University Hospitals Bristol
NHS Foundation Trust

Adult Congenital Heart Disease

Evidence Update
June 2018 (Quarterly)

Contents

Current Journals: Tables of Contents

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If you require full articles please email: library@uhbristol.nhs.uk

Lunchtime Drop-in Sessions

June (12.00-13.00)

20th (Wed) **Interpreting Statistics**

28th (Thurs) **Literature Searching**

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5th (Thu) **Critical Appraisal**

9th (Mon) **Statistics**

19th (Thu) **Literature Searching**

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16th (Thu) **Critical Appraisal**

22nd (Wed) **Statistics**

30th (Thu) **Literature Searching**

Your Outreach Librarian Sarah Barrett

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Library Clinic

Stop by and find out more about our services. We will be here to answer any questions you may have!

June 19th: **Welcome Centre, BRI** *10.00-16.00*

July 3rd: **Welcome Centre, BRI** *10.00-16.00*

July 4th: **Canteen (Level 9, BRI)** *12.00-14.00*

August 8th: **Foyer, Education Centre** *12.00-14.00*

August 29th: **Foyer, St Michael's Hospital** *12.00-14.00*

September 5th: **Canteen (Level 9, BRI)** *12.00-14.00*

September 11th: **Welcome Centre, BRI** *10.00-16.00*

October 3rd: **Terrace (Level 4, Education Centre)** *12.00-14.00*

November 7th: **Canteen (Level 9, BRI)** *12.00-14.00*

December 5th: **Foyer, Education Centre** *12.00-14.00*

December 11th: **Welcome Centre, BRI** *10.00-16.00*

Latest Evidence

NICE National Institute for Health and Care Excellence

Structural heart defects

Everything NICE has said on structural heart defects in an interactive flowchart

NICE Pathway Published November 2013 Last updated May 2018

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Medical management of cyanotic congenital heart disease in adults

Author: Heidi M Connolly, MD, FASE

Section Editor: John K Triedman, MD

Deputy Editor: Susan B Yeon, MD, JD, FACC

Contributor Disclosures

All topics are updated as new evidence becomes available and our peer review process is complete.

Literature review current through: May 2018. | **This topic last updated:** Mar 01, 2018.

Anesthesia for adults with congenital heart disease undergoing noncardiac surgery

Authors: Edmund Jooste, MB, ChB; Kelly Machovec, MD, MPH

Section Editors: Jonathan B Mark, MD; Lena S Sun, MD; Heidi M Connolly, MD, FASE

Deputy Editors: Nancy A Nussmeier, MD, FAHA; Susan B Yeon, MD, JD, FACC

Contributor Disclosures

All topics are updated as new evidence becomes available and our peer review process is complete.

Literature review current through: May 2018. | **This topic last updated:** May 16, 2018.

Management of patients post-Fontan procedure

Authors: Jonathan Johnson, MD; Heidi M Connolly, MD, FASE

Section Editor: Gruschen Veldtman, MBChB, FRCP, Dip Obst

Deputy Editor: Susan B Yeon, MD, JD, FACC

Contributor Disclosures

All topics are updated as new evidence becomes available and our peer review process is complete.

Literature review current through: May 2018. | **This topic last updated:** Jan 10, 2018.

Management of Eisenmenger syndrome

Author: Heidi M Connolly, MD, FASE

Section Editor: John K Triedman, MD

Deputy Editor: Susan B Yeon, MD, JD, FACC

Contributor Disclosures

All topics are updated as new evidence becomes available and our peer review process is complete.

Literature review current through: May 2018. | **This topic last updated:** Apr 26, 2018.

Recent Database Articles

Below is a selection of articles recently added to the healthcare databases.

If you would like any of the following articles in full text, or if you would like a more focused search on your own topic, then get in touch: library@uhbristol.nhs.uk

1. Risque thromboembolique des adultes ayant une cardiopathie congenitale: revue de la litteratureRisk of thromboembolic complications in adult congenital heart disease: A literature review

Author(s): Karsenty C.; Marijon E.; Ladouceur M.; Zhao A.

Source: Archives of Cardiovascular Diseases; 2018

Publication Date: 2018

Publication Type(s): Article In Press

Abstract: Adult congenital heart disease (ACHD) is a constantly expanding population with challenging issues. Initial medical and surgical treatments are seldom curative, and the majority of patients still experience late sequelae and complications, especially thromboembolic events. These common and potentially life-threating adverse events are probably dramatically underdiagnosed. Better identification and understanding of thromboembolic risk factors are essential to prevent long-term related morbidities. In addition to specific situations associated with a high risk of thromboembolic events (Fontan circulation, cyanotic congenital heart disease), atrial arrhythmia has been recognized as an important risk factor for thromboembolic events in ACHD. Unlike in patients without ACHD, thromboembolic risk stratification scores, such as the CHA2DS2-VASc score, may not be applicable in ACHD. Overall, after a review of the scientific data published so far, it is clear that the complexity of the underlying congenital heart disease represents a major risk factor for thromboembolic events. As a consequence, prophylactic anticoagulation is indicated in patients with complex congenital heart disease and atrial arrhythmia, regardless of the other risk factors, as opposed to simple heart defects. The landscape of ACHD is an ongoing evolving process, and specific thromboembolic risk scores are needed, especially in the setting of simple heart defects; these should be coupled with specific trials or long-term follow-up of multicentre cohorts.

2. Incidence and Mortality of Adults With Pulmonary Hypertension and Congenital Heart Disease.

Author(s): Schwartz, Sara Søndergaard; Madsen, Nicolas; Laursen, Henning Bækgaard; Hirsch, Russel; Olsen, Morten Smærup

Source: The American journal of cardiology; Jun 2018; vol. 121 (no. 12); p. 1610-1616

Publication Date: Jun 2018

Publication Type(s): Journal Article

Abstract: Reports on pulmonary hypertension (PH) in the aging congenital heart disease (CHD) population are few and focused on arterial PH and patients with systemic-to-pulmonary shunts. Our objective was to estimate incidence and mortality of adult-onset PH in the CHD population. Using Danish nationwide registries, we identified all patients diagnosed with CHD from 1963 to 1974 and 1977 to 2012. Patients were matched 1:10 by birth year and gender with general population subjects. Between 1977 and 2013 adults >18 years of age were followed up until PH diagnosis, death, or emigration, whichever came first, using data from the Danish National Registry of Patients.

We computed cumulative incidences of PH. Using Cox regression, we compared the mortality rate between CHD subjects with and without PH matched by gender and birth year. We identified 14,860 patients with CHD. At 70 years of age, their overall cumulative incidence of PH was 7.2% (8.3% in those with systemic-to-pulmonary shunts and 5.3% in those without) compared with 0.4% in the general population. The 1-, 5-, and 10-year mortality for adults with CHD and PH was 24%, 44%, and 52%, respectively. This represented a 4-fold (95% confidence interval 3.3 to 5.6) increase in mortality compared with adults with CHD without PH after adjusting for gender, birth year, CHD severity, and presence of extracardiac defects. In conclusion the incidence of PH was substantially increased in adults with CHD relative to the general population. Of note, the increased incidence was not limited to those with a history of systemic-to-pulmonary shunts. PH was associated with increased mortality.

3. Feasibility and Outcomes in a Pilot Randomized Controlled Trial of a Psychosocial Intervention for Adults With Congenital Heart Disease.

Author(s): Kovacs, Adrienne H; Grace, Sherry L; Kentner, Amanda C; Nolan, Robert P; Silversides, Candice K; Irvine, M Jane

Source: The Canadian journal of cardiology; Jun 2018; vol. 34 (no. 6); p. 766-773

Publication Date: Jun 2018

Publication Type(s): Journal Article

Abstract:BACKGROUNDNorth American adults with congenital heart disease (CHD) are known to be at elevated risk of mood and anxiety disorders. This was the first trial of a group psychosocial intervention targeting this patient population.METHODSWithin this feasibility study, we conducted a 2-arm pilot randomized controlled trial (RCT) in which patients were randomized to Usual Care or an 8-session group psychosocial intervention (Adult Congenital Heart Disease-Coping and Resilience [ACHD-CARE]). Here, we report feasibility outcomes in accordance with published recommendations: (1) process, (2) resources, (3) management, (4) acceptability of the intervention, and (5) scientific outcomes (for which the primary outcome measures were anxiety and depression symptoms).RESULTSForty-two patients were randomized in the pilot RCT. The study was executable within a realistic timeline and revealed no significant human and data-management problems. The intervention was determined to be acceptable and highly valued by participants who participated in the ACHD-CARE program. The main challenges were practical barriers (eg, transportation, scheduling group sessions in-person given competing schedules) and retention. With regard to scientific outcomes, there were no adverse outcomes, and treatment fidelity was confirmed. Although not powered to test efficacy, there was a medium effect size (in favour of the intervention group) for depression symptoms.CONCLUSIONSWe determined it would be feasible to conduct a full-scale trial of a psychosocial intervention targeting adults with CHD, although with modifications to address practical barriers to participation. Should this intervention prove effective, a manualized intervention could be made be available.

4. Heart failure in grown-up congenital heart disease.

Author(s): Faccini, Alessia; Micheletti, Angelo; Negura, Diana G; Giugno, Luca; Butera, Gianfranco; Carminati, Mario; Giamberti, Alessandro; Chessa, Massimo

Source: Minerva cardioangiologica; Jun 2018; vol. 66 (no. 3); p. 329-336

Publication Date: Jun 2018

Publication Type(s): Journal Article

Abstract: The increasing survival to adulthood of patients with congenital heart disease (CHD) has changed the epidemiology of adult CHD (ACHD) patients and has led to an increment in hospitalization rates due to heart failure (HF). ACHD patients hospitalized for HF have a five-fold higher risk of death than those compensated. HF occurs predominantly in patients with tetralogy of Fallot, single ventricles, and after the Mustard operation for transposition of the great arteries. Diagnostic strategies applied in acquired HF patients are usually used to evaluate ACHD patients, but sometimes this can postpone the identification of HF that can become manifest with unusual and peculiar signs or symptoms. In the same way, therapeutic management resembles the acquired HF one, even if no large randomized clinical trials have been conducted in ACHD patients. Therefore, a close monitoring in dedicated units is mandatory in order to identify in time HF manifestations and manage them adequately.

5. Red cell distribution width in adults with congenital heart disease: A worldwide available and low-cost predictor of cardiovascular events.

Author(s): Baggen, Vivan J M; van den Bosch, Annemien E; van Kimmenade, Roland R; Eindhoven, Jannet A; Witsenburg, Maarten; Cuypers, Judith A A E; Leebeek, Frank W G; Boersma, Eric; Roos-Hesselink, Jolien W

Source: International journal of cardiology; Jun 2018; vol. 260 ; p. 60-65

Publication Date: Jun 2018

Publication Type(s): Journal Article

Abstract:BACKGROUNDRed cell distribution width (RDW) is a standard component of the automated blood count, and is of prognostic value in heart failure and coronary heart disease. We investigated the association between RDW and cardiovascular events in patients with adult congenital heart disease (ACHD).METHODS AND RESULTSIn this prospective cohort study, 602 consecutive patients with ACHD who routinely visited the outpatient clinic were enrolled between 2011 and 2013. RDW was measured in fresh venous blood samples at inclusion in 592 patients (median age 33 [IQR 25-41] years, 58% male, 90% NYHA I) and at four annual follow-up visits. During 4.3 [IQR 3.8-4.7] years of follow-up, the primary endpoint (death, heart failure, hospitalization, arrhythmia, thromboembolic events, cardiac intervention) occurred in 196 patients (33%). Median RDW was 13.4 (12.8-14.1)% versus 12.9 (12.5-13.4)% in patients with and without the primary endpoint (P < 0.001). RDW was significantly associated with the endpoint when adjusted for age, sex, clinical risk factors, CRP, and NT-proBNP (HR 1.20; 95% CI 1.06-1.35; P = 0.003). The C-index of the model including RDW was slightly, but significantly (P = 0.005) higher than the model without (0.74, 95% CI 0.70-0.78 versus 0.73, 95% CI 0.69-0.78). Analysis of repeated RDW measurements (n = 2449) did not show an increase in RDW prior to the occurrence of the endpoint.CONCLUSIONSRDW is associated with cardiovascular events in patients with ACHD, independently of age, sex, clinical risk factors, CRP, and NT-proBNP. This readily available biomarker could therefore be considered as an additive biomarker for risk stratification in these patients.

6. Morbidity After Cardiac Surgery in Patients With Adult Congenital Heart Disease in Comparison With Acquired Disease.

Author(s): Karangelis, Dimos; Mazine, Amine; Narsupalli, Sreekanth; Mendis, Shamarli; Veldtman, Gruschen; Nikolaidis, Nicolas

Source: Heart, lung & circulation; Jun 2018; vol. 27 (no. 6); p. 739-744

Publication Date: Jun 2018

Publication Type(s): Journal Article

Abstract:BACKGROUNDDue to the advancements in congenital cardiac surgery and interventional cardiology in the last 5 decades, more than 85% of congenital heart patients now survive to adulthood.METHODSThis retrospective study included 135 Adult Congenital Heart Disease (ACHD) patients, who had cardiac surgery at Southampton General Hospital over 3 consecutive years. We also included 42 patients with a structurally normal heart who had cardiac surgery for acquired cardiac conditions as a control group. Preoperative, intraoperative and postoperative data were analysed in both groups to identify risk factors for morbidity and mortality.RESULTSIn the ACHD group, in hospital mortality was 0.7%. In the control group no deaths were observed. Fifty-eight per cent of the ACHD patients had significantly higher perioperative morbidity with arrhythmias (26%), bleeding (3%), prolonged ventilation (11.3%) and renal replacement therapy 1.5%. In the non ACHD control group 32% (p=0.003) developed perioperative complications with arrhythmias (9.8%), bleeding (2.5%), prolonged ventilation (4.3%) and renal replacement therapy (2.5%). In ACHD patients total in-hospital stay was longer in patients with longer cardiopulmonary bypass (CPB) time (p=0.005), aortic cross clamp time (p=0.013) and higher preoperative alkaline phosphatase level (p=0.005). Early postoperative complications were higher in ACHD patients with longer cardiopulmonary bypass time (p=0.04) and presence of pulmonary artery hypertension (p=0.012).CONCLUSIONSEven though the preoperative and operative characteristics are similar to both groups, the morbidity is more in ACHD group. Longer CBP time, aortic cross clamp time and presence of pulmonary hypertension are risk factors for higher morbidity in this group.

7. Validation of the grown-ups with congenital heart disease score.

Author(s): Hörer, Jürgen; Roussin, Régine; LeBret, Emanuel; Ly, Mohamed; Abdullah, Jarrah; Marzullo, Rafaella; Pabst von Ohain, Jelena; Belli, Emre

Source: Heart (British Cardiac Society); Jun 2018; vol. 104 (no. 12); p. 1019-1025

Publication Date: Jun 2018

Publication Type(s): Journal Article

Available at Heart - from BMJ Journals - NHS

Available at Heart - from BMJ Journals

Abstract:OBJECTIVESAdults with congenital heart disease in need of heart surgery frequently present with significant comorbidity. Furthermore, additional technical difficulties often related to redo operations increase the risk for postoperative mortality and morbidity. Hence, next to the type of the procedure, additional procedure-dependent and procedure-independent factors have to be considered for risk evaluation. The recently proposed grown-ups with congenital heart disease (GUCH) mortality and morbidity scores account for these additional risk factors. We sought to validate their predictive power in a large population operated in a single centre.METHODSData of all consecutive patients aged 18 years or more, who underwent surgery for congenital heart disease between 2005 and 2016, were collected. Mortality was defined as hospital mortality or mortality within 30 days following surgery. Morbidity was defined as occurrence of one or more of the following complications: renal failure requiring dialysis, neurologic deficit persisting at discharge, atrioventricular block requiring permanent pacemaker implantation, mechanical circulatory support, phrenic nerve injury and unplanned reoperation. The discriminatory power of the GUCH scores was assessed using the area under the receiver operating characteristics curve (c-index, including 95% CI).RESULTSEight hundred and twenty-four operations were evaluated. Additional procedure-dependent and procedure-independent factors, as defined in the GUCH scores, were present in 165 patients (20.0%) and 544 patients (66.0%), respectively. Hospital mortality and morbidity was 3.4% and 10.0%, respectively. C-index for GUCH mortality score was 0.809 (0.742-0.877). C-index for

GUCH morbidity score was 0.676 (0.619-0.734).CONCLUSIONSWe could confirm the good predictive power of the GUCH mortality score for postoperative mortality in a large population of adults with congenital heart disease.

8. Illness identity: A novel predictor for healthcare use in adults with congenital heart disease

Author(s): Van Bulck L.V.; Goossens E.; Apers S.; Moons P.; Luyckx K.; Oris L.

Source: Journal of the American Heart Association; Jun 2018; vol. 7 (no. 11)

Publication Date: Jun 2018

Publication Type(s): Article

Available at Journal of the American Heart Association - from Wiley Online Library Free Content - NHS

Available at Journal of the American Heart Association - from HighWire - Free Full Text

Available at Journal of the American Heart Association - from Europe PubMed Central - Open Access

Available at Journal of the American Heart Association - from ahajournals.org

Abstract: Background--To optimize healthcare use of adults with congenital heart disease, all important predictors of healthcare utilization should be identified. Clinical and psychological characteristics (eg, age and depression) have been found to be associated with healthcare use. However, the concept of illness identity, which assesses the degree to which congenital heart disease is integrated into one's identity, has not yet been investigated in association with healthcare use. Hence, the purpose of the study is to examine the predictive value of illness identity for healthcare use. Methods and Results--In this ambispective analytical observational cohort study, 216 adults with congenital heart disease were included. The self-reported Illness Identity Questionnaire was used to assess illness identity states: engulfment, rejection, acceptance, and enrichment. After 1 year, self-reported healthcare use for congenital heart disease or other reasons over the past 6 months was assessed including hospitalizations; visits to general practitioner; visits to medical specialists; and emergency room visits. Binary logistic and negative binomial regression analyses were conducted, adjusting for age, sex, disease complexity, and depressive and anxious symptoms. The more profoundly the heart defect dominated one's identity (ie, engulfment), the more likely this person was to be hospitalized (odds ratio=3.76; 95% confidence interval=1.43-9.86), to visit a medical specialist (odds ratio=2.32; 95% confidence interval=1.35-4.00) or a general practitioner (odds ratio=1.78; 95% confidence interval=1.01-3.17), because of their heart defect. Conclusions--Illness identity, more specifically engulfment, has a unique predictive value for the occurrence of healthcare encounters. This association deserves further investigation, in which the directionality of effects and the contribution of illness identity in terms of preventing inappropriate healthcare use should be determined.

9. Efficacy and Safety of Low-Dose Amiodarone Therapy for Tachyarrhythmia in Congenital Heart Disease

Author(s): Iwasawa S.; Uyeda T.; Saito M.; Ishii T.; Inage A.; Hamamichi Y.; Yazaki S.; Yoshikawa T.

Source: Pediatric Cardiology; Jun 2018; vol. 39 (no. 5); p. 1016-1022

Publication Date: Jun 2018

Publication Type(s): Article

Abstract: Amiodarone (AMD) is a class III anti-arrhythmic drug that is highly effective for tachyarrhythmia treatment. AMD is widely used in adults with congenital heart disease (CHD); however, higher doses of AMD (> 200 mg/day) can cause various non-cardiac side effects. The

purpose of this study was to assess the efficacy, safety, and adverse events of low-dose AMD (<= 200 mg/day) for tachyarrhythmia in patients with CHD. We retrospectively studied 80 patients with CHD and tachyarrhythmia who received oral low-dose AMD (<= 200 mg/day) from January 2004 to March 2016. Low-dose AMD therapy was used to treat supraventricular tachycardia (SVT) in 51 patients and ventricular tachycardia (VT) in 29 patients. After a mean follow-up of 2.9 years for SVT and 3.2 years for VT, 36% and 65% of the patients with SVT and VT, respectively, were free from a first tachyarrhythmia recurrence for 3 years. The incidence of AMD-induced side effects was 23%, and all these cases consisted of thyroid dysfunction. Low-dose AMD was effective for the treatment of tachyarrhythmia in patients with CHD and had a relatively low incidence of side effects. These findings suggest that low-dose AMD is useful and effective for decreasing the frequency of tachyarrhythmia in patients with CHD and has a low incidence of side effects.

10. Outcomes after cardiac transplantation for adult congenital heart disease in the modern era

Author(s): Zeng X.; Vela R.; Pruszynski J.; Martinez J.; Ring S.; Peltz M.

Source: American Journal of Transplantation; Jun 2018; vol. 18 ; p. 651-652

Publication Date: Jun 2018

Publication Type(s): Conference Abstract

Abstract: Background: More patients with congenital heart disease (CHD) are surviving into adulthood and require heart transplantation. These patients historically experienced worse survival early after transplantation. The purpose of this study was to review outcomes in a center with a multidisciplinary approach to cardiac transplantation of patients with CHD. Methods: CHD patients at our center are evaluated by a multi-disciplinary team of adult and CHD trained cardiologists and surgeons. A retrospective review of all transplants between 9/2011 and 8/2017 was performed. Outcomes of patients with CHD were compared to all other diagnoses. Kaplan-Meier plots were generated and survival was compared by the log rank test. Other characteristics were analyzed using the Kruskal-Wallis and Fisher's exact tests. A p-value < .05 was considered signifi cant. Results: 14 patients with CHD out of 195 total transplants performed were identified. CHD patients underwent 2.23 (range 1-4) prior cardiac operations. There were 3 single ventricle physiology cases all treated with heart transplantation alone. 57% of CHD patients required additional reconstructive procedures at the time of transplant. Cardiopulmonary bypass and aortic cross clamp times were longer in patients with CHD consistent with the increased surgical complexity. Warm and total ischemic times were also prolonged in CHD patients but this difference did not reach statistical signifi cance. Survival was not different between groups. (Figure presented) Conclusions: Cardiac transplantation in adult patients with CHD can be technically challenging often requiring additional reconstructive procedures. However, with a specialized team, excellent outcomes are achievable.

11. Posterior Cerebellar Volume and Executive Function in Young Adults With Congenital Heart Disease.

Author(s): Semmel, Eric S; Dotson, Vonetta M; Burns, Thomas G; Mahle, William T; King, Tricia Z

Source: Journal of the International Neuropsychological Society : JINS; May 2018 ; p. 1-10

Publication Date: May 2018

Publication Type(s): Journal Article

Abstract:OBJECTIVESAs the number of adolescents and young adults (AYAs) surviving congenital heart disease (CHD) grows, studies of long-term outcomes are needed. CHD research documents poor executive function (EF) and cerebellum (CB) abnormalities in children. We examined whether

AYAs with CHD exhibit reduced EF and CB volumes. We hypothesized a double dissociation such that the posterior CB is related to EF while the anterior CB is related to motor function. We also investigated whether the CB contributes to EF above and beyond processing speed.METHODSTwenty-two AYAs with CHD and 22 matched healthy controls underwent magnetic resonance imaging and assessment of EF, processing speed, and motor function. Volumetric data were calculated using a cerebellar atlas (SUIT) developed for SPM. Group differences were compared with t tests, relationships were tested with Pearson's correlations and Fisher's r to z transformation, and hierarchical regression was used to test the CB's unique contributions to EF.RESULTSCHD patients had reduced CB total, lobular, and white matter volume (d=.52-.99) and poorer EF (d=.79-1.01) compared to controls. Significant correlations between the posterior CB and EF (r=.29-.48) were identified but there were no relationships between the anterior CB and motor function nor EF. The posterior CB predicted EF above and beyond processing speed (ps<.001).CONCLUSIONSThis study identified a relationship between the posterior CB and EF, which appears to be particularly important for inhibitory processes and abstract reasoning. The unique CB contribution to EF above and beyond processing speed alone warrants further study. (JINS, 2018, 24, 1-10).

12. Risk of thromboembolic complications in adult congenital heart disease: A literature review.

Author(s): Karsenty, Clement; Zhao, Alexandre; Marijon, Eloi; Ladouceur, Magalie

Source: Archives of cardiovascular diseases; May 2018

Publication Date: May 2018

Publication Type(s): Journal Article Review

Abstract: Adult congenital heart disease (ACHD) is a constantly expanding population with challenging issues. Initial medical and surgical treatments are seldom curative, and the majority of patients still experience late sequelae and complications, especially thromboembolic events. These common and potentially life-threating adverse events are probably dramatically underdiagnosed. Better identification and understanding of thromboembolic risk factors are essential to prevent long-term related morbidities. In addition to specific situations associated with a high risk of thromboembolic events (Fontan circulation, cyanotic congenital heart disease), atrial arrhythmia has been recognized as an important risk factor for thromboembolic events in ACHD. Unlike in patients without ACHD, thromboembolic risk stratification scores, such as the CHA2DS2-VASc score, may not be applicable in ACHD. Overall, after a review of the scientific data published so far, it is clear that the complexity of the underlying congenital heart disease represents a major risk factor for thromboembolic events. As a consequence, prophylactic anticoagulation is indicated in patients with complex congenital heart disease and atrial arrhythmia, regardless of the other risk factors, as opposed to simple heart defects. The landscape of ACHD is an ongoing evolving process, and specific thromboembolic risk scores are needed, especially in the setting of simple heart defects; these should be coupled with specific trials or long-term follow-up of multicentre cohorts.

13. Electrophysiology and structural interventions in adults with congenital heart disease: Comparison of combined versus separate procedures.

Author(s): Lindsay, Ian; Nik-Ahd, Farnoosh; Aboulhosn, Jamil A; Moore, Jeremy P

Source: Journal of cardiovascular electrophysiology; May 2018

Publication Date: May 2018

Publication Type(s): Journal Article

Abstract:BACKGROUNDElectrophysiologic (EP) and structural interventions in adult congenital heart disease (ACHD) are typically completed during separate hospital encounters. With planning/coordination, these cases can be combined.OBJECTIVESWe hypothesized that this integrated approach would yield patient and health system benefits.METHODSConsecutive ACHD patients undergoing combined interventions were matched to controls with identical but separate procedures. Primary endpoints of total hospital length of stay and cost were compared.RESULTSSixty-six combined cases and 120 controls were identified (45% male, mean age 36.2 ± 14.2 years). The most common diagnoses were Fontan (27%), tetralogy of Fallot (23%), and transposition complexes (20%). The most common EP procedure was catheter ablation (n = 30) followed by electrophysiologic study (n = 13); the most common structural intervention was transcatheter valve replacement (n = 16) followed by angioplasty/stenting (n = 14). Compared to controls, cases showed shorter anesthesia duration (323 [IQR 238-405] vs 355 minutes [270-498], p = 0.06), smaller contrast dose (130 [50-189] vs 177 mL [94-228], p = 0.045), fewer venipunctures (4 [3-4] vs 6 [5-7], p < 0.001), and fewer work days missed (2 [2-5] vs 4 [4-6], p < 0.001). There was shorter hospital stay (30 [19-35] vs 38 hours [26-50], p = 0.023) and a 37% reduction in hospital charges ($117,894 vs $187,648; p = 0.039) and 27% reduction in payments ($65,757 vs $88,859; p = 0.016), persisting after adjustment for group differences. There were no significant differences in number of complications or efficacy.CONCLUSIONSThere appear to be advantages to combining ACHD interventional procedures that include reductions in hospital length of stay and cost, without detectable difference in procedural outcome.

14. Arrhythmias in adult patients with congenital heart disease and pulmonary arterial hypertension.

Author(s): Drakopoulou, Maria; Nashat, Heba; Kempny, Aleksander; Alonso-Gonzalez, Rafael; Swan, Lorna; Wort, Stephen J; Price, Laura C; McCabe, Colm; Wong, Tom; Gatzoulis, Michael A; Ernst, Sabine; Dimopoulos, Konstantinos

Source: Heart (British Cardiac Society); May 2018

Publication Date: May 2018

Publication Type(s): Journal Article

Available at Heart (British Cardiac Society) - from BMJ Journals - NHS

Available at Heart (British Cardiac Society) - from BMJ Journals

Abstract:OBJECTIVESApproximately 5%-10% of adults with congenital heart disease (CHD) develop pulmonary arterial hypertension (PAH), which affects life expectancy and quality of life. Arrhythmias are common among these patients, but their incidence and impact on outcome remains uncertain.METHODSAll adult patients with PAH associated with CHD (PAH-CHD) seen in a tertiary centre between 2007 and 2015 were followed for new-onset atrial or ventricular arrhythmia. Clinical variables associated with arrhythmia and their relation to mortality were assessed using Cox analysis.RESULTSA total of 310 patients (mean age 34.9±12.3 years, 36.8% male) were enrolled. The majority had Eisenmenger syndrome (58.4%), 15.2% had a prior defect repair and a third had Down syndrome. At baseline, 14.2% had a prior history of arrhythmia, mostly supraventricular arrhythmia (86.4%). During a median follow-up of 6.1 years, 64 patients developed at least one new arrhythmic episode (incidence 3.47% per year), mostly supraventricular tachycardia or atrial fibrillation (78.1% of patients). Arrhythmia was associated with symptoms in 75.0% of cases. The type of PAH-CHD, markers of disease severity and prior arrhythmia were associated with arrhythmia during follow-up. Arrhythmia was a strong predictor of death, even after adjusting for demographic and clinical variables (HR 3.41, 95% CI 2.10 to 5.53, p<0.0001).CONCLUSIONSArrhythmia is common in PAH-CHD and is associated with an adverse long-term outcome, even when managed in a specialist centre.

15. It's like balancing on a slackline - A description of how adults with congenital heart disease describe themselves in relation to physical activity.

Author(s): Bay, Annika; Lämås, Kristina; Berghammer, Malin; Sandberg, Camilla; Johansson, Bengt

Source: Journal of clinical nursing; May 2018

Publication Date: May 2018

Publication Type(s): Journal Article

Abstract:AIMS AND OBJECTIVESTo illuminate how adults with CHD describe themselves in relation to physical activity.BACKGROUNDSeveral studies have shown that adults with congenital heart disease (CHD) have reduced exercise capacity and do not reach the recommended daily level of physical activity. With this in view, it is of immense importance to investigate how this population experiences physical activity.DESIGNQualitative study with semi-structured interviews analysed with qualitative content analysis.METHODSSemi-structured interviews were individually performed with fourteen adults (women=7, age 19-68 years) with complex CHD. Patients were purposively recruited from the clinic waiting list, based on a scheduled follow-up and diagnosis.RESULTSThe overall theme, It's like balancing on a slackline, illustrates how adults with CHD described themselves in relation to physical activity. This overall theme consisted of four subthemes: (1) Being an adventurer- enjoying the challenges of physical activity; (2) Being a realist- adapting to physical ability; (3) Being a non-doer- lacking prerequisites for physical activity; and (4) Being an outsider- feeling excluded depending on physical ability.CONCLUSIONSAdults with CHD seem to have a diverse relationship to physical activity and it involves various aspects throughout the lifespan. The findings point out factors that might constitute as obstacles for being physically active, specific for people with chronic conditions like CHD. This highlights the importance of further exploring the hindering and facilitating factors for being physically active in order to get a deeper understanding of how to support adults with CHD to be physically active.RELEVANCE TO CLINICAL PRACTICEGiven the diverse relationship to physical activity, nurses have to further investigate the patients' relationship to physical activity, in order to support a healthy lifestyle. Nurses and allied health professionals should offer individualized exercise prescriptions and education about suitable physical activities in relation to physical ability.

16. Pulmonary arterial hypertension in adult congenital heart disease.

Author(s): Brida, Margarita; Gatzoulis, Michael A

Source: Heart (British Cardiac Society); May 2018

Publication Date: May 2018

Publication Type(s): Journal Article Review

Available at Heart (British Cardiac Society) - from BMJ Journals - NHS

Available at Heart (British Cardiac Society) - from BMJ Journals

Abstract: Pulmonary arterial hypertension (PAH) is commonly associated with congenital heart disease (CHD) and relates to type of the underlying cardiac defects and repair history. Large systemic to pulmonary shunts may develop PAH if untreated or repaired late. PAH, when present, markedly increases morbidity and mortality in patients with CHD. Significant progress has been made for patients with Eisenmenger syndrome in pathophysiology, prognostication and disease-targeting therapy (DTT), which needs to be applied to routine patient care. Patients with PAH-CHD and systemic to pulmonary shunting may benefit from late defect closure if pulmonary vascular resistance (PVR) is still normal or near normal. Patients with PAH and coincidental defects, or previous repair of CHD should be managed as those with idiopathic PAH. Patients with a Fontan

circulation, despite not strictly fulfilling criteria for PAH, may have elevated PVR; recent evidence suggests that they may also benefit from DTT, but more data are required before general recommendations can be made. CHD-PAH is a lifelong, progressive disease; patients should receive tertiary care and benefit from a proactive DTT approach. Novel biomarkers and genetic advances may identify patients with CHD who should be referred for late defect closure and/or patients at high risk of developing PAH despite early closure in childhood. Ongoing vigilance for PAH and further controlled studies are clearly warranted in CHD.

17. An Interagency Registry for Mechanically Assisted Circulatory Support (INTERMACS) analysis of hospitalization, functional status, and mortality after mechanical circulatory support in adults with congenital heart disease.

Author(s): Cedars, Ari; Vanderpluym, Christina; Koehl, Devin; Cantor, Ryan; Kutty, Shelby; Kirklin, James K

Source: The Journal of heart and lung transplantation : the official publication of the International Society for Heart Transplantation; May 2018; vol. 37 (no. 5); p. 619-630

Publication Date: May 2018

Publication Type(s): Journal Article

Abstract:BACKGROUNDAdult congenital heart disease (ACHD) prevalence is increasing worldwide, with advanced heart failure (HF) as a leading cause of death. Limited data are available on durable mechanical circulatory support (MCS) in ACHD patients.METHODSACHD patients from the Interagency Registry for Mechanically Assisted Circulatory Support (INTERMACS) database were identified and propensity matched with non-ACHD patients using risk factors from the INTERMACS Seventh Annual Report. We compared these groups for the primary outcome of post-MCS mortality. We also investigated adverse event rates, functional status, and health-related quality of life.RESULTSACHD (n = 128) and non-ACHD (n = 512) patients were appropriately matched by baseline characteristics. ACHD patients had a longer length of stay at MCS implant (24 vs 19 days, p = 0.006) but similar rates of post-MCS adverse events and hospitalization. There were similar improvements in functional status and health related quality of life post-MCS in both groups. ACHD patients had significantly higher mortality post-MCS exclusively during the first 5 months after implant (p = 0.003) and a lower probability of receiving a transplant (p = 0.003). Risk factors for early mortality were biventricular or total artificial heart device implant and age > 50 years.CONCLUSIONSACHD patients experience a higher early mortality after MCS but have similar adverse event rates and similar improvements in functional capacity and quality of life compared with non-ACHD patients. These data support expansion of MCS use in selected ACHD patients.

18. Mechanical deformation in adult patients with unrepaired aortic coarctation.

Author(s): Avendaño-Pérez, Leonel; Soto, María Elena; Ávila-Vanzzini, Nydia; Bracamontes-Castelo, Guillermo; Ruán-Díaz, José Carlos; Alexanderson-Rosas, Erick; Espinola-Zavaleta, Nilda

Source: The international journal of cardiovascular imaging; May 2018; vol. 34 (no. 5); p. 735-741

Publication Date: May 2018

Publication Type(s): Journal Article

Abstract: Aortic coarctation is a congenital heart disease that causes an increased left ventricular afterload, resulting in increased systolic parietal tension, compensatory hypertrophy, and left ventricular systolic and diastolic dysfunction. The speckle tracking is a new echocardiographic technique that allows the detection of subclinic left ventricular systolic dysfunction. The aim of this

study was to detect early left ventricular dysfunction using mechanical deformation by echocardiography in adults with un-repaired aortic coarctation. A total of 41 subjects were studied, 20 patients with aortic coarctation and 21 control subjects, 21 women (51.2%), with an average age of 30 ± 10 years. All patients with aortic coarctation had systemic arterial hypertension ($p < 0.001$). Seventy percent (14/20) of the patients had bicuspid aortic valve. Statistically significance ($p < 0.005$) were found in left ventricular mass index, E/e ratio, pulmonary artery systolic pressure and peak velocity and maximum gradient of the aortic valve. The global longitudinal deformation of the left ventricle in patients with aortic coarctation was significative decreased, $p < 0.001$. The ejection fraction and the global longitudinal deformation of the left ventricle were significantly lower in patients with aortic coarctation compared to the control group, $p < 0.003$, $p < 0.001$, respectively. The subgroup of patients with coarctation and left ventricular ejection fraction < 55% had a marked decrease in global longitudinal strain (- 15.9 ± 4%). The radial deformation was increased in patients with aortic coarctation and showed a trend to be significant ($r = 0.421$; $p < 0.06$). A significant negative correlation was observed between the global longitudinal deformation and left ventricular mass index ($r = 0.54$; $p = 0.01$) in the aortic coarctation group. The patients with aortic coarctation and left ventricular hypertrophy had marked reduction of left ventricular global longitudinal deformation (- 16%, $p < 0.05$). In our study patients with normal left ventricular ejection fraction had abnormal global longitudinal deformation and also the increased left ventricular mass was related with a decreased left ventricular global longitudinal deformation as a sign of subclinical systolic dysfunction.

19. Evaluation of the Adult Congenital Heart Surgery Mortality Score at Two European Centers.

Author(s): Hörer, Jürgen; Belli, Emre; Roussin, Régine; LeBret, Emanuel; Ly, Mohamed; Abdullah, Jarrah; Marzullo, Raffaella; Strbad, Martina; Cleuziou, Julie; Pabst von Ohain, Jelena; Lange, Rüdiger

Source: The Annals of thoracic surgery; May 2018; vol. 105 (no. 5); p. 1441-1446

Publication Date: May 2018

Publication Type(s): Journal Article

Abstract:BACKGROUNDThe adult congenital heart surgery (ACHS) score was derived from The Society of Thoracic Surgeons Congenital Heart Surgery Database. The score was validated with data for 1,603 operations and reached a good predictive power. We sought to evaluate its predictive power for 1,654 operations performed in two European centers.METHODSData of all consecutive patients aged 18 years or more who underwent surgery for congenital heart disease between 2004 and 2013 at center 1 (n = 830) and between 2005 and 2016 at center 2 (n = 824) were collected. Mortality was defined as hospital mortality or mortality within 30 days after surgery. The discriminatory power of the ACHS score was assessed using the area under the receiver-operating characteristics curve (c-index).RESULTSDuring the examined 13-year period, 1,639 operations of 43 different procedural groups were eligible for scoring. The most frequent procedures were closure of atrial septal defect (n = 175, 10.7%), repair of partial anomalous pulmonary venous connection (n = 117, 7.1%), and aortic valve replacement (n = 112, 6.8%). Hospital mortality was 3.1%. The procedures with the highest mortality were heart transplantation (3 of 11, 27.3%), mitral valve replacement (9 of 39, 23.1%), and systemic venous stenosis repair (2 of 9, 22.2%). The c-index for the ACHS mortality score was 0.760 (0.750 in center 1 and 0.772 in center 2).CONCLUSIONSThe ACHS score reached similar, good predictive power in two different centers. The score is a useful tool to analyze surgical outcomes and to support individual decision making.

20. Congenital heart disease in adults: Assessmentof functional capacity using cardiopulmonary exercise testing.

Author(s): Aguiar Rosa, Sílvia; Agapito, Ana; Soares, Rui M; Sousa, Lídia; Oliveira, José Alberto; Abreu, Ana; Silva, Ana Sofia; Alves, Sandra; Aidos, Helena; Pinto, Fátima F; Ferreira, Rui Cruz

Source: Revista portuguesa de cardiologia : orgao oficial da Sociedade Portuguesa de Cardiologia = Portuguese journal of cardiology : an official journal of the Portuguese Society of Cardiology; May 2018; vol. 37 (no. 5); p. 399-405

Publication Date: May 2018

Publication Type(s): Journal Article

Abstract:AIMThe aim of the study was to compare functional capacity in different types of congenital heart disease (CHD), as assessed by cardiopulmonary exercise testing (CPET).METHODSA retrospective analysis was performed of adult patients with CHD who had undergone CPET in a single tertiary center. Diagnoses were divided into repaired tetralogy of Fallot, transposition of the great arteries (TGA) after Senning or Mustard procedures or congenitally corrected TGA, complex defects, shunts, left heart valve disease and right ventricular outflow tract obstruction.RESULTSWe analyzed 154 CPET cases. There were significant differences between groups, with the lowest peak oxygen consumption (VO2) values seen in patients with cardiac shunts (39% with Eisenmenger physiology) (17.2±7.1ml/kg/min, compared to 26.2±7.0ml/kg/min in tetralogy of Fallot patients; p<0.001), the lowest percentage of predicted peak VO2 in complex heart defects (50.1±13.0%) and the highest minute ventilation/carbon dioxide production slope in cardiac shunts (38.4±13.4). Chronotropism was impaired in patients with complex defects. Eisenmenger syndrome (n=17) was associated with the lowest peak VO2 (16.9±4.8 vs. 23.6±7.8ml/kg/min; p=0.001) and the highest minute ventilation/carbon dioxide production slope (44.8±14.7 vs. 31.0± 8.5; p=0.002). Age, cyanosis, CPET duration, peak systolic blood pressure, time to anaerobic threshold and heart rate at anaerobic threshold were predictors of the combined outcome of all-cause mortality and hospitalization for cardiac cause.CONCLUSIONAcross the spectrum of CHD, cardiac shunts (particularly in those with Eisenmenger syndrome) and complex defects were associated with lower functional capacity and attenuated chronotropic response to exercise.

21. Lateral thoracotomy for epicardial pacemaker placement in patients with congenital heart disease.

Author(s): Haight, Paulina J; Stewart, Robert E; Saarel, Elizabeth V; Pettersson, Gosta B; Najm, Hani K; Aziz, Peter F

Source: Interactive cardiovascular and thoracic surgery; May 2018; vol. 26 (no. 5); p. 845-851

Publication Date: May 2018

Publication Type(s): Journal Article

Abstract:OBJECTIVESOur institution adopted a lateral thoracotomy approach to epicaridal pacemaker implantation with the objective of avoiding epicardial scar tissue and to achieve adequate lead pacing and sensing. We sought to assess the short-term outcomes of this approach.METHODSA single-centre review was conducted in paediatric patients and adults with congenital heart disease or inherited arrhythmia syndromes who underwent a lateral thoracotomy for epicardial pacemaker placement from August 2010 to January 2016. Patient histories were recorded along with outcomes including complications, lead and generator performance.RESULTSTwenty-one operations were performed in 20 patients (median age 17 years, range 3 months-49 years), including 19 pacemakers and 2 implantable-cardioverter defibrillators (ICDs). Prior epicardial pacemakers had been placed in 11 (55%) patients, including 3 with multiple

pacemakers. Most patients had undergone at least 1 prior cardiac operation, with a mean of 3.2 (range 0-7) prior cardiac operations. Through our lateral thoracotomy approach, 17 of the 19 attempted atrial leads (89%) and 20 of the 20 attempted ventricular leads (100%) were successfully implanted with acceptable pacing thresholds. Complications included 1 (5%) bleeding, 2 (10%) pacemaker pocket revisions and 1 late death at 6 months unrelated to the pacemaker. There were no lead failures at a mean follow-up period of 27.5 months (range of 0.7-56.1 months).CONCLUSIONSThe lateral thoracotomy is a useful approach for epicardial pacemaker implantation in patients with congenital heart disease or inherited arrhythmia syndromes including those with multiple prior operations.

22. Risk of Dementia in Adults With Congenital Heart Disease: Population-Based Cohort Study.

Author(s): Bagge, Carina N; Henderson, Victor W; Laursen, Henning B; Adelborg, Kasper; Olsen, Morten; Madsen, Nicolas L

Source: Circulation; May 2018; vol. 137 (no. 18); p. 1912-1920

Publication Date: May 2018

Publication Type(s): Journal Article

Abstract:BACKGROUNDMore children with congenital heart disease (CHD) are surviving to adulthood, and CHD is associated with risk factors for dementia. We compared the risk of dementia in CHD adults to that of the general population.METHODSIn this cohort study, we used medical registries and a medical record review covering all Danish hospitals to identify adults with CHD diagnosed between 1963 and 2012. These individuals with CHD were followed from January 1, 1981, 30 years of age, or date of first CHD registration (index date for matched members of the general population cohort) until hospital diagnosis of dementia, death, emigration, or end of study (December 31, 2012). For each individual with CHD, we identified 10 members of the general population utilizing the Danish Civil Registration System matched on sex and birth year. We computed cumulative incidences and hazard ratios (HRs) of dementia, adjusting for sex and birth year.RESULTSThe cumulative incidence of dementia was 4% by 80 years of age in 10 632 adults with CHD (46% male). The overall HR comparing adults with CHD with the general population cohort was 1.6 (95% confidence interval [CI], 1.3-2.0). The HR among individuals with CHD without extracardiac defects was 1.4 (95% CI, 1.1-1.8). Adults with mild-to-moderate CHD had an HR of 1.5 (95% CI, 1.1-2.0), whereas the HR was 2.0 (95% CI, 1.2-3.3) for severe CHD, including univentricular hearts. The HR for early onset dementia (<65 years of age) was 2.6 (95% CI, 1.8-3.8), whereas the late-onset HR was 1.3 (95% CI, 1.0-1.8).CONCLUSIONSCHD was associated with an increased risk of dementia compared with the general population, in particular for early onset dementia. Further understanding of dementia risk in the population with CHD is a potential target for future investigation.

23. Safety and efficacy of dronedarone in adults with moderate or complex congenital heart disease

Author(s): Zimmerman F.J.; Foster A.; Gamboa D.; Shetty I.

Source: Heart Rhythm; May 2018; vol. 15 (no. 5)

Publication Date: May 2018

Publication Type(s): Conference Abstract

Abstract: Background: Antiarrhythmic therapy for adults with congenital heart disease (CHD) is often limited due to the risk of proarrhythmia or increased mortality. Dronedarone is a class III antiarrhythmic agent that has been effective for treatment of atrial fbrillation. However, there is an

increased risk of stroke, heart failure and death in select patient populations. As such, safety concerns and limited experience have led to cautious use in those with CHD. Objective: The purpose of this study was to assess the safety and effcacy of dronedarone for the treatment of atrial arrhythmias in adults with moderate or complex CHD. Methods: Adults with moderate or complex CHD treated with dronedarone for atrial arrhythmias from 2014-2017 at our center were included in this study. Arrhythmia burden, functional status (NYHA class), ECG, ECHO, and transaminase levels were evaluated before, during and after medical therapy Results: Thirteen adults (ages 21-66 yrs, mean 37 yrs, 8 females) received dronedarone (400mg BID) for an average of 1.8 yrs (range 3 mos-4 yrs) and total follow-up of 23 pt-years. Initial CHD diagnosis was single ventricle s/p Fontan (5), tetralogy of Fallot (3), dTGA s/p Mustard (2), dTGA/VSD s/p arterial switch (1), complex VSD (1) and Ebstein's anomaly (1). Arrhythmia diagnosis was non-sustained atrial tachycardia (7), intra-atrial reentry tachycardia (3), non-sustained atrial fbrillation (2) and AV node reentry tachycardia (1). Prior to starting medication, systemic ventricular function was normal in 11 and mildly reduced in 2 The functional status was NYHA class 1 in 11 and NYHA class 2 in 2. At last follow-up or at the time of stopping the medication, there was no signifcant change in systemic ventricular function, ECG parameters (PR, QRS or QTc intervals), functional status or transaminase levels. There were no exacerbations of heart failure and no deaths Atrial arrhythmia burden was decreased in 11/13 and unchanged in 2/13. There was no increase or new occurrence of atrial or ventricular arrhythmias. Conclusion: Dronedarone use in a select group of adults with moderate or complex congenital heart disease was safe and moderately effective in controlling atrial arrhythmia burden with no occurrence of proarrhythmia, exacerbation of heart failure or death.

24. Long-term outcomes of ICD implants in adult congenital heart disease patients: A single centre experience

Author(s): Sawhney V.; Whittaker-Axon S.; Daw H.; Cullen S.; VonKlemperer K.; Pandya B.; Walker F.; Lowe M.; Ezzat V.

Source: Heart Rhythm; May 2018; vol. 15 (no. 5)

Publication Date: May 2018

Publication Type(s): Conference Abstract

Abstract: Background: Sudden Cardiac Death (SCD) due to ventricular arrhythmias (VA) accounts for nearly a third of all deaths in the adult congenital heart disease (ACHD) population. Implantable cardioverter defbrillators (ICD) are effective in preventing SCD. However, there is little evidence to establish the safety and effcacy of ICDs in the ACHD population. Objective: We reviewed the indications and long-term outcomes of ICD implants in our ACHD patients. Methods: Retrospective analyses of all ACHD patients undergoing ICD implants at a single centre. All procedural data, complications and follow-up were prospectively recorded. Appropriate and inappropriate device therapy was recorded over the follow-up period. Results: Over a 5-year period, 30 patients with ACHD had ICD implants. 73% male, mean age 43 (22-67) yrs. Mean age at implant was 41 yrs. Underlying etiology was repaired tetralogy of Fallot (TOF) in 30% patients, Mustard for transposition of great arteries (TGA) in 30%, tricuspid atresia in 7% and other (ASD/VSD/coarctation of aorta/AS) in 33%. Vast majority (63%) had secondary prevention devices. Of these 5 patients had OOHCA, 3 presented with syncope and 11 had sustained VA. Acute procedural success was 90% with failed DFT requiring new lead implant in 1, failed CS lead in 1 and heamatoma due to inadvertent arterial puncture leading to a contralateral implant in 1 patient. Over a mean follow-up of 3 years, 8 patients (27%) received appropriate ICD shocks for VA. These included 4 TGA, 2 TOFs, 1 aortic coarctation and 1 Ebsteins' patient. Atrial arrhythmias were logged in 63% patients, however, the rate of inappropriate device therapy was small (1 of 30 patients). Late complications were seen in 3 patients (2 A-lead and 1-V displacements requiring re-positioning). All cause mortality over the follow-up

period was 30%. Conclusion: At our centre, the majority of ICD implants in the ACHD population are for secondary prevention and in patients with TOF or TGA. Rate of inappropriate therapy is low(<3%) and incidence of major complications (20%) smaller than that reported in the current literature. ICD implants in ACHD population come with a modest risk of complications but are effcacious with a low rate of inappropriate therapy in a carefully selected group of patients.

25. Utility of ripple mapping in complex adult congenital heart disease (ACHD) patients with intra-atrial reentry tachycardia (IART)

Author(s): Saeed Y.; Bhaskaran A.; Nayyar S.; Ha A.; Nair K.

Source: Heart Rhythm; May 2018; vol. 15 (no. 5)

Publication Date: May 2018

Publication Type(s): Conference Abstract

Abstract: Background: Electro-anatomic mapping of complex arrhythmias in ACHD patients is extremely challenging because of multiple wave fronts. These maps can be susceptible to annotation and interpolation errors Ripple Mapping (RM) is a new technique that displays data incorporating voltage and timing simultaneously as dynamic bars on the 3-D maps to overcome these limitations. Objective: To assess the utility of RM in ACHD patients with complex Intra-atrial reentry tachycardia. Methods: CARTO (Biosense Webster) mapping system was used. RM was reterospectively applied to ACHD patients with IART to create Ripple maps on the offine CARTO system. Experienced CARTO users with no RM training was presented and asked to diagnose the tachycardia circuit and identify the critical isthmus for ablation The users were blinded to conventional EP data, patient diagnosis and outcome of ablation Results: CARTO maps of IART were studied in the RM format in 7 ACHD patients The diagnoses of IART were previously confrmed and all these patients had undergone successful ablation procedure with termination of IART in all cases. Experienced CARTO users (N=5) were shown the Ripple maps of the IART frst to diagnose and localize the critical isthmus/site of ablation. They were then shown the local activation time (LAT) and propagation maps All maps were annotated using the CONFIDENSE algorithm, points were manually checked and window of interest applied with De-ponti method The assessors reached the correct diagnosis in 25/35 (72%) using RM only maps compared to 27/35 (77%) with activation mapping using LAT and propagation maps (p = NS). Combination of RM with activation mapping using LAT and propagation maps improve the diagnostic accuracy to 32/35 (91%) ($p < 0.05$). Conclusion: Combination of RM with local activation time and propagation maps can help in improving the diagnostic accuracy of arrhythmia mapping and potentially aid in catheter ablation in ACHD patients.

26. Incidence and factors associated with post-operative arrhythmias in adults with congenital heart disease undergoing cardiac surgery

Author(s): Sganga D.; Lui G.K.; Dubin A.M.; Romfh A.W.; Rogers I.S.; Axelrod D.M.; Kwiatkowski D.M.; Fernandes S.M.; Motonaga K.S.; Haeffele C.; Viswanathan M.N.; Maeda K.; Hanley F.L.; Ceresnak S.R.

Source: Heart Rhythm; May 2018; vol. 15 (no. 5)

Publication Date: May 2018

Publication Type(s): Conference Abstract

Abstract: Background: Arrhythmias are common in adults undergoing cardiac surgery, though there are limited data on the incidence and factors associated with the development of post-operative tachyarrhythmias in adults with congenital heart disease (CHD). As the number of adults with CHD continues to rise, there is a need to better identify those at risk for post-operative tachyarrhythmias.

Objective: To determine the incidence and factors associated with post-operative tachyarrhythmias in ACHD patients undergoing cardiac surgery. Methods: A retrospective cohort study of adults (>= 18 years) with CHD undergoing cardiac surgery at a single large center from 2010-2016 was performed. Patients undergoing a MAZE procedure were excluded. Pre, peri, and post-op data were obtained. Signifcant post-op tachyarrhythmias were defned as atrial futter (AFL), atrial fbrillation (AFIB), SVT, VT and NSVT. Comparisons were made between patients who developed a post-op arrhythmia (ARR) and those who did not develop an arrhythmia (NoARR). Results: 307 adult patients with CHD were included [142 males (46%), mean age 33 +/- 13 years and weight 72 +/- 18 kg] The most common diagnoses were: TOF 70 (22%), aortic valve disease 39 (13%) and Ebstein's anomaly 24 (8%). There were 9 non-arrhythmia related deaths (2.8%). There were 47 ARR patients (15%) including: 17 NSVT (6%), 13 AFIB (4%), 8 AFL (3%), 7 VT (2%), and 2 SVT (13%). Factors associated with arrhythmia development included age (ARR 42 +/- 15 vs. NoARR 31 +/- 12 years; p < 0.001), diabetes (13% vs. 2%; p < 0.01), and a prior history of arrhythmias (47% vs. 28%; p = 0.01). There was no difference in bypass time, single ventricle status, hypertension, gender, or coronary artery disease between groups. On multivariate logistic regression, only age >= 35 years (OR 3.90, CI 1.96-7.75, p < 0.01) was associated with development of a signifcant post-op tachyarrhythmia. Conclusion: Post-operative tachyarrhythmias occurred in 15% of adults undergoing surgery for CHD, with NSVT being most common (6%). Age >= 35 years was the most important factor associated with development of a signifcant post-op tachyarrhythmia.

27. Ventricular tachycardia ablation in adult congenital heart disease reduces the burden of anti-arrhythmic drugs, particularly amiodarone

Author(s): Brunnquell M.J.; Yang J.; Santangeli P.; Liang J.J.; Kay J.; Collins K.K.; Sauer W.H.; Tzou W.S.; Nguyen D.T.

Source: Heart Rhythm; May 2018; vol. 15 (no. 5)

Publication Date: May 2018

Publication Type(s): Conference Abstract

Abstract: Background: Ventricular tachycardia is one of the leading causes of morbidity and mortality for adult patients with congenital heart disease (ACHD). As ACHD patients tend to be younger, adverse effects of anti-arrhythmic drugs (AADs) may contribute signifcantly to morbidity. RF ablation for VT in ACHD is effective and may reduce the morbidity associated with AADs. Objective: We sought to evaluate the effectiveness of VT ablation at reducing the burden of AAD use in ACHD patients. Methods: In a multicenter prospective study, 48 consecutive ACHD patients undergoing VT ablation were followed. Baseline characteristics including number and type of anti-arrhythmic drugs (AAD) were collected. Procedural details, and post-procedural care, including AAD use at 6 months to 1 year post ablation, were collected. Results: Patients had a mean age of 41.5 and were 77% male. Prior to ablation, 22 out of 48 patients were on AAD (not including beta-blockers). 14 of these 22 patients were able to stop all AAD after ablation. Patients went from a mean of 0.63 AAD prior to ablation to 0.25 AAD post ablation (p = 0.008). Importantly, 8 patients were able to stop amiodarone after ablation. There was no correlation between discontinuation of AAD and recurrence of VT. Conclusion: VT ablation is safe and effective in this complex population. In the largest multicenter study of VT ablation in ACHD to date, we demonstrate that ablation can reduce the burden of AAD use, particularly allowing for the discontinuation of amiodarone. Especially vital to this young population, decreasing AAD use through ablation could decrease drug related morbidity. [Figure Presented].

28. Characterization of the ventricular tachycardia substrate in adult congenital heart disease: Results from a multicenter registry

Author(s): Yang J.; Brunnquell M.; Frankel D.S.; Tzou W.S.; Marchlinski F.E.; Liang J.J.; Nguyen D.; Sauer W.H.; Liu B.; Ruckdeschel E.S.; Santangeli P.

Source: Heart Rhythm; May 2018; vol. 15 (no. 5)

Publication Date: May 2018

Publication Type(s): Conference Abstract

Abstract: Background: Adult patients with congenital heart disease (ACHD) are at high risk of ventricular tachycardia (VT). There is limited data on the electrophysiologic characteristics in these patients. Objective: To describe the electrophysiologic characteristics in ACHD patients undergoing radiofrequency catheter ablation. Methods: We included consecutive patients with ACHD and VT ablation between 2000 and 2017 The electrophysiologic characteristics and substrate for VT were analyzed. Results: A total of 48 patients undergoing a mean of 1.21+/-0.46 ablation procedures were included. Overall, 77 different VTs were induced at initial/repeat ablation. Forty VTs were hemodynamically stable and amenable to entrainment mapping, while the remaining 37 VTs were targeted based on substrate-and pace-mapping since they were either nonsustained or hemodynamically unstable The isthmuses in Tetralogy of Fallot, ventricular septal defect and double outlet ventricle shared similarities, and substrate/linear ablation targeted 5 different isthmuses (Figure): (i) ventriculotomy-to-tricuspid annulus (10.87%); (ii) ventriculotomy-to-septal patch (13.04%); (iii) ventriculotomy-to-pulmonary annulus (26.09%); (iv) septal patch to pulmonary annulus (23.91%); (v) septal patch to tricuspid annulus (26.09%). Complete procedural success in initial ablation was achieved in 34/48 (70.8%) patients. There were no major complications. Conclusion: VTs in ACHD patients were mainly reentry-mediated and related to ventriculotomy scars or patch repairs associated with prior surgeries. Ablation in ACHD is safe and effective.

29. Atrial arrhythmias and thromboembolic complications in adults after univentricular or biventricular repair of severe congenital heart disease

Author(s): Wan D.; Tsui C.; Cheung C.; Grewal J.; Barlow A.; Kiess M.; Human D.; Krahn A.D.; Chakrabarti S.

Source: Heart Rhythm; May 2018; vol. 15 (no. 5)

Publication Date: May 2018

Publication Type(s): Conference Abstract

Abstract: Background: Adults with surgically corrected/palliated severe congenital heart disease represent one of the fastest growing groups of patients in contemporary cardiology practice. Although atrial arrhythmias (AA) are relatively common in these individuals, comprehensive data about etiology and associated thromboembolic complications in this cohort is sparse. Objective: To compare the development and progression of atrial arrhythmias and associated thromboembolic complications post-biventricular and univentricular repair of severe complex congenital heart disease in adults. Methods: Single centre observational cohort study including all consecutive patients >= 18 years of age with univentricular physiology (Fontan) and biventricular repaired Tetralogy of Fallot (ToF) who have been followed for at least 1 year at our adult congenital heart disease clinic. Results: 353 patients met inclusion criteria for this study. While diabetes and hypertension were more common in the ToF group, treatment with warfarin, digoxin and ACE inhibitors were more common in the Fontan population. Although prevalence of atrial arrhythmias (OR 3.93; 95% CI [2.15-7.19]) was more frequent in the Fontan group, there was no signifcant difference in individual type of AA: atrial fbrillation (14% vs. 10%), atrial futter (17.2% vs. 14.6%) and

ectopic atrial tachycardia (17.2% vs. 15.8%). Age (OR 1.08; 95% CI [1.05-1.11]) and treated heart failure (OR 3.94; 95% CI [1.22-12.7]) were signifcant predictors of atrial arrhythmia in multivariate analysis in either cohort. A high prevalence of thromboembolic complications (stroke, TIA, pulmonary embolism, atrial thrombus) was noted in the study population; age (OR 1.03; 95% CI [1.00-1.06]) and the presence of Fontan circulation (OR 3.62; 95% CI [1.65-7.96]) were signifcant predictors of thromboembolic events in univariate and multivariate analyses. Conclusion: Adult patients with univentricular physiology (Fontan) represent a younger population that is more severely affected by atrial arrhythmia and associated systemic thromboembolism when compared to patients with repaired biventricular physiology (ToF) despite having less hypertension or diabetes.

30. Long-term lead survival in adult congenital heart disease patients: A retrospective analysis using clinical correspondence data mining

Author(s): Stokes A.; Karadakhy O.; Kirkwood G.

Source: Heart Rhythm; May 2018; vol. 15 (no. 5)

Publication Date: May 2018

Publication Type(s): Conference Abstract

Abstract: Background: Advances in clinical management have improved life expectancy in the ACHD population; as a result, implantable cardiac devices remain in situ for longer periods than ever before. These patients pose unique challenges, including; young age at implant, multiple device procedures during their lifetime, complex venous and cardiac anatomy, and co-morbidities which may predispose to infection. In the non-ACHD population, there is useful long-term research which helps to guide clinical decision-making; however, small numbers and incomplete outcome data has lead to limited comparability against ACHD patients. Objective: This study aimed to examine 20 years of pacing experience in a large ACHD centre, using novel data-mining techniques to generate comprehensive lead survival data. Methods: A retrospective analysis was performed of pacing and defbrillator leads implanted between June 1996 and December 2016 in a 173 patient ACHD cohort. Automated text searching algorithms with manual review of over 100000 pieces of clinical correspondence was used to maximise identifcation of outcomes. Lead-specifc complications, non-elective removal and overall lead survival were compared for surgical and transvenous leads in patients with complex and non-complex ACHD anatomy. Results: A total of 340 leads with complete implant and follow-up data were identifed of which 53% had complex ACHD anatomy and 15% had tricuspid valve abnormalities. Median lead survival was 13.6 and 15.9 years in transvenous leads and surgical leads (p= n.s.), respectively. Transvenous leads were associated with a higher risk of major infection than surgically-implanted leads (11.5% vs 0% p<0.05), while surgical leads had a higher rate of pacing failure or lead damage (25.9% vs 8.0% p<0.001). Complication rates were not affected by complex ACHD anatomy or abnormal tricuspid valve function. Conclusion: In this large cohort of patients with ACHD, lead survival appears to be reasonable regardless of whether a transvenous or surgical approach is used. A higher incidence of infection in transvenous leads is offset by a higher electrical failure rate in surgical leads. Further work examining large cohorts is required to tailor pacing strategies to patient-specifc anatomy.

31. Safety and outcomes after atrial fibrillation ablation among adults with congenital heart disease of different degrees of complexity: A six-center registry study

Author(s): Liang J.J.; Frankel D.S.; Parikh V.K.; Lakkireddy D.R.; Mohanty S.; Natale A.; Szilagyi J.; Gerstenfeld E.P.; Moore J.P.; Kay J.D.; Sauer W.H.; Nguyen D.T.

Source: Heart Rhythm; May 2018; vol. 15 (no. 5)

Publication Date: May 2018

Publication Type(s): Conference Abstract

Abstract: Background: Catheter ablation is an effective treatment modality for patients with congenital heart disease (CHD). While rhythm control is often felt to be of greater importance among patients with more complex CHD, safety and outcomes of ablation in these patients remain unknown. Objective: To compare safety and effcacy of AF ablation among patients with different degrees of CHD complexity. Methods: A multicenter retrospective analysis was performed of CHD patients undergoing AF ablation. Clinical data were collected including AF and CHD type, and procedural characteristics Patients were divided into 3 groups based on CHD complexity, as defned by 2014 PACES/HRS CHD Consensus Statement: simple, moderate, and severe. One-year procedural success was defned as freedom from AF, in the absence of drugs (complete) or on previously failed drugs (partial). Results: A total of 84 CHD patients undergoing AF ablation were included (32 simple; 41 moderate, 11 severe CHD complexity). There were no signifcant differences in baseline characteristics among groups (Table). One-year complete (and including partial) success rates were 51.7% (75.8%) for simple, 55.3% (71.1%) for moderate, and 37.5% (62.5%) for severe CHD complexity (p=0.66 for complete and p=0.74 for partial freedom between groups). There were no major procedural complications. Conclusion: There are dramatic differences in the degree of CHD complexity among patients referred for AF ablation. However, when performed at experienced centers, AF ablation is both safe and effective even among patients with the most complex forms of CHD.

32. Outcome of catheter ablation of arrhythmias in adult congenital heart disease (ACHD) with and without automatic annotation with multi-electrode mapping

Author(s): Saeed Y.; Bhaskaran A.; Porta-Sanchez A.; Benson L.; Chauhan V.S.; Downar E.; Nair K.

Source: Heart Rhythm; May 2018; vol. 15 (no. 5)

Publication Date: May 2018

Publication Type(s): Conference Abstract

Abstract: Background: Catheter ablation in ACHD is challenging because of the need to map complex arrhythmias in dilated cardiac chambers. Automatic annotation with multi-electrode mapping (MEM) may be useful in these patients (pts) in terms of decreasing the procedure time and improving outcomes Objective: To compare the outcomes of catheter ablation in complex ACHD pts with and without automatic annotation of activation with MEM. Methods: Retrospective analysis of ACHD pts undergoing electrophysiological (EP) study in a single tertiary centre from 1 st November 2014 till 1 st November 2016 was performed. Two groups were compared. Group (GP-A) include ACHD pts in which automated MEM was performed. In GP-A, activation maps were created with a 20-pole PentaRay(R) catheter using the CARTO CONFIDENSE TM (Biosense Webster, Diamond Bar, CA, USA) module. Group B (GP-B) include ACHD pts with only multi-electrode mapping without the use of automated software. The CARTO mapping system was used. Results: GP-A: N=18. Mean Age 48 +/- 15 years, Male 80%. GP-B: N=15. Mean Age 44+/-14 years, Male 63%. Procedures performed included ablation of atypical atrial futter (15 pts in GP-A and 11 pts in GP-B) and ventricular tachycardia (3 pts in GP-A, 4 pts in GP-B). 33 arrhythmias were induced during the EP study in 18 pts in GP-A and 19 arrhythmias were induced in 13 pts in GP-B. 20 mapping procedures were performed with CA attempted in 19 cases in GP-A and 15 mapping procedure with CA attempted in 13 cases in GP-B The acute success rate after a single CA (defned as non inducibility of all organized arrhythmias, clinical and non-clinical) was 87.5% in GP-A and 63% in GP-B ($p < 0.05$). Mean mapping time per arrhythmia was 26 +/- 20 min in GP-A. Mean procedure time was 233 +/- 80 min in GP-A and 265 +/- 76 min in GP-B ($p < 0.05$), mean fuoroscopy time 35 +/- 13 min in GP-A and 36 +/- 25 min in GP-B (p

= NS) and mean ablation time 23 +/- 14 min in GP-A and 17 +/- 10 min in GP-B (p = NS). Arrhythmia free survival rate at follow up in all patients (17 +/- 5 months in GP-A, 24+/- 8 in GP-B) 77% in GP-A and 56% in GP-B (p < 0.05). Conclusion: Catheter ablation with an automatic wave front annotation mapping algorithm offered a signifcantly higher acute success rate, lower procedure time and higher arrhythmia free survival as compared to ablation without the use of this algorithm.

33. The peak-to-end interval of the T-wave: A risk factor for sudden cardiac death in adults with congenital heart disease

Author(s): Vehmeijer J.T.; Koyak Z.; Mulder B.; De Groot J.R.

Source: Heart Rhythm; May 2018; vol. 15 (no. 5)

Publication Date: May 2018

Publication Type(s): Conference Abstract

Abstract: Background: Adult congenital heart disease (ACHD) patients are at risk of sudden cardiac death (SCD). However, current risk stratifcation methods are not yet well-defned The Tpeak-Tend interval (TpTe) is a measure of dispersion of ventricular repolarization, and an established risk factor for SCD in non-ACHD patients. Objective: We analyzed the predictive value of TpTe for SCD in ACHD patients. Methods: From an international multicenter cohort of 25,790 ACHD patients, we identifed 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 and maximum TpTe of all 12 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 in cases vs. controls was 70+/-16 vs. 63+/-10, the maximum 97+/-2 vs. 84+/-17 and dispersion 51+/-22 vs. 41+/-16 The OR for SCD of each variable is shown in the table The OR for mean TpTe, adjusted for QRS width, heart failure symptoms and systemic ventricular impairment, was 1.7 (1.3-2.3, p <0.001) per 10 ms increase. Conclusion: Tpeak-Tend interval predicts sudden cardiac death in adults with congenital heart disease The mean TpTe and maximum TpTe and the TpTe dispersion appear to be of importance, and may add to current risk stratifcation methods for SCD in this young patient group. [Figure Presented].

34. Cardiopatia congenita em adultos: avaliacao da capacidade funcional por prova de esforco cardiorrespiratoriaCongenital heart disease in adults: Assessmentof functional capacity using cardiopulmonary exercise testing

Author(s): Aguiar Rosa S.; Agapito A.; Soares R.M.; Sousa L.; Oliveira J.A.; Abreu A.; Silva A.S.; Alves S.; Ferreira R.C.; Aidos H.; Pinto F.F.

Source: Revista Portuguesa de Cardiologia; May 2018; vol. 37 (no. 5); p. 399-405

Publication Date: May 2018

Publication Type(s): Article

Abstract: Aim: The aim of the study was to compare functional capacity in different types of congenital heart disease (CHD), as assessed by cardiopulmonary exercise testing (CPET). Methods: A retrospective analysis was performed of adult patients with CHD who had undergone CPET in a single tertiary center. Diagnoses were divided into repaired tetralogy of Fallot, transposition of the great arteries (TGA) after Senning or Mustard procedures or congenitally corrected TGA, complex defects, shunts, left heart valve disease and right ventricular outflow tract obstruction. Results: We

analyzed 154 CPET cases. There were significant differences between groups, with the lowest peak oxygen consumption (VO2) values seen in patients with cardiac shunts (39% with Eisenmenger physiology) (17.2+/-7.1 ml/kg/min, compared to 26.2+/-7.0 ml/kg/min in tetralogy of Fallot patients; p<0.001), the lowest percentage of predicted peak VO2 in complex heart defects (50.1+/-13.0%) and the highest minute ventilation/carbon dioxide production slope in cardiac shunts (38.4+/-13.4). Chronotropism was impaired in patients with complex defects. Eisenmenger syndrome (n=17) was associated with the lowest peak VO2 (16.9+/-4.8 vs. 23.6+/-7.8 ml/kg/min; p=0.001) and the highest minute ventilation/carbon dioxide production slope (44.8+/-14.7 vs. 31.0+/- 8.5; p=0.002). Age, cyanosis, CPET duration, peak systolic blood pressure, time to anaerobic threshold and heart rate at anaerobic threshold were predictors of the combined outcome of all-cause mortality and hospitalization for cardiac cause. Conclusion: Across the spectrum of CHD, cardiac shunts (particularly in those with Eisenmenger syndrome) and complex defects were associated with lower functional capacity and attenuated chronotropic response to exercise.

35. Pulmonary vasodilator therapy is associated with greater survival in Eisenmenger syndrome

Author(s): Arnott C.; Celermajer D.S.; Strange G.; Bullock A.; Kirby A.C.; O'Donnell C.; Radford D.J.; Grigg L.E.

Source: Heart; May 2018; vol. 104 (no. 9); p. 732-737

Publication Date: May 2018

Publication Type(s): Article

Available at Heart - from BMJ Journals - NHS

Available at Heart - from BMJ Journals

Abstract: Objective Eisenmenger syndrome (ES) is a severe form of pulmonary hypertension in adults with congenital heart disease (CHD) and has a poor prognosis. We aimed to understand factors associated with survival in ES and particularly to assess the potential benefits of advanced pulmonary vasodilator therapy (AT). Methods From January 2004, when AT became generally available for patients with ES, we followed 253 ES adults from 12 adult congenital heart disease centres across Australia and New Zealand. Demographic, medical and outcome data were collected and analysed prospectively and retrospectively. Results The patients with ES were predominantly female (60%), aged 31 (SD 12) years. At diagnosis of ES, 64% were WHO functional class >=3. The most common underlying lesion was ventricular septal defect (33%) with 21% having a complex' anatomy. Over a median follow-up time of 9.1 years, the majority (72%) had been prescribed at least one AT (49% single agent), mostly bosentan (66%, 168 patients). The mean time on AT was 6 (SD 3.6) years. Those on AT were more functionally impaired at presentation (69% WHO >=3 vs 51%, p=0.007) and more likely to have been prescribed anticoagulation (47% vs 27%, p=0.003). The risk of death/transplant was 4.8 %/year in AT exposed versus 8.4% in those never exposed. On multivariable analysis, exposure to AT was independently associated with greater survival (survival HR 2.27, 95% CI 1.49 to 3.45; p<0.001). WHO >=3 at presentation was associated with a worse prognosis (mortality HR 1.82, 95% CI 1.19 to 2.78; p=0.006). Conclusion Treatment with AT was independently associated with greater survival in patients with ES, even though they were comparatively sicker prior to treatment.

36. Transition Intervention for Adolescents With Congenital Heart Disease.

Author(s): Mackie, Andrew S; Rempel, Gwen R; Kovacs, Adrienne H; Kaufman, Miriam; Rankin, Kathryn N; Jelen, Ahlexxi; Yaskina, Maryna; Sananes, Renee; Oechslin, Erwin; Dragieva, Dimi;

Mustafa, Sonila; Williams, Elina; Schuh, Michelle; Manlhiot, Cedric; Anthony, Samantha J; Magill-Evans, Joyce; Nicholas, David; McCrindle, Brian W

Source: Journal of the American College of Cardiology; Apr 2018; vol. 71 (no. 16); p. 1768-1777

Publication Date: Apr 2018

Publication Type(s): Journal Article

Available at Journal of the American College of Cardiology - from ProQuest (Hospital Premium Collection) - NHS Version

Abstract:BACKGROUNDThere is little evidence regarding the efficacy of interventions to prepare adolescents with congenital heart disease (CHD) to enter adult care.OBJECTIVESThe goal of this study was to evaluate the impact of a nurse-led transition intervention on lapses between pediatric and adult care.METHODSA cluster randomized clinical trial was conducted of a nurse-led transition intervention for 16- to 17-year-olds with moderate or complex CHD versus usual care. The intervention group received two 1-h individualized sessions targeting CHD education and self-management skills. The primary outcome was excess time to adult CHD care, defined as the interval between the final pediatric and first adult cardiology appointments, minus the recommended time interval, analyzed by using Cox proportional hazards regression accounting for clustering. Secondary outcomes included scores on the MyHeart CHD knowledge survey and the Transition Readiness Assessment Questionnaire.RESULTSA total of 121 participants were randomized to receive the intervention (n = 58) or usual care (n = 63). At the recommended time of first adult appointment (excess time = 0), intervention participants were 1.8 times more likely to have their appointment within 1 month (95% confidence interval: 1.1 to 2.9; Cox regression, p = 0.018). This hazard increased with time; at an excess time of 6 months, intervention participants were 3.0 times more likely to have an appointment within 1 month (95% confidence interval: 1.1 to 8.3). The intervention group had higher scores at 1, 6, 12, and 18 months on the MyHeart knowledge survey (mixed models, p < 0.001) and the Transition Readiness Assessment Questionnaire self-management index (mixed models, p = 0.032).CONCLUSIONSA nurse-led intervention reduced the likelihood of a delay in adult CHD care and improved CHD knowledge and self-management skills. (Congenital Heart Adolescents Participating in Transition Evaluation Research [CHAPTER 2]; NCT01723332).

37. Causes of death in a contemporary adult congenital heart disease cohort.

Author(s): Yu, Christopher; Moore, Benjamin M; Kotchetkova, Irina; Cordina, Rachael L; Celermajer, David S

Source: Heart (British Cardiac Society); Apr 2018

Publication Date: Apr 2018

Publication Type(s): Journal Article

Available at Heart (British Cardiac Society) - from BMJ Journals - NHS

Available at Heart (British Cardiac Society) - from BMJ Journals

Abstract:OBJECTIVEThe life expectancy of patients with congenital heart disease (CHD) has significantly improved with advances in their paediatric medical care. Mortality patterns are changing as a result. Our study aims to describe survival and causes of death in a contemporary cohort of adult patients with CHD.METHODSWe reviewed 3068 patients in our adult CHD database (age ≥16 years, seen at least once in our centre between 2000 and 2015), and documented the number and causes of death, via Australia's National Death Index. Survival and mortality patterns were analysed by complexity of CHD and by underlying congenital diagnosis.RESULTSOur cohort comprised 3068 adult patients (53% male). The distribution of patients (per the Bethesda classification) was 47% simple, 34% moderate and 18% complex (1% not classifiable). Over a median

follow-up of 6.2 years (IQR 3.5-10.4), 341 patients (11%) died with an incidence of 0.4 deaths/100 patient years (py). Survival was significantly worse with increasing complexity of CHD (p<0.001); mortality rate in the simple group was 0.3 deaths/100 py with a median age of death 70 years, and in the complex group was 1.0 death/100 py with a median age of death 34 years. Overall, non-cardiac causes of death outnumbered cardiac causes, at 54% and 46%, respectively. The leading single cause of death was heart failure (17%), followed by malignancy (13%). Simple adult CHD patients mostly died due to non-cardiac causes such as malignancy. Perioperative mortality only accounted for 5% of deaths.CONCLUSIONSPremature death is common in adults with CHD. Although heart failure remains the most common cause of death, in the contemporary era in a specialist CHD centre, non-cardiac related deaths outnumber cardiac deaths, particularly in those with simple CHD lesions.

38. Prognostic utility of MELD-XI in adult congenital heart disease patients undergoing cardiac transplantation.

Author(s): Adams, Evan D; Jackson, Nicholas J; Young, Tim; DePasquale, Eugene C; Reardon, Leigh C

Source: Clinical transplantation; Apr 2018 ; p. e13257

Publication Date: Apr 2018

Publication Type(s): Journal Article

Abstract:BACKGROUNDModel of End-Stage Liver Disease eXcluding INR (MELD-XI) at cardiac transplant has demonstrated prognostic survival utility, but has not been specifically validated in adult congenital heart disease (ACHD) in a registry study.METHODSAdults undergoing first-time orthotopic heart transplant from 2005 to 2015 in the United Network for Organ Sharing (UNOS) registry were examined in parallel: ACHD (n = 543), ischemic-dilated cardiomyopathy (IDCM, n = 6954) and valvular heart disease (VHD, n = 355). Our primary endpoint was a composite of death, graft failure, and retransplantation assessed at 3 months (early), and those with freedom from early endpoint were reassessed at 5 years (late). Interactions between hepatorenal indices and waitlist time were examined. Secondary outcomes relating to long-term morbidity were assessed at late endpoint. Freedom from endpoint analysis in ACHD at clinically relevant endpoints was also conducted.RESULTSModel of End-Stage Liver Disease eXcluding INR score at transplant associated with an increased risk of early endpoint in all cohorts. At late endpoint, bilirubin level associated with increased risk uniquely in ACHD.CONCLUSIONSModel of End-Stage Liver Disease eXcluding INR holds prognostic application to ACHD in early time points and demonstrates unique waitlist interactions. Transplant bilirubin level may hold significance in long-term risk stratification of the ACHD population. Time on waitlist is an important consideration to contextualize these values.

39. Oral anticoagulant therapy in adults with congenital heart disease and atrial arrhythmias: Implementation of guidelines.

Author(s): Yang, H; Heidendael, J F; de Groot, J R; Konings, T C; Veen, G; van Dijk, A P J; Meijboom, F J; Sieswerda, G Tj; Post, M C; Winter, M M; Mulder, B J M; Bouma, B J

Source: International journal of cardiology; Apr 2018; vol. 257 ; p. 67-74

Publication Date: Apr 2018

Publication Type(s): Journal Article

Abstract:BACKGROUNDCurrent guidelines on oral anticoagulation (OAC) in adults with congenital heart disease (ACHD) and atrial arrhythmias (AA) consist of heterogeneous and divergent recommendations with limited level of evidence, possibly leading to diverse OAC management and

different outcomes. Therefore, we aimed to evaluate real-world implementation and outcome of three guidelines on OAC management in ACHD patients with AA.METHODSThe ESC GUCH 2010, PACES/HRS 2014 and ESC atrial fibrillation (AF) 2016 guidelines were assessed for implementation. ACHD patients with recurrent or sustained non-valvular AA from 5 tertiary centers were identified using a national ACHD registry. After two years of prospective follow-up, thromboembolism, major bleeding and death were assessed.RESULTSIn total, 225 adults (mean age 54±15years, 55% male) with various defects (simple 43%; moderate 37%; complex 20%) and AA were included. Following the most strict indication (OAC is recommended in all three guidelines), one should treat a mere 37% of ACHD patients with AA, whereas following the least strict indication (OAC is recommended in any one of the three guidelines), one should treat 98% of patients. The various guidelines were implemented in 54-80% of patients. From all recommendations, Fontan circulation, CHA2DS2-VASc≥1 and AF were independently associated with OAC prescription. Superiority of any guideline in identifying outcome (n=15) could not be demonstrated.CONCLUSIONSThe implementation of current guidelines on OAC management in ACHD patients with AA is low, probably due to substantial heterogeneity among guidelines. OAC prescription in daily practice was most consistent in patients with AF and CHA2DS2-VASc≥1 or Fontan circulation.

40. Adult Congenital Heart Disease with Pregnancy.

Author(s): Niwa, Koichiro

Source: Korean circulation journal; Apr 2018; vol. 48 (no. 4); p. 251-276

Publication Date: Apr 2018

Publication Type(s): Journal Article Review

Available at Korean Circulation Journal - from Europe PubMed Central - Open Access

Available at Korean Circulation Journal - from PubMed Central

Abstract: The number of women with congenital heart disease (CHD) at risk of pregnancy is growing because over 90% of them are grown-up into adulthood. The outcome of pregnancy and delivery is favorable in most of them provided that functional class and systemic ventricular function are good. Women with CHD such as pulmonary hypertension (Eisenmenger syndrome), severe left ventricular outflow stenosis, cyanotic CHD, aortopathy, Fontan procedure and systemic right ventricle (complete transposition of the great arteries [TGA] after atrial switch, congenitally corrected TGA) carry a high-risk. Most frequent complications during pregnancy and delivery are heart failure, arrhythmias, bleeding or thrombosis, and rarely maternal death. Complications of fetus are prematurity, low birth weight, abortion, and stillbirth. Risk stratification of pregnancy and delivery relates to functional status of the patient and is lesion specific. Medication during pregnancy and post-delivery (breast feeding) is a big concern. Especially prescribing medication with teratogenicity should be avoidable. Adequate care during pregnancy, delivery, and the postpartum period requires a multidisciplinary team approach with cardiologists, obstetricians, anesthesiologists, neonatologists, nurses and other related disciplines. Caring for a baby is an important issue due to temporarily pregnancy-induced cardiac dysfunction, and therefore familial support is mandatory especially during peripartum and after delivery. Timely pre-pregnancy counseling should be offered to all women with CHD to prevent avoidable pregnancy-related risks. Successful pregnancy is feasible for most women with CHD at relatively low risk when appropriate counseling and optimal care are provided.

41. Percutaneous pulmonary valve implantation in grown-up congenital heart disease patients: Insights from the Zurich experience.

Author(s): Oechslin, Luca; Corti, Roberto; Greutmann, Matthias; Kretschmar, Oliver; Gaemperli, Oliver

Source: Journal of interventional cardiology; Apr 2018; vol. 31 (no. 2); p. 251-260

Publication Date: Apr 2018

Publication Type(s): Journal Article

Abstract:OBJECTIVESThe aim of the study was to assess indications, procedural success, complications, echocardiographic, and clinical outcomes of percutaneous pulmonary valve implantation (PPVI) in adult patients with congenital heart disease (CHD).BACKGROUNDPPVI offers a non-surgical treatment option for failing prosthetic conduits in pulmonary position. However, efficacy and clinical outcomes after PPVI are still underreported.METHODSFrom January 2008 to March 2016, 25 adult CHD patients with right ventricular outflow tract (RVOT) stenosis and/or pulmonary regurgitation underwent PPVI in our institution. Clinical and echocardiographic data was collected at baseline, at 12 months of follow-up and yearly afterwards.RESULTSTetralogy of Fallot and repaired pulmonary atresia were among the most prevalent underlying congenital defects. Twenty-one (84%) received a Medtronic Melody® and four (16%) patients an Edwards Sapien valve prosthesis. The PPVI procedure was successful in all 25 patients. Pre-stenting was performed in all but two (8%) patients. PPVI reduced peak-to-peak pulmonary valve gradient from 43 (IQR 28-60) mmHg to 16 (IQR 14-22) mmHg ($P < 0.001$). Periprocedural complications occurred in two (8%) patients (tricuspid valve damage, pulmonary artery perforation). Over a median follow-up of 43 (IQR 18-58) months all patients were alive. Only two (8%) required re-operation and two (8%) developed stent fractures (one of them had not undergone pre-stenting). NYHA functional class improved significantly, with 20 (80%) patients in NYHA class I on follow-up.CONCLUSIONSPPVI with Medtronic Melody or Edwards Sapien valve conduits is safe and provides effective relief from right ventricular outflow tract obstruction or pulmonary regurgitation.

42. Association of Albuminuria With Major Adverse Outcomes in Adults With Congenital Heart Disease: Results From the Boston Adult Congenital Heart Biobank.

Author(s): Rajpal, Saurabh; Alshawabkeh, Laith; Almaddah, Nureddin; Joyce, Caroline M; Shafer, Keri; Gurvitz, Michelle; Waikar, Sushrut S; Mc Causland, Finnian R; Landzberg, Michael J; Opotowsky, Alexander R

Source: JAMA cardiology; Apr 2018; vol. 3 (no. 4); p. 308-316

Publication Date: Apr 2018

Publication Type(s): Journal Article

Abstract: ImportanceAlbuminuria is associated with adverse outcomes in diverse groups of patients, but the importance of albuminuria in the emerging population of increasingly complex adults with congenital heart disease (ACHD) remains unknown.ObjectiveTo assess the prevalence, risk factors, and prognostic implications of albuminuria in ACHD.Design, Setting, and ParticipantsThis prospective study assessed a cohort of ambulatory patients aged 18 years and older who were examined at an ACHD referral center and enrolled in the Boston ACHD Biobank between May 17, 2012, to August 5, 2016. Albuminuria was defined as an urine albumin-to-creatinine (ACR) ratio of 30 mg/g or more.Main Outcomes and MeasuresDeath or nonelective cardiovascular hospitalization, defined as overnight admission for heart failure, arrhythmia, thromboembolic events, cerebral hemorrhage, and/or disease-specific events.ResultsWe measured the ACR of 612 adult patients with CHD (mean [SD] age, 38.6 [13.4] years; 308 [50.3%] women). Albuminuria was present in 106 people (17.3%) and

was associated with older age (patients with ACR <30 mg/g: mean [SD]: 37.5 [13.2] years; vs patients with ACR ≥30 mg/g: 43.8 [13.1] years; P < .001), presence of diabetes mellitus (ACR <30 mg/g: 13 of 506 [2.6%]; vs ≥30 mg/g: 11 of 106 [10.4%]; P < .001), lower estimated glomerular filtration rate (ACR <30 mg/g: median [interquartile range (IQR)]: 103.3 [90.0-116.4] mL/min/1.73 m2; ACR ≥30 mg/g: 99.1 [78.8-108.7] mL/min/1.73 m2; P = .002), and cyanosis (ACR <30 mg/g: 23 of 506 [5.1%]; vs ACR ≥30 mg/g: 21 of 106 [22.6%]; P < .001). After a mean (SD) follow-up time of 270 (288) days, 17 patients (2.5%) died, while 68 (11.1%) either died or experienced overnight inpatient admission. Albuminuria predicted outcome, with 30 of 106 patients with albuminuria (28.3%) affected vs 38 of 506 patients without albuminuria (7.5%; hazard ratio [HR], 3.0; 95% CI, 1.9-4.9; P < .001). Albuminuria was also associated with increased mortality (11 of 106 [10.4%]; vs 6 of 506 [1.2%] in patients with and without albuminuria, respectively; HR, 6.4; 95% CI, 2.4-17.3; P < .001). Albuminuria was associated with the outcomes only in patients with a biventricular circulation (HR, 4.5; 95% CI, 2.5-8.0) and not those with single-ventricle circulation (HR, 1.0; 95% CI, 0.4-2.8; P = 0.01 compared with biventricular circulation group). Among 133 patients (21.7%) in NYHA functional class 2, albuminuria was strongly associated with death or nonelective hospitalization.Conclusions and RelevanceAlbuminuria is common and is associated with increased risk for adverse outcome in patients with ACHD with biventricular circulation. Albuminuria appears especially useful in stratifying risk in patients categorized as NYHA functional class 2.

43. Factors associated with coronary artery disease and stroke in adults with congenital heart disease.

Author(s): Bokma, Jouke P; Zegstroo, Ineke; Kuijpers, Joey M; Konings, Thelma C; van Kimmenade, Roland R J; van Melle, Joost P; Kiès, Philippine; Mulder, Barbara J M; Bouma, Berto J

Source: Heart (British Cardiac Society); Apr 2018; vol. 104 (no. 7); p. 574-580

Publication Date: Apr 2018

Publication Type(s): Journal Article

Available at Heart (British Cardiac Society) - from BMJ Journals - NHS

Available at Heart (British Cardiac Society) - from BMJ Journals

Abstract:OBJECTIVETo determine factors associated with coronary artery disease (CAD) and ischaemic stroke in ageing adult congenital heart disease (ACHD) patients.METHODSWe performed a multicentre case-control study, using data from the national CONgenital CORvitia (CONCOR) registry to identify ACHD patients within five participating centres. Patients with CAD were matched (1:2 ratio) with ACHD patients without CAD on age, CHD defect group and gender. Patients with ischaemic stroke (or transient ischaemic attack) were matched similarly. Medical charts were reviewed and a standardised questionnaire was used to determine presence of risk factors.RESULTSOf 6904 ACHD patients, a total of 55 cases with CAD (80% male, mean age 55.1±12.4 years) and 56 cases with stroke (46% male, mean age 46.9±15.2) were included and matched with control patients. In multivariable logistic regression analysis, traditional atherosclerotic risk factors (hypertension (OR 2.45; 95% CI 1.15 to 5.23), hypercholesterolaemia (OR 3.99; 95% CI 1.62 to 9.83) and smoking (OR 2.25; 95% CI 1.09 to 4.66)) were associated with CAD. In contrast, these risk factors were not associated with ischaemic stroke. In multivariable analysis, stroke was associated with previous shunt operations (OR 4.20; 95% CI 1.36 to 12.9), residual/unclosed septal defects (OR 2.38; 95% CI 1.03 to 5.51) and left-sided mechanical valves (OR 2.67; 95% CI 1.09 to 6.50).CONCLUSIONSTraditional atherosclerotic risk factors were associated with CAD in ACHD patients. In contrast, ischaemic stroke was related to factors (previous shunts, septal defects, mechanical valves) suggesting a cardioembolic aetiology. These findings may inform surveillance and prevention strategies.

44. Self-efficacy as a predictor of patient-reported outcomes in adults with congenital heart disease.

Author(s): Thomet, Corina; Moons, Philip; Schwerzmann, Markus; Apers, Silke; Luyckx, Koen; Oechslin, Erwin N; Kovacs, Adrienne H

Source: European journal of cardiovascular nursing : journal of the Working Group on Cardiovascular Nursing of the European Society of Cardiology; Apr 2018 ; p. 1474515118771017

Publication Date: Apr 2018

Publication Type(s): Journal Article

Abstract:OBJECTIVESelf-efficacy is a known predictor of patient-reported outcomes in individuals with acquired diseases. With an overall objective of better understanding patient-reported outcomes in adults with congenital heart disease, this study aimed to: (i) assess self-efficacy in adults with congenital heart disease, (ii) explore potential demographic and medical correlates of self-efficacy and (iii) determine whether self-efficacy explains additional variance in patient-reported outcomes above and beyond known predictors.METHODSAs part of a large cross-sectional international multi-site study (APPROACH-IS), we enrolled 454 adults (median age 32 years, range: 18-81) with congenital heart disease in two tertiary care centres in Canada and Switzerland. Self-efficacy was measured using the General Self-Efficacy (GSE) scale, which produces a total score ranging from 10 to 40. Variance in the following patient-reported outcomes was assessed: perceived health status, psychological functioning, health behaviours and quality of life. Hierarchical multivariable linear regression analysis was performed.RESULTSPatients' mean GSE score was 30.1 ± 3.3 (range: 10-40). Lower GSE was associated with female sex (p = 0.025), not having a job (p = 0.001) and poorer functional class (p = 0.048). GSE positively predicted health status and quality of life, and negatively predicted symptoms of anxiety and depression, with an additional explained variance up to 13.6%. No associations between self-efficacy and health behaviours were found.CONCLUSIONSGSE adds considerably to our understanding of patient-reported outcomes in adults with congenital heart disease. Given that self-efficacy is a modifiable psychosocial factor, it may be an important focus for interventions targeting congenital heart disease patients' well-being.

45. A model for assessment of cathetrization risk in adult (CRISA) patients

Author(s): Taggart N.; Du W.; Forbes T.; Kobayashi D.; Nykanen D.; Du Y.; Reeves J.

Source: Catheterization and Cardiovascular Interventions; Apr 2018; vol. 91

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Publication Type(s): Conference Abstract

Abstract: Background: There are limited data about the safety of cardiac catheterization on adults with CHD in pediatric catheterization laboratories. We sought to develop a multifactorial, pre-catheterization risk scoring system for adults with CHD. Methods: Data was prospectively collected using a multicenter registry of the Congenital Cardiovascular Interventional Study Consortium (CCISC). The occurrence of serious adverse events (SAE) was compared to 12 pre-defined, demographic, physiologic and procedural variables to assess the relative contribution of each factor to risk of SAE. A composite score for Catheterization RISk in Adult patients (CRISA), graded from 0-22, was derived from the relative contributions of these variables to overall risk. CRISA score was compared to the American Society of Anesthesiology (ASA) score and a consensus-derived, 20-point risk score originally applied to the CCISC registry. Results: From June 2008-September 2017, a total of 484 SAE's occurred in 300 out of a total of 7317 catheterization procedures (overall SAE rate 4.1%)

performed in adults over 18 years of age at 27 contributing centers. Eleven of the 12 variables significantly predicted SAE in univariate analysis. Three other variables were ultimately excluded from the final CRISA risk score, due to inter-correlation with other, more predictive variables. Increasing CRISA score was found to predict increasing risk of SAE and was superior to ASA or the original 20-point risk score. Minimal (CRISA score 0-2), low (3-7), moderate (8-10) and high (>=11) risk categories were identified, corresponding to 0.5%, 3.2%, 7.9%, and 16.7% risk of SAE, respectfully. Conclusion: CRISA score reliably predicts risk of SAE among adults with congenital heart disease undergoing cardiac catheterization. CRISA can be used for more individualized pre-procedural planning and patient education.

46. Adult congenital heart catheterization: Pediatric or adult facility? Trends across hospital models

Author(s): Boyer P.; Seckeler M.D.

Source: Catheterization and Cardiovascular Interventions; Apr 2018; vol. 91

Publication Date: Apr 2018

Publication Type(s): Conference Abstract

Abstract: Background: Adult survivors of congenital heart disease (ACHD), often require additional interventions. Outcomes for ACHD surgeries have been studied in Pediatric vs Adult hospitals, but caths have not. The purpose of this study was to compare outcomes and costs for ACHD caths in different hospital settings. Methods: Vizient Clinical Database/Resource Manager was queried from 10/2013 - 10/2015 for patients >=18 years with ICD-9 codes for CHD and a procedure code for diagnostic or interventional caths. Hospitals were divided into Pediatric, Adult and Mixed. Demographics, LOS, ICU admission rate, complications, in-hospital mortality and costs were compared between hospital groups using t-test and chi2. Additional comparisons were made for the most common interventional procedures: transcatheter pulmonary valve replacement, ASD closure and angioplasty. Results: Demographics, outcomes, costs and procedural comparisons are shown in the Table. Mixed hospitals had longer LOS and higher complication rates than Pediatric hospitals, but no difference from Adult hospitals. Adult hospitals had lower costs than the other groups, but there was a decreasing proportion of interventional procedures performed from Pediatric to Mixed to Adult hospitals (42%>36%>25%, respectively). Conclusion: ACHD caths can be safely performed across a variety of hospital settings, however, more complex interventions are currently performed at Pediatric hospitals. It is important to have the appropriate infrastructure to allow for safe ACHD caths. (Table Presented).

47. Transplant center volume impacts survival among ACHD patients undergoing heart transplantation - An analysis of the UNOS registry

Author(s): Menachem J.N.; Lindenfeld J.; Schlendorf K.; Shah A.S.; Bichell D.P.; Brinkley D.; Danter M.; Frischhertz B.; Keebler M.; Mettler B.; Sacks S.B.; Wigger M.; Zalawadiya S.; Book W.; Kogon B.; Rossano J.; Young T.

Source: Journal of Heart and Lung Transplantation; Apr 2018; vol. 37 (no. 4)

Publication Date: Apr 2018

Publication Type(s): Conference Abstract

Abstract: Purpose: Adult congenital heart disease (ACHD) patients undergoing heart transplant (HT) have inferior outcomes compared to non-ACHD patients. The impact of transplant center volume on outcomes in this area has not been well studied. Methods: Using the UNOS data registry, we

analyzed ACHD patients undergoing HT between 1/2000 and 12/2015. Excluding transplants in patients <18 years retransplants, and history of any solid organ transplant, center volumes were stratified into three groups: low-volume (LVC) (ref. category; <14 HT/year, n= 221), medium-volume (MVC) (14-38 HT/year, n = 362), and high-volume (HVC) (>38 HT/year, n= 244). Kaplan-Meier survival and multivariable Cox-proportional hazard analyses were performed to assess 1-year risk of mortality of transplanted ACHD patients across groups. Results: 827 ACHD patients were identified (age 36+/-13years; 60% men, 84% Caucasian). HT patients at LVC were younger (30+/-12years), had lower body mass index (23.6+/-4.9 kg/m2) and higher estimated glomerular filtration rate (eGFR 117+/-67) compared to MVC (age: 38+/-12years, BMI:25.2+/-5.5 kg/m2 and eGFR:95+/-72) and HVC (age:38+/-13years, BMI:25.2+/-5.5, eGFR:91+/-39) (p<0.01 for all). Donors at LVC- were younger (25+/-11years) with lower creatinine (1.1+/-0.7mg/dL) compared to MVC (age:28+/-11years, creatinine:1.3+/-1) (Figure presented) (Table presented) and HVC (age:31+/-12years, creatinine:1.3+/-1.3) (p<0.05 for all). Compared to LVC, patients transplanted at MVC and HVC had lower risk of 1-year mortality (adjusted hazard ratio:0.61 for MVC, 95% confidence interval (CI):0.42-0.88, p= 0.008; aHR:0.57 for HVC, 95% CI:0.38-0.87, p= 0.009). Conclusion: 1-year survival in ACHD patients undergoing HT is influenced, in part, by transplant center volume, with LVC performing poorly relative to MVC and HVC. 1-year survival of ACHD HT at MVC and HVC is approaching that of non-ACHD patients. Strategies to improve transplant outcomes in ACHD patients are needed, such as development of regional ACHD transplant centers.

48. Panel reactive antibodies and outcomes following heart transplantation in adults with congenital heart disease

Author(s): Alshawabkeh L.; Singh T.P.; Opotowsky A.R.; Landzberg M.J.; Cherikh W.S.; Kucheryavaya A.Y.; Curry M.A.; Rossano J.W.; Givertz M.M.

Source: Journal of Heart and Lung Transplantation; Apr 2018; vol. 37 (no. 4)

Publication Date: Apr 2018

Publication Type(s): Conference Abstract

Abstract: Purpose: The distribution of panel reactive antibodies (PRA) in adults with congenital heart disease (CHD) and its impact on post-transplant outcomes have not been studied. Methods: This was a retrospective cohort study from the ISHLT registry on adults (>= 18 years old) with data on PRA who underwent heart alone transplantation between July 1, 2004 and June 30, 2015. The outcome was the composite of mortality, graft failure, or acute rejection within 1 year of transplantation. Propensity score matching on age in CHD (N= 535) and non-CHD (N= 564) was performed, and rates of the composite outcome were compared across both diagnosis and PRA categories using the contingency table method with Cochran-Mantel-Haenszel test. Multivariable logistic regression was used to examine the association between the composite outcome, diagnosis group, and PRA in the presence of other independent variables (restricted cubic splines were used for continuous variables). We used multiple imputation for missing values (N= 30 imputations). Results: Among 24,236 non-CHD and 762 CHD recipients 17.7% vs. 27.5% had a PRA > 10%, respectively (p < 0.001). The rates of the composite outcome are shown in the Table. Interaction between PRA and CHD diagnosis group was not significant (p = 0.14). Multivariable analysis identified an inverse relationship between the composite outcome and adult center volume, recipient age (both p < 0.01), and donor/recipient weight ratio (p = 0.04). The risk increased with donor age, ischemic time (both p < 0.01), and serum creatinine (p = 0.02). CHD diagnosis and female gender were associated with 27% and 14% increase in the risk of the composite outcome, respectively (both p < 0.01). However, PRA was not an independent risk factor (p = 0.83). Conclusion: Although there is a higher prevalence of elevated PRA among heart transplant recipients with CHD, elevated PRA itself does not portend higher risk of the composite outcome of m(Table presented).

49. Impact of pre-procedural planning with 3D printed models on patient outcomes for ventricular assist device placement in adults with congenital heart disease: Rationale and design of a multicenter prospective registry

Author(s): Farooqi K.M.; Chelliah A.; Chai P.J.; Bacha E.A.; Saeed O.; Jorde U.P.; Einstein A.J.

Source: Journal of Heart and Lung Transplantation; Apr 2018; vol. 37 (no. 4)

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Abstract: Purpose: Mechanical circulatory support (MCS) in adults with congenital heart disease (ACHD) and heart failure (HF) remains underutilized, in part due to the highly variable anatomy and complex physiology. We describe the design and rationale of a prospective multicenter registry aimed at collecting clinical data to be compared with retrospective control data to measure changes in outcomes and utility for surgeons when 3D printed models are used for planning VAD placement. Methods: ACHD-HF patients undergoing VAD placement will be identified and clinical variables will be collected at enrollment, after VAD placement, at 30 day, 90 day and 1 year follow up. 3D imaging data will be obtained from a contrast enhanced CT scan or cardiac MRI for model creation. We have previously created 3D models for lesions most likely to develop HF including those with a systemic right ventricle (L-Transposition of the Great Arteries (TGA), Figure 1, or D-TGA after an atrial switch procedure) and Fontan palliation. Results: To date, ten centers have agreed to enroll patients. Lesion specific clinical data collected at the aforementioned time-points will be compared with retrospective data from the INTERMACS registry in ACHD-HF patients after VAD placement. To elicit improvement in the surgical experience in VAD placement using printed 3D models, questionnaires will be completed at three time points: after reviewing imaging data, after reviewing 3D model, and after VAD implantation. Conclusion: This study is designed to identify improvements in clinical parameters as compared to historical controls as well as pinpoint specific factors which are beneficial for surgical experience in VAD placement as compared to using imaging studies alone. Demonstrating these advantages may promote VAD use and potentially improve outcomes in this growing population in which MCS is underutilized (Figure presented).

50. Comparison of heart transplantation outcomes between adult congenital heart disease and matched adult cardiac patients in a single quaternary reference centre

Author(s): Kinsella A.; Alvarez J.S.; Ribeiro R.V.; Yu F.; Badiwala M.; Rao V.; Heggie J.

Source: Journal of Heart and Lung Transplantation; Apr 2018; vol. 37 (no. 4)

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Abstract: Purpose: Adult Congenital Heart Disease (ACHD) heart transplantation has historically been associated with very high perioperative risk. The perception that long-term conditional outcomes are superior to patients in other diagnostic groups has been cited as justification for their access to donor organs. We sought to compare late survival between ACHD and a cohort of non-ischemic, non-congenital adults comparable in age and year of transplantation. Methods: We retrospectively reviewed our heart transplant database and selected all congenital heart transplants performed between 1988 and 2016 (ACHD group). Thirty-five ACHD patients were transplanted during this period. Outcomes were compared to 81 patients matched by age, gender and transplanted year (control group). Ischemic patients were excluded. Results: There were no significant differences in donor age, cause of death, gender and blood type between groups. Likewise, no difference was seen

in waiting list times. Controls were more likely to be bridged to transplant with a VAD. ACHD transplantation was usually a 3rd sternotomy and 90% were either single ventricle Fontan physiology (n= 15) or sub-aortic RV physiology (n= 13). Ischemic times did not differ between groups. Length of stay was also similar. Peri-operative (30 day) survival was 93.8% for controls and 70.6% in ACHD (p= 0.001). Overall, longterm survival was also significantly lower in congenital patients (Control 1y 90%, 3y 85%, 5y 77% and 10y 63%; ACHD 1y 65%, 3y 54%, 5y 54% and 10y 54%; Log-rank test p= 0.018). Overall conditional survival 1 year post-transplant, however, was not different between groups (Control 3y 94%, 5y 86%, 10y 70%; ACHD 3y 83%, 5y 83%, 10y 83%; Log-rank test p= 0.893). Graft failure (31%) was the most common cause of death in controls, while multiple organ failure (31%) was more common in ACHD (p= 0.261). Conclusion: Transplantation in ACHD patients is notoriously high-risk, but with appropriate team structure, survival can be favourable in the modern era. In our series, patients with ACHD demonstrated an overall significantly worse short- and long-term survival when compared to a matched cohort. ACHD patients represent a known high risk population for cardiac transplantation; however, long-term conditional survival appears to be similar to matched transplant patients.

51. Marijuana in pediatric and adult congenital heart transplant listing: A survey of provider practices and attitudes

Author(s): Phillips K.A.; Thrush P.T.; Lal A.; Kindel S.J.; Castleberry C.; Sparks J.; Daly K.P.; Johnson J.N.

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Abstract: Purpose: Despite increasing legalization and use of marijuana for medical purposes, there is no consensus among pediatric heart transplant institutions or providers regarding users' eligibility for cardiac transplant. Pediatric heart transplant providers may care for both pediatric and adult congenital heart disease (ACHD) patients. Methods: We sent an anonymous, web-based survey to pediatric and ACHD transplant providers, including physicians, surgeons, transplant coordinators, and pharmacists. Survey questions focused on current institutional policies and personal opinions about marijuana use in patients being considered for heart transplantation. Results: Of the respondents, 84% practice in the U.S. and Canada, with the remaining from Europe (12%), Asia (3%) and South America (1%). Most providers (80%) care for both pediatric and ACHD patients. Respondents included cardiologists (77%), surgeons (11%), with the remaining being coordinators and pharmacists. Most providers (73%) reported their institution had no policy regarding marijuana use in heart transplant candidates. The mode of consumption is considered in listing decisions by 18% of institutions, with 87% and 53% approving of oral and transdermal routes respectively and only 7% approving of vaporized or smoked routes. While 73% of providers would consider illegal marijuana use an absolute/relative contraindication to heart transplant listing, the number decreases to 57% for legal recreational users and 21% for legal medical users. There were no significant differences in responses between centers that transplant ACHD patients and those who did not. Most providers personally believe marijuana to be physically and mentally/emotionally harmful to pediatric patients (68% and 73% respectively). Conclusion: Many institutions lack a policy regarding marijuana use in pediatric and ACHD heart transplant listing candidates, and there is considerable disagreement among providers on the best practice. With increasing legalization and use of marijuana, each institution will have to address this issue thoughtfully to continue to provide high-quality, consistent, and equitable care for pediatric and ACHD transplant candidates.

52. Outcomes in heart transplantation for adults with congenital heart disease-contemporary analysis and development of a risk model-ReDoT score

Author(s): Robinson M.R.; Oliveira G.H.; Rossano J.; Cherikh W.; Kucheryavaya A.; Stehlik J.

Source: Journal of Heart and Lung Transplantation; Apr 2018; vol. 37 (no. 4)

Publication Date: Apr 2018

Publication Type(s): Conference Abstract

Abstract: Purpose: Adults with congenital heart disease (ACHD) have traditionally been considered poor heart transplant candidates due to higher mortality peri-operatively. This early disadvantage may be counterbalanced by excellent survival past the first year after heart transplantation (HT). Yet, deciding which patients with ACHD will benefit most from HT remains challenging. In a contemporary cohort, we evaluated clinical outcomes in patients with ACHD compared to adults with ischemic (ICM) & nonischemic cardiomyopathy (NICM). We also developed & validated a risk model score to predict mortality outcomes in the ACHD population Methods: 22 070 adults in the ISHLT registry database who underwent HT between July 1 2005 and June 30 2012 were evaluated. The cohort was stratified into ACHD30%, NICM 41%,ICM 55%. Unadjusted patient survival rates within 10 years were computed by diagnosis group. The association between diagnosis & mortality within 10 years in the presence of other risk factors was examined using a multivariable Cox model. The final (ReDoT) risk score incorporates Recipient, Donor & Transplant characteristics and is a cumulative percentage scale from 0 to 100%, higher scores reflect a higher risk of 10-year mortality Results: Median age in ACHD group was 34 years; 62% males. At the time of HT, 52% were hospitalized/in ICU, 44% required inotropic support, 5% required invasive ventilation & 7% required mechanical support. The median ischemic time was 3.5 hours. ACHD and NICM had similar 10-year unadjusted survival rates (63% [58%, 67%] vs. 61% [60%, 62%], p= 0.9), but ACHD had a significantly better 10-year survival than ICM (53% [51%, 54%], p< 0.0001). This mortality difference persisted when adjustment for recipient diagnosis was made to the Cox model.Significant continuous risk factors contributing to mortality included recipient age, BMI, eGFR, donor age, & ischemic time.These significant risk factors were incorporated into the ReDoT score, which reflects hazard ratios for mortality over 10 years Conclusion: For ACHD, contemporary long-term outcomes survival post HT is comparable to those with patients with NICM and better than for ICM. The ReDoT risk score allows for risk-stratification of ACHD patients from the perspective of long-term survival at the time decisions are made regarding transplant eligibility in these patients.

53. Prognostic value of serial N-terminal pro-B-type natriuretic peptide measurements in adults with congenital heart disease

Author(s): Baggen V.J.M.; van den Bosch A.E.; Eindhoven J.A.; Witsenburg M.; Cuypers J.A.A.E.; Roos-Hesselink J.W.; Boersma E.; Baart S.J.

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Abstract: Background--A single NT-proBNP (N-terminal pro-B-type natriuretic peptide) measurement is a strong prognostic factor in adult congenital heart disease. This study investigates NT-proBNP profiles within patients with adult congenital heart disease and relates these to cardiovascular events. Methods and Results--In this prospective cohort, 602 patients with adult congenital heart disease were enrolled at the outpatient clinic (years 2011-2013). NT-proBNP was measured at study inclusion in 595 patients (median age 33 [IQR 25-41] years, 58% male, 90% NYHA I) and at subsequent annual visits. The primary end point was defined as death, heart failure, hospitalization, arrhythmia, thromboembolic event, or cardiac intervention; the secondary end point as death or heart failure. Repeated measurements were analyzed using linear mixed models and joint models. During a median follow-up of 4.4 [IQR 3.8-4.8] years, a total of 2424 repeated measurements were collected. Average NT-proBNP increase was 2.9 pmol/L the year before the primary end point (n=199, 34%) and 18.2 pmol/L before the secondary end point (n=58, 10%), compared with 0.3 pmol/L in patients who remained end point-free (P-value for difference in slope 0.006 and < 0.001, respectively). In patients with elevated baseline NTproBNP (> 14 pmol/L, n=315, 53%), repeated measurements were associated with the primary end point (HR per 2-fold higher value 2.08; 95% CI 1.31-3.87; P < 0.001) and secondary end point (HR 2.47; 95% CI 1.13-5.70; P=0.017), when adjusted for the baseline measurement. Conclusions--NT-proBNP increased before the occurrence of events, especially in patients who died or developed heart failure. Serial NT-proBNP measurements could be of additional prognostic value in the annual follow-up of patients with adult congenitive heart disease with an elevated NT-proBNP.

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