOF) is an important cause of adverse right ventricular remodelling (RVM) and dysfunction. No medical therapies have been proven to be effective after dysfunction occurs. Pre-surgical, surgical and environmental factors contribute to the risk of RVM in TOF, but little is known about the genetic predictors and molecular pathways associated with RVM.

Methods: We performed whole genome sequencing on 501 TOF patients after surgical repair, and identified rare and low-frequency variants (MAF < 0.05) that were predicted to be damaging. Enrichment of genes for rare and low-frequency variants was evaluated using gene collapsing burden analysis, Kernel-Based Adaptive Cluster (KBAC) and Sequence Kernel Association Test for survival traits (SKAT) between 294 cases with adverse RVM [defined as moderate or severe RV systolic dysfunction (MRI or TTE), moderate or severe RV dilatation (MRI or TTE) and/or need for pulmonary valve replacement (PVR)] and 207 controls with no or mild RVM. We also did a subgroup analysis in patients with early RVM i.e. RVM <17 years post repair (n=137) versus controls i.e. no RVM in patients with at least 17 years of follow-up after repair (n=64).

Results: The median age at repair was 16 months (range:6-44), and the median age at PVR (n=159) was 24 years (range:16.0-38.0). Freedom from adverse RVM was 87.4% (95% CI:83.7-90.2) and 57.3% (95% CI: 52.0-62.2) at 10 and 20 years respectively. The median follow-up duration was 16 years (range:4-28). We identified significant enrichment of damaging variants in genes associated with cytoskeleton organisation, inflammation, fatty acid metabolism and mitochondrial metabolism in cases compared to controls (p-value <10-3). Furthermore, a significant enrichment of variants in pathways of apoptosis signalling and growth processes (p-value< 10-3) was observed in TOF patients without RVM.

Conclusion: Our study identified genes and biological pathways that are associated with adverse RVM in TOF patients. Knowledge of genetic susceptibility can be utilized for early identification of TOF patients at risk for adverse RVM who may benefit from earlier TOF repair. These pathways may also provide new pharmacological targets for developing drugs aimed at preventing or ameliorating adverse RVM.

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PATIENT-REPORTED OUTCOMES IN TETRALOGY OF FALLOT: BASELINE RESULTS FROM A PROSPECTIVE, INTERNATIONAL, MULTI-SITE STUDY

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Objectives: In addition to understanding causes of morbidity and mortality following tetralogy of Fallot repair (rTOF), the importance of patient-reported outcomes (PROs) such as quality of life is increasingly being recognized. We hypothesized that PROs in rTOF would be associated with selected sociodemographic factors, functional status, and/or clinical variables.

Methods: As part of a prospective study of patients ≥12 years with rTOF and significant pulmonary regurgitation, participants completed PRO surveys: SF-12 Health Status (physical component summary [PCS] and mental component summary [MCS] t-scores), EQ-5D (5 dimensions: mobility, self-care, usual activities, pain/discomfort, and anxiety/depression), and a 0-100 quality of life linear analogue scale (QOL-LAS). With multivariable regression analysis, we determined the association between PROs and selected predictors.

Results: We enrolled 627 participants (55% male, 26±13 years, 75% adult) at 14 sites from North America, Europe, and Asia. The median age at repair was 1.6 years (IQR 0.6, 4.2). In general, scores on the SF-12 and QOL-LAS suggested good outcomes: SF-12 PCS <18 years 52±6 and ≥18 years 51±9; SF-12 MCS <18 years 52±9 and ≥18 years 49±10; QOL-LAS <18 years 83±14 and ≥18 years 78±16. On the EQ-5D, difficulties were reported with usual activities (16%), pain (25%) and anxiety/depression (38%). In multivariable regression analysis, younger age at enrollment emerged as an independent predictor of better physical health status although worse mental health status ($p=0.0005$ and $p=0.029$, respectively) but was not related to quality of life. Better NYHA functional class was a predictor of better physical health status ($p<0.0005$), mental health status ($p=0.002$), and quality of life ($p<0.0001$). Other factors (including complexity of underlying TOF anatomy, previous shunt palliation and need for arrhythmia intervention) were not associated with PROs.

Conclusion: Age and NYHA functional class at enrollment were found to be predictors of PROs in rTOF. While global measures of health status and quality of life (SF-12 and QOL-LAS) suggest relatively good PROs, this study demonstrates the importance of inquiring about specific health problems (such as using the EQ-5D) to achieve a richer understanding of PROs in order to potentially maximize patient well-being.

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CAST A WIDE NET: CATCHING ACHD REFERRALS IN A SEA OF PROVIDERS WITHIN A LARGE, MULTIHOSPITAL AND MULTISTATE PRIVATE HEALTH SYSTEM - A CONCEPT PROPOSAL

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Background: Many adults with congenital heart disease are not receiving the specialized care they deserve despite the fact that referral to an ACHD center is associated with lower mortality1. In the context of the increasing centralization that is happening in health care, with hospitals and clinics joining into large regional health systems, we have an opportunity to standardize the ACHD referral processes within these systems and improve access to care. Providence Saint Joseph Health (PSJH) is a large, private, Catholic health system that consists of 50 hospitals in seven states. The Providence Adult and Teen Congenital Heart Program (PATCH) is the only ACHD program in that system and has been accredited as an ACHA ACHD Comprehensive Care Center. We have developed a referral process map that is being promoted throughout the PSJH system with the aim to organize the care of ACHD patients, both at the local level with ACHD Lead Physicians, and in referrals to Spokane for complex consultations, procedures, or surgeries.

Method: PATCH team members and PSJH leadership created an ACHD Referral Process Map that is currently being disseminated throughout our health system. We are building a team of ACHD Lead Physicians throughout the system who will serve as local experts who can consult with, or refer to, the main ACHD base in Spokane. We additionally have created quarterly system-wide teleconferences in which any PSJH provider can present his or her own cases to the PATCH team for discussion and education, which may also increase referrals. In the coming years, we plan to survey our referring providers to ask how they initiated a referral to our program. Specifically, we would like to know if, and in what ways, this referral process map was used in that process. We recognize that tracking this data will be challenging, but we hope to use what we learn to improve access to specialized ACHD care.

Results: This is currently a concept proposal in its early stages.

Conclusions: We believe that our referral process map can translate into an increase in referrals to our center, improve care at the local level at sites away from Spokane, and perhaps serve as a model for other large health systems.

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HAPLOINSUFFICIENCY OF VASCULAR ENDOTHELIAL GROWTH FACTOR RELATED SIGNALING GENES IS ASSOCIATED WITH TETRALOGY OF FALLOT

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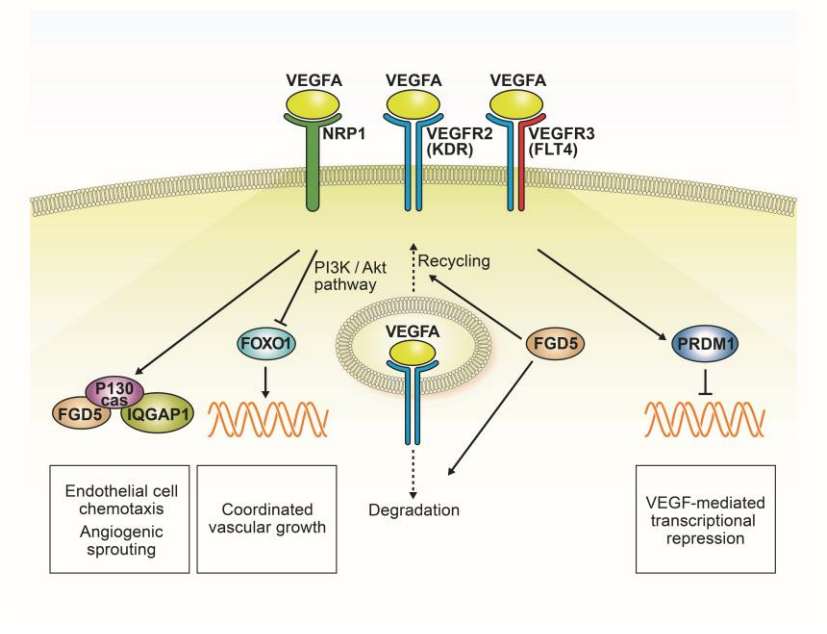
Background: The purpose of the study was to determine disease-associated variants in conotruncal defects, particularly single gene defects in tetralogy of Fallot (TOF).

Methods: We used whole genome sequencing to analyze for variants in FLT4 (VEGFR3) and other genes in the vascular endothelial growth factor (VEGF) pathway, in 231 adults with TOF (n=175) or related cardiac defects from a single site.

Results: We identified nine TOF patients with novel FLT4 variants (5.1%, 9/175: two predicted damaging and seven loss-of-function, including an 8 kb deletion). In 10 other TOF patients we found likely disruptive variants in VEGF-related genes: KDR (VEGFR2; two stopgain and two non-synonymous variants), VEGFA, FGD5, BCAR1, IQGAP1, FOXO1, and PRDM1. Detection of VEGF-related variants (19/175, 10.9%) was associated with an increased prevalence of absent pulmonary valve (26.3% versus 3.4%, p<0.0001) and right aortic arch (52.6% versus 29.1%, p=0.029). Extracardiac anomalies were rare. In an attempt to replicate findings, we identified three loss-of-function or damaging variants in FLT4, KDR, and IQGAP1 in ten independent families with TOF.

Conclusions: Loss-of-function variants in FLT4 and KDR contribute substantially to the genetic basis of TOF. Our findings support the hypothesis that dysregulated VEGF signaling contributes to the pathogenesis of TOF.



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ECHOCARDIOGRAPHIC PREDICTORS OF ELEVATED LEFT VENTRICULAR END DIASTOLIC PRESSURE IN ADOLESCENT AND ADULT PATIENTS WITH REPAIRED TETRALOGY OF FALLOT

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Background: Elevated left ventricular end diastolic pressure (LVEDP) is a risk factor for ventricular arrhythmias in patients with repaired tetralogy of Fallot (TOF). Recent, invasive hemodynamic data is often unavailable and therefore, reliable non-invasive assessment of left ventricular diastolic function is necessary. This retrospective study aims to identify echocardiographic measures associated with LVEDP >12 mmHg in patients with repaired TOF.

Methods: Patients age >13 years with repaired TOF who underwent a left heart catheterization at our institution between 2001-2017 were evaluated. Patients were included if they had an echocardiogram performed within 7 days prior to the catheterization. Demographic, medical history, cardiac catheterization, and echocardiographic data were collected on each patient. Echocardiographic assessment of diastolic function included mitral early filling velocity (E), late filling velocity (A), and tissue Doppler imaging (TDI) variables, such as e' and E/e'. Univariate comparison was made in echocardiographic and clinical variables between patients with LVEDP >12 vs. ≤12 mmHg. Optimal cutoffs of the echocardiographic variables to predict LVEDP >12 mmHg were identified using receiver operating characteristic (ROC) curves.

Results: This study included 94 patients (54% male) with a median age at catheterization of 24.6 years (IQR 17.5-43.0). Of these, 34 (36%) had an LVEDP >12 mmHg. Patients with LVEDP >12mmHg were older (median 32.9 vs. 24.0 years, p=0.02), more likely to have a history of an aortopulmonary shunt (62% vs. 38%, p=0.03), and have a diagnosis of hypertension (24% vs. 7%, p=0.03) compared to those with LVEDP ≤12 mmHg. There were no significant differences in mitral valve E/A ratio, septal or lateral e' velocity, or E/e' ratio between patients with LVEDP >12 vs. ≤12 mmHg. Patients with LVEDP >12mmHg had a larger left ventricular internal diameter at end diastole (LVIDd; mean 4.8 vs. 4.5 cm, p=0.02), larger left atrial area (mean 17.7 vs. 14.0 cm2, p=0.03), and a larger left atrium anterior-posterior diameter (mean 36.0 vs. 30.6, p=0.004). Optimal cutoffs associated with LVEDP > 12 mmHg were LVIDd ≥ 4.4 cm (area under the curve [AUC] 0.63; OR 2.84, 95% CI 1.11-8.01), left atrial area ≥ 17 cm2 (AUC 0.65; OR 5.03, 95% CI 1.64-17.1) and left atrium anterior-posterior diameter ≥ 27 mm (AUC 0.67; OR 15.4, 95% CI 2.93-285).

Conclusions: Typical echocardiographic measures of left ventricular diastolic dysfunction including mitral valve E/A, tissue Doppler e', and E/e' ratio were not associated with LVEDP >12 mmHg in TOF patients. The strongest predictors of LVEDP >12 mmHg were measurements of left atrium size. Therefore, assessing risk for ventricular arrhythmias related to elevated LVEDP using traditional echocardiographic measures of diastolic function may not be reliable. Prospective studies with the use of novel echocardiographic measures are needed.

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CHARACTERIZATION OF THE SURGICALLY MODIFIED RIGHT VENTRICULAR OUTFLOW TRACT USING MAGNETIC RESONANCE ANGIOGRAPHY IN ADULTS LATE AFTER TETRALOGY OF FALLOT REPAIR.

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Background: Surgical repair of tetralogy of Fallot (rTOF) typically involves extensive remodeling of the right ventricular (RV) outflow tract (RVOT) and pulmonary arteries (PA). To date, there has been a paucity of literature focused on delineation of RVOT-PA anatomy in rTOF, despite increasing interest in development of newer prosthetic pulmonary valves suitable for percutaneous implantation in a wider range of anatomies. Our aims were therefore to (1) characterize the surgically modified RVOT-PA anatomies using cardiovascular magnetic resonance imaging (CMR) and (2) explore potential associations between RVOT-PA findings and clinical characteristics.

Methods: Adults were identified from a prospective observational registry of CMR eligible adults with pulmonary regurgitation (PR) late after TOF repair. Subjects were included if available CMR included a contrast-enhanced MR angiogram (MRA) and were excluded if an RV-PA conduit or prosthetic pulmonary valve was implanted prior to CMR. Contemporary clinical data were reviewed (including electrocardiograms, echocardiograms and cardiopulmonary exercise studies). Cardiac chamber sizes, severity of PR and systolic function were calculated using CMR. Measurements of the RVOT-PA were made using the MRA dataset and morphologies were classified into 5 types.

Results: One hundred consecutively recruited adults (65% male) were identified. Median age at repair was 3 years (IQR 1-6) and was 31 years at CMR (IQR 21-41). Mean RV end-diastolic volume index (RVEDVi) was 168±47mL/m2, RV ejection fraction was 44±6% and pulmonary regurgitation (PR) fraction was 39±16%. RVOT-PA morphologies from most to least frequent were: convergent (45%), concave (17%), straight (15%), divergent (14%) and convex (9%). Mean RVOT-PA diameters were 32±6mm proximally and 29±7mm distally; RVOT-PA length was 42±9mm. Correlations were observed between indexed RVOT-PA diameters and RVEDVi ($r=0.487$, $p<0.0001$), RV end-sytolic volume index ($r=0.418$, $p<0.0001$), RV mass index ($r=0.496$, $p<0.0001$) and PR fraction ($r=0.402$, $p<0.0001$) but not with RVEF. Patients with a divergent morphology had the largest RVOT-PA average diameters and volumes; this morphology was associated with lower NYHA functional class ($p=0.028$) and worse ventilatory efficiency (VE/VCO2) during exercise ($p=0.009$).

Conclusion: Convergent RVOT-PA morphology was evident in nearly half of patients studied. Correlations were observed between RVOT-PA measurements and RV volumes, RV mass and PR severity. Divergent morphology was associated with decreased NYHA functional classification and worse ventilatory efficiency. Further study is required to evaluate the potential clinical implications of these findings

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PREMATURE MORTALITY IN A GENETIC SUBTYPE OF TETRALOGY OF FALLOT

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Background: The 22q11.2 deletion, associated with 22q11.2 deletion syndrome (22q11.2DS), is the underlying cause of tetralogy of Fallot (TOF) in a significant portion of patients with this congenital cardiac disease. Because data are limited on possible effects of genetic subtype on adult patients, we investigated the impact of a 22q11.2 deletion on mortality in an adult TOF population.

Methods: The cohort studied comprised 603 patients with TOF recruited as adults through specialty clinics at the Toronto General Hospital: 72 with and 531 without 22q11.2DS. We used available outcome data (up to 20 years post-recruitment) to evaluate the impact of a 22q11.2 deletion on all-cause mortality, and a regression model to account for other possible factors.

Results: The 22q11.2DS group was younger (mean age 33.2, SD 10.1, vs 43.3, SD 13.7, years; p<.0001) and had a higher proportion of individuals with pulmonary atresia (25.0% vs. 11.3%; p=0.0012) than the non-22q group. A greater proportion in the 22q11.2DS group had died (18.1% vs 6.8%, p<0.0013), on average 17 years younger (at median 37, range 19 to 58, years) than those in the non22q group (at 54, range 18 to 80, years, p=0.0098). The presence of a 22q11.2 deletion was a significant predictor of mortality in TOF (HR= 5.79, p<.0001), in a model that accounted for pulmonary atresia, sex, decade of birth, and age at ascertainment

Conclusion: The 22q11.2 deletion significantly contributes to premature mortality in adults with TOF, mediated only in part by innate cardiac complexity. The results suggest that genetic subtyping is a clinically relevant factor in predicting the late outcome of TOF.

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PROGRESSIVE RIGHT VENTRICULAR ENLARGEMENT DUE TO PULMONARY REGURGITATION: CLINICAL CHARACTERISTICS OF A "LOW RISK" GROUP

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Background: The optimal interval between serial cardiac magnetic resonance imaging (CMRI) scans for monitoring progressive right ventricular (RV) enlargement is unknown because of limited data. The purpose of this study was to: (1) Determine the annual change in RV volume on serial CMRI scans; (2) Identify the risk factors associated with rapid progression of RV enlargement (RP-RVE).

Methods: Retrospective study of adults with native PR and ≥2 CMRI scans at Mayo Clinic Rochester, 2000-2015. RP-RVE (rapidly progressing right ventricular enlargement) was defined as first upper quartile of annual increase in RV end-diastolic volume index (RVEDVi) for the cohort.

Results: Of the 63 patients (age 36±9 years) in the study, 43 (68%) had tetralogy of Fallot while 20 (32%) had valvular pulmonic stenosis. RV outflow tract interventions that resulted in PR were balloon pulmonary valvuloplasty (n=4, 7%), transannular patch repair (n=30, 58%), and non-transannular patch repair (n=18, 35%). Interval between baseline and second CMRI was 2 (1-4) years. In comparison to baseline CMRI, RVEDVi increased from 130 (109-141) ml/m2 to 135 (126-155) ml/m2 and median annual change in RVEDVi was 3.1 (1.7-5.9) ml/m2. Univariate risk factors for RP-RVE (annual increase in RVEDVi >6 ml/m2) were ≥moderate tricuspid regurgitation and RVEDVi >130 ml/m2. Among the 24 patients without these risk factors (low risk subgroup), RVEDVi increased by only 3 (0 - 7) ml/m2 over 7 (5 - 9) years.

Conclusion: Patients without risk these factors (RVEDVi >130 ml/m2 and ≥ moderate tricuspid regurgitation) represent a low risk subgroup of patient that may potentially require much less aggressive imaging follow-up

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COMPREHENSIVE HEMODYNAMIC ASSESSMENT WITH QUANTITATIVE 4D FLOW MRI POST REPAIR FOR TETRALOGY OF FALLOT

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Background: Repair for tetralogy of Fallot (rTOF) is usually undertaken in the first year of life. Advances in surgical techniques have led to improvement in long-term survival of patients. Cardiac MRI is routinely used to monitor patients post rTOF as they are known to develop pulmonary regurgitation (PR), residual/recurrent pulmonary stenosis and resultant right ventricular (RV) remodeling. Cardiac MRI derived RV volumes, RV ejection fraction and PR are being used to guide further management. 4D flow MRI has been emerging as a useful technique for evaluation of congenital heart disease (CHD). The purpose of our study was (a) To compare 4D with 2D flow MRI for PR assessment in pediatric patients post rTOF; (b) To compare 4D flow MRI derived pulmonary helicity and vorticity in rTOF with controls.

Methods: In this prospective study, whole heart 4D flow MRI was performed at 1.5T using a respiratory navigator-gated sequence in patients post rTOF (n=11, 64% female, 13.6±3.2 years) and controls (n=6, 50% female, 12.3±2.1 years). 2D cine phase contrast MRI and multi-planar cine steady-state free precession imaging was also performed. Controls were CHD patients with normal right ventricular outflow tracts (RVOT) but left-sided pathology such as bicuspid aortic valve and coarctation of aorta. Cardiac time-resolved flow, vorticity and helicity were calculated from 4D flow MRI over manually segmented regions comprising the RVOT, main, right and left pulmonary arteries (MPA, RPA, LPA). Statistical analysis included dependent or independent samples t-test as appropriate, Bland-Altman analysis and Pearson correlation.

Results: There was no significant difference between regurgitant fraction (RF) measured by 4D or 2D flow MRI in the MPA (37.9±11.4% vs. 36.3±11.0%, p=0.08), RPA (35.6±18.7% vs. 34.6±19.2%, p=0.80) or LPA (27.2±11.0% vs. 30.3±11.5%, p=0.17), with excellent correlation between techniques, $r \geq 0.94$, $p<0.01$. On Bland-Altman analysis, there was mild positive bias for 4D compared to 2D flow MRI for calculation of MPA RF with good precision (bias=1.53%, 95%CI [-2.88, 5.93]). There was no significant difference in average or peak helicity between rTOF and controls in MPA, RPA, and LPA. Compared to controls, average and peak vorticity were significantly higher in MPA and LPA, all $p<0.01$. There was strong correlation between average diastolic vorticity and MPA RF, $r=0.88$, $p<0.01$; moderate correlation between peak systolic RVOT vorticity and RV ejection fraction, $r=0.68$, $p=0.02$.

Conclusion: In patients post rTOF, there is no significant difference in PR calculation by 4D or 2D flow MRI. Patients post rTOF demonstrate elevated vorticity in the pulmonary artery related to systolic right ventricular function and PR.

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CATHETER AND SURGICAL INTERVENTIONS IN ADULT PATIENTS WITH REPAIRED TETRALOGY OF FALLOT

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Background: Tetralogy of Fallot (TF) is presently one of the most common conditions among adults with congenital heart disease (ACHD). Cardiac interventions late after primary repair have made possible longer life expectancy and better quality of life. OBJECTIVES Retrospective study to determine the prevalence, type and outcomes of surgical and percutaneous interventions performed in a population of ACHD patients with repaired TF.

Methods: Medical records of 2 specialized centers caring ACHD with surgical, hemodynamic and electrophysiological (EP) facilities were reviewed. All patients admitted in the study had repaired TF and were 16 years-old or older. The outcomes and type of procedures were analyzed. Follow-up time extended since the date of the initial repair to the last time seen.

Results: There were 183 patients with repaired TF at a median age of 3.0 years (range 1-32) and a median follow-up of 29 years (range 1-55). Pulmonary valve (PV) atresia and PV agenesia were found in 6 (3.3%) patients each. The median age was 34 years (range 16-68), 95 (51.9%) were female. Twenty nine women had had 49 full or near term pregnancies. Genetic syndromes were present in 18 (9.8%) patients. Four patients had infective endocarditis (IE). Seventy two (39.2%) patients required one or more interventions totaling 89 procedures during their follow-up. Open heart surgery for PV incompetence or stenosis, right ventricular outflow tract aneurysm, residual ventricular septal defect and miscellaneous causes was performed 49 times in 44 (24.0%) cases. Percutaneous catheter interventions to address pulmonary artery branch or conduit stenoses mostly with stents and for PV replacement were carried out 28 times in 21 (11.5%) patients. In one patient a patent foramen ovale was device closed after occurrence of paradoxical embolism to a retinal artery. Electro physiologic (EP) treatment and pacemaker or cardioverter defibrillator (CD) implantation were performed in 32 circumstances in 24 (13.1%) cases for radiofrequency ablation, symptomatic bradycardia and primary or secondary prevention of sudden cardiac death (SD). A CD was implanted in 14 (7.6%) patients with 1 aborted SD. There were no major adverse cardiac events and just one surgical death (2% mortality for surgical reintervention patients). In the whole cohort, including patients without reintervention, 6 (3.3%) deaths occurred at a mean age of 53 years, 4 cardiac related (congestive heart failure 2, IE 1, surgery 1).

Conclusions: Long term results for repaired TF depend not only of the center experience, patient load and surgeon's expertise but on the original heart morphology. For these reasons, residua and sequelae are not infrequently seen in the long term follow-up of these patients. Almost 40% of our sample required surgical, catheter intervention or an EP procedure for symptomatology or prevention from later deterioration. There was only one death in the group of surgical patients which were usually very challenging. MACE not present in the remaining patients. Taking into accounts these good results, these interventions should not be delayed whenever there is an indication.

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TETRALOGY OF FALLOT: LONG-TERM OUTCOME IN ADOLESCENT AND ADULTS AFTER SURGICAL REPAIR.

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Background: Adult survivors with repaired Tetralogy of Fallot (TOF) are a growing population. Many patients require reinterventions during follow-up.

Objective: We aimed to identify the incidence and characteristics of dyspnea and asthma-like symptoms before and after ASD repair.

Methods/Results: Methods and results: 182 patients (p) with repaired TOF were studied from 1988 to 2016, mean age 25.3years, median follow-up 20.2years. Right ventriculotomy was performed in 76p(42%) and transannular patch in 73p(40%). NYHA functional class was I: 112p, II: 57p and III: 13p. Pulmonary regurgitation (PR) was moderate: 56p and severe: 40p. Complete right bundle branch block with QRS duration 160-200ms was present in 52p and >200ms in 6p. Tachyarrhythmias were present in 27 patients (15%), non-sustained ventricular tachycardia (VT): 8p sustained VT: 7p, atrial fibrillation (AF) or flutter: 11p. VT + AF: 5p.Some haemodynamic substrate was present in 66% of them .Seventynine p (43%) underwent one or more re-interventions at a mean of 11.6 years from repair: It was surgery in 56p and transcatheter intervention in 23p. In 69%, the major cause of reoperation was PR. Ten p. developed infective endocarditis, 5 of them required surgery. There were 43 pregnancies in 16 women with 36 alive newborns. Overall survival at 10, 15, 25 and 35 years was: 99%, 98%, 94% and 82%, respectively. Mortality rate was 3.3%: 6p died at a mean age of 32.4 years, after a median time from surgery of 22.5 years. By univariate analysis, the mortality was associated with arrhythmia(p 0.001), QRS duration>160 mseg(p 0.001), severe PR (p 0.004) reoperations(p 0.05) and infective endocarditisp(0.001).

Conclusion: The long- term survival of repaired TOF is excellent, with good functional class in most patients. At a mean time of 11.6 years after repair, 43% of patients underwent one or more reinterventions. Tachyarrhythmias were present in 15% of patients, 66% of them with some hemodynamic substrate. Late mortality was 3.3%, associated to arrhythmia, severe PR, QRS duration and infective endocarditis.

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RISK FACTORS FOR PULMONIC VALVE REPLACEMENT IN POST-OPERATIVE TETRALOGY OF FALLOT.

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Background: Sudden cardiac death is a major cause of mortality in adults with congenital heart disease (ACHD). Despite extensive guidelines on implantable cardioverter-defibrillator (ICD) implantation in acquired heart disease, indications for ICD in ACHD remain unclear. We aim to assess indications and outcome of ICD implantation in ACHD in the current literature.

Methods: From 1997 to 2016, total 651 patients underwent TOF-repair at Faculty of Medicine Siriraj Hospital, Thailand. 403 patients (62%) were included. Follow up data as well as re-operation data were identified and analyzed.

Results: 134 patients (33%) underwent palliative shunt before total repair. At the time of total repair, the median age was 4.41 years (0.9-55.3) and median weight was 13.58 kgs (5.5-68). The median of pre-operative PV annulus z-score and McGoon ratio were -1.98 (-8.95, 2.3) and 2.02 (1, 4.8), respectively. The cardiac bypass and aortic cross clamp time were 110 (48, 396) and 69 (28, 218) min. 199 patients (49.4%) underwent total correction with transannular patch (TAP) technique. During the median follow-up period of 9.1 years, with a maximum of 21.1 years, 80 patients (19.8%) underwent postoperative re-catheterization. Transcatheter interventions were needed in 65 patients. In our institution, we sent the patients for cardiovascular magnetic resonance (CMR) only based on clinical symptoms, QRS duration on ECG and echocardiographic findings that showed the significant pulmonary regurgitation. Of 65 patients who were evaluated by CMR, the means of right ventricular (RV) end diastolic and systolic volume index were 169.8 ml/m2 and 93.0 ml/m2, respectively. The RV ejection fraction were 46.88 % with pulmonary regurgitation fraction of 44.9 %. Considering the recommendation for pulmonary valve replacement (PVR) in patients with repaired TOF proposed by Tal Geva, 41 patients reached the indications. The median time of indicated-PVR following total correction was 12.2 years (6.2-18.9). On multivariable analysis, the risk factors for PVR post-repaired TOF were the reconstruction with TAP [HR=3.61 (1.72, 7.55) p-value < -2.5; [HR=5.07 (2.33, 11.03), p-value= 0.001], postoperative PV z-scores of > 2 [HR=7.79 (1.51, 40.38), p-value= 0.014] and longer cardiac bypass time (p-value=0.035). Age, BW and palliative shunt before surgery did not affect the risk of PVR.

Conclusions: Time to re-operation with PVR was comparable to previous studies. The small native PV and the needed of TAP are the risk factors of PVR. If TAP is indicated, the appropriate post-operative PV z-score should be less than 2. However, more aggregated data for long term outcomes is warranted.

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LONG TERM OUTCOMES IN PATIENTS WITH TURNER SYNDROME: A 68-YEAR FOLLOW-UP

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Background/Objective: Turner syndrome (TS) is the most common sex chromosome abnormality in women, and is associated with increased morbidity and mortality. We describe long term outcomes in a large cohort of patients with TS.

Methods: Retrospective review of patients with TS followed at Mayo Clinic Rochester from 1950-2017 was performed. Clinical, imaging, surgical, and genetic data were collected and analyzed. Survival analysis was performed with Kaplan Meier method using age-matched Olmsted County residents as the reference group.

Results: The study cohort comprised 317 patients with TS. Average age at diagnosis was 9 (2-12) years, genetic testing was performed in 202 (64%), and monosomy X was present in 75 (37%). Congenital heart disease occurred in 131 (41%), with most frequent lesions being bicuspid aortic valve and coarctation of the aorta. Ascending aortic dilation was common, with mean aortic root size index 2 cm/m2, and aortic dissection occurred in 6 (2%) patients. The average follow-up was 11 (2-26) years yielding 3,898 patient-years, and during this period 46 (14%) patients died, with mean age at the time of death was 53±17 years. TS patients had reduced survival compared to the control group (82% vs 94% at 30 years, $p<0.001$), and the leading causes of death were cardiovascular disease, liver disease, and malignancy.

Conclusions: Patients with TS have reduced survival compared to age-matched controls, and cardiovascular disease is the major cause of death. Further studies are required to determine if targeted cardiovascular risk factor modification will result in improved survival in this population.

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CARDIOVASCULAR SURGERY IN PATIENTS WITH TURNER SYNDROME: THE MAYO CLINIC EXPERIENCE

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Background: Cardiovascular disease is the leading cause of death in patients with Turner syndrome (TS), and cardiovascular surgery is frequently required in the management of these patients. This study describes the long-term surgical outcomes of patients with TS.

Methods: Retrospective review of TS patients who had cardiovascular surgery at Mayo Clinic Rochester, 1977-2017. Clinical, imaging, and surgical data were analyzed. Survival analysis was performed with Kaplan Meier method.

Results: The study cohort comprised 51 TS patients, average age at the time of first surgery at Mayo Clinic was 28 (8 - 41) years, and 23 (45%) patients were under the age of 18. The most common procedures were coarctation of aorta repair in 14 (28%) patients, aortic valve replacement in 6 (12%) patients, and composite aortic valve/root/ascending aorta replacement in 7 (14%) patients, with 7 (14%) patients undergoing repair of more than one lesion. There were 5 surgical interventions for aortic dissection (3 type A and 2 type B), and 1 of the 5 patients (20%) died prior to hospital dismissal. Average hospital length of stay was 6 ± 2 days. There were 4 (8%) early surgical deaths and 47 (92%) patients survived to hospital dismissal. Freedom from death was 97% and 89% at 10 and 20 years, and the freedom from reoperation was 93% and 81% at 10 and 20 years.

Conclusions: Cardiovascular surgery was associated with 8% early mortality in TS patients, and the patients who survived to hospital dismissal had good long-term survival. The potential need for late reoperations underscores the importance of lifelong cardiovascular follow-up.

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TEN YEAR FOLLOW-UP OF AORTIC DILATATION IN PATIENTS WITH A SMAD3 MUTATION

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Background: Aneurysm-Osteoarthritis Syndrome (AOS) caused by a SMAD3 mutation has only been recently discovered as one of the more aggressive genetic syndromes. It is characterized by fast and unpredictable aneurysm development. Since it is a rare disease little data on follow-up of SMAD3 patients is available, so although still a small cohort, it is the largest cohort with the longest follow-up. The objective of this study is to evaluate the progression rate of aortic dilatation and to describe cardiovascular events in patients with AOS caused by a SMAD3 mutation in a patient cohort.

Methods: Methods All patients were monitored according to our clinical AOS protocol, including a yearly ECG-gated full-body CT scan. Patients with at least two CT scans between December 2003 and January 2018 were included in this study. Two observers, blinded to each other, measured thoracic and abdominal aortic diameters at eight different levels. Evaluation was done by multi planar reconstruction; vessel diameters were measured perpendicular to the aorta. To account for the multiple measurements within each patient, average time trends were estimated with Linear Mixed models. These models use both fixed effects and random effects for intercept and slope (time trend). This allows us to model an overall (population) intercept and slope, and deviations for each patient with respect to the intercept and the slope. For measurements on ANN, SIN, STJ, and ASC only observations before the operation were included in the models. The remaining measurements utilize all observations. Graphical representations of the data are provided by individual profile plots. These plots connect all observations within the same patient with a line. The measurements after the operation are left out.

Results: Among the 28 SMAD3 patients seen in our center, aged between 24 and 73 (47±15.3 years), no mortality occurred during the median follow-up duration of 10 years (range: 3-14years). However, 14 (50%) patients needed elective valve sparing root replacement (VSRR), their median age at time of operation was 40.6 years (IQR: 22.76). Six of these patients had only one pre-operative scan. In the other eight operated patients, at least two pre-operative scans were available, their mean time from inclusion to operation was 3.4 years (±2.7years). When considering all patients, all levels show an increase of aortic diameter (figure 1A). In addition to the 14 VSSRs an additional 14 vascular events other than the fourteen VSSR occurred, most often coiling of an aneurysm. These fourteen procedures were conducted in 9 patients, 5 of which also had a VSSR. The mean age at the time of the first arterial event was 42 years (±12.6 years), three patients experienced two or more arterial events. When these events were taken into account 18 patients (64%) experienced a cardiovascular event during the duration of this study (figure 1B).

Conclusions: Growth occurs at all levels of the aorta in this patient group, leading to fourteen operations and an equal number of vascular interventions. Aneurysm growth in AOS patients can be fast and unpredictable, warranting extensive and frequent cardiovascular monitoring.

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PREGNANCY OUTCOMES IN WOMEN WITH CONGENITAL HEART DISEASE: RESULTS FROM THE COLORADO ADOLESCENT AND ADULT CHD SURVEILLANCE SYSTEM (COCHD)

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Background: Advances in the diagnosis and treatment of congenital heart disease (CHD) have led to a growing population of adults with CHD; a majority of women being childbearing age. The normal hemodynamic changes during pregnancy can trigger cardiac dysfunction in women with CHD, leading to an increased risk of adverse cardiac events. Additionally, utero-placental flow is impaired in women with CHD which may result in an increased risk of adverse offspring outcomes. Therefore, the aim of our study was to examine if severity of maternal CHD is associated with adverse maternal and offspring events.

Methods: Using data from the state-wide CHD surveillance system in adolescents and adults in Colorado, 926 pregnant women with CHD were identified that resulted in 1031 live births (including twins and successive births) between 2011 and 2013. Maternal CHD type was dichotomized as severe (n=149) and moderate/minor (total n=882) based on presence of a CHD ICD-9 diagnosis code in the mothers electronic health record (EHR). Adverse maternal cardiac events were obtained from the EHR and adverse offspring events were obtained from the infant's birth certificate. The risk of adverse maternal and offspring events were calculated out of the total number of pregnancies in each severity category.

Results: Primary maternal cardiac events include cardiovascular mortality, arrhythmia, heart failure, thromboembolic events (pulmonary embolism, valve thrombosis or deep venous thrombosis), vascular events (stroke, myocardial infarction or dissection) and endocarditis, which occurred in 24% and 9% of women with severe and moderate/minor CHD, respectively. Premature birth (delivery <37 weeks) and/or low birth weight (<2,500 grams) occurred in 31% of the women with severe CHD and 21% of women with moderate/minor CHD. The risk of offspring congenital anomalies was higher among women with severe compared to moderate/minor CHD (40.5 vs. 14.8 per 1000 live births, respectively). The most common congenital anomalies among infants born to mothers with severe CHD were circulatory/respiratory (15%), musculoskeletal/integumentary (4%), cyanotic congenital heart disease (4%) and Down syndrome (3%). The most common congenital anomalies among infants born to mothers with moderate/mild CHD were circulatory/respiratory (6%), urogenital anomalies (1%), and Down syndrome (0.8%).

Conclusion: Population-level surveillance of CHD in Colorado provides novel assessment of the substantial risk of maternal and neonatal adverse events associated with pregnancy in women with CHD.

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PREGNANCY OUTCOMES IN WOMEN WITH TETRALOGY OF FALLOT. DATA FROM THE EUROPEAN SOCIETY OF CARDIOLOGY REGISTRY OF PREGNANCY AND CARDIAC DISEASE (ROPAC)

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Background: Pregnancy outcomes in women with Tetralogy of Fallot (ToF) are not intensively investigated.

Methods: Within the international, prospective, observational Registry On Pregnancy And Cardiac disease (ROPAC), we describe cardiac, obstetric and fetal outcomes of pregnancy within patients with ToF and identify predictors of adverse cardiac outcome.

Results: In the 240 included ToF patients (mean age 28.7 +/- 5.1 years) no maternal mortality occurred during or directly after pregnancy. In 18 pregnancies (8%), at least one adverse cardiac event occurred, of which heart failure was the most common complication (n = 11, 5%). Ventricular tachyarrhythmias complicated 7 pregnancies (3%) and supraventricular tachyarrhythmias occurred in 2% (n = 5). One patient (0.5%) suffered from thrombosis of a valvular prosthesis. Seven patients (3%) had not undergone complete correction of ToF, of which 4 patients (2%) had only undergone either a surgical correction of the VSD or a surgical valve replacement. At least one obstetric adverse event occurred in 10 patients (4%), with postpartum hemorrhage in 4 patients (2%) and pre-eclampsia in 3 patients (1%). There were 3 miscarriages in the total cohort (1%) and 4 patients suffered from pregnancy-induced hypertensive disorder (2%). Fetal adverse events occurred in 40 patients (17%), with 3 cases of late fetal mortality (1%). Preterm birth (<37 weeks) and low apgar score occurred in 34 patients (14%) and 9 patients (4%) respectively. In univariable analysis, medication use and pulmonary insufficiency were predictors of fetal complications.

Conclusions: Most women with ToF tolerate pregnancy well, and can safely embark on pregnancy. However fetal complications were not uncommon, with medication use and pulmonary insufficiency as predictors of adverse fetal outcome.

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CARDIAC OUTPUT ASSESSMENT DURING AND AFTER PREGNANCY IN WOMEN WITH HEART DISEASE AS COMPARED WITH NORMAL CONTROLS USING CARDIOVASCULAR MAGNETIC RESONANCE IMAGING (MOMS HEART STUDY)

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Background: Mothers with heart disease are at increased risk of pregnancy-related complications. Insufficient cardiac output (CO)/cardiac index (CI) in maternal cardiac disease has been implicated in poor maternal/fetal outcomes. Phase contrast flow analysis using cardiovascular magnetic resonance imaging (CMR) is the reference standard for non-invasive measurement of CO/CI. We devised a pilot study to evaluate the feasibility of antepartum measurement of CO/CI with comparison to postpartum measurements in women with heart disease and matched controls. Hypothesis: Antepartum assessment of CO/CI using CMR will be feasible and will highlight differences in flows in women with heart disease as compared with controls.

Methods: Pregnant women with moderate or severe valvular/ventricular disease were matched with healthy controls. Participants were scanned using a 1.5T Siemens scanner during the third trimester of pregnancy (corresponding to peak CO) and six months postpartum (surrogate for baseline). Phase contrast CMR was used to quantify CO/CI (summation of superior vena cava and descending aorta flows). Calculation of CI in pregnancy was achieved using pre-pregnancy body surface area for both groups. Comparison of groups was achieved using the paired Student's t-test.

Results: Twelve women with heart disease (mean age 34±4 years) and 12 controls (mean age 33±4 years) were included. Heart disease included left ventricular (LV) systolic dysfunction n=4 (LV ejection fraction [LVEF] 40-50% n=2 and LVEF <40% n=2), systemic right ventricle n=1, ≥moderate/severe aortic regurgitation n=4, ≥moderate mitral regurgitation n=1, severe tricuspid regurgitation n=1, and ≥moderate pulmonic regurgitation n=1. Cardiovascular medications (beta-blocker/diuretics) were used in n=10. Cardiovascular complications in the women with heart disease included new-onset arrhythmia n=2 and heart failure n=2. The CO/CI values are shown. There was no statistical difference in CO/CI in the antepartum period of women with heart disease versus controls (p=0.686 for CO and p=0.520 for CI). The magnitude of adaptive change in CO (difference between ante-partum and postpartum) was greater in women with heart disease versus controls (p=0.04) although CI was not significantly different. No fetal or neonatal complications occurred in either group.

Conclusion: Antepartum CMR hemodynamic assessment of CO/CI is feasible in women with cardiac disease. Despite significant ventricular and valvular dysfunction, the magnitude of adaptive change in CO is larger in women with heart disease as compared with controls. Further study of a larger population of women with a wider range of cardiac lesions may provide greater insights into adaptations to pregnancy in women with heart disease.

Table 1: CMR evaluation of CO/CI by phase contrast imaging in pregnancy and postpartum for women with heart disease and controls.

	Antepartum		Postpartum		Delta antepartum-postpartum	
	CO (+/- SD)	CI (+/- SD)	CO (+/- SD)	CI (+/- SD)	CO (+/- SD)	CI (+/- SD)
Women with heart disease	7.3 (+/-1.1)	4 (+/-0.6)	4.9 (+/-1.4)	2.6 (+/-0.6)	2.4 (+/-1.2)*	1.4 (+/-0.8)**
Controls	7.0 (+/-1.5)	4.2 (+/-0.8)	5.6 (+/-1.2)	3.3 (+/-0.5)	1.4 (+/-0.9)	0.8 (+/-0.6)

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PROGNOSTIC VALUE OF ECHOCARDIOGRAPHIC AND BLOOD BIOMARKERS IN PATIENTS WITH A SYSTEMIC RIGHT VENTRICLE

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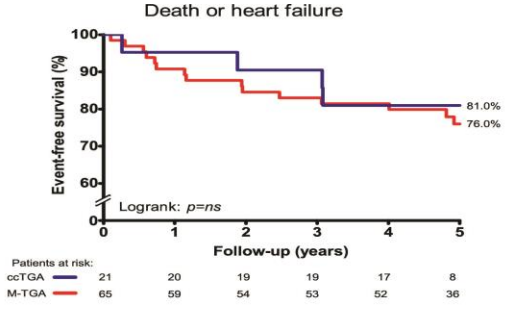
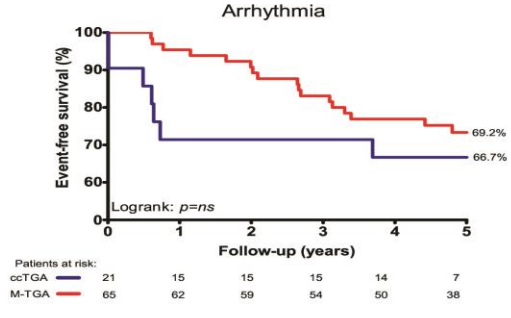
Background: Currently used prognostic factors struggle to identify patients with a systemic right ventricle(sRV) who are at risk for late cardiac complications. This study aims to identify prognostic factors, derived from echocardiographic and blood biomarkers, that are associated with cardiovascular events.

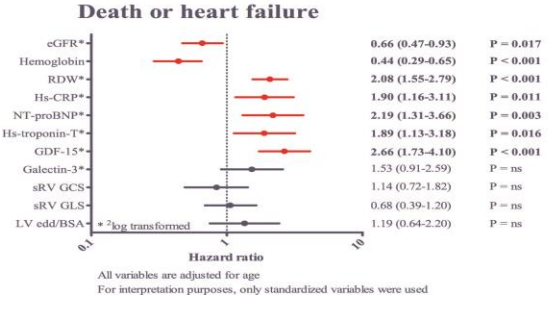
Methods: Prospectively recruited clinically stable patients with congenitally corrected transposition of the great arteries(ccTGA) and patients who underwent an atrial switch operation(M-TGA), were included between 2011-2013. All patients underwent venous blood sampling and echocardiography. The primary endpoint was a composite of death and heart failure, the secondary endpoint was arrhythmia. Associations between variables and endpoints were assessed with Cox-regression analysis (standardized variables were used for interpretation purposes), event-free survival with Kaplan-Meier curves.

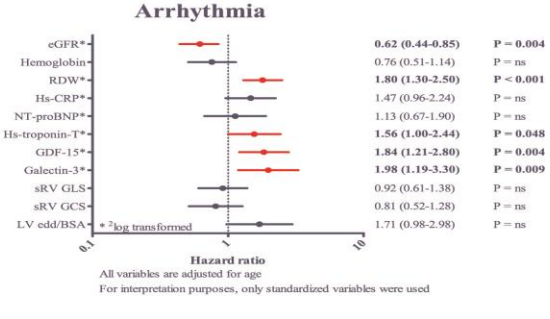
Results: Sixty-five patients with M-TGA and 21 ccTGA patients were included(age: 37±9 years, 65% male). both groups were equal regarding age and sex, except the loss of sinus rhythm, less in the M-TGA group(77% vs 43%, p-value: 0.004). Median follow-up time was 5.3[4.8-5.7] years. The primary and secondary endpoint occurred 21 and 26 times, respectively. For both the primary and secondary endpoint, event -free survival did not differ between ccTGA and M-TGA patients. Cox-regression analysis, adjusted for age, showed that al biomarkers were significantly associated with death and heart failure. However none of the echocardiographic markers, including strain analysis, were associated with the primary endpoint. Regarding the secondary endpoint, arrhythmia, several blood biomarkers were significantly associated, but hemoglobin, hsCRP and NT-proBNP were not. None of the echocardiographic parameters were significantly associated with both endpoints.

Conclusions: Echocardiographic parameters were not able to identify patients at risk for death and heart failure, or arrhythmia, whereas blood biomarkers from several pathophysiological axes were. Also of note, there was no difference in death, mortality or arrhythmia between patients with ccTGA and M-TGA.









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SYSTEMIC RIGHT VENTRICULAR FUNCTION: TEMPORAL TRENDS AND RISK FOR EVENTS

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Background: In patients with congenitally corrected transposition of the great arteries (ccTGA) and transposition of the arteries after Mustard/Senning repair (TGA), systemic right ventricular (SRV) dysfunction is a major complication. Current guidelines recommend annual follow-up preferably with imaging of the SRV. Data regarding the natural course of SRV function throughout life as assessed with gold-standard cardiovascular magnetic resonance (CMR) imaging are lacking. Purpose: We aimed to evaluate temporal trends in SRV ejection fraction (EF) and determine its impact on clinical outcome.

Method: This multicenter study included adult TGA (n=42) and ccTGA (N=16) patients from a prospective trial (n=88) between 2006 and 2009 and from whom serial CMR data were available. Primary endpoint was time-to-first-event, defined as arrhythmia, heart failure, tricuspid valve surgery, heart transplantation or death. We assessed serial CMR imaging using linear mixed modelling and performed regression analysis for risk of events.

Result: Sixty-eight patients (65% male; 28% ccTGA, age 32±8 years) were included. Median duration between first and last CMR imaging was 4.3 (interquartile range 3.0-7.3) years. There was no significant change in SRVEF; mean decline was 0.18% (95%CI -0.41% to +0.06%) per year (Figure). The individual course varied considerably, but males seemed to have a faster decline than females. No other relevant predictors of faster decline were found. Forty-one (60%) patients experienced events (arrhythmia n=38, heart failure n=15, tricuspid valve surgery n=6, death n=6). The probability of events increased with advancing age (Figure). Older patients with reduced EF experienced more episodes of heart failure. However, young patients with normal EF appeared also at high risk for particularly arrhythmias.

Conclusion: In ccTGA and TGA patients, systemic right ventricular EF remains quite stable over time. Risk for events increased with advancing age. Yet, normal EF at younger age was also associated with significant morbidity. Thus, regular follow-up is required in all patients with SRV. Figure. Relation between age versus systemic right ventricular ejection fraction and age versus probability of events.

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THROMBOEMBOLISM AND BLEEDING IN ADULTS WITH CONGENITAL HEART DISEASE USING NON-VITAMIN K ANTAGONIST ORAL ANTICOAGULANTS (NOACS) FOR THROMBOEMBOLIC PREVENTION: A PROSPECTIVE WORLDWIDE OBSERVATIONAL STUDY

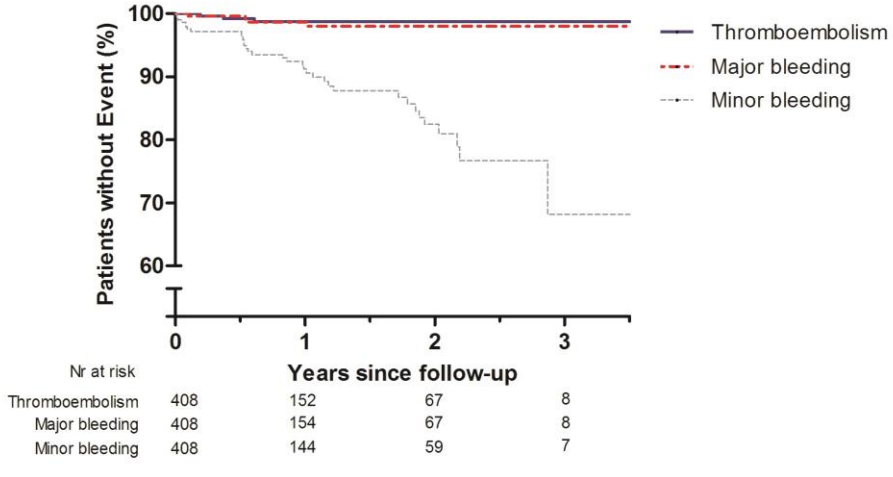
Hayang Yang; Josephine Heidendael; Thelma Konings; Gertjan Sieswerda; Folkert Meijboom; Markus Post; Arie van Dijk; Werner Budyts; Marielle Morissens; Tobias Rutz; Judith Bouchardy; Matthias Greutmann; Giancarlo Scognamiglio; Mikael Dellborg; Christina Christersson; Thomas Kronvall; Lina Gumbiene; Paul Khairy; Jamil Aboulhosn; Gruschen Veldtman; Craig Broberg; Alexander Opotowsky; Shane Tsai; Tabitha Moe; Koichiro Niwa; Alexander Egbe; Jonathan Buber; Rafel Alonso-Gonzalez; W. Davidson; H. Singh; M. Lipczynska; S. Chakrabarti; Berto Bouma; Barbara Mulder; *Academic Medical Center; Amsterdam, NL*

Background: Adults with congenital heart disease (ACHD) and atrial arrhythmias (AA) have high annual incidence of bleeding (4.4%) under the use of vitamin K antagonists (VKA). Non-vitamin K antagonist oral anticoagulants (NOACs) may be attractive alternatives, however, data on safety are lacking and adherence is of great concern due to lack of monitoring. The NOTE registry was designed to evaluate safety and adherence of NOACs in ACHD patients.

Methods: This is an international multicenter prospective study of ACHD using NOACs. Follow-up took place at 6 months and/or yearly thereafter. Primary endpoints were thromboembolic events and major bleeding. Secondary endpoints were minor bleeding and sufficient adherence, measured with pharmacy interrogation (≥80% medication refill rate) and Morisky-8 questionnaire (≥6 out of 8).

Results: In total, 408 ACHD patients(mean age 45±16 years;55% male;46% complex;43% moderate) using NOACs(34% apixaban;6% edoxaban;11% dabigatran;49% rivaroxaban) were included. Indications of NOACs were mostly AA (88%). During 1.3±0.8years, three thromboembolic events (annual incidence 0.8% [95%CI 0.2-2.2]), all in complex defects (mean CHA_2DS_2-VASc 2 [range 0-3]) and three major bleedings (annual incidence 0.8% [95%CI 0.2-2.1]) in moderate/complex defects (mean HASBLED 0.6 [range0-1]) occurred. Annual incidence of minor bleeding was 10.1% [95%CI 7.1-13.8] (n=35), mostly occurring in complex defects (mean HASBLED 0.4[range 0-3]). Adherence was sufficient at 1- and 2-year follow-up in 95% (n=41) and 93% (n=28) of patients by pharmacy interrogation and in 80% (n=69) and 91% (n=49) of patients by Morisky-8.

Conclusions: Annual incidence of thromboembolism and major bleeding were lower under NOACs than previously reported in ACHD using VKA. Adherence to NOACs was good and similar to previously reported VKA adherence. Risk of thromboembolism and bleeding seemed to be mostly related to defect severity.



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LIVER EXTRACELLULAR VOLUME FRACTION (ECV) IS ELEVATED IN PATIENTS WITH A FONTAN PALLIATION: A CONVENIENT MARKER OF HEPATIC FIBROSIS OBTAINED DURING CARDIAC SCANNING

Lidija McGrath, Ryan Van Workom; Stephen Heitner; Craig Broberg; *OHSU, Oregon, USA*

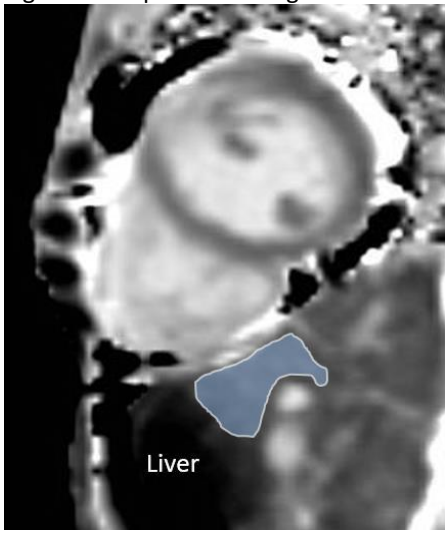
Background: Extracellular volume (ECV) fraction, a marker of tissue remodeling due to excessive collagen deposition, can be calculated using contrast-enhanced T1 mapping with cardiac magnetic resonance. Liver ECV can be measured on cardiac studies without requiring additional imaging. We hypothesized that patients with a Fontan palliation have higher liver ECV, reflecting a higher degree of liver fibrosis or congestion.

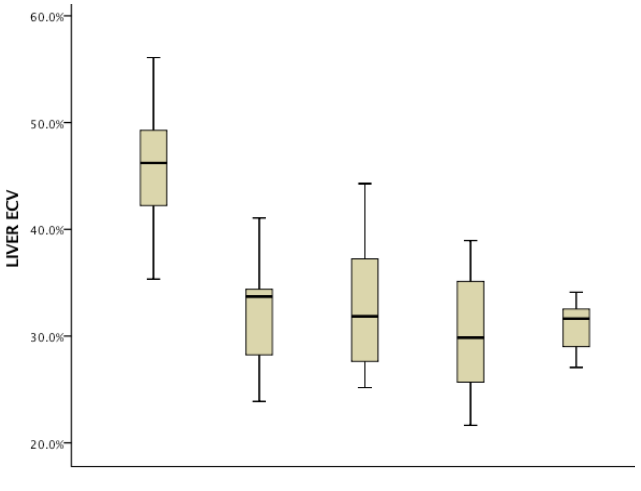
Methods: Using a 3T cardiac scanner (Siemens Tim Trio), adult Fontan patients prospectively underwent cardiac MRI including T1 mapping of the myocardium in a single short axis plane. These images also contain a section of the left lobe of the liver. Thus, a region of interest of the liver, excluding blood vessels, was contoured to quantify T1 before and 3 minutes, 7 minutes, and 15 minutes after gadolinium administration. ECV fraction was calculated using T1 values and hematocrit values collected at the time of the MRI. Cardiac catheterization was performed in all patients following the MRI. For comparison, liver ECV was also calculated using identical methods in patients with cancer (n=8), hypertrophic cardiomyopathy (HCM; n=12), tetralogy of Fallot (ToF; n=14), and healthy control subjects (n=11) who had undergone similar cardiac MRI at either 3T or 1.5T.

Methods: A total of 17 Fontan patients participated (mean age 28.5±7.4 years, 7 [41%] female). The mean liver ECV fraction for Fontan patients was 45.9±6.4%. Using one-way ANOVA, the mean liver ECV was significantly higher ($p < 0.001$) in Fontan patients compared to all other patient groups (31.5±3.3% for healthy control subjects, 32.2±5.3% for cancer patients, 32.7±6.0% for HCM patients, and 30.7±5.5% for ToF patients, FIGURE). By Bonferroni post-hoc analysis, Fontan liver ECV was significantly higher than all other groups, whereas there was no statistically significant difference in mean liver ECV between the other patients groups. Fontan patients were younger than the other clinical groups studied. Amongst Fontan patients, hepatic ECV correlated with cardiac ECV ($R=0.542$, $p=0.025$), but was not associated with age, PA pressure, or ventricular end-diastolic pressure.

Conclusions: Calculated liver ECV using contrast-enhanced T1 mapping obtained for cardiac ECV measurement is convenient to obtain and significantly higher in patients with a Fontan palliation as compared to controls without liver disease. This is consistent with a higher degree of hepatic fibrosis and/or congestion. Liver ECV may be an alternative method of fibrosis detection, though further studies are needed to determine its clinical utility in liver health surveillance.

Figure: Example of liver region interest on a mid-ventricle short axis image (left). Comparison of liver ECV by group (right)





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THROMBO-PROPHYLAXIS IN ADULTS WITH ATRIO-PULMONARY FONTAN

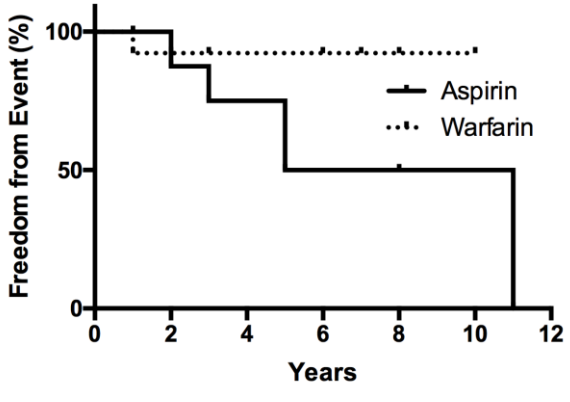
Adam Small; Jamil Aboulhosn; Gentian Lluri; *Ahmanson/UCLA Adult Congenital Heart Disease Center, California, USA*

Background: Although aspirin has been compared to warfarin for thromboembolic prophylaxis in the general Fontan population, little is known about the optimal preventative strategy for the atrio-pulmonary (RA-PA) Fontan particularly. We hypothesized that given the severity of right atrial dilation and consequent blood stasis in the RA-PA Fontan, warfarin would be associated with fewer thromboembolic events than aspirin.

Methods: : A retrospective cohort study of adult patients included in the Ahmanson/UCLA Adult Congenital Heart Disease Center database between 2004 and 2017 was performed. Patients with history of RA-PA Fontan operation and use of either aspirin or warfarin as most recent primary prophylaxis against thromboembolism were included in the final analysis. Patients with a history of thromboembolism that were not taking aspirin or warfarin prophylaxis prior to the event were excluded, as were patients who were taking aspirin or warfarin for secondary prevention. Primary outcome was incident thromboembolism, defined as space-occupying lesion on imaging consistent with thrombus within the Fontan or pulmonary arterial circuit. Secondary outcomes were death, transplantation, Fontan conversion, and bleeding requiring either transfusion or invasive intervention. Follow-up was terminated upon achievement of a primary outcome or achievement of a secondary outcome other than bleeding. Kaplan-Meier analysis of freedom from thrombosis was performed.

Results: One hundred and twenty-four adult patients with Fontan operations were identified in the database, of whom 52 had RA-PA Fontan (42%). Among those with RA-PA Fontan, all had documented atrial tachyarrhythmia at some point (19 with chronic atrial arrhythmia requiring invasive intervention) and none had known history of thromboembolism prior to receiving their care at the study center. Twenty-six patients met inclusion criteria by using aspirin or warfarin monotherapy as most recent prophylaxis. Among the excluded patients, prophylaxis varied. Five received other regimens: one received aspirin plus warfarin, one received apixaban, one received dabigatran, and two received clopidogrel. Two additional patients had prophylaxis stopped, one for lymphoma and one out of concern for medication interactions. None of the excluded patients achieved the primary outcome. Six additional patients had inadequate follow-up and one more had incomplete medical records. Of the 26 included in the final analysis, thirteen received aspirin as most recent primary prophylaxis (50%) and 13 received warfarin (50%). Tricuspid atresia was the most common underlying diagnosis (42%), followed by double-inlet left ventricle (38%). Median age at Fontan operation was 8.2 years; median age at prophylaxis initiation was 25.9 years. After six years from initiation of prophylaxis, the aspirin group had 50 +/- 35% freedom from thrombosis, the warfarin group 92 +/- 8% (p = 0.15). Incidences of secondary outcomes were not significantly different between the groups.

Conclusions: In this cohort of long-term Fontan survivors with RA-PA Fontan, the risk of thromboembolic complications is high, especially in those taking aspirin rather than warfarin. Larger studies are needed to confirm these findings.



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SHOULD THE NORMAL RANGE OF PULMONARY VASCULARE RESISTANCE BE RE-DEFINED IN PATIENTS WITH FONTAN CIRCULATION?

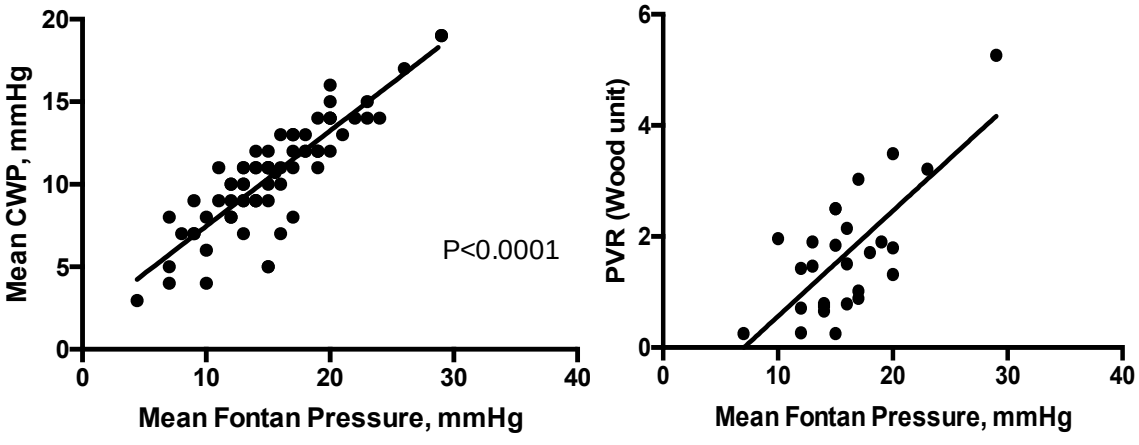
Ashish Shah; Kelly Rohan; Graeme Kirkwood; Vaikom Mahadevan; Ashley Stokes; Ozhin Brigante; James Carmichael; Guy Kendall; Raveenjot Nagra; Tasnime Yearoo; Mehul Patel; Bawan Hama; Amaran Gill; Arjamand Shauq; Andreas Hoschtitzky; Petra Jenkins; Jaspal Dua; Purvi Shah; Pradeepkumar Charla; Heiko Schneider; *St Boniface Hospital, University of Manitob, Winnipeg, Canada*

Background: The Fontan operation provides a palliative cure to those born with anatomical or physiologically single ventricle. The long-term outcomes are poor and survivors are at a high risk of constellation of medical problems, described as "Fontan failure". Adult patients with Fontan circulation have limited exercise capacity, mainly due to inability to augment the cardiac output, predominantly secondary to limited venous return. Studies have suggested that pulmonary vasodilator therapy results in marked improvement in hemodynamics. Normal range of pulmonary vascular resistance (PVR) is defined in those with bi-ventricular physiology, and pulsatile pulmonary flow. We evaluated observed PVR in a cohort of patients with Fontan circulation from a tertiary adult congenital heart disease centre.

Method: We retrospectively reviewed data from a large tertiary adult congenital heart disease centres in UK. Clinical and procedural details were obtained by reviewing electronic charts, with special emphasis on haemodynamic data.

Results: From a cohort of 4454 patients with complex ACHD conditions, 154 had Fontan circulation; of whom 70 patients with failing Fontan were investigated by cardiac catheterization. Thirty-four (48.6%) were male, mean age of 30.1±6.2 years (17-43), and mean body mass index of 24.1±5.3 kg/m^2 (16.6-47.7). Mean Fontan pressure was 16±4 mmHg (7-29), mean wedge capillary pressure was 11±3 mmHg (4-19), and mean trans-pulmonary gradient (TPG) was 5±3 mmHg (0-15). Mean cardiac output was 4.1±2.7 L/min and calculated PVR was 1.7±1.2 Wood units (0.25-5.3). Although cardiac output was well maintained, Fontan pressure was significantly correlated with capillary wedge pressure (P<0.0001) and PVR (P<0.0001). Rise in Fontan pressure was observed earlier than increase in PVR; however, 3/4th of patients with failing Fontan were noted to have normal PVR.

Conclusion: In this large series of patients with Fontan circulation, rise in PVR above the normally accepted range was observed only after moderate rise in Fontan pressure. In patients without sub-pulmonic ventricle and passively filling pulmonary circulation, range of normally accepted PVR should be re-defined, as patients can be treated in a timely fashion.



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MID-TERM OUTCOME AFTER FONTAN CONVERSION COMPARED WITH PRIMARY TOTAL CAVOPULMONARY CONNECTION.

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Background: The indication criteria of Fontan conversion from atriopulmonary connection (APC) to total cavopulmonary connection (TCPC) is not well defined, especially for patients without any late Fontan complications. In our hospital, Fontan conversion has been somehow prophylactically offered to patients after APC. The aim of this study was to evaluate mid-term outcome of adult patients after Fontan conversion compared with those after primary TCPC.

Methods: This is a single-centre retrospective study including Fontan patients who underwent cardiac catheterization at > 18 years of age between July 2005 and February 2018. They were divided into two groups according to the type of first Fontan surgery (APC group and TCPC group). Fontan conversion was undertaken in all of APC group but two cases who were excluded. Two groups were compared in terms of catheterization data, arrhythmias and late complications.

Results: There were 26 cases in APC group and 15 cases in TCPC group. The mean age at Fontan surgery was 6.6 ± 3.7 years in APC group and 13.5 ± 6.5 years in TCPC group ($p<0.01$). Patients in APC group underwent Fontan conversion at the mean age of 22.2 ± 5.3 years, 15.6 ± 3.65 years after first Fontan surgery. Out of 26 cases, 10 cases (38%) had atrial tachyarrhythmia before Fontan conversion. Antiarrhythmic surgery was added in 25 cases, where 6 cases (24%) developed sinus node dysfunction immediately after the procedure. Four of them required permanent pacemaker implant. Cardiac catheterization was performed at the mean age of 26.4 ± 6.0 years in APC group and 28.6 ± 4.8 years in TCPC group (n.s.), 4.2 ± 2.0 years and 15.2 ± 3.6 years after conversion or TCPC ($p<0.01$), respectively. The mean SVC pressure and ventricular end-diastolic pressure (EDP) was 13 ± 3 mmHg, 12 ± 3 mmHg in APC group and 15 ± 6 mmHg, 14 ± 6 mmHg in TCPC group (both n.s.). The median BNP at the time of catheterization was 13.7 pg/μL (range 5 - 59.2) in APC group and 30.2 pg/μL (range 6.2 - 323.0) in TCPC group (n.s.). The mean age at the last follow-up was 28.9 ± 5.9 years in APC group and 30.8 ± 5.1 years in TCPC group. NYHA class was I in 22 cases (85%) in APC group and 13 cases (80%) in TCPC group. There was no death or heart transplant in both groups. Recurrence of tachyarrhythmia was seen in a case in APC group immediately after surgery. Two patients in TCPC group newly developed atrial flutter who were complicated with either severe aortic stenosis or atrioventricular regurgitation, requiring surgical valve replacement.

Conclusion: After Fontan conversion with antiarrhythmic surgery, atrial tachyarrhythmia seemed to be well controlled in APC patients, though a few cases were complicated with sinus node dysfunction necessitating pacemaker implant. Hemodynamics and ADL were similar between APC after conversion and TCPC in their early 30s.

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FIBROSURE AND ELASTOGRAPHY POORLY PREDICT THE SEVERITY OF LIVER FIBROSIS IN FONTAN ASSOCIATED LIVER DISEASE

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Background: Hepatic fibrosis is common in Adults born with Single Ventricle Physiology (SVP) corrected with a Fontan circulation. Non-invasive methods, FibroSURE and Elastography (TE or SWE), have been studied to predict hepatic fibrosis although the benefits remain undefined. A liver biopsy is the gold standard, albeit an imperfect test. Friedrich-Rust et al (2008) suggested FibroSURE and Elastography may be useful in children. Kutty et al (2013) demonstrated SWE correlated with liver fibrosis in children and adults. Wu et al (2017) demonstrated FibroSURE did not correlate with liver fibrosis in adults. Our aim was to determine if FibroSURE, SWE, hepatic Duplex sonography alone or in combination predicts the presence or severity of liver fibrosis on biopsy in adults with SVP.

Methods: 14 consecutive adults undergoing routine evaluation for liver disease agreed to participate. FibroSURE (LabCorp), duplex US followed by SWE (GE-LOGIQ E9) and a liver biopsy were obtained as well as routine labs. The radiologist was blinded to clinical and liver biopsy results. Congestive Hepatic Fibrosis score (Surrey et al 2016) was determined by a single hepatopathologist blinded to all data. Finally, we analyzed our results comparing none/mild fibrosis (F0-2) vs advanced fibrosis (F3-4).

Results: 5 females/9 males had a mean age± SD of 26.4±7.5 (range 19-43yo); mean years since surgery to liver biopsy 24.7±6.2; AHA class 1 in 12/14, two AHA class 2. Seven subjects had a history of an arrhythmia. FibroSURE tests were obtained 3.14±1.75 months and SWE 8.23±4.61 weeks prior to liver biopsy. All liver biopsy samples were considered adequate (length 25±8mm, portal tracts 14±8). 100% of subjects had liver fibrosis, 2 (F1), 7 (F2), and 5 (F3-4). FibroSURE staging agreed with liver biopsies in only 5 cases (35.7%), underestimated in 7 and overestimated in 2 cases (kappa = 0.016). SWE agreed with liver fibrosis staging in 0/14 cases, underestimated 4 cases and overestimated 10 cases (kappa = -0.21). Comparing F0-2 to F3-4, FibroSURE, SWE, and Duplex measurements were all statistically insignificant (Table).

Conclusion: FibroSURE, Elastography and duplex US did not accurately predict the presence or degree of liver fibrosis. Future studies are warranted to identify novel liver fibrosis biomarkers in adults with SVP.

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PREVALENCE OF MENTAL ILLNESS IN ADOLESCENTS AND ADULTS WITH CONGENITAL HEART DISEASE: RESULTS FROM POPULATION-LEVEL COLORADO SURVEILLANCE SYSTEM OF CHD

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Background: Prior studies have shown high prevalence of mood and anxiety disorders in adults with congenital heart disease (CHD) seen at referral centers, yet little is known about the spectrum and population-based prevalence of mental illness in adults with CHD.

Methods: We analyzed the prevalence of a broad spectrum of mental illnesses using data from the Colorado CHD Surveillance System in Adolescents (age 11-17 years) and Adults (age 18-64) from 2011 to 2013. We also examined associations between complexity of CHD diagnosis, cardiac health care utilization, and underlying genetic syndromes on presence of mental illness using logistic regression.

Results: Of 3,215 adolescents with CHD, 19% received care for a mental illness during our 3-year surveillance period. The most prevalent categories of mental illness in adolescents were developmental disorders (6%), anxiety disorders (5%), externalizing disorders (attention-deficit/hyperactivity disorders, conduct disorders or disruptive behavior disorders) (5%), and mood disorders (4%). Of 9,197 adults with CHD, 25% received care for a mental illness over the 3-year period. The most prevalent categories of mental illness in adults were mood disorders (16%), anxiety disorders (14%), substance-related disorders (4%), and alcohol-related disorders (3%). Compared to simple lesions, severe lesion complexity was positively associated with presence of developmental disorders in both adolescents and adults (OR 2.97, 95%CI 1.93-4.57 and OR 3.62, 95%CI 2.41-5.46, respectively). In adults, compared to simple lesion complexity, severe lesion complexity was also positively associated with presence of externalizing disorders (OR 2.06, 95%CI 1.12-3.80). In adolescents and adults, compared to patients who underwent no cardiac procedures, undergoing three or more cardiac procedures was positively associated with anxiety disorders and mood disorders (OR 2.31, 95%CI 1.77-3.46 and OR 1.99, 95%CI 1.25-3.06, respectively). Having a diagnosis of Down syndrome, Turner syndrome, 22q11.2 deletion syndrome, other chromosomal anomalies, or other specified congenital anomalies was positively associated with a developmental disorder and anxiety disorder diagnosis (OR 3.84, 95%CI 2.76-6.43 and OR 2.17, 95%CI 1.63-3.81, respectively) compared to those without a genetic disorder diagnosis.

Conclusion: In a population-based cohort, a broad spectrum of mental illness is highly prevalent among adolescents and adults with CHD. Higher CHD complexity, higher cardiac healthcare utilization, and genetic syndromes are associated with higher prevalence of specific categories of mental illness.

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CARDIAC REHABILITATION IN ADULT CONGENITAL HEART DISEASE PATIENTS: EXTENDING BEYOND THE TYPICAL PATIENT POPULATION

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Background: the adult population with congenital heart disease continues to grow. These patients require complex medical care. Cardiac rehabilitation (CR) is a multifaceted program to optimize physical, psychological and social functioning in patients with cardiovascular disease. CR has been proven to significantly decrease morbidity and mortality in adults after cardiovascular events. The application, efficacy and safety of CR in adults with congenital heart disease (ACHD) have not been established and CR has been underutilized for ACHD patients nationwide. In the Children's Hospital of Michigan, CR program was launched as a quality improvement project in 2014. During the effort of system development, a multidisciplinary approach was used to reduce the gap in the delivery of CR to ACHD patients. CR team consisted of cardiovascular surgeons, ACHD specialists, transplant cardiologists and regional rehabilitation center. OBJECTIVES: The primary objective was to describe our 4-year experience of single centered CR program for ACHD patients. The secondary objective was to evaluate the efficacy and safety of CR as well as the barriers to CR referral in ACHD patients.

Methods: This was a retrospective study. The inclusion criteria were patients aged 16 years who were referred to regional CR centers for chronic heart failure, post-cardiac transplant and post-cardiac surgery. Through the referral process, barriers in referring ACHD patients were identified. The detail reports of CR were collected to assess the efficacy of CR on exercise capacity and adverse events. Qualitatively, all patients were interviewed at completion and 6-month post CR.

Results: Among our cohort of 36 patients referred to CR, 23 completed CR, 1 is presently undergoing CR, 11 are in the referral process, 1 died before CR was initiated. The median age of patients was 22 years (range 15 to 55). The primary indication for CR was post-surgical 56%, followed by chronic heart failure 38% and post-transplant 6%. Over the 4 years, a total of 23 regional CR centers were identified in the greater Detroit area. Post-CR, metabolic equivalent increased by 1.6 (95% confidence interval 1.0 to 2.2, $p<0.001$), maximal heart rate during session increased by 13 beats/min (2 to 25, $p=0.026$), CR exercise time increased by 5.7 min (0.1 to 11, $p=0.047$), treadmill speed increased by 0.7 mile/hr (0.3 to 1.0, $p=0.007$). There was no difference in body weight, resting blood pressure or resting heart rate. There have been no serious adverse events during CR sessions. All the patients (n=23) who completed CR remain alive at the median follow up of 17 months (range 5 to 45). Qualitatively, all patients perceived CR favorably on interviews. Common barriers to CR included issues associated with accessibility (transportation, CR center location), availability of session time, returning to work/school and cost for phase III rehabilitation.

Conclusion: The multidisciplinary quality improvement program successfully built a referral system for ACHD patients to CR. In our limited cohort of patients, CR was effective and safe, well received by patients.

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THE PROGNOSTIC VALUE OF ST2 IN ADULTS WITH CONGENITAL HEART DISEASE

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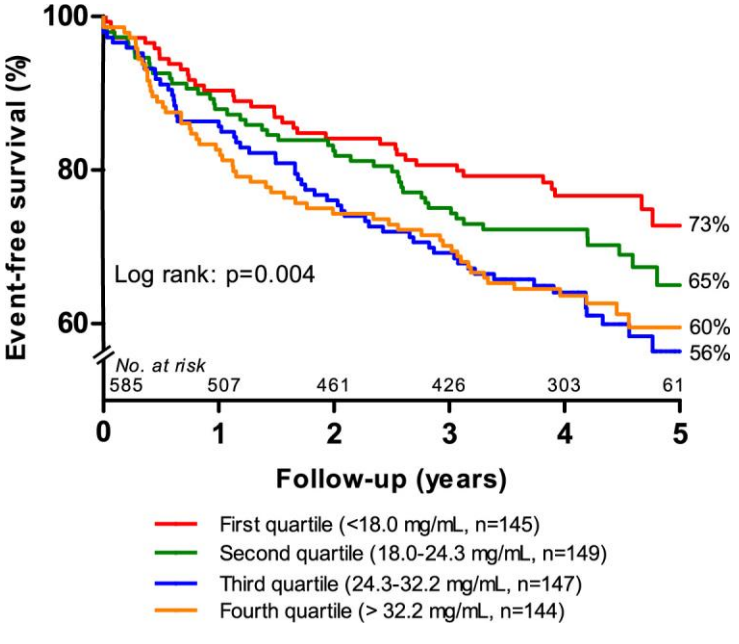
Background: Soluble suppression of tumorigenicity 2 (ST2) is upregulated as a response to myocardial stress and is associated with adverse events in chronic heart failure patients. This study aimed to investigate the association between ST2 and adverse cardiovascular events in adults with congenital heart disease (CHD).

Objectives: Aim of this study was to report on maternal outcome of pregnancy in women with diagnosis of CoA.

Methods: This is a prospective cohort study. ST2 and NT-proBNP was measured for research purposes in consecutive stable adults with CHD visiting the outpatient clinic between April 2011 and April 2013. Patients were prospectively followed for adverse cardiac events by annual evaluation at the outpatient clinic. The study endpoint was a composite of mortality, heart failure, hospitalization for cardiac reasons, arrhythmia, thromboembolic events and cardiac re-intervention. We performed multivariable Cox proportional-hazards regression to identify the association between ST2 level and the study endpoint, adjusted for age, sex and NT-proBNP level.

Results: We included 590 patients with a ST2 measurement (median age 33 [IQR 25-41] years, 42% female, 90% NYHA class 1),with tetralogy of Fallot (n=176), congenital aortic stenosis (n=133), aortic coarctation (n=109), arterial switch operation (n=24), Mustard operation (n=65), congenitally corrected transposition of the great arteries (n=20), Fontan (n=36) or other (n=27). Follow-up data was available in 585 patients and 195 reached the study endpoint (median follow-up 48.9 [IQR 31.7-55.8] months). ST2 was strongly associated with the incidence of the study endpoint after 5 years of follow-up: 27% of the patients with ST2 24.3 mg/mL (3rd and 4th quartile) (Figure). A twofold-increase in ST2 was associated with a 32% increased risk of the study endpoint (adjusted HR 1.32, 95%CI 1.06-1.65, p=0.014).

Conclusions: Serum ST2 level is associated with adverse cardiovascular events in adults with CHD and provides incremental prognostic information beyond the established biomarker NT-proBNP. Figure legend: Event-free survival for ST2 quartiles in adults with congenital heart disease.



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DEVELOPMENT OF A CLINICAL MODEL TO PREDICT SURVIVAL WITHOUT MAJOR ICU COMPLICATIONS FOR PATIENTS WITH ADULT CONGENITAL HEART DISEASE UNDERGOING CARDIAC SURGERY

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Background: Adult congenital heart disease (ACHD) patients needing cardiac surgery have comorbidities associated with lifelong cardiac disease. We sought to develop a population-specific risk model to predict in-hospital survival free of major complications for ACHD patients undergoing cardiac surgery. The same model was used to predict survival at one year.

Methods: Data was acquired from 4 prospective databases and individual chart review of 783 consecutive adults (age > 16 years) undergoing surgery for congenital heart disease during 2004-2015 in a single center. Only pre-surgical data was considered. Social, anatomic and physiological factors were used to build our prediction model including liver function (MELD XI score = Mayo End Stage Liver score-modified), pulmonary function and anatomic factors. The primary outcome is a composite adverse outcome (CAO) of in hospital mortality, prolonged ventilation (exceeding 7 days), and acute kidney injury requiring dialysis. The secondary outcome is one-year mortality. Univariate (UV) analysis identified the unadjusted association of variables with the primary outcome. Risk factors as identified by a stepwise model selection were used in the multivariable (MV) regression to develop our predictive model.

Results: A total of 783 patients are included in the study. The primary CAO occurred in 54/783 patients (6.9%), in-hospital mortality was 33/783 (4.2%), kidney injury requiring dialysis 26/783 (3.3%), and prolonged ventilation 40/783 (5.1%) After candidate variables were selected, multivariate regression analysis demonstrated that MELD XI score, cognitive impairment, number of chest wall incisions, BMI and anatomical diagnosis other than a 2-ventricle heart with AV concordance were predictors of the primary outcome. 657 out of 783 patients had follow up data beyond one year. One-year mortality was at best 5.1% (40/783) or at worst 6% (40/657). By multivariate analysis, risk factors for death at 1 year are chronic anti-coagulation (1.75), higher MELD-XI score (0.19/ I unit increase) & morphology other than 2-V heart with AV concordance.

Conclusions: Our model that incorporates 5 pre-operative variables predicts the risk of in-hospital survival without major complications. Two of these variables, as well as anticoagulation, are predictive of survival to 1 year following surgery.

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PROLONGED TPEAK-TEND INTERVAL IS A RISK FACTOR FOR SUDDEN CARDIAC DEATH IN ADULTS WITH CONGENITAL HEART DISEASE

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Background: Adult congenital heart disease (ACHD) patients are at risk of sudden cardiac death (SCD). However, current risk stratification methods are not yet well-defined. The Tpeak-Tend interval (TpTe) is a measure of dispersion of ventricular repolarization, and a risk factor for SCD in non-ACHD patients. We analyzed the predictive value of TpTe for SCD in ACHD patients.

Methods: From an international multicenter cohort of 25,790 ACHD patients, we identified 165 SCD cases. Cases were matched to 310 controls by age, gender, congenital defect and (surgical) intervention. TpTe was measured in one T-wave of each ECG lead on the last ECG before death in cases, and the ECG at the same age in controls. The mean (TpTe-mean) and maximum TpTe (TpTe-max) of all twelve ECG leads and TpTe dispersion were measured. Odds ratios (OR) were calculated using conditional logistic regression analysis.

Results: ECGs were available for 146 cases (median age at death 33.5 years (quartiles 26.2, 48.0), 66% male) and 302 controls. The mean TpTe-max was 97±24 ms in cases vs. 84±17ms in controls, TpTe-mean was 70±16 vs. 63±10, and dispersion 51±22 vs. 41±16, respectively. Assessing each ECG lead separately, TpTe in lead aVR predicted SCD most accurately. The mean TpTe in aVR was 71±23 in SCD cases vs 61±13ms in controls. At a cutoff of 80ms, the adjusted odds ratio of SCD of a longer TpTe in aVR was 5.8 (95% CI 2.7-12.4, $p<0.001$).

Conclusions: Tpeak-Tend interval predicts sudden cardiac death in adults with congenital heart disease. Particularly TpTe in lead aVR, the mean TpTe, the maximum TpTe and the TpTe dispersion appear to be of importance, and may add to current risk stratification methods for SCD in this young patient group.

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TRENDS IN IN-HOSPITAL MORTALITY AND HOSPITAL OUTCOMES AMONG MECHANICALLY VENTILATED ADULT PATIENTS WITH CONGENITAL HEART DISEASE IN THE UNITED STATES

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Background: There are an increasing number of adult patients with congenital heart disease (ACHD). The impact of ACHD patients on critical care services remains unknown. The primary objective of this study was to evaluate patient characteristics and trends in mortality of mechanically ventilated patients with ACHD. Secondary objectives included the evaluation of cost and length of hospital stay.

Methods: We evaluated mechanically ventilated patients with ACHD using the National Inpatient Sample (NIS) from 2004-2013. We isolated all patients greater than 18 years of age who had International Classification of Diseases, Ninth Edition (ICD-9) diagnosis codes of ACHD and procedure codes of mechanical ventilation. The NIS is a public all-payer inpatient United States database that contains a stratified sample of 20% of hospital discharges and includes approximately 8 million discharges per year. Estimates were obtained using complex survey methods and adjustments were made to account for differences in sampling method after the NIS re-design in 2012.

Results: A total of 77,316,763 discharges, representing 368,889,400 hospitalizations were evaluated using the NIS trend weights between 2004-2013. 9,734 of these discharges, representing 47,013 hospitalizations had ACHD, were mechanically ventilated and were greater than 18 years of age. The mean age of these patients was 56.8 years (standard error [SE] 0.3), and the mean Charlson comorbidity index was 2.10 (SE 0.3). 46% of patients were female. Average cost was USD $61,805 (SE $1,153). Mean length of stay was 16.2 days (SE 0.3). 56% of these patients were admitted with Angus definition of severe sepsis and septic shock. 60% of these patients also had acute respiratory distress syndrome (ARDS). 24.7% of patients died in-hospital. Over the years, it appears that hospital costs and length of stay were increasing ($p < 0.05$). Mortality in these patients over the years appeared to be unchanged ($p=0.35$).

Conclusion: Mechanically ventilated ACHD patients appear to have increasing burden of hospital consumption over the years. Severe sepsis and ARDS appear to be leading causes of admission. In-hospital mortality of these patients appears to remain constant.

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BAFFLE LEAKS IN ADULTS AFTER ATRIAL SWITCH FOR TRANSPOSITION OF THE GREAT ARTERIES: PRESENCE AT REST AND DURING EXERCISE

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Background: Recent single-center results revealed that baffles often showed leaks in unselected, asymptomatic patients with transposition of the great arteries after atrial switch (Mustard/Senning) when routinely evaluated with contrast-enhanced transthoracic echocardiography (CE-TTE) at rest. The aim of the present study was to evaluate presence of baffle leaks during rest and exercise.

Methods: Consecutive adult Mustard/Senning patients underwent contrast-enhanced semisupine bicycle echocardiography with agitated saline injection at rest and during exercise in two tertiary medical centers. Baffle leaks, diagnosed as right-to-left shunting, were analyzed at rest, with Valsalva, and at peak exercise. Clinical characteristics in patients with and without leaks were compared.

Results: 18 Mustard/Senning patients (14 male, median age 40.6[IQR 34.1-46.1]years, median age at AtrSO 1.1[IQR 0.6-2.2]years, 15 Mustards and 3 Sennings performed at 6 tertiary centers, 5 previous baffle reintervention) were included. None had clinically known baffle leaks. Contrast-enhanced echocardiography showed baffle leaks (2 severe, 4 moderate, 4 mild) in 10 patients. Right-to-left shunting at rest was present in 5 patients, in 3 only during Valsalva and exercise, and in 2 only at peak exercise. Although 1 patient with severe right-to-left shunting had a saturation of 94% at rest which decreased to 90% at peak exercise, median saturations at rest (97%[IQR 94-99] vs 97%[IQR 97-97]) and peak exercise (95%[IQR 94-96] vs 96%[IQR 92-97]), as well as additional clinical characteristics, were not significantly different between patients with and without leaks. Only one patient with a previous baffle intervention currently had a baffle leak (p=0.060). Median time since last baffle intervention was 37[IQR 31-42]years in patients with and 33[IQR 9-40]years in patients without leaks (p=0.29). Heart failure medication was used by 7 patients with and 3 patients without leaks (p=0.17).

Conclusion: Routine contrast enhanced transthoracic echocardiography revealed clinically unsuspected baffle leaks in 50% of the adults with Mustard/Senning. Half of patients with leaks had right-to-left shunting both at rest and during exercise. No significant differences between clinical characteristics of patients with and without baffle leaks were detected. The clinical significance of these unsuspected baffle leaks is still unclear and needs further investigation.

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DRUG THERAPY IN ADULT CONGENITAL HEART DISEASE: THE BURDEN OF POLYPHARMACY

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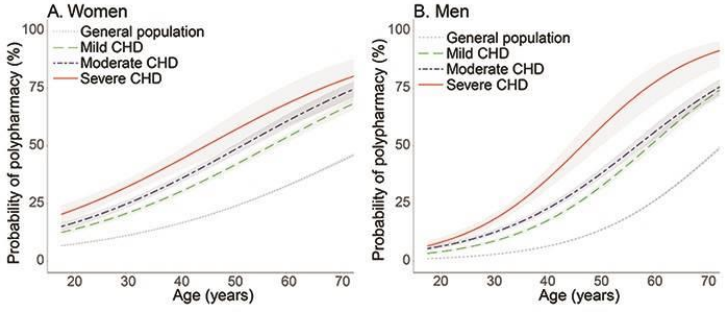
Background: Adult congenital heart disease (ACHD) increasingly includes frail individuals with more severe defects, with sequelae and comorbidities often requiring medical treatment. Half of ACHD patients use chronic medication, which may cumulate over time. This study investigated the prevalence, risk factors, and contributing drugs to polypharmacy in ACHD, and its association with mortality.

Methods: We identified patients from our nationwide ACHD registry and age- and sex-matched referents in a 1:10-ratio in the national Dispensed Drug Register and Cause of Death Register for the years 2006-2014. Drugs were classified according to the Anatomical Therapeutic Chemical classification, aggregated per year. Generalized estimating equations were used to determine associations between clinical characteristics and polypharmacy, defined as ≥5 different dispensed drug types per year. Associations between baseline polypharmacy and mortality were analyzed in patients surviving their baseline year using multivariable Cox regression.

Results: Overall, 14138 ACHD patients (49% male, median age 35 years, median follow-up 8 years) were included, of which 29% had polypharmacy at inclusion, compared to 13% of referents (P<0.001). Cardiovascular drugs were most prevalent(42% in patients vs 13% in referents, P<0.001), particularly antithrombotics (25% vs 4%, P<0.001), beta-blockers (21% vs 6%, P<0.001) and renin-angiotensin-aldosterone-system inhibitors (18% vs 5%, P<0.001). Polypharmacy decreased marginally from 2006 to 2014 (OR=0.97/year[0.96-0.98]). ACHD severity (OR=2.51[95%CI 2.40-2.61] for mild, OR=3.22[95%CI 3.06-3.40] for moderate, and OR=4.87[95%CI 4.41-5.38] for severe defects), was strongly associated polypharmacy, independent of sex and age. Polypharmacy risk was already 30% at age 40 in ACHD patients, compared to age 60 in referents. Young female patients already had relative high risk of polypharmacy: 24% of female patients <40 had polypharmacy at baseline (vs 12% of female referents <40), with high use of antibiotics (42% vs 23%) and sex hormones (44% vs 34%). In men, polypharmacy showed a steeper increase with age than in women (Pinteraction<0.001). 41% of male patients ≥40 had polypharmacy at baseline (vs 18% of male referents >40) with high use of antithrombotics (46% vs 12%), betablockers (37% vs 11%), and renin-angiotensin-aldosterone-system inhibitors (38% vs 14%). During 7[IQR 5-8] years of follow-up, 595 patients (4%) and 2375 referents (2%) died. Adjusted for age, sex, and defect severity, baseline polypharmacy was independently associated with mortality in ACHD (HR=2.90[95%CI 2.42-3.49], P<0.001).

Conclusions: Polypharmacy is as common in 40-year old ACHD patients as in 60-year old referents from the general population. Below the age of 40, especially female patients are at risk, with high use of non-cardiovascular drugs. Male patients are especially at risk over the age of 40, with high use of cardiovascular drugs. Polypharmacy is strongly associated with defect severity and with all-cause mortality. These findings underscore the importance of specialist care, weighing risks and benefits of polypharmacy in this population.

Figure: Probability of polypharmacy by age during the study stratified by sex and congenital heart defect (CHD) severity.



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VASOPRESSOR MAGNITUDE PREDICTS POOR OUTCOME IN ADULTS WITH CONGENITAL HEART DISEASE AFTER CARDIAC SURGERY

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Background: High levels of vasoactive inotrope support (VIS) after congenital heart surgery are predictive of morbidity in pediatric patients. We sought to discern if this relationship applies to adults with congenital heart disease (ACHD).

Methods: We retrospectively studied adult patients (≥18 years old) admitted to the intensive care unit after cardiac surgery for congenital heart disease from 2002-2013 at Mayo Clinic. Vasoactive medication dose values within 96 hours of admission were examined to determine the relationship between VIS score and poor outcome of early mortality, early morbidity or complication related morbidity.

Results: Overall, 1040 ACHD patients had cardiac surgery during the study timeframe; 243 (23.4%) met study inclusion criteria. Sixty-two patients (25%), experienced composite poor outcome [including 8 deaths within 90 days of hospital discharge (3%)]. Thirty-eight patients (15%) endured complication related early morbidity. The maximum VIS (maxVIS) score area under the curve was 0.92 (95% CI:0.86-0.98) for in-hospital mortality; and 0.82 (95% CI: 0.76-0.89) for combined poor clinical outcome. On univariate analysis, maxVIS score ≥3 was predictive of composite adverse outcome (OR:14.2, 95% CI: 7.2-28.2; p <0.001), prolonged ICU LOS ICU LOS (OR:19.2; 95% CI: 8.7-42.1; P < .0001), prolonged mechanical ventilation (OR:13.6; 95% CI:4.4-41.8; P <.0001) and complication related morbidity (OR: 7.3; 95% CI: 3.4-15.5; P <.0001).

Conclusions: MaxVIS score strongly predicted adverse outcomes and can be used as a risk prediction tool to facilitate early intervention that may improve outcome and assist with clinical decision making for ACHD patients after cardiac surgery.

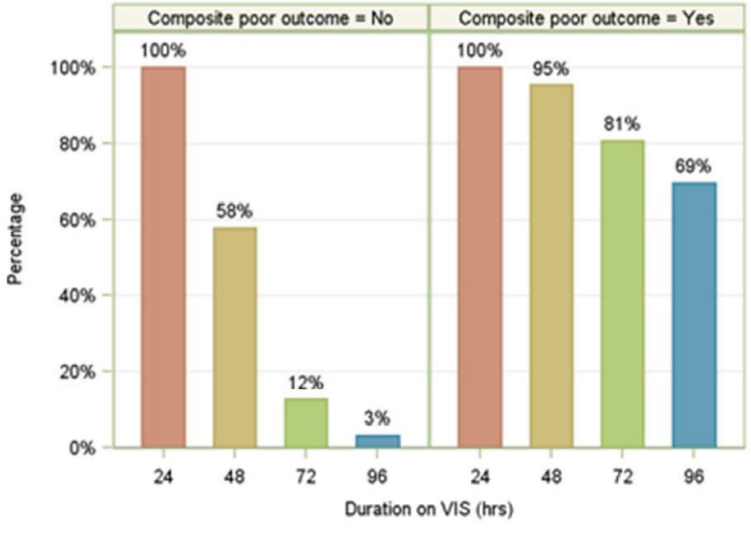

Figure 2
Figure Title: Frequency of VIS values within each 24-hour period
Figure caption: After 48 hours, only 12% of those patients who did not have any poor outcome were still requiring vasoactive support, comparing to 79% patients who did experience at least one poor outcome (Percentage is based on the total number of patients in each outcome group).

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PREDICTING OUTCOMES IN PARTURIENTS WITH UNDERLYING CARDIAC DISEASE: AN IRISH PERSPECTIVE

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Background: The incidence of pregnancy in women with underlying cardiac disease is increasing and this is now the leading cause of maternal death in the UK and Ireland. The reasons for this include the increased incidence of women with congenital heart disease reaching child-bearing age, the general trend towards increased maternal age, as well as increased incidence of chronic diseases which increase cardiac risk, such as diabetes mellitus and hypertension. Pregnancy is associated with well-described haemodynamic changes which cause increased risk to parturients with underlying cardiac disease. These include increased plasma volume, physiological anaemia, increased pulse rate, increased cardiac output, and decreased systemic vascular resistance. The magnitude of complications in pregnancy varies depending on the underlying disease. As in many western countries, congenital heart disease is the biggest cause of maternal heart disease in Ireland.

Objective: The Rotunda Hospital in Dublin is the oldest continuously operating stand-alone maternity hospital in the world and in combination with the Mater Misericordiae University Hospital provides obstetric care to parturients with underlying cardiac disease. We wanted to assess outcomes for all women who were deemed to be at increased risk, and who accordingly were discussed in the the Joint Obstetric Cardiac multidisciplinary meeting. Indeed, a lack of co-location of cardiac and maternity care is perceived as one of the major barriers to improving care of parturient with underlying cardiac disease, hence we wanted to compare outcome data with published figures.

Methods: : We retrospectively assessed the women who were discussed in the Joint Obstetric Cardiac multidisciplinary meeting using cardiac risk stratification tools for the period 2015-2017. We included all women who were discussed in the MDT, identifying them using the minutes from each meeting held between 2015 and 2017. Much of the antenatal information required was obtained in this way. We subsequently pulled the paper charts for these women to determine

Results: 200 patients were discussed in the Cardiac MDT. 9% of these were for valvular heart disease, 55% for congenital heart disease, 6% for cardiomyopathy, 4% for coronary artery disease, and 20% for arrhythmias. We assessed CARPREG and ZAHARA scores, depending on their underlying cardiac disease, and then compared predicted outcomes to actual outcomes.

Conclusions: Our results so far show that the workload from an obstetric cardiac viewpoint in our unit is similar to what would be seen in other westernised healthcare systems with the main cause of obstetric cardiac disease being congenital in origin. We will be presenting outcome data for the patients concerned.

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CONGENITAL HEART SURGERY IN PATIENTS OLDER THAN 16 YEARS IN A PUBLIC PEDIATRIC INSTITUTION: SCENARIO AND PROFILE OF A GROWN POPULATION.

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Background: Major advances and refinement in the diagnosis and surgical treatment of congenital heart defects in the last four decades has resulted in an increasing number of adult survivors. It should be noted that congenital heart surgery is "reparative" and not curative.Many patients (p) will develop residual lesions and sequelae requiring reinterventions.

Methods/ Results: From 2013 to 2015, 56 patients with a median age of 22 years (range 16.1-27) were operated with extracorporeal circulation. 35/56 were reoperations. 13% of the lesions were simple and 87% complex. Distribution by number of surgeries per year, reinterventions and type of lesion are resumed in chart 1 41p had previous surgicalinterventions: The interventions were performed: on right ventricle outflow tract: 14p, left ventricle outflow tract: 7p, atrioventricular valve: 7p, partial cavopulmonary connection (Glenn): 1p, Fontan (extracardiac conduit):3p, reconversion to extracardiac conduit + Maze-Cox procedure: 2p, atrial septal defect closure: 4p, scimitar syndrome: 1p, unifocalization and right ventricle to pulmonary artery conduit: 1p. Median time of hospitalization in the cardiovascular criticalcareunitwas 4 days (range: 2-21 d). Mortality rate was 1.7 % (1p), and was related to RACHS score IV (p 0.0000)

Conclusion: 73% of patients had previous interventions. The congenital heart defects was moderate to severe in 87% of the cases Mortality rate was 1.7%, related to greatcomplexitycongenitalheartdiseasewithpoor prognosis. The operated group conform a growing population in quantity and complexity. Most of them are followed by the pediatric cardiac team. Transference to adult public institution is challenging.

Chart 1: Distribution by number of surgeries per year, reinterventions and type of lesion.

Period	Surgeries	Reoperations	Rachs1	Rachs2	Rachs3	Rachs4
2013	15	11	2	1	12	0
2014	21	13	1	1	18	1
2015	20	17	2	0	18	0

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IMPACT OF MACITENTAN ON 6MWT IN TRISOMY 21 PATIENTS WITH EISENMEGER SYNDROME

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Background: Adult patients with Eisenmenger syndrome (ES) develop slow progressive hypoxemia with central cyanosis and limited exercise capacity. ES occurs as a consequence of pulmonary arteriolar remodeling secondary to chronic unprotected left-to-right shunting. A disproportionate percentage of adult patients with Trisomy 21 have ES due to historical concerns regarding risk of surgical repair. Macitentan is a novel endothelin receptor antagonist (ERA) which has been used in Eisenmenger patients. The aim of our study was to evaluate the impact of Macitentan on 6-minute walk test (6MWT) data in Trisomy 21 patients with ES.

Methods: Data was collected retrospectively from all our ES patients. Patients who did not have a six minute walk test (6MWT) both pre and post initiation of Macitentan were excluded

Results: Nine patients had a 6 MWT pre and post initiation of Macitentan. Six patients were naïve to ERA therapy and three of them had no previous exposure to pulmonary hypertension therapy. All patients tolerated therapy with no adverse drug reaction. Six patients demonstrated an improvement to WHO Functional Class II (slight limitation of physical activity) after treatment initiation. There was a significant improvement in the median 6MWD from pre-initiation (301 m; 95% confidence interval [CI] 181.5-438) to post-initiation (380 m; 95% CI 240-406.9) (P=0.0007). The mean distance improved from 264.78 m to 323.56 m Baseline mean resting oxygen saturation also improved between pre- and post-treatment initiation from 82% to 84% (P=0.032).

Conclusions: Although our sample size is small, there are measurable clinical improvements with Macitentan therapy in T21 patients with Eisenmenger Syndrome. This specific subgroup may warrant further evaluation to determine if this effect is sustained in a larger cohort of patients.

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PULMONARY ARTERIAL HYPERTENSION IN A MULTI-ETHNIC ASIAN POPULATION - CHARACTERISTICS, SURVIVAL AND MORTALITY PREDICTORS FROM A 12 YEAR FOLLOW-UP STUDY.

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Background: Pulmonary arterial hypertension (PAH) is a rare and fatal disease. Data from Asia is lacking compared to the West. We aim to describe disease characteristics in an ethnically-diverse Asian population across 12 years and assess predictors for survival.

Methods: We consecutively enrolled patients with PAH referred to our pulmonary hypertension specialty centre from January 2003 to December 2016. Baseline characteristics and survival were analysed. Based on a forward predictor-selection procedure, a multi-level structural equation model was applied to identify predictors associated with mortality.

Results: Out of 148 patients, 77% were female and mean age was 50.8±15.9 years. Racial distribution consisted of 60.1% Chinese, 27.0% Malay, 9.5% Indian and 3.4% others. The most common aetiologies were congenital heart disease (35.8%), idiopathic PAH (29.7%), then connective tissue disease (24.3%). Most presented in WHO Functional Class (FC) II (48.6%), followed by FC III (28.8%). Majority (54.1%) were on phosphodiesterase inhibitor monotherapy. Survival rates were 85.8% at first year, 70.9% at 3 years, 66.9% at 5 years, and 55.4% at 10 years. The REVEAL score was established as the best predictor of mortality, a score>6 was identified as a cut off. Other predictors include mean right atrial pressure, heart rate, aetiology, age and NT pro-brain natriuretic peptide.

Conclusions: I n this first registry study from a multi-ethnic Asian cohort, the most prevalent aetiology was PAH associated with congenital heart disease. Our survival rates are comparable with other national registries. The REVEAL score is validated in our population to be a good predictor of mortality.

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DO WE HAVE THE ACHD PHYSICIAN RESOURCES WE NEED TO CARE FOR THE BURGEONING ACHD POPULATION?

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Background: Available workforce of adult congenital heart disease (ACHD) cardiologists is too small for the population. US data is limited and our population estimates are extrapolated from Canadian data. In 2008, Marelli et al determined that US had an estimate of 1.5 million ACHD patients in the US, of which approximately 50% had moderate-complex lesions. This estimate determined there would be a 4.5% per annum increase, leading to a current estimate of 2.3 million, and 1.15 million moderate-complex patients. We proposed to evaluate the adequacy of ACHD physician:patient ratios in the US at both national and regional levels.

Methods/Overview: : Data from the Adult Congenital Heart Association (ACHA) website, a publicly available database of self-reported information, along with metropolitan area and statewide population data from 2016 US Census Bureau estimates was analyzed. Physicians listed on ACHA website were cross-referenced with ABIM data to verify ACHD board certification status.

Results: There are 115 self-identified programs in the ACHA website, with 110,112 ACHD patient visits per year. Programs represent 78 Metropolitan Statistical Areas (MSA). Three MSA with > 1 million residents do not have an ACHD program (Providence RI, Richmond VA, and Buffalo NY). The ACHA directory includes 418 physicians self-identified as being ACHD physicians, of which only 159 are board-certified in ACHD. There are 320 Board-Certified ACHD Cardiologists in the US today, including 161 not listed in the ACHA website. Fourteen programs in the ACHA directory do not list a single ACHD-certified physician. Regarding ratios of ACHD-certified physicians to patients, the worst-served MSA is Riverside-San Bernardino-Ontario, CA, with 4.5 million residents:physician. The best-served MSA with > 1 million residents is Raleigh-Cary, NC, with 325,000 residents:physician. The best-served State is Washington, D.C., with 340,585 residents:physician. The worst-served State with any ACHD-certified physician is Indiana with 6.3 million residents:physician. Nine states with ACHA programs do not have a single ACHD-certified physician and 8 states have no program at all. For an estimated US population of 326,929,143. with the recommendation to have a regional program with 3 physicians for every 2 million people, there should be 163 programs and 490 ACHD physicians. Additionally, an estimated 1.15 million moderate to severe complexity ACHD patients in the US, with the recommendation of a physician:patient ratio of 1 :1000 moderate-to-complex ACHD patients, there should be 1,150 ACHD physicians in the US. In order to bring the ACHD patient:physician ratio to 1000:1 a bare minimum of 170 additional ACHD-boarded physicians are needed today.

Conclusions: An estimated 10% of moderate-complex patients appear to be "in care" at an ACHD center in US. The ACHD population is anticipated to plateau in 2040, and the looming national physician shortage is likely to greatly impact the ACHD population as senior pediatric and ACHD cardiologists retire. In order to meet the minimum criteria based on ACC/AHA guidelines for the Care of Adults with Congenital Heart Disease, 170 board-certified ACHD physicians are needed immediately, and a succession plan needs to be laid in place.