



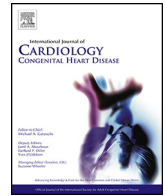
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20 top abstracts from the 31st International Symposium on ACHD, October 2021

Oral presentations

AORTOPATHY AND ENERGY LOSS IN ADULTS WITH TRANSPOSITION OF THE GREAT ARTERIES AFTER THE JATENE PROCEDURE

Yumi Shiina^{1,2}, Kei Inai^{1,2}, Michinobu Nagao^{1,2}. ¹St.Luke's International Hospital, Tokyo, Japan² Tokyo Women's Medical University Hospital, Tokyo, Japan

Purpose: Dilated aortic root is a well-known sequela after arterial switch operation (ASO) in adults with transposition of the great arteries (TGA). This aortopathy in TGA is multifactorial. 4D flow MRI enables the quantification of instantaneous viscous energy loss (EL), which is a marker of prominent secondary aortic flow structural abnormality and cardiac workload. Elevated EL suggests potential ventriculo-arterial decoupling, which is a novel concept of fluid dynamics.

We aimed to assess quantitatively aortic flow profiles and EL in TGA adults after ASO (Jatene procedure with LeCompte maneuver) using 4D flow MRI.

Methods: Prospectively, 9 consecutive adults (30.2 ± 6.6 years) after ASO (Jatene operation with LeCompte technique), 13 consecutive adults (34.3 ± 7.2 years) after the atrial switch operation with Senning procedure, and 8 age-matched control patients, who underwent turbo field echo (TFE) echo planner imaging (EPI) 4D flow MRI, were enrolled. Abnormal aortic structure (the diameter ratio of the sinus of Valsalva and the ascending aorta, and acute aortic arch angle), abnormal aortic flow profiles, and EL were evaluated.

Results: TGA after ASO showed a markedly dilated sinus of Valsalva, compared to TGA after atrial switch operation (26.6 ± 4.9 vs. 18.6 ± 1.5 mm/cm²). Energy loss/Cardiac index (EL/CI) and helicity in the aortic root in TGA after Jatene procedure were the greatest among the three. EL was relevant to the diameter ratio of the sinus of Valsalva and the AAO (r=0.78, P=0.03).

Conclusions: Abnormal aortic structure as well as histologically vulnerable neo-aortic root after Jatene procedure may play an adjunctive role in promoting aortopathy and elevating EL. The cause and effect relationship is always challenging to discuss between abnormal aortic profile and aortopathy; however, it is highly possible that abnormal flow dynamics aggravate aortopathy secondarily.

Oral presentations

EXERCISE CAPACITY CORRELATES WITH THORACIC SKELETAL MUSCLE ABNORMALITIES IN ADULTS WITH CONGENITAL HEART DISEASE

Jacob Steffen, Seth Garton, Ravi Ashwath, Jennifer Maldonado, Krista Young, Colleen Lancial, Osamah Aldoss, Prashob Porayette. University of Iowa Hospitals and Clinics, Iowa City, United States

Background: Sarcopenia, the reduction in skeletal muscle mass, is common in adults with congenital heart disease (ACHD). We hypothesized ACHD patients have reduced exercise capacity in correlation with thoracic skeletal muscle area (SMA).

Material and Methods: This single-center retrospective study reviewed ACHD patients between the ages of 18-50 years who underwent cardiac pulmonary exercise test (CPET) and chest MRI/CT within one year of each other between January 2010-December 2019. SMA(cm²) was measured on CT/MRI derived images by drawing ROI manually at T10-T12 vertebral levels and compared with previously published reference healthy US population data. SMI:SMA indexed to patients' height squared. SMRA from CT:mean skeletal muscle radiation attenuation in Hounsfield Units(HU) (inversely related to muscle fat content).

Results: Forty-two patients (55% males;mean age 28.2±8.1years) met inclusion

criteria. The most common CHD diagnosis was tetralogy of Fallot (29%). Males but not females had significantly lower SMA and SMI at T10 (p=0.002; p=0.007, respectively) and T11 level (p=0.007;p=0.02, respectively) compared to healthy population.

Peak O₂ pulse significantly correlated with SMA at T10 (r=0.68,R²=0.46,p<0.0001), T11 (r=0.73,R²=0.54,p<0.0001), and T12 (r=0.86,R²=0.74,p=0.001) levels (Figure). Similar correlation existed between peak O₂ pulse and SMI. Peak VO₂ correlated with SMA at T11 (r=0.37, R²=0.14,p=0.04) and SMRA(HU) at T11 (r=0.87,R²=0.77,p<0.001) and T12 (r=0.67,R²=0.45,p=0.04) levels. SMA at T10 (r=-0.31, R²=0.1,p=0.049) and T11 (r=-0.44,R²=0.2,p=0.01) had weak negative correlation with VE/VCO₂.

Upon subgroup analysis, patients with D-transposition of the great arteries and Mustard operation had significantly lower peak VO₂ (24.8±4.6 vs 37.6±8.2ml/kg/min;p=0.02) and predicted VO₂ (59.8±7.5 vs 84.3±11.3%;p=0.006) despite similar skeletal muscle values to those with arterial switch operation.

Conclusions: Decreased thoracic SMA and increased skeletal muscle fat content correlates with lower exercise parameters in ACHD patients. Routine measurement of thoracic SMA might help prevent and treat SMA loss with nutrition and/or exercise training to improve exercise capacity and thereby outcomes in ACHD patients.

Oral presentations

EXPERIENCE OF ADULTS WITH CONGENITAL HEART DISEASE DURING THE COVID-19 PANDEMIC

Mikyla Janzen, Karen LeComte, Gnalini Sathananthan, Marla Kiess, Santabhanu Chakrabarti, Jasmine Grewal. University of British Columbia, Vancouver, Canada

Background: The COVID-19 pandemic has brought unprecedented challenges for those with pre-existing conditions such as adults with congenital heart disease (ACHD). This study sought to understand the experiences of ACHD throughout the pandemic. Objectives were to determine 1) the prevalence of COVID-19 symptoms, testing, and infection; 2) the prevalence of psychological distress; and 3) changes to day-to-day functioning.

Methods: A web-based survey was distributed to patients between December 2020-January 2021. Patients reported on whether they had experienced symptoms of COVID-19 and/or sought testing, their psychological distress across 5 categories (the STOP-D survey; depression, anxiety, stress, anger, and lack of social support) at the beginning of the pandemic and the time of the survey, and changes to their work and social behaviours. Survey results were linked to clinical data and patients were classified by anatomical and physiological status.

Results: Five hundred seventy-nine survey responses were received, of which 555 were linked to clinical data. Patients were 45 ± 15 years old. COVID-19 symptoms were reported by 145 (25%) patients. One hundred eighty-two (31%) patients sought testing and 10 (1.7%) tested positive. Psychological distress was reported in 27-42% of patients when reflecting on the two weeks prior to completing the survey (Figure 1). The greatest number of patients reported depression (42%) and/or anxiety (53%). More patients reported depression (42% vs 21%), anxiety (53% vs. 34%), and lack of social support (29% vs. 10%) during the pandemic compared to before. There were no strong trends across increasing anatomic complexity and physiologic status. Fifty (9%) patients lost employment due to a COVID-19 related reason. Most patients were either only going out for essential visits (233, 40%) or socializing with less than 6 people (149, 26%).

Conclusions: Few patients with ACHD contracted COVID-19, but a substantial

proportion reported significant psychological distress during the pandemic.

Oral presentations

INCIDENCE OF INFECTIVE ENDOCARDITIS IN ADULTS WITH CONGENITAL HEART DISEASE AND DIAGNOSED 22Q11.2 DELETION SYNDROME

Adonis Ng^{1,2}, Tracy Heung^{1,2}, Erwin Oechslin^{2,3}, Jessica Patzer³, Anne Bassett^{1,2,3,4,5}. ¹ *Clinical Genetics Research Program, Centre for Addiction and Mental Health, Toronto, Canada* ² *The Dalglish Family 22q Clinic, University Health Network, Toronto, Canada* ³ *Toronto Congenital Cardiac Centre for Adults, Division of Cardiology at the Peter Munk Cardiac Centre, Department of Medicine, University Health Network, University of Toronto, Toronto, Canada* ⁴ *Department of Psychiatry, University of Toronto, Toronto, Canada* ⁵ *Toronto General Research Institute and Campbell Family Mental Health Research Institute, Toronto, Canada*

Background: 22q11.2 deletion syndrome, formerly known as DiGeorge syndrome, is characterized by the 22q11.2 microdeletion. Conotruncal cardiac defects are commonly associated with 22q11.2 deletion patients. We aimed to determine the effect of 22q11.2 deletion syndrome on infective endocarditis (IE) incidence among adults with congenital heart disease (ACHD), with additional focus on ToF patients.

Methods: In this study, we examined the data available for 192 patients (aged 18-69 years) with ACHD and a 22q11.2 deletion confirmed through standard molecular genetic testing, followed over 6,377 person-years at the Toronto Dalglish Family 22q Clinic. We compared these data with those from a national registry involving a population of ACHD patients in the Netherlands, not tested for 22q11.2 deletions (n=10,210). A 2-Sample Poisson Rate test determined if there is a significant difference in the incidence rate of IE between populations.

Results: Within the 22q11.2 deletion-ACHD cohort (n=192), 20 (10%) patients developed IE over 6,377 person-years. Of the 192 patients, 89 (46%) have ToF and 12 (13%) of 89 developed IE. The incidence rate of IE in the 22q11.2DS-ACHD sample was significantly higher than in the Netherlands-ACHD sample (314 vs 106 per 100,000 person-years, p<0.0001). Restricting to adults with ToF, the 22q11.2 population incidence rate is significantly greater than in the Netherlands population (426 vs 110 per 100,000 person-years, p=0.0021).

Conclusions: Results indicate that the 22q11.2 microdeletion is associated with a substantially increased incidence of IE in ACHD patients and reinforces the importance of considering clinical testing for the 22q11.2 deletion in ACHD patients. We believe the 22q11.2 microdeletion is a significant additional risk factor for IE in ACHD patients. Further research will compare data from this 22q11.2 cohort to our local center's incidence of IE in all ACHD patients and determining the additional risk factors of 22q11.2 microdeletion patients that predispose them to IE.

Oral presentations

INCREASED RIGHT VENTRICULAR ENERGY EFFICIENCY BY 4DMR AFTER HARMONY VALVE IMPLANTATION

Jennifer Woo¹, Melody Dong², Fanwei Kong³, Doff McElhinney⁴, Nicole Schiavone², Frandics Chan⁵, George Lui¹, Francois Haddad¹, Daniel Bernstein⁶, Alison Marsden². ¹ *Department of Medicine, Division of Cardiovascular Medicine, Stanford University School of Medicine, Stanford, United States* ² *Department of Bioengineering, Stanford University, Stanford, United States* ³ *Department of Mechanical Engineering, University of California, Berkeley, Berkeley, United States* ⁴ *Department of Cardiothoracic Surgery, Stanford University School of Medicine, Stanford, United States* ⁵ *Department of Radiology & Cardiovascular Imaging, Stanford University School of Medicine, Stanford, United States* ⁶ *Department of Pediatrics, Division of Pediatric Cardiology, Stanford University School of Medicine, Stanford, United States*

Background: In patients with repaired tetralogy of Fallot (rTOF), it is not known how transcatheter pulmonary valve replacement (PVR) affects right ventricular (RV) kinetic energy (KE) and flow. We performed a pilot study to evaluate RV energy by 4D flow magnetic resonance (4DMR) before and after PVR.

Methods: Three rTOF patients underwent transcatheter Harmony PVR and 4DMR within 7 days before and 6 months after PVR. 4DMR velocity was corrected for phase offsets using Arterys, (San Francisco, CA). Custom Python scripts were used to obtain hemodynamic metrics by automatically reconstructing RV geometry for each timepoint in the cardiac cycle using a deep-learning based approach and computing spatially averaged turbulent KE and rate of viscous energy loss over the entire RV.

Results: Mean age was 24±10 years. Mean pulmonary regurgitant flow (PRF) at

baseline was 39±11% and improved after PVR to 0%. RV systolic and diastolic peak KE's were lower after PVR (systolic KE 0.053±0.006 vs. 0.041±0.017 mJ/ml3 and diastolic KE 0.029±0.014 vs. 0.012±0.005 mJ/ml3). There was no change in systolic peak rate of viscous energy loss; however, diastolic peak rate of viscous energy loss was less after PVR (0.027±0.006 vs. 0.018±0.012 mJ/ml3). After PVR, there was higher pulmonary cardiac output (CO) (6.1±0.9 vs. 5.2±1.3 L/min). The inverse relationship between CO and RV KE suggests that after PVR, the RV is more efficient - using less energy to generate a larger CO. RV vorticity was unchanged by PVR (13±0.04% vs 12±0.01%).

Conclusions: In this first study describing changes in RV energetics after transcatheter PVR, we demonstrate increased RV energy efficiency. Although future studies with a larger cohort are needed, RV energy by 4DMR may be useful in determining optimal timing of PVR.

Oral presentations

OUTCOMES OF BALLOON VALVULOPLASTY VERSUS SURGICAL VALVOTOMY IN CONGENITAL AORTIC STENOSIS: A SYSTEMATIC REVIEW AND META-ANALYSIS

Takhliq Amir¹, Hatim Alraddadi², David Barron^{3,4}, Osami Honjo^{3,4}. ¹ *Michael G. DeGroot School of Medicine, McMaster University, Hamilton, Canada* ² *Division of Cardiac Surgery, McMaster University, Hamilton, Canada* ³ *Division of Cardiovascular Surgery, The Labatt Family Heart Centre, The Hospital for Sick Children, Toronto, Canada* ⁴ *Department of Surgery, University of Toronto, Toronto, Canada*

Background: The debate regarding the best therapeutic approach for congenital aortic stenosis (AS) has persisted for decades due to equivocal evidence. Historically, the selection of interventions, primarily surgical aortic valvotomy (SAV) versus balloon aortic valvuloplasty (BAV), has depended on individual/institutional preferences. Recent single-centre studies demonstrate better outcomes with SAV; however, their results were attributed to improved surgical techniques. In this study, we perform a contemporary systematic review and meta-analysis to compare the effects of BAV and SAV on patient morbidity and mortality.

Material & Methods: We searched MEDLINE and EMBASE from inception to March 2021 for studies comparing BAV versus SAV for critical congenital AS in patients younger than age 18. We performed title and abstract screening, full-text review, risk of bias assessment using the CLARITY tool, and data collection independently and in duplicate. We pooled data using the random-effects model and the Mantel-Haenszel statistical method. We evaluated the overall quality of evidence using the GRADE framework.

Results: 12 studies (n=1214) comparing outcomes for BAV and SAV in the pediatric population were included. Differences in mortality at 30-day and at longest follow-up were not statistically significant between the two groups (RR 0.66, 95% CI [0.34, 1.27] and 0.74, 95% CI [0.35, 1.56], respectively). Post-procedural aortic insufficiency was more common after BAV but failed to reach statistical significance (RR 1.5, 95% CI [0.99 – 2.99]). Reintervention favoured surgical repair at the longest follow-up but did not reach statistical significance (RR 1.11, 95% CI [0.89, 1.38]).

Conclusion: In pediatric patients with congenital AS, surgical valvotomy did not offer additional advantages over balloon valvuloplasty. There was a trend towards increased post-procedural AI and rate of reintervention with BAV. However, the quality of evidence is very low; an appropriately powered study is required to adequately address the risks and benefits of the two interventions.

Oral presentations

SYSTEMIC VENOUS HYPERTENSION AND LOW OUTPUT ARE PREVALENT AT CATHETERIZATION IN ADULTS WITH PULMONARY ATRESIA AND INTACT VENTRICULAR SEPTUM REGARDLESS OF REPAIR STRATEGY

C. Charles Jain, Alexander Egbe, Elizabeth Stephens, Heidi Connolly, Donald Hagler, Moira Hilscher, William Miranda. Mayo Clinic, Rochester, United States

Background: Patients with pulmonary atresia and intact ventricular septum (PA-IVS) require intervention early in life, and most survive to a definitive procedure of either Fontan circulation or right ventricle to pulmonary artery (RV-PA) repair. It remains unknown how surgical strategy impacts hemodynamics and comorbidities in adults.

Material and Methods: Retrospective analysis of adults (age >18 years) with PA-IVS undergoing hemodynamic catheterization at Mayo Clinic, MN between