

Adult Congenital Heart Disease Evidence Update



February 2018
(Quarterly)

Respecting everyone
Embracing change
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Lunchtime Drop-in Sessions

All sessions last one hour

March (13.00-14.00)

| | |
|------------|----------------------|
| 8th (Thu) | Statistics |
| 12th (Mon) | Literature Searching |
| 20th (Tue) | Critical Appraisal |
| 28th (wed) | Statistics |

April (12.00-13.00)

| | |
|------------|----------------------|
| 5th (Thu) | Literature Searching |
| 9th (Mon) | Critical Appraisal |
| 17th (Tue) | Statistics |
| 25th (Wed) | Literature Searching |

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Contents

| | |
|---|----|
| New Additions to NICE, the Cochrane Library and UpToDate® | 3 |
| Recent Database Articles | 5 |
| Journals: Tables of Contents | 46 |
| Library Clinics | 47 |
| Library Opening Times | 48 |

New Additions to NICE, the Cochrane Library, and UpToDate®

NICE National Institute for
Health and Care Excellence

 **Cochrane**
Library

UpToDate®

OpenAthens login required. Register here: <https://openathens.nice.org.uk/>

[Pregnancy in women with congenital heart disease: Specific lesions](#)

Authors: [Carol A Waksmonski, MD](#); [Michael R Foley, MD](#)

Literature review current through: Feb 2018. | **This topic last updated:** Nov 03, 2017.



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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. A Qualitative Assessment of Adolescent Girls' Perception of Living with Congenital Heart Disease: Focusing on Future Pregnancies and Childbirth.

Author(s): Nakamura, Mayumi; Kita, Sachiko; Kikuchi, Ryota; Hirata, Yoichiro; Shindo, Takahiro; Shimizu, Nobutaka; Inuzuka, Ryo; Oka, Akira; Kamibeppu, Kiyoko

Source: Journal of pediatric nursing; 2018; vol. 38 ; p. e12

Publication Date: 2018

Publication Type(s): Journal Article

PubMedID: 29153935

Abstract: PURPOSE Congenital heart disease (CHD) is the most common birth anomaly in Japan, occurring in approximately 10.6 of every 1,000 live births. Advancements in medical and surgical care have increased births by women diagnosed with CHD. The study's purpose was to examine the perceptions of pregnancy and childbirth among adolescent girls with CHD. DESIGN AND METHOD Twelve semi-structured interviews were conducted, and the data were analyzed using a modified grounded-theory approach. RESULT Three categories and 16 subcategories were extracted. Adolescent girls with CHD reported feelings of distress and anxiety while struggling with their disease, and feared how their disease might negatively influence their future pregnancy. These concerns were related to a desire to become familiar with CHD. The girls also explored how their disease would be managed during pregnancy and childbirth. Overall, these perceptions were influenced by the girls' acceptance of their disease, and support from family, friends, and healthcare professionals. CONCLUSION Healthcare professionals might assess adolescent girls' awareness of their disease before discussing pregnancy and childbirth risks. To encourage them to understand and cope with their disease, healthcare professionals might provide interventions tailored to the timing, stage, and degree of pregnancy and childbirth awareness. This could allow safer life planning, especially concerning pregnancy and childbirth decisions. PRACTICE IMPLICATION To address adolescent girls' needs, healthcare professionals should continuously assess their awareness of pregnancy and childbirth as well as their psychological status, alongside CHD issues.

Database: Medline

2. Maternal and fetal outcome in operated vs non-operated cases of congenital heart disease cases in pregnancy.

Author(s): Yadav, Vikas; Sharma, J B; Mishra, S; Kriplani, A; Bhatla, Neerja; Kachhawa, Garima; Kumari, Rajesh; Karthik; Kriplani, Isha

Source: Indian heart journal; 2018; vol. 70 (no. 1); p. 82-86

Publication Date: 2018

Publication Type(s): Journal Article

PubMedID: 29455793

Available at [Indian Heart Journal](#) - from ScienceDirect

Abstract:OBJECTIVE To study pregnancy outcomes in operated vs non-operated cases of congenital heart disease cases during pregnancy. MATERIALS AND METHODS A total of 55 patients of congenital heart disease who delivered in the authors unit in last 10 years were taken in this retrospective study. These were divided into two groups Group 1: 29 (52.7%) patient who had no cardiac surgery and Group 2: 26 (47.2%) who had cardiac surgery to correct their cardiac defect before pregnancy. All patients were evaluated for cardiac complications and outcome during pregnancy. Obstetric complications, mode of delivery and fetal outcome was compared in the two groups using statistical analysis. RESULT The commonest lesion was atrial septal defect (ASD) seen in 22 (40%) patients followed by ventricular septal defect (VSD) in 16 (29%). Congenital valvular disease 8 (14.5%) and patent ductus arteriosus in 4 (7.2%) cases. The mean age was 25.9 ± 3.15 years in Group 1 and 26.3 ± 4.53 years in Group 2. The baseline characteristics were similar in the two groups. There was no difference in cardiac complications, NYHA deterioration and need of cardiac drugs in the two groups. Obstetric complications and mode of delivery were also similar in the two groups. Mean birth weight was 2516.65 ± 514.04 gm in Group 1 and 2683.00 ± 366.00 gm in Group 2 and was similar. APGAR < 8, stillbirth rate and other neonatal complications were also similar in two groups. CONCLUSION The maternal and fetal outcome was excellent in patients with congenital heart disease and was similar in unoperated and operated cases.

Database: Medline

3. Frequency and Consequences of Influenza Vaccination in Adults With Congenital Heart Disease

Author(s): Bare I.; Pon K.; Dehghani P.; Crawford J.; Farida N.

Source: American Journal of Cardiology; 2018

Publication Date: 2018

Publication Type(s): Article In Press

Available at [The American Journal of Cardiology](#) - from ProQuest (Hospital Premium Collection) - NHS Version

Abstract: Immunization against influenza is a critical, but perhaps underappreciated prevention of morbidity and mortality in the cardiac population. The purpose of the present study is to examine influenza vaccination rates in adults with congenital heart disease (ACHD). A secondary purpose is to explore whether there is an association between demographic, medical, and behavioral variables and receipt of the influenza vaccination. Of the 183 consecutive ACHD patients who were contacted, 123 responded to our telephone survey. Mean age was 38.4 ± 14.7 , with the most common type of lesion complexity being moderate (65.3%), followed by simple (21.0%) and severe (13.7%). Overall, 53 respondents reported undergoing influenza vaccination in the previous season. Fifty-two percent of all subjects claimed they were notified of the benefits of vaccination by their physician. Univariate analysis revealed that older age ($p = 0.006$), female gender ($p = 0.027$), perceived susceptibility to influenza illness ($p < 0.001$), perceived severity of the influenza illness ($p < 0.001$), perceived benefits of the influenza vaccination ($p < 0.001$), side effects from previous immunization ($p = 0.006$), and physician recommendation ($p = 0.008$) were predictors of receipt of influenza vaccination. On multivariate analyses, however, only side effects from previous immunization was a predictor (odds ratio = 0.34 [95% confidence interval 0.13 to 0.91]), whereas physician recommendation was numerically, but not statistically, significant (odds ratio 2.01 [95% confidence interval 0.85 to 4.78]). Our study demonstrated that less than 50% of ACHD population receives influenza vaccination. We believe educating both the patients about the side effects of vaccination and the physicians about their role in counseling ACHD patients will increase the vaccination rates in this high-risk population.

Database: EMBASE

4. Advanced Cardiovascular Magnetic Resonance Techniques in Grown-Up Congenital Heart Disease

Author(s): Valbuena-Lopez S.; Refoyo E.; Rosillo S.; Guzman G.

Source: Current Cardiovascular Imaging Reports; Apr 2018; vol. 11 (no. 4)

Publication Date: Apr 2018

Publication Type(s): Review

Abstract: Purpose: Congenital heart diseases comprise a complex population in which functional and anatomic evaluation with cardiac magnetic resonance (CMR) acquires utmost importance in the decision-making process. The use of new CMR technologies can help us understand the physiopathology and improve risk stratification. Recent Findings: Efforts towards a better understanding of the processes that underlie the occurrence of adverse events in these patients, mainly heart failure and arrhythmia, have been made in the last years. Early identification of subclinical myocardial involvement by means of detection of changes at the cellular level (fibrosis and extracellular expansion) or impaired ventricular mechanics (deformation techniques) is now possible thanks to late gadolinium enhancement/T1 mapping and strain respectively. Summary: Preliminary studies suggest that the presence of fibrosis, either replacement or diffuse fibrosis, and the detection of impaired deformation parameters or significant dyssynchrony are related to outcomes. Assessment of intracardiac and great vessels flow with 4D flow imaging is an attractive research tool that offers a comprehensive evaluation of hemodynamic status and the possibility of obtaining new promising functional parameters.

Database: EMBASE

5. Outcomes of hospitalization in adults with Fontan palliation: The Mayo Clinic experience

Author(s): Egbe A.; Khan A.R.; Al-Otaibi M.; Connolly H.M.; Said S.M.

Source: American Heart Journal; Apr 2018; vol. 198 ; p. 115-122

Publication Date: Apr 2018

Publication Type(s): Article

Abstract: The outcomes of hospitalization in the Fontan population have not been specifically studied. The purpose of this study was to describe outcomes of hospitalization (frequency and indications for hospitalization, and in-hospital mortality) in this population and to determine how these outcomes differ from those of other adults with congenital heart disease (CHD). Methods: This was a retrospective study of adult Fontan patients hospitalized at Mayo Clinic Rochester in 1990-2015. We selected age- and gender-matched control group of patients with repaired CHD and biventricular circulation hospitalized within the study period. Results: A total of 367 Fontan patients (age 31 +/- 7 years and 259 [71%] with atriopulmonary Fontan) had 853 hospital admissions in 4 years (58 hospitalizations per 100 patient-years). The most common indications were arrhythmia (n = 188, 22%), heart failure (n = 169, 20%), and cardiac surgery (n = 133, 16%). Overall in-hospital mortality was 4% (n = 38), and the highest in-hospital mortality occurred in patients hospitalized for cardiac surgery (n = 15, 11%) and heart failure (n = 13, 8%). In comparison to the repaired CHD and biventricular circulation group, the Fontan group had more frequent hospitalizations (22 vs 58 per 100 patient-years, P < .001) and higher overall in-hospital mortality (1% vs 5%, P < .001), mortality after cardiac surgery (2% vs 11%, P = .01), and mortality for heart failure-related hospitalizations (2% vs 8%, P = .04). Conclusions: Adults with Fontan palliation had more frequent hospitalization and in-hospital mortality compared to the rest of the CHD population. Arrhythmia and heart failure were the most common indications for hospitalization. Perhaps optimal management of heart failure and arrhythmia may improve outcomes in this population. **Database:** EMBASE

6. Clinical Outcomes in Adolescents and Adults After the Fontan Procedure.

Author(s): Dennis, Mark; Zannino, Diana; du Plessis, Karin; Bullock, Andrew; Disney, Patrick J S; Radford, Dorothy J; Hornung, Tim; Grigg, Leanne; Cordina, Rachael; d'Udekem, Yves; Celermajer, David S

Source: Journal of the American College of Cardiology; Mar 2018; vol. 71 (no. 9); p. 1009-1017

Publication Date: Mar 2018

Publication Type(s): Journal Article

PubMedID: 29495980

Abstract:BACKGROUND Long-term outcomes of Fontan patients who survive to age ≥ 16 years have not been well characterized. The Australian and New Zealand Fontan Registry (ANZFR) provides a unique opportunity to understand survival and complication rates in Fontan patients who transition to adult congenital heart disease centers. OBJECTIVE This study sought to describe the survival and complications of adult patients who have had a Fontan procedure. METHOD The study analyzed outcomes in patients ≥ 16 years of age who were prospectively enrolled in the ANZFR. RESULT Data from all 683 adult survivors from the ANZFR were analyzed. Mortality status was confirmed from the National Death Index. There were 201 atriopulmonary (AP) connections and 482 total cavopulmonary connections (249 lateral tunnels and 233 extracardiac conduits). For these subjects, the survival rate at age 30 years was 90% (95% CI: 87% to 93%), and it was 80% (95% CI: 75% to 87%) at 40 years of age. Survival at age 30 years was significantly worse for the patients with AP connections ($p = 0.03$). At latest follow-up, only 53% of patients were in New York Heart Association functional class I. After the age of 16 years, 136 (20%) had experienced at least 1 new arrhythmia, 42 (6%) required a permanent pacemaker, 45 (7%) had a thromboembolic event, and 135 (21%) required a surgical reintervention. Only 41% (95% CI: 33% to 51%) of Fontan patients were free of serious adverse events at 40 years of age. CONCLUSION This comprehensively followed cohort showed that a variety of morbid complications is common in Fontan adults, and that there is a substantial incidence of premature death, particularly in patients with AP connections.

Database: Medline

7. Incidence and clinical characteristics of sudden cardiac death in adult congenital heart disease.

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

Source: International journal of cardiology; Mar 2018; vol. 254 ; p. 101-106

Publication Date: Mar 2018

Publication Type(s): Journal Article

PubMedID: 29224925

Abstract:BACKGROUND The life expectancy of adults with congenital heart disease (CHD) has significantly improved in recent decades, with non-cardiovascular causes of death now competing with traditional cardiovascular causes. The risk of sudden cardiac death (SCD), a devastating event, still remains elevated above that of the general population. METHOD We reviewed 2935 patients in our adult CHD database (age ≥ 16 years, seen at least once in our centre) and documented all cases of SCD between 2000-2015. Incidence and characteristics of SCD cases by congenital defect and complexity of disease were determined. RESULT We documented 35 cases of SCD, with an incidence of 0.4 deaths/1000 patient years (py). Incidence in simple, moderate and complex congenital categories was 0.04/1000py, 0.57/1000py and 2.0/1000py respectively. The highest risk category was Eisenmenger syndrome, with an incidence of 4.8 deaths/1000py. Moderate risk lesions included

transposition of the great arteries (atrial switch surgery or congenitally corrected) and Fontan circulations. Repaired tetralogy, atrial septal defect and left ventricular outflow tract lesions were all relatively low risk. We observed a high prevalence of atrial arrhythmias (43%) and QRS prolongation (mean 132ms) in our SCD cases. **CONCLUSION** The adult CHD population remains at an elevated risk for SCD, particularly in the setting of complex underlying defects. Moderate to high risk lesions include Eisenmenger syndrome, transposition of the great arteries (atrial switch or congenitally corrected) and Fontan circulations.

Database: Medline

8. Invasive and noninvasive hemodynamic assessment in adults with Fontan palliation.

Author(s): Egbe, Alexander C; Connolly, Heidi M; Taggart, Nathaniel W; Al-Otaibi, Mohamad; Borlaug, Barry A

Source: International journal of cardiology; Mar 2018; vol. 254 ; p. 96-100

Publication Date: Mar 2018

Publication Type(s): Journal Article

PubMedID: 29229372

Abstract: **BACKGROUND/OBJECTIVES** Although echocardiographic-Doppler cardiac index (CI) assessment is widely used to guide heart failure management in patients with biventricular circulation, this application has not been studied in the Fontan population. The objective of this study was to: (1) determine the correlation between Doppler and cardiac catheterization CI calculation; (2) determine the association between Doppler CI and the occurrence of Fontan failure. **METHODS** Retrospective review of adult Fontan patients followed at Mayo Clinic Adult Congenital Heart Disease program, 1994-2015. Inclusion criteria were: systemic left ventricle and echocardiogram and cardiac catheterization performed within the same week. Fontan failure was defined as a composite of all-cause mortality, heart transplantation listing, and palliative care. **RESULTS** 59 patients (age 29 ± 6 years; men 32 [54%]) underwent 97 studies. Of the 59, 41 [69%] had atriopulmonary Fontan and 12 (20%) had cirrhosis. Compared to patients without cirrhosis, patients with cirrhosis had higher Doppler CI (3.6 ± 0.6 vs 2.8 ± 0.4 L/min \cdot m², $p=0.039$); Fick CI (3.3 [2.5-3.7] vs 2.4 [1.6-3.1] L/min/m², $p=0.028$); lower systemic vascular resistance (20 ± 3 vs 25 ± 4 WU \cdot m², $p=0.04$). There was a positive correlation between Doppler and Fick CI ($r=0.52$; $p<0.0001$). Fontan failure occurred in 13 patients (22%) within 7.5 ± 2.1 years. In patients without cirrhosis, Fick CI and Doppler CI <2.5 L/min/m² were associated with Fontan failure (odds ratio [OR] 1.58, $p=0.046$) and (OR 1.43, $p=0.051$) respectively. **CONCLUSIONS** Doppler CI assessment is feasible in a selected group of Fontan patients and it is predictive of clinical outcomes. The application of this concept in systemic right ventricles deserves further research.

Database: Medline

9. Retrospective UK multicentre study of the pregnancy outcomes of women with a Fontan repair.

Author(s): Cauldwell, Matthew; Steer, Philip J; Bonner, Samantha; Asghar, Omar; Swan, Lorna; Hodson, Kenneth; Head, Catherine E G; Jakes, Adam Daniel; Walker, Nicola; Simpson, Margaret; Bolger, Aidan P; Siddiqui, Farah; English, Katherine M; Maudlin, Lucy; Abraham, Dilip; Sands, Andrew J; Mohan, Aarthi R; Curtis, Stephanie L; Coats, Louise; Johnson, Mark R

Source: Heart (British Cardiac Society); Mar 2018; vol. 104 (no. 5); p. 401-406

Publication Date: Mar 2018

Publication Type(s): Journal Article

PubMedID: 28954835

Available at [Heart](#) - from BMJ Journals - NHS

Available at [Heart](#) - from BMJ Journals

Abstract:BACKGROUNDThe population of women of childbearing age palliated with a Fontan repair is increasing. The aim of this study was to describe the progress of pregnancy and its outcome in a cohort of patients with a Fontan circulation in the UK.METHODSA retrospective study of women with a Fontan circulation delivering between January 2005 and November 2016 in 10 specialist adult congenital heart disease centres in the UK.RESULTS50 women had 124 pregnancies, resulting in 68 (54.8%) miscarriages, 2 terminations of pregnancy, 1 intrauterine death (at 30 weeks), 53 (42.7%) live births and 4 neonatal deaths. Cardiac complications in pregnancies with a live birth included heart failure (n=7, 13.5%), arrhythmia (n=6, 11.3%) and pulmonary embolism (n=1, 1.9%). Very low baseline maternal oxygen saturations at first obstetric review were associated with miscarriage. All eight women with saturations of less than 85% miscarried, compared with 60 of 116 (51.7%) who had baseline saturations of $\geq 85\%$ ($p=0.008$). Obstetric and neonatal complications were common: preterm delivery (n=39, 72.2%), small for gestational age (<10th percentile, n=30, 55.6%; <5th centile, n=19, 35.2%) and postpartum haemorrhage (n=23, 42.6%). There were no maternal deaths in the study period.CONCLUSIONWomen with a Fontan circulation have a high rate of miscarriage and, even if pregnancy progresses to a viable gestational age, a high rate of obstetric and neonatal complications.

Database: Medline

10. Prognostic value of galectin-3 in adults with congenital heart disease.

Author(s): Baggen, Vivan J M; van den Bosch, Annemien E; Eindhoven, Jannet A; Menting, Myrthe E; Witsenburg, Maarten; Cuypers, Judith A A E; Boersma, Eric; Roos-Hesselink, Jolien W

Source: Heart (British Cardiac Society); Mar 2018; vol. 104 (no. 5); p. 394-400

Publication Date: Mar 2018

Publication Type(s): Journal Article

PubMedID: 28942393

Available at [Heart](#) - from BMJ Journals - NHS

Available at [Heart](#) - from BMJ Journals

Abstract:OBJECTIVEGalectin-3 is an emerging biomarker for risk stratification in patients with heart failure. This study aims to investigate the release of galectin-3 and its association with cardiovascular events in patients with adult congenital heart disease (ACHD).METHODSIn this prospective cohort study, 602 consecutive patients with ACHD who routinely visited the outpatient clinic were enrolled between 2011 and 2013. Galectin-3 was measured in thaw serum by batch analysis. The association between galectin-3 and a primary endpoint of all-cause mortality, heart failure, hospitalisation, arrhythmia, thromboembolic events and cardiac interventions was investigated using multivariable Cox models. Reference values and reproducibility were established by duplicate galectin-3 measurements in 143 healthy controls.RESULTSGalectin-3 was measured in 591 (98%) patients (median age 33 (25-41) years, 58% male, 90% New York Heart Association (NYHA) class I). Median galectin-3 was 12.7 (range 4.2-45.7) ng/mL and was elevated in 7% of patients. Galectin-3 positively correlated with age, cardiac medication use, NYHA class, loss of sinus rhythm, cardiac dysfunction and N-terminal pro-B-type natriuretic peptide (NT-proBNP). During a median follow-up of 4.4 (IQR 3.9-4.8) years, the primary endpoint occurred in 195 patients (33%). Galectin-3 was significantly

associated with the primary endpoint in the univariable analysis (HR per twofold higher value 2.05; 95% CI 1.44 to 2.93, $p < 0.001$). This association was negated after adjustment for NT-proBNP (HR 1.04; 95% CI 0.72 to 1.49, $p = 0.848$). **CONCLUSIONS** Galectin-3 is significantly associated with functional capacity, cardiac function and adverse cardiovascular events in patients with ACHD. Nevertheless, the additive value of galectin-3 to a more conventional risk marker such as NT-proBNP seems to be limited.

Database: Medline

11. Do not forget the parents-Parents' concerns during transition to adult care for adolescents with congenital heart disease.

Author(s): Bratt, E L; Burström, Å; Hanseus, K; Rydberg, A; Berghammer, M; On behalf on the STEPSTONES-CHD consortium

Source: Child: care, health and development; Mar 2018; vol. 44 (no. 2); p. 278-284

Publication Date: Mar 2018

Publication Type(s): Journal Article

PubMedID: 28980341

Abstract: **BACKGROUND** Growing up with congenital heart disease (CHD) often means transfer to adult care and lifelong medical follow-up. An optimal transition process usually involves a multipart collaboration between the patient, their parents and other family members, and the healthcare providers. Taking an active role while knowing when it is time to step aside can be difficult for all the concerned parties, even the healthcare professionals. The aim of the present study therefore, was to explore parents' expectations and needs during their adolescent's transition to adult care. **METHODS** Semi-structured interviews were conducted with 18 parents of 16 adolescents (aged 13-18 years) with CHD in 4 pediatric cardiology settings in Sweden. The interviews were analysed with qualitative content analysis. **RESULTS** The analysis resulted in 2 main themes: (a) Feeling secure- the importance of being prepared and informed. This theme focused on the need to be prepared and informed about transition and future transfer to adult care. (b) Recognizing when to hand over at the right time. This theme addressed the process of handing over the responsibility from the parent to the adolescents and contained handing over from pediatric care to adult care. **CONCLUSION** Being prepared and informed about the upcoming transition process was essential. The parents underlined the importance of being involved in the transition planning for gradually handing over responsibility to the adolescent. They also considered establishing contact with the adult healthcare team before transfer as important and needed to be assured that CHD-related information of importance for the young person's daily life would be given.

Database: Medline

12. Exercise self-efficacy in adults with congenital heart disease

Author(s): Bay A.; Sandberg C.; Johansson B.; Wadell K.; Thilen U.

Source: IJC Heart and Vasculature; Mar 2018; vol. 18 ; p. 7-11

Publication Date: Mar 2018

Publication Type(s): Article

Available at [International journal of cardiology. Heart & vasculature](#) - from Europe PubMed Central - Open Access

Available at [International journal of cardiology. Heart & vasculature](#) - from PubMed Central

Abstract:Background Physical activity improves health, exercise tolerance and quality of life in adults with congenital heart disease (CHD), and exercise training is in most patients a high-benefit low risk intervention. However, factors that influence the confidence to perform exercise training, i.e. exercise self-efficacy (ESE), in CHD patients are virtually unknown. We aimed to identify factors related to low ESE in adults with CHD, and potential strategies for being physically active. Methods Seventy-nine adults with CHD; 38 with simple lesions (16 women) and 41 with complex lesions (17 women) with mean age 36.7 +/- 14.6 years and 42 matched controls were recruited. All participants completed questionnaires on ESE and quality of life, carried an activity monitor (Actiheart) during four consecutive days and performed muscle endurance tests. Results ESE in patients was categorised into low, based on the lowest quartile within controls, (≤ 29 points, $n = 34$) and high (> 29 points, $n = 45$). Patients with low ESE were older (42.9 ± 15.1 vs. 32.0 ± 12.4 years, $p = 0.001$), had more complex lesions (65% vs. 42%, $p = 0.05$) more often had New York Heart Association functional class III (24% vs. 4%, $p = 0.01$) and performed fewer shoulder flexions (32.5 ± 15.5 vs. 47.7 ± 25.0 , $p = 0.001$) compared with those with high ESE. In a logistic multivariate model age (OR; 1.06, 95% CI 1.02-1.10), and number of shoulder flexions (OR; 0.96, 95% CI 0.93-0.99) were associated with ESE. Conclusion In this study we show that many adults with CHD have low ESE. Age is an important predictor of low ESE and should, therefore, be considered in counselling patients with CHD. In addition, muscle endurance training may improve ESE, and thus enhance the potential for being physically active in this population.

Database: EMBASE

13. Risks and outcomes of direct current cardioversion in children and young adults with congenital heart disease.

Author(s): Morello, Melissa L; Khoury, Philip R; Knilans, Timothy K; Veldtman, Gruschen; Spar, David S; Anderson, Jeffery B; Czosek, Richard J

Source: Pacing and clinical electrophysiology : PACE; Feb 2018

Publication Date: Feb 2018

Publication Type(s): Journal Article

PubMedID: 29476621

Abstract:OBJECTIVESEvaluate the efficacy, outcomes and complications associated with DCCV in the treatment of arrhythmias in pediatric and adult CHD populations and identify patient and procedural characteristics associated with adverse events.BACKGROUNDPediatric and adult patients with congenital heart disease (CHD) are at risk of atrial arrhythmias. Direct current cardioversion (DCCV) is effective but is associated with potential complications.METHODSIn this single-center retrospective series, patients undergoing DCCV Jan 2010-May 2015 were identified and categorized as pediatric (< 18 years). Records were reviewed for demographic, arrhythmic and CHD-specific characteristics; acute efficacy; and 3-month arrhythmia recurrence. Complications were categorized as life-threatening (LT) or non-life-threatening (NLT). Univariate followed by multiple-variable and logistic regression analyses were used to identify characteristics associated with complications.RESULTSWe identified 104 patients with 152 discrete DCCV events, median age 17.4 years (0.15-62.2). DCCV efficacy was 89% with 3-month recurrence of 46%. There were 52 complications amongst 24 patients, median 17.7 years (0.15-49). Risks associated with NLT complications: moderate-severe systolic dysfunction (8/152 encounters, $p = 1$ shock per DCCV encounter ($p = 18$ and associated NLT complication. Adults had more frequent arrhythmia recurrence within 3 months than children ($p < 0.01$).CONCLUSIONS DCCV is effective for arrhythmias but is associated with frequent recurrence particularly in adult patients. Complications

associated with DCCV may be greater than previously reported. Additional support and precautions should be in place for those at greatest risk.

Database: Medline

14. Clinical Characteristics of Adult Patients With Congenital Heart Disease Hospitalized for Acute Heart Failure.

Author(s): Negishi, Jun; Ohuchi, Hideo; Miyazaki, Aya; Tsuda, Etsuko; Shiraishi, Isao; Kurosaki, Kenichi

Source: Circulation journal : official journal of the Japanese Circulation Society; Feb 2018; vol. 82 (no. 3); p. 840-846

Publication Date: Feb 2018

Publication Type(s): Journal Article

PubMedID: 29311517

Abstract:BACKGROUNDHeart failure (HF) is an important complication in adults with congenital heart disease (CHD), but because only a few studies have focused on acute HF hospitalization in adults with CHD, we study aimed to define the clinical characteristics of such patients and examine the differences in acute HF between adults with CHD and acquired heart disease.Methods and Results:We retrospectively evaluated 50 adults with CHD admitted for treatment of acute HF and compared their data with those from Japanese HF registries. Patient mean age was 37±15 years and 58% were male. In total, 86% of the patients had complex forms of CHD and 62% had undergone corrective surgery, including the Fontan procedure; 66% of patients showed right heart hemodynamic abnormality. In-hospital mortality was 4%, which was comparable to the Japanese HF registries. Survival rate was 93% at 1 year and 75% at 3 years, which was similarly poor to the rates of HF secondary to acquired heart disease.CONCLUSIONSWe clarified the clinical characteristics of adults with CHD requiring HF hospitalization. Young adults with complex CHD were hospitalized for management of acute right HF. Short-term and mid-term outcomes were similarly poor compared with acute HF secondary to acquired heart disease.

Database: Medline

15. A multinational observational investigation of illness perceptions and quality of life among patients with a Fontan circulation.

Author(s): Holbein, Christina E; Fogleman, Nicholas D; Hommel, Kevin; Apers, Silke; Rassart, Jessica; Moons, Philip; Luyckx, Koen; Sluman, Maayke A; Enomoto, Junko; Johansson, Bengt; Yang, Hsiao-Ling; Dellborg, Mikael; Subramanyan, Raghavan; Jackson, Jamie L; Budts, Werner; Kovacs, Adrienne H; Morrison, Stacey; Tomlin, Martha; Gosney, Kathy; Soufi, Alexandra; Eriksen, Katrine; Thomet, Corina; Berghammer, Malin; Alday, Luis; Callus, Edward; Fernandes, Susan M; Caruana, Maryanne; Menahem, Samuel; Cook, Stephen C; Rempel, Gwen R; White, Kamila; Khairy, Paul; Kutty, Shelby; Veldtman, Gruschen; APPROACH-IS consortium and the International Society for Adult Congenital Heart Disease (ISACHD)

Source: Congenital heart disease; Feb 2018

Publication Date: Feb 2018

Publication Type(s): Journal Article

PubMedID: 29457362

Abstract:OBJECTIVEFirst, to compare QOL and illness perceptions between patients with a Fontan circulation and patients with anatomically simple defects (ie, atrial septal defects [ASD] or ventricular septal defects [VSD]). Second, to explore illness perceptions as a mediator of the association between congenital heart disease (CHD) diagnosis and QOL.DESIGNCross-sectional observational study.SETTINGTwenty-four cardiology centers from 15 countries across five continents.PATIENTSFour hundred thirty-five adult patients with congenital heart disease (177 Fontan and 258 ASD/VSD) ages 18-83 years.OUTCOME MEASURESQOL and illness perceptions were assessed by the Satisfaction With Life Scale and the Brief Illness Perceptions Questionnaire, respectively.RESULTSPatients with a Fontan circulation reported lower QOL (Wald $Z = -3.59$, $p < .001$) and more negative perceptions of their CHD (Wald $Z = -7.66$, $p < .001$) compared with patients with ASD/VSD. After controlling for demographics, anxiety, depressive symptoms, and New York Heart Association functional class, path analyses revealed a significant mediation model, $\alpha\beta = 0.15$, $p = .002$, 95% CI = 0.06-0.25, such that CHD diagnosis was indirectly related to QOL through illness perceptions.CONCLUSIONSThe Fontan sample's more negative perceptions of CHD were likely a reflection of life with a more complex defect. Illness perceptions appear to account for unique differences in QOL between groups of varying CHD complexity. Psychosocial screening and interventions may be important treatment components for patients with CHD, particularly those with Fontan circulations.

Database: Medline

16. Adolescents and Young Adults Living with Congenital Heart Disease: Coping, Stress Reactivity, and Emotional Functioning.

Author(s): Monti, Jennifer D; Jackson, Jamie L; Vannatta, Kathryn

Source: Journal of clinical psychology in medical settings; Feb 2018

Publication Date: Feb 2018

Publication Type(s): Journal Article

PubMedID: 29455367

Abstract:Living with congenital heart disease (CHD) presents survivors with numerous stressors, which may contribute to emotional problems. This study examined (a) whether coping with CHD-related stress predicts symptoms of depression and anxiety, and (b) whether associations between coping and emotional distress are moderated by involuntary stress reactivity. Adolescents and young adults diagnosed with CHD ($M_{age} = 26.4$) were recruited from pediatric and adult outpatient cardiology clinics. Participants ($N = 168$) completed online self-report measures. Hierarchical multiple regression analyses revealed that secondary control coping (e.g., cognitive restructuring, positive thinking) predicted lower depression and anxiety. Primary control coping (e.g., problem-solving) and stress reactivity (e.g., rumination, emotional numbing) interacted to predict depression and anxiety: the higher individuals were in involuntary stress reactivity, the stronger was the association between primary control coping and lower depression and anxiety. These results can inform clinical efforts to prevent or reduce emotional distress among CHD survivors.

Database: Medline

17. Anemia in Adolescents and Young Adult Patients With Congenital Heart Disease.

Author(s): Rodríguez-Hernández, Juan L; Rodríguez-González, Fayna; Martínez-Quintana, Efrén

Source: Journal of pediatric hematology/oncology; Feb 2018

Publication Date: Feb 2018

Publication Type(s): Journal Article

PubMedID: 29432304

Abstract:INTRODUCTIONDespite anemia in acquired heart disease being a common problem, little is known in patients with congenital heart disease (CHD).METHODSIn total, 544 consecutive stable noncyanotic CHD patients were studied to determine demographic, clinical, and analytic parameters. Anemia was defined as a condition in which hemoglobin concentration was II/IV (odds ratio [OR], 8.37; 95% confidence interval [CI], 1.69-41.35), N-terminal proB-type natriuretic peptide levels >125 pg/mL (OR, 7.90; 95% CI, 2.88-21.69), and apoferritin levels below 15 ng/mL (OR, 0.21; 95% CI, 0.09-0.50). The Kaplan-Meier survival analysis showed no significant differences in mortality between anemic and nonanemic CHD patients (P=0.143).CONCLUSIONSThe incidence of anemia in CHD patients is similar to that of the normal population and iron deficiency anemia accounts for most of the cases. There were no significant differences in mortality between CHD patients with and without anemia.

Database: Medline

18. 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; Feb 2018

Publication Date: Feb 2018

Publication Type(s): Journal Article

PubMedID: 29410185

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 of 1,603 operations and reached a good predictive power. We sought to evaluate its predictive power on 1,654 operations performed in 2 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 following 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 (3/11, 27.3%), mitral valve replacement (9/39, 23.1%) and systemic venous stenosis repair (2/9, 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 2 different centers. The score is a useful tool to analyze surgical outcomes and to support individual decision making.

Database: Medline

19. Managing congenital heart disease in the obstetric patient.

Author(s): Connolly, Heidi M

Source: Seminars in perinatology; Feb 2018; vol. 42 (no. 1); p. 39-48

Publication Date: Feb 2018

Publication Type(s): Journal Article Review

PubMedID: 29463390

Abstract:OBJECTIVE Cardiovascular disease is the major cause of pregnancy-related maternal mortality in the United States, and congenital heart disease (CHD) is the most common form of structural heart disease affecting women of childbearing age. Most females born with CHD will reach childbearing age and consider pregnancy. Adult CHD and maternal-fetal medicine (MFM) specialists managing women with CHD should provide preconception counseling, cardiovascular risk assessment prior to pregnancy that estimates maternal and fetal risk, management during pregnancy, and in the peripartum period and also know the potential complications and special circumstances that may occur in the post-partum period. This chapter will review the population at risk, patient risk prior to pregnancy, management during pregnancy, management in the peripartum and post-partum periods, and outline specific cardiovascular complications. The chapter will also briefly review some common or high-risk congenital cardiovascular lesions commonly encountered. CONCLUSION Management of patients with most forms of CHD encountered during pregnancy requires a multidisciplinary approach and careful team-based care to facilitate safe and appropriate management and pregnancy success.

Database: Medline

20. Phenotype, management and predictors of outcome in a large cohort of adult congenital heart disease patients with heart failure.

Author(s): Van De Bruaene, Alexander; Hickey, Edward J; Kovacs, Adrienne H; Crean, Andrew M; Wald, Rachel M; Silversides, Candice K; Redington, Andrew N; Ross, Heather J; Alba, Ana Carolina; Billia, Filio; Nair, Krishnakumar; Benson, Lee; Horlick, Eric; Osten, Mark; Colman, Jack; Heggie, Jane; Oechslin, Erwin N; Roche, S Lucy

Source: International journal of cardiology; Feb 2018; vol. 252 ; p. 80-87

Publication Date: Feb 2018

Publication Type(s): Journal Article

PubMedID: 29103857

Abstract:OBJECTIVE Although heart failure (HF) is the leading cause of premature death in adult congenital heart disease (ACHD), little population-specific data exist. This study reports early experience from a dedicated, sub-specialty adult congenital heart disease-heart failure (ACHD-HF) clinic, aiming to identify risk factors for adverse outcome. METHODS Between 2012 and 2015, 126 patients (57% male) attended the ACHD-HF clinic. Baseline and follow-up data were analysed and compared across 4 anatomical/physiological subgroups: cyanotic ACHD, Fontan circulation (1V), biventricular circulation with a subaortic right ventricle (2V-RV) and biventricular circulation with a subaortic left ventricle (2V-LV). Predictors of the composite primary outcome: death, transplant or ventricular assist device (VAD) were identified using multivariable Cox proportional hazard models. RESULTS Mean age at first visit was 38±13 years. Patients were grouped as follows: cyanotic ACHD 10%, 1V 24%, 2V-RV 29% and 2V-LV 37%. During a median follow-up of 1.7 (IQR 0.8-2.9) years, 38 patients (30%) reached the primary outcome. Event-free survival was 89%, 78% and 63% at 1, 2 and 3 years. Forty (31.7%) patients experienced 69 HF hospitalisations. Between-group differences were noted for systolic function, valvular regurgitation, pacing prevalence and invasive hemodynamics. Multivariable analysis revealed 2V-RV subgroup (p=0.001), NYHA class (p=0.002) B-type natriuretic peptide >164pg/ml (p=0.003) and sodium <136mmol/L (p=0.036) as independently associated with death, transplant or VAD. CONCLUSIONS Our young ACHD-HF patients experienced

high adverse event rates during a short period of follow-up. The prognostic markers identified will aid clinicians to stratify short-term risk and thereby guide advanced HF management decisions in ACHD.

Database: Medline

21. Extracardiac Conduit Fontan - Outcome Data in Early Adulthood.

Author(s): Zentner, Dominica; Cheshire, Caitlin; Grigg, Leeanne

Source: Heart, lung & circulation; Feb 2018; vol. 27 (no. 2); p. 254-259

Publication Date: Feb 2018

Publication Type(s): Journal Article

PubMedID: 28545820

Abstract:BACKGROUND To describe the survival and health outcome status of young adults with an extracardiac Fontan procedure performed either as a primary or conversion (secondary) Fontan surgery. METHOD The database of the Adult Congenital Heart disease service at the Royal Melbourne Hospital was interrogated to identify all adults who had undergone a primary extracardiac conduit Fontan (n=29) or a Fontan conversion with this procedure (n=8). We then determined vital status, age, original anatomy and functional status in early adulthood in both groups. RESULTS Adults with an ECC Fontan procedure report reasonable NYHA functional class (84% NYHA I or II) though, objectively, exercise testing demonstrates a reduced exercise capacity, and desaturation on exertion is frequent. The majority (86%) have completed secondary education. Most (78%) are managed on warfarin and there is a preponderance of ACE inhibition use (62%). Atrial arrhythmias have been documented in 5 of the 29 primary ECC groups (17%); in 3 patients this preceded primary ECC and 2 patients developed post primary ECC (between 6 and 14 years postoperatively). At a lesser time of follow-up [median 4.5 years (IQR 3.3-6)], conversion to an ECC as a secondary Fontan procedure has successfully treated atrial arrhythmias in the 7 (of 8) patients where this was the surgical indication for conversion. CONCLUSION Though long-term data will require decades to establish, in young adulthood the functional outcomes of a primary ECC Fontan operation are encouraging. Secondary ECC conversion successfully mitigates atrial arrhythmias in the short to medium term.

Database: Medline

22. Surgical treatment for adult congenital heart disease: consideration for indications and procedures.

Author(s): Matsuo, Kozo; Kabasawa, Masashi; Asano, Soichi; Tatenno, Shigeru; Kawasoe, Yasutaka; Okajima, Yoshitomo; Hayashida, Naoki; Murayama, Hirokazu

Source: General thoracic and cardiovascular surgery; Feb 2018; vol. 66 (no. 2); p. 57-64

Publication Date: Feb 2018

Publication Type(s): Journal Article Review

PubMedID: 29119450

Abstract: The number of the adult patients with congenital heart diseases (ACHD) continues to grow owing to improvement of surgical results and medical management. Corrective surgery for complex CHD does not always mean complete cure. It is not rare that the patients will visit the cardiology institutes because of secondary lesions due to residua or sequela in adults. Some patients with CHD remain unrepairable with different degree of heart failure and pulmonary arterial hypertension.

Association of arrhythmias is common in ACHD patients and sometimes critical. We experienced 265 surgical procedures for ACHD patients at our center between 1999 and 2015. Of these procedures, palliative surgery was performed in 3%, palliation to corrective surgery in 6%, primary repair in 57%, and redo surgery in 34%. Hospital mortality within 30 days in this period was 1.1%. Surgery for ACHD patients is safe, beneficial and low-risk treatment; however, tailored procedures for the individual patient are essential to obtain the optimal quality. A comprehensive multidisciplinary approach is required to fulfill this goal.

Database: Medline

23. Variation in practice patterns in device closure of atrial septal defects and patent ductus arteriosus: An analysis of data from the IMproving Pediatric and Adult Congenital Treatment (IMPACT) registry.

Author(s): O'Byrne, Michael L; Kennedy, Kevin F; Rome, Jonathan J; Glatz, Andrew C

Source: American heart journal; Feb 2018; vol. 196 ; p. 119-130

Publication Date: Feb 2018

Publication Type(s): Journal Article

PubMedID: 29421004

Abstract:Practice variation is a potentially important measure of healthcare quality. The IMPACT registry provides a representative national sample with which to study practice variation in transcatheter interventions for congenital heart disease. **METHODS**We studied cases for closure of atrial septal defect (ASD) and patent ductus arteriosus (PDA) in IMPACT between January 1, 2011, and September 30, 2015, using hierarchical multivariate models studying (1) the distribution of indications for closure and (2) in patients whose indication for closure was left (LVVO) or right ventricular volume overload (RVVO), the factors influencing probability of closure of a small defect (either in size or in terms of the magnitude of shunt). **RESULTS**Over the study period, 5233 PDA and 4459 ASD cases were performed at 77 hospitals. The indications for ASD closure were RVVO in 84% and stroke prevention in 13%. Indications for PDA closure were LVVO in 57%, endocarditis prevention in 36%, and pulmonary hypertension in 7%. There was statistically significant variability in indications between hospitals for PDA and ASD procedures (median rate ratio (MRR): 1.3 and 1.1; both $P < .001$). The proportion of cases for volume overload with a Qp:Qs $< 1.5:1$ decreased with increasing PDA and ASD procedural volume ($P = .04$ and 0.05). For ASD, the proportion was higher at hospitals with a larger proportion of adult cases ($P = .0007$). There was significant variation in practice in the risk of closing PDA < 2 mm for LVVO (MRR: 1.4, $P < .001$). **CONCLUSION**There is measurable variation in transcatheter closure of PDA and ASD. Further research is necessary to study whether this affects outcomes or resource utilization.

Database: Medline

24. Atrial Fibrillation Burden in Young Patients With Congenital Heart Disease.

Author(s): Mandalenakis, Zacharias; Rosengren, Annika; Lappas, Georg; Eriksson, Peter; Gilljam, Thomas; Hansson, Per-Olof; Skoglund, Kristofer; Fedchenko, Maria; Dellborg, Mikael

Source: Circulation; Feb 2018; vol. 137 (no. 9); p. 928-937

Publication Date: Feb 2018

Publication Type(s): Academic Journal

PubMedID: 29092907

Abstract:Background: Patients with congenital heart disease (CHD) are assumed to be vulnerable to atrial fibrillation (AF) as a result of residual shunts, anomalous vessel anatomy, progressive valvulopathy, hypertension, and atrial scars from previous heart surgery. However, the risk of developing AF and the complications associated with AF in children and young adults with CHD have not been compared with those in control subjects.Methods: Data from the Swedish Patient and Cause of Death registers were used to identify all patients with a diagnosis of CHD who were born from 1970 to 1993. Each patient with CHD was matched by birth year, sex, and county with 10 control subjects from the Total Population Register in Sweden. Follow-up data were collected until 2011.Results: Among 21 982 patients (51.6% men) with CHD and 219 816 matched control subjects, 654 and 328 developed AF, respectively. The mean follow-up was 27 years. The risk of developing AF was 21.99 times higher (95% confidence interval, 19.26-25.12) in patients with CHD than control subjects. According to a hierarchical CHD classification, patients with conotruncal defects had the highest risk (hazard ratio, 84.27; 95% confidence interval, 56.86-124.89). At the age of 42 years, 8.3% of all patients with CHD had a recorded diagnosis of AF. Heart failure was the quantitatively most important complication in patients with CHD and AF, with a 10.7% (70 of 654) recorded diagnosis of heart failure.Conclusions: The risk of AF in children and young adults with CHD was 22 times higher than that in matched control subjects. Up to the age of 42 years, 1 of 12 patients with CHD had developed AF, and 1 of 10 patients with CHD with AF had developed heart failure. The patient groups with the most complex congenital defects carried the greatest risk of AF and could be considered for targeted monitoring.

Database: CINAHL

25. Poorly Understood Maternal Risks of Pregnancy in Women With Heart Disease.

Author(s): Florio, Karen; Daming, Tara Banaszek; Grodzinsky, Anna

Source: Circulation; Feb 2018; vol. 137 (no. 8); p. 766-768

Publication Date: Feb 2018

Publication Type(s): Academic Journal

PubMedID: 29459460

Abstract:The article discusses the maternal risks of pregnancy in women with heart disease, focusing on lack of understanding about the risks. It states that significant improvements in treating congenital heart disease have led to more women with congenital cardiac malformations reaching reproductive age and desiring fertility, and highlights the inherent risk of cardiac events during gestation along with risks to the fetus. It notes the need of improving risk stratification and management.

Database: CINAHL

26. Evaluation and Management of Maternal Congenital Heart Disease: A Review

Author(s): Hopkins M.K.; Kuller J.A.; Goldstein S.A.; Ward C.C.

Source: Obstetrical and Gynecological Survey; Feb 2018; vol. 73 (no. 2); p. 116-124

Publication Date: Feb 2018

Publication Type(s): Review

Abstract:Objective Congenital heart defects represent the most common major congenital anomalies. The objective of this review was to define the most common forms of congenital heart disease (CHD) in pregnancy, outline preconception counseling, discuss the associated morbidity and

mortality of each lesion, and review current recommendations for management of CHD in pregnancy. Evidence Acquisition A MEDLINE search of "congenital heart disease in pregnancy" and specific conditions in pregnancy including "ventricular septal defect," "atrial septal defect," "left outflow obstruction," "right outflow obstruction," "tetralogy of Fallot," and "transposition of the great vessels" was performed. Results The evidence included in the review contains 18 retrospective studies, 8 meta-analyses or systematic reviews or expert opinions, 5 case reports including surgical case reports, 2 prospective studies, and 2 clinical texts. Conclusions Given advances in surgical and medical management, women with a history of congenital cardiac defects are more frequently reaching childbearing age and requiring obstetric care. Many women with CHD can have successful pregnancies, although there are a few conditions that confer significant maternal risk, and pregnancy may even be contraindicated. Appropriate care for women with CHD requires a knowledge of cardiac physiology in pregnancy, the common lesions of CHD, and coordinated care from cardiology and maternal-fetal medicine specialists. Target Audience Obstetricians and gynecologists, family physicians. Learning Objectives After completing this activity, the learner should be better able to compare pregnancy risks among varying types of the most common maternal CHD; apply criteria of relative and absolute contraindications to pregnancy when performing preconception counseling to women with CHD; interpret for patients and other health providers the prognosis, management, and expectations of pregnancies with women with the most common types of CHD; and select relevant imaging studies, tests, and appropriate consultations and referrals when caring for women with maternal CHD.

Database: EMBASE

27. The creation of stakeholder-based clinical and mobile based programs to assist in transition and transfer to adult care for a diverse group of congenital heart disease adolescents

Author(s): Lopez K.N.; O'Connor M.; Fordis M.; King J.; Challman M.; Alexander D.; Smith A.; Fawcett E.; Franklin W.J.; Thompson D.I.; Goodly N.; Lovick D.K.

Source: Journal of Adolescent Health; Feb 2018; vol. 62 (no. 2)

Publication Date: Feb 2018

Publication Type(s): Conference Abstract

Abstract: Purpose: Congenital heart diseases (CHDs) are the most common type of birth defects. Improvements in CHD care have led to roughly 1.4 million survivors reaching adulthood. Unfortunately, <30% of adults with CHD are seen by adult CHD physicians, and few formal CHD transition-to-adult programs exist. The objective of this project was to first complete a stakeholder needs assessment to inform the knowledge gaps, transition readiness, and delivery of transition education for a diverse group of adolescents with CHD. Second, to develop the workflow, educational content, and electronic medical record (EMR) components for a clinical and mobile-based education with the assistance of an adolescent CHD expert panel. Methods: We conducted a literature search for best practices and CHD guidelines in transition medicine. We then conducted individual interviews with pediatric cardiologists, adult CHD specialists, parents and their CHD adolescents to understand acceptable delivery of education both in clinic and via a mobile application. We assessed knowledge via CHD questionnaires we created, and transition readiness via the Transition Q. We then partnered with clinic staff, 2 adolescent CHD expert panels, transition experts, and electronic medical record teams to further determine educational content, delivery as well as clinical and mobile application design. Results: We completed 327 individual interviews with CHD adolescents ages 15-22 years. 78.2% had moderate or severe CHD complexity. 41.6% of CHD adolescents were female, 12.7% were African American, and 35.8% were Latino. 36.5% of patients had public insurance. 42.2% of patients aged 15-17 years and 47.9% aged 18-22 years had minimal

understanding of their CHD, but an interest in learning. Transition readiness scores reflected an average of 49.4% readiness for those aged 15-17 and 58.6% for those aged 18-22. Clinical educational requests were accomplished including same-day delivery at the cardiology appointment, and transition education to be delivered by a transition nurse and social worker. EMR requests included tracking of education and a deliverable medical summary, which we built. For the mobile application, adolescents expressed the need for a tailored application to their specific CHD, question-answer exchange, a blog-type CHD forum, and a transition checklist. We concomitantly built a mobile application incorporating educational content, assessments of transition readiness and knowledge, a CHD diagram, a medical summary, a blog, checklist, and question/answer space. Conclusions: Based on our data of average CHD knowledge and transition readiness scores, CHD adolescents are largely not prepared for transition and transfer to adult care. The vast majority of adolescents found education in clinic and via a mobile application acceptable. We were able to create a mobile application and a clinical education program with our stakeholders including CHD adolescents to aid in the CHD transition process. Next steps are to begin use of EMR educational and medical summary templates, and to further build CHD educational content, to conduct mobile application usability testing, and to use focus groups to refine our current mobile application. Sources of Support: This project was supported by the National Institute of Health, National Heart Lung and Blood institute Grant 5K23HL127164-02 (Lopez) and an institutional grant (Lopez).

Database: EMBASE

28. Congenital heart disease and reported self knowledge

Author(s): Katz A.J.; Srivastava N.; Farrell A.; Wilson S.; Blythe M.J.; Shew M.L.

Source: Journal of Adolescent Health; Feb 2018; vol. 62 (no. 2)

Publication Date: Feb 2018

Publication Type(s): Conference Abstract

Abstract: Purpose: For adolescent and young adult women with congenital heart disease (CHD), knowledge about their cardiac lesion and how it may affect reproductive health is important. This study examines whether these women with CHD can identify their cardiac lesion, understand their risk for genetic transmission, and identify contraindications to their use of any methods of contraception or pregnancy. Methods: A convenience sample of women (N = 61) who could independently complete an on-line questionnaire, ages 14 to 21 years, were recruited from university associated cardiology clinics. Participants completed a survey that assessed their reproductive health knowledge and needs as it relates to their cardiac lesion(s). For each participant enrolled, an independent questionnaire was generated for the study cardiologist. The cardiologist reviewed the patient's medical records to identify the cardiac lesion(s) and determine contraindications to contraceptive use and pregnancy and risk for genetic transmission. Contraindications to pregnancy and contraception were established through review of the literature, which included published guidelines and cardiologists' consensus reports. Participants' reports were then compared to those from the cardiologist. Results: Mean age was 17.1 years (SD 2.3); 90.2% (N = 55) were white. Cardiac lesions highly varied, but the most common lesions included ventricular septal defect, atrial septal defect, coarctation of the aorta, and transposition of the great arteries. In comparison with the cardiologist report, 19 (31.1%) could identify their lesion(s) correctly; 28 (45.9%) were unable to identify or did not know their lesion(s). For the remainder of the women (N = 14), most had multiple lesions (12/14) and could partially identify the lesion(s) correctly. Fourteen (23%) reported specifically discussing hormonal birth control and their cardiac lesion with a health care provider and 20 (32.8%) women reported that they were or had used birth control. Twenty-two women had contraindications to estrogen (N = 20) or IUD (N = 2). Of the 20 women reporting use of

oral contraceptives, six had contraindications to the use of estrogen. Of the four women who carried specific contraindications to pregnancy, only one reported having conversations about transmission with a health care provider. Twenty-two women (36.1%) reported having discussed risk of genetic transmission to a fetus with a health care provider, yet 50.8% (31) thought their cardiac lesion could be transmitted to the fetus. Only 21 (34.4%) of these adolescents were at risk for fetal transmission by cardiology report. Conclusions: Less than half of young women with CHD could correctly identify their cardiac lesion(s). Discussion about reproductive health issues with CHD patients were limited. Targeted clinical interventions for this group are needed to potentially reduce poor outcomes in reproductive health with this population of young women. Sources of Support: General division funds of Adolescent Medicine.

Database: EMBASE

29. Clinical features and peripartum outcomes in pregnant women with cardiac disease: a nationwide retrospective cohort study in Japan

Author(s): Isogai T.; Matsui H.; Yasunaga H.; Tanaka H.; Kohyama A.; Fushimi K.

Source: Heart and Vessels; Feb 2018 ; p. 1-13

Publication Date: Feb 2018

Publication Type(s): Article In Press

Abstract: Although the number of pregnancies in women with cardiac disease is increasing worldwide, there are few data concerning their clinical characteristics and peripartum outcomes. Using the Diagnosis Procedure Combination database between 2008 and 2014 in Japan, we retrospectively identified pregnant women who underwent high-risk delivery due to obstetric or non-obstetric comorbidities. We classified eligible women into those with pre-existing cardiac disease (cardiac disease group) and those with non-cardiac comorbidities (non-cardiac disease group) and compared their characteristics and peripartum outcomes. Of 94,364 women undergoing high-risk delivery at 556 hospitals, 857 (0.91%) had pre-existing cardiac disease (302, congenital heart disease; 190, arrhythmia; 176, valvular heart disease; 120, ischemic heart disease; 65, cardiomyopathy; 4, pericardial disease). Women in the cardiac disease group were more likely to be treated at university hospitals (51.1 versus 28.6%; $p < 0.001$) and in intensive care units (33.5 versus 18.8%; $p < 0.001$) than those in the non-cardiac disease group. The proportion of cesarean deliveries was 69.4% (emergency, 28.4%; elective, 41.1%) in the cardiac disease group and 73.4% (emergency, 38.4%; elective, 35.0%) in the non-cardiac disease group. Epidural analgesia during vaginal delivery was used significantly more frequently in the cardiac disease than non-cardiac disease group (15.6 versus 2.3%; $p < 0.001$). Heart failure occurred more frequently in the cardiac disease than the non-cardiac disease group (10.2 versus 0.3%; $p < 0.001$). In cardiac subgroup comparisons, heart failure occurred more frequently in women with congenital heart disease (12.3%), valvular heart disease (12.5%), or cardiomyopathy (12.3%) than in women with arrhythmia (6.3%) or ischemic heart disease (5.8%). Multivariable logistic regression analysis showed a significant positive association between pre-existing cardiac disease and risk of heart failure (adjusted odds ratio, 24.7; 95% confidence interval, 17.6-34.6; $p < 0.001$). No woman in the cardiac disease group died, whereas 18 women (0.02%) in the non-cardiac disease group did ($p = 1.000$). These findings suggest that pregnant women with pre-existing cardiac disease are at a higher risk of heart failure during the peripartum period than those with non-cardiac comorbidities.

Database: EMBASE

30. Cardiac resynchronization therapy in adults with congenital heart disease

Author(s): Koyak Z.; De Groot J.R.; MacKay T.M.; Bouma B.J.; Mulder B.J.M.; Krimly A.; Silversides C.K.; Oechslin E.N.; Harris L.; Hoke U.; Van Erven L.; Budts W.; Van Gelder I.C.

Source: Europace; Feb 2018; vol. 20 (no. 2); p. 315-322

Publication Date: Feb 2018

Publication Type(s): Article

Abstract: Aims In adults with congenital heart disease (CHD) heart failure is one of the leading causes of morbidity and mortality but experience with and reported outcome of cardiac resynchronization therapy (CRT) is limited. We investigated the efficacy of CRT in adults with CHD. Methods and results This was a retrospective study including 48 adults with CHD who received CRT since 2003 in four tertiary referral centres. Responders were defined as patients who showed improvement in NYHA functional class and/or systemic ventricular ejection fraction by at least one category. Ventricular function was assessed by echocardiography and graded on a four point ordinal scale. Median age at CRT was 47 years (range 18-74 years) and 77% was male. Cardiac diagnosis included tetralogy of Fallot in 29%, (congenitally corrected) transposition of great arteries in 23%, septal defects in 25%, left sided lesions in 21%, and Marfan syndrome in 2% of the patients. The median follow-up duration after CRT was 2.6 years (range 0.1-8.8). Overall, 37 out of 48 patients (77%) responded to CRT either by improvement of NYHA functional class and/or systemic ventricular function. There were 11 non-responders to CRT. Of these, three patients died and four underwent heart transplantation. Conclusion In this cohort of older CHD patients, CRT was accomplished with a success rate comparable to those with acquired heart disease despite the complex anatomy and technical challenges frequently encountered in this population. Further studies are needed to establish appropriate guidelines for patient selection and long term outcome.

Database: EMBASE

31. Clinical and Psychological Drivers of Perceived Health Status in Adults With Congenital Heart Disease

Author(s): Ko J.M.; Tecson K.M.; Rashida V.A.; Sodhi S.; Saef J.; Mufti M.; Ludbrook P.A.; White K.S.; Cedars A.M.

Source: American Journal of Cardiology; Feb 2018; vol. 121 (no. 3); p. 377-381

Publication Date: Feb 2018

Publication Type(s): Article

Available at [The American Journal of Cardiology](#) - from ProQuest (Hospital Premium Collection) - NHS Version

Abstract: The factors having the greatest impact on self-reported health status in adults with congenital heart disease (ACHD) remain incompletely studied. We conducted a single-site, cross-sectional study of ACHD patients followed at the Center for ACHD at Washington University School of Medicine, including retrospectively gathered clinical data and psychometric and health status assessments completed at the time of enrollment. To identify primary drivers of perceived health status, we investigated the impact of the demographic, clinical, and psychological variables on self-reported health status as assessed using the Rand 36-Item Short Form Health Survey. Variables with significant associations within each domain were considered jointly in multivariable models constructed via stepwise selection. There was domain-specific heterogeneity in the variables having the greatest effect on self-reported health status. Depression was responsible for the greatest amount of variability in health status in all domains except physical functioning. In the physical functioning domain, depression remained responsible for 5% of total variability, the third most

significant variable in the model. In every domain, depression more strongly influenced health status than did any cardiac-specific variable. In conclusion, depression was responsible for a significant amount of heterogeneity in all domains of self-perceived health status. Psychological variables were better predictors of health status than clinical variables.

Database: EMBASE

32. Limited Accuracy of Administrative Data for the Identification and Classification of Adult Congenital Heart Disease.

Author(s): Khan, Abigail; Ramsey, Katrina; Ballard, Cody; Armstrong, Emily; Burchill, Luke J; Menashe, Victor; Pantely, George; Broberg, Craig S

Source: Journal of the American Heart Association; Jan 2018; vol. 7 (no. 2)

Publication Date: Jan 2018

Publication Type(s): Journal Article

PubMedID: 29330259

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

Abstract:BACKGROUNDAdministrative data sets utilize billing codes for research and quality assessment. Previous data suggest that such codes can accurately identify adults with congenital heart disease (CHD) in the cardiology clinic, but their use has yet to be validated in a larger population.METHODS AND RESULTSAll administrative codes from an entire health system were queried for a single year. Adults with a CHD diagnosis code (International Classification of Diseases, Ninth Revision, (ICD-9) codes 745-747) defined the cohort. A previously validated hierarchical algorithm was used to identify diagnoses and classify patients. All charts were reviewed to determine a gold standard diagnosis, and comparisons were made to determine accuracy. Of 2399 individuals identified, 206 had no CHD by the algorithm or were deemed to have an uncertain diagnosis after provider review. Of the remaining 2193, only 1069 had a confirmed CHD diagnosis, yielding overall accuracy of 48.7% (95% confidence interval, 47-51%). When limited to those with moderate or complex disease (n=484), accuracy was 77% (95% confidence interval, 74-81%). Among those with CHD, misclassification occurred in 23%. The discriminative ability of the hierarchical algorithm (C statistic: 0.79; 95% confidence interval, 0.77-0.80) improved further with the addition of age, encounter type, and provider (C statistic: 0.89; 95% confidence interval, 0.88-0.90).CONCLUSIONSICD codes from an entire healthcare system were frequently erroneous in detecting and classifying CHD patients. Accuracy was higher for those with moderate or complex disease or when coupled with other data. These findings should be taken into account in future studies utilizing administrative data sets in CHD.

Database: Medline

33. Executive Function and Internalizing Symptoms in Adolescents and Young Adults With Congenital Heart Disease: The Role of Coping.

Author(s): Jackson, Jamie L; Gerardo, Gina M; Monti, Jennifer D; Schofield, Kyle A; Vannatta, Kathryn

Source: Journal of pediatric psychology; Jan 2018

Publication Date: Jan 2018

Publication Type(s): Journal Article

PubMedID: 29329440

Abstract:Objective Executive functioning deficits have been documented among congenital heart disease (CHD) survivors and may contribute to emotional distress. Little research has investigated the role of coping in this association. This study examined the role of coping in accounting for the association between self-reported executive function problems and internalizing symptoms among adolescents and emerging adults (AEAs), as well as young adults (YAs) with CHD. Participants included 74 AEA (M age = 19.32 ± 3.47 years, range 15-25 years) and 98 YA CHD survivors (M age = 32.00 ± 3.69 years, range 26-39 years), recruited from pediatric and adult outpatient cardiology clinics. Participants completed self-report measures of executive function problems, coping (primary control, secondary control, and disengagement coping), and internalizing symptoms. Lesion severity classification and functional impairment due to symptoms of heart failure were determined from medical chart review. Significant problems in executive function were reported by 5% of AEA and 13% of YA. Coping was not associated with executive function problems or internalizing symptoms for AEA. However, among YA, less use of adaptive coping strategies and more maladaptive coping responses was associated with both more executive function problems and internalizing symptoms. An indirect effect of executive function problems on internalizing symptoms via secondary control coping emerged for YA. Executive function problems may disrupt the ability to use important adaptive coping skills, such as cognitive reappraisal, positive thinking, and acceptance, thereby resulting in greater emotional distress among YA CHD survivors.

Database: Medline

34. Outcomes following implantation of mechanical circulatory support in adults with congenital heart disease: An analysis of the Interagency Registry for Mechanically Assisted Circulatory Support (INTERMACS).

Author(s): VanderPluym, Christina J; Cedars, Ari; Eghtesady, Pirooz; Maxwell, Bryan G; Gelow, Jill M; Burchill, Luke J; Maltais, Simon; Koehl, Devin A; Cantor, Ryan S; Blume, Elizabeth D

Source: The Journal of heart and lung transplantation : the official publication of the International Society for Heart Transplantation; Jan 2018; vol. 37 (no. 1); p. 89-99

Publication Date: Jan 2018

Publication Type(s): Journal Article

PubMedID: 28365175

Abstract:BACKGROUNDAdults with congenital heart disease represent an expanding and unique population of patients with heart failure (HF) in whom the use of mechanical circulatory support (MCS) has not been characterized. We sought to describe overall use, patient characteristics, and outcomes of MCS in adult congenital heart disease (ACHD).METHODSAll patients entered into the Interagency Registry for Mechanically Assisted Circulatory Support (INTERMACS) between June 23, 2006, and December 31, 2015, were included. Patients with ACHD were identified using pre-operative data and stratified by ventricular morphology. Mortality was compared between ACHD and non-ACHD patients, and multivariate analysis was performed to identify predictors of death after device implantation.RESULTSO f 16,182 patients, 126 with ACHD stratified as follows: systemic morphologic left ventricle (n = 63), systemic morphologic right ventricle (n = 45), and single ventricle (n = 17). ACHD patients were younger (42 years ± 14 vs 56 years ± 13; p < 0.0001) and were more likely to undergo device implantation as bridge to transplant (45% vs 29%; p < 0.0001). A higher proportion of ACHD patients had biventricular assist device (BiVAD)/total artificial heart (TAH)

support compared with non-ACHD patients (21% vs 7%; $p < 0.0001$). More ACHD patients on BiVAD/TAH support were INTERMACS profile 1 compared with patients on systemic left ventricular assist device (LVAD) support (35% vs 15%; $p = 0.002$). ACHD and non-ACHD patients with LVADs had similar survival; survival was worse for patients on BiVAD/TAH support. BiVAD/TAH support was the only variable independently associated with mortality (early phase hazard ratio 4.4; 95% confidence interval, 1.8-11.1; $p = 0.001$). For ACHD patients receiving MCS, ventricular morphology was not associated with mortality. **CONCLUSIONS** ACHD patients with LVADs have survival similar to non-ACHD patients. Mortality is higher for patients requiring BiVAD/TAH support, potentially owing to higher INTERMACS profile. These outcomes suggest a promising role for LVAD support in ACHD patients as part of the armamentarium of therapies for advanced HF.

Database: Medline

35. Initial validation of a healthcare needs scale for young people with congenital heart disease.

Author(s): Chen, Chi-Wen; Ho, Ciao-Lin; Su, Wen-Jen; Wang, Jou-Kou; Chung, Hung-Tao; Lee, Pi-Chang; Lu, Chun-Wei; Hwang, Be-Tau

Source: Journal of advanced nursing; Jan 2018; vol. 74 (no. 1); p. 223-231

Publication Date: Jan 2018

Publication Type(s): Journal Article

PubMedID: 28702947

Abstract: **AIM** To validate the initial psychometric properties of a Healthcare Needs Scale for Youth with Congenital Heart Disease. **BACKGROUND** As the number of patients with congenital heart disease surviving to adulthood increases, the transitional healthcare needs for adolescents and young adults with congenital heart disease require investigation. However, few tools comprehensively identify the healthcare needs of youth with congenital heart disease. **DESIGN** A cross-sectional study was employed to examine the psychometric properties of the Healthcare Needs Scale for Youth with Congenital Heart Disease. **METHOD** The sample consisted of 500 patients with congenital heart disease, aged 15-24 years, from paediatric cardiology departments and covered the period from March-August 2015. The patients completed the 25-item Healthcare Needs Scale for Youth with Congenital Heart Disease, the questionnaire on health needs for adolescents and the WHO Quality of Life-BREF. Reliability and construct, concurrent, predictive and known-group validity were examined. **RESULTS** The Healthcare Needs Scale for Youth with Congenital Heart Disease includes three dimensions, namely health management, health policy and individual and interpersonal relationships, which consist of 25 items. It demonstrated excellent internal consistency and sound construct, concurrent, predictive and known-group validity. **CONCLUSION** The Healthcare Needs Scale for Youth with Congenital Heart Disease is a psychometrically robust measure of the healthcare needs of youth with congenital heart disease. It has the potential to provide nurses with a means to assess and identify the concerns of youth with congenital heart disease and to help them achieve a successful transition to adult care.

Database: Medline

36. Management of acute heart failure in adult patients with congenital heart disease.

Author(s): Van De Bruaene, Alexander; Meier, Lukas; Droogne, Walter; De Meester, Pieter; Troost, Els; Gewillig, Marc; Budts, Werner

Source: Heart failure reviews; Jan 2018; vol. 23 (no. 1); p. 1-14

Publication Date: Jan 2018

Publication Type(s): Journal Article Review

PubMedID: 29277859

Abstract:Heart failure is an increasing reason for hospitalization and the leading cause of death in patients with adult congenital heart disease (ACHD). Recently, the European Society of Cardiology and the American Heart Association published consensus documents on the management of chronic heart failure in ACHD patients. However, little data and/or guidelines are available for the management of (sub)acute heart failure. The ACHD population is heterogeneous by definition and often has complex underlying anatomy, which could pose a challenge to the physician confronted with the ACHD patient in (sub)acute heart failure. Recognizing the underlying anatomy and awareness of the possible complications related would result in better treatment, avoid unnecessary delays, and improve outcomes of the ACHD patient with (sub)acute heart failure. This review focuses on the management of (sub)acute heart failure in ACHD with specific attention to lesion-specific issues.

Database: Medline

37. Infective endocarditis in adults with congenital heart disease remains a lethal disease.

Author(s): Tutarel, Oktay; Alonso-Gonzalez, Rafael; Montanaro, Claudia; Schiff, Renee; Uribarri, Aitor; Kempny, Aleksander; Grübler, Martin R; Uebing, Anselm; Swan, Lorna; Diller, Gerhard-Paul; Dimopoulos, Konstantinos; Gatzoulis, Michael A

Source: Heart (British Cardiac Society); Jan 2018; vol. 104 (no. 2); p. 161-165

Publication Date: Jan 2018

Publication Type(s): Journal Article

PubMedID: 28754810

Available at [Heart](#) - from BMJ Journals - NHS

Available at [Heart](#) - from BMJ Journals

Abstract:OBJECTIVEInfective endocarditis (IE) is associated with significant morbidity and mortality. Patients with adult congenital heart disease (ACHD) have an increased risk of developing IE. The aim of this study is to describe the incidence, predictors of outcome and mortality associated with IE in ACHD in a contemporary cohort.METHODSAll episodes of IE in adults with congenital heart disease referred to our tertiary centre between 1999 and 2013 were included in the study. Patients were identified from the hospital database. The diagnosis of endocarditis was established according to the modified Duke criteria. The primary endpoint of the study was endocarditis-associated mortality.RESULTSThere were 164 episodes of IE in 144 patients (male 102, 70.8%). Mean age at presentation was 32.3±22.7 years. Out of these, 43% had a simple, 23% a moderate and 32% a complex lesion. It was at least the second bout of IE in 37 episodes (23%). A predisposing event could be identified in only 26.2% of episodes. Surgical intervention during the same admission was performed in 61 episodes (37.2%). During a median follow-up of 6.7 years (IQR 2.9-11.4), 28 (19.4%) patients died. Out of these, 10 deaths were related to IE (IE mortality 6.9%). On univariate regression analysis, the development of an abscess (OR: 7.23; 95% CI 1.81 to 28.94, p<0.01) and age (OR: 1.05; 95% CI 1.01 to 1.10, p=0.03) were the only predictors of IE-associated mortality. There was no increase in IE cases at our centre during the period of the study.CONCLUSIONSIE-associated morbidity and mortality in a contemporary cohort of ACHD patients is still high in the current era.

Database: Medline

38. Establishing a successful transition care plan for the adolescent with congenital heart disease.

Author(s): Talluto, Christopher

Source: Current opinion in cardiology; Jan 2018; vol. 33 (no. 1); p. 73-77

Publication Date: Jan 2018

Publication Type(s): Journal Article

PubMedID: 29028634

Abstract:**PURPOSE OF REVIEW**In this review we explore the aspects needed to develop a successful transition program for adolescents with congenital heart disease and the barriers that exist to the development of such a program.**RECENT FINDINGS**We review the literature including more recent publications which support the development of a transition program in order to facilitate transfer from the pediatric to the adult healthcare system.**SUMMARY**With the development of a successful transition program patients will hopefully receive high-quality, continuous life-long care in the appropriate setting.

Database: Medline

39. Overview of adult congenital heart transplants.

Author(s): Bryant, Roosevelt; Morales, David

Source: Annals of cardiothoracic surgery; Jan 2018; vol. 7 (no. 1); p. 143-151

Publication Date: Jan 2018

Publication Type(s): Journal Article

PubMedID: 29492392

Available at [Annals of Cardiothoracic Surgery](#) - from Europe PubMed Central - Open Access

Available at [Annals of Cardiothoracic Surgery](#) - from PubMed Central

Abstract:Transplantation for adult patients with congenital heart disease (ACHD) is a growing clinical endeavor in the transplant community. Understanding the results and defining potential high-risk patient subsets will allow optimization of patient outcomes. This review summarizes the scope of ACHD transplantation, the mechanisms of late ventricular dysfunction, the ACHD population at risk of developing heart failure, the indications and potential contraindications for transplant, surgical considerations, and post-transplant outcomes. The findings reveal that 3.3% of adult heart transplants occur in ACHD patients. The potential mechanisms for the development of late ventricular dysfunction include a morphologic systemic right ventricle, altered coronary perfusion, and ventricular noncompaction. The indications for transplant in ACHD patients include systemic ventricular failure refractory medical therapy, Fontan patients failing from chronic passive pulmonary circulation, and progressive cyanosis leading to functional decline. Transplantation in ACHD patients can be quite complex and may require extensive reconstruction of the branch pulmonary arteries, systemic veins, or the aorta. Vasoplegia, bleeding, and graft right ventricular dysfunction can complicate the immediate post-transplant period. The post-transplant operative mortality ranges between 14% and 39%. The majority of early mortality occurs in ACHD patients with univentricular congenital heart disease. However, there has been improvement in operative survival in more contemporary studies. In conclusion, the experience with cardiac transplantation for ACHD patients with end-stage heart failure is growing, and high-risk patient subsets have been defined. Significant strides have been made in developing evidence-based guidelines of indications for transplant, and the intraoperative management of complex reconstruction has evolved. With proper patient selection, more aggressive use of mechanical circulatory support, and earlier referral of patients with failing Fontan physiology, outcomes should continue to improve. **Database:** Medline

40. Short-term outcomes following implementation of a dedicated young adult congenital heart disease transition program.

Author(s): Vaikunth, Sumeet S; Williams, Roberta G; Uzunyan, Merujan Y; Tun, Han; Barton, Cheryl; Chang, Philip M

Source: Congenital heart disease; Jan 2018; vol. 13 (no. 1); p. 85-91

Publication Date: Jan 2018

Publication Type(s): Journal Article

PubMedID: 29152906

Abstract:OBJECTIVE Transition from pediatric to adult care is a critical time for patients with congenital heart disease. Lapses in care can lead to poor outcomes, including increased mortality. Formal transition clinics have been implemented to improve success of transferring care from pediatric to adult providers; however, data regarding outcomes remain limited. We sought to evaluate outcomes of transfer within a dedicated transition clinic for young adult patients with congenital heart disease. DESIGN, SETTING, AND PATIENTS We performed a retrospective analysis of all 73 patients seen in a dedicated young adult congenital heart disease transition clinic from January 2012 to December 2015 within a single academic institution that delivered pediatric and adult care at separate children's and adult hospitals, respectively. INTERVENTION AND OUTCOME MEASURES Demographic characteristics including congenital heart disease severity, gender, age, presence of comorbidities, presence of cardiac implantable electronic devices, and type of insurance were correlated to success of transfer. Rate of successful transfer was evaluated, and multivariate analysis was performed to determine which demographic variables were favorably associated with transfer. RESULTS Thirty-nine percent of patients successfully transferred from pediatric to adult services during the study period. Severe congenital heart disease (OR 4.44, 95% CI 1.25-15.79, $P = .02$) and presence of a cardiac implantable electronic device (OR 4.93, 95% CI 1.18-20.58, $P = .03$) correlated with transfer. Trends favoring successful transfer with presence of comorbidities and private insurance were also noted. CONCLUSIONS Despite a dedicated transition clinic, successful transfer rates remained relatively low though comparable to previously published rates. Severity of disease and presence of implantable devices correlated with successful transfer. Other obstacles to transfer remain and require combined efforts from pediatric and adult care systems, insurance carriers, and policy makers to improve transfer outcomes.

Database: Medline

41. Body composition, appetite-related hormones, adipocytokines, and heart failure in adult patients with congenital heart disease: A preliminary study.

Author(s): Shiina, Yumi; Murakami, Tomoaki; Matsumoto, Noriko; Okamura, Daisuke; Takahashi, Yuta; Nishihata, Yosuke; Komiyama, Nobuyuki; Niwa, Koichiro

Source: Congenital heart disease; Jan 2018; vol. 13 (no. 1); p. 79-84

Publication Date: Jan 2018

Publication Type(s): Journal Article

PubMedID: 29181895

Abstract:OBJECTIVE To assess body composition and relationships among body composition, appetite-related hormones, adipocytokines, and heart failure (HF) in adult patients with congenital heart disease (CHD). PATIENTS This prospective study enrolled 46 consecutive adult patients with CHD and 12 age-matched healthy controls. The patients and control subjects were divided into four groups: 13 patients with Fontan circulation (group A), 16 patients with cyanosis (group B), 17

patients who previously underwent biventricular repair (group C), and 12 age-matched healthy controls. DESIGN Body composition was measured using InBody730, and levels of appetite-related hormones (ghrelin and leptin) and adipocytokines (leptin, interleukin-6, and tumor necrosis factor- α) were determined. Relationships of these measurements between severe HF, defined as New York Heart Association functional class III-IV and/or recent repeated unscheduled hospitalizations due to HF, were examined using univariate logistic analysis. RESULTS Mean patient age was 32.1 ± 7.4 years. The skeletal muscle mass was significantly decreased in groups A and B compared with that in controls. Interestingly, ghrelin levels in groups A and B were also significantly lower than those in controls. Univariate logistic analysis revealed that ghrelin level, percent body fat, and pulse oximetric oxygen saturation were significantly associated with severe HF. CONCLUSIONS Patients with Fontan circulation and those with cyanosis might be at a risk of sarcopenia. Despite the decreased skeletal muscle mass and increased body fat, ghrelin levels in these patients were decreased. These changes might have a negative impact on HF in these patients.

Database: Medline

42. Identification of adults with congenital heart disease of moderate or great complexity from administrative data.

Author(s): Steiner, Jill M; Kirkpatrick, James N; Heckbert, Susan R; Habib, Asma; Sibley, James; Lober, William; Randall Curtis, J

Source: Congenital heart disease; Jan 2018; vol. 13 (no. 1); p. 65-71

Publication Date: Jan 2018

Publication Type(s): Journal Article

PubMedID: 28736836

Abstract: INTRODUCTION There is relatively sparse literature on the use of administrative datasets for research in patients with adult congenital heart disease (ACHD). The goal of this analysis is to examine the accuracy of administrative data for identifying patients with ACHD who died. METHODS A list of the International Classification of Diseases codes representing ACHD of moderate- or great-complexity was created. A search for these codes in the electronic health record of adults who received care in 2010-2016 was performed, and used state death records to identify patients who died during this period. Manual record review was completed to evaluate performance of this search strategy. Identified patients were also compared with a list of patients with moderate- or great-complexity ACHD known to have died. RESULTS About 134 patients were identified, of which 72 had moderate- or great-complexity ACHD confirmed by manual review, yielding a positive predictive value of 0.54 (95% CI 0.45, 0.62). Twenty six patients had a mild ACHD diagnosis. Thirty six patients had no identified ACHD on record review. Misidentifications were attributed to coding error for 19 patients (53%), and to acquired ventricular septal defects for 11 patients (31%). Diagnostic codes incorrect more than 50% of the time were those for congenitally corrected transposition, endocardial cushion defect, and hypoplastic left heart syndrome. Only 1 of 21 patients known to have died was not identified by the search, yielding a sensitivity of 0.95 (0.76, 0.99). CONCLUSION Use of administrative data to identify patients with ACHD of moderate or great complexity who have died had good sensitivity but suboptimal positive predictive value. Strategies to improve accuracy are needed. Administrative data is not ideal for identification of patients in this group, and manual record review is necessary to confirm these diagnoses.

Database: Medline

43. Systemic Right Ventricle in Adults With Congenital Heart Disease: Anatomic and Phenotypic Spectrum and Current Approach to Management.

Author(s): Brida, Margarita; Diller, Gerhard-Paul; Gatzoulis, Michael A.

Source: Circulation; Jan 2018; vol. 137 (no. 5); p. 508-518

Publication Date: Jan 2018

Publication Type(s): Academic Journal

PubMedID: 29378757

Abstract:The systemic right ventricle (SRV) is commonly encountered in congenital heart disease representing a distinctly different model in terms of its anatomic spectrum, adaptation, clinical phenotype, and variable, but overall guarded prognosis. The most common clinical scenarios where an SRV is encountered are complete transposition of the great arteries with previous atrial switch repair, congenitally corrected transposition of the great arteries, double inlet right ventricle mostly with previous Fontan palliation, and hypoplastic left heart syndrome palliated with the Norwood-Fontan protocol. The reasons for the guarded prognosis of the SRV in comparison with the systemic left ventricle are multifactorial, including distinct fibromuscular architecture, shape and function, coronary artery supply mismatch, intrinsic abnormalities of the tricuspid valve, intrinsic or acquired conduction abnormalities, and varied SRV adaptation to pressure or volume overload. Management of the SRV remains an ongoing challenge because SRV dysfunction has implications on short- and long-term outcomes for all patients irrespective of underlying cardiac morphology. SRV dysfunction can be subclinical, underscoring the need for tertiary follow-up and timely management of target hemodynamic lesions. Catheter interventions and surgery have an established role in selected patients. Cardiac resynchronization therapy is increasingly used, whereas pharmacological therapy is largely empirical. Mechanical assist device and heart transplantation remain options in end-stage heart failure when other management strategies have been exhausted. The present report focuses on the SRV with its pathological subtypes, pathophysiology, clinical features, current management strategies, and long-term sequelae. Although our article touches on issues applicable to neonates and children, its main focus is on adults with SRV.

Database: CINAHL

44. Cardiovascular morbidity and mortality in pregnancy: Management challenges in pregnancies complicated by maternal heart disease are complex, requiring ongoing involvement of a team of specialists.

Author(s): BRICKNER, BETH

Source: Contemporary OB/GYN; Jan 2018; vol. 63 (no. 1); p. 12-18

Publication Date: Jan 2018

Publication Type(s): Academic Journal

Available at [Contemporary OB/GYN](#) - from ProQuest (Hospital Premium Collection) - NHS Version

Available at [Contemporary OB/GYN](#) - from EBSCO (CINAHL with Full Text)

Abstract:The article offers insights into the link between cardiovascular disease (CVD) and maternal morbidity and mortality. Topics mentioned include the types of CVD that prevail during pregnancy, the need of coordinated care during pregnancy by involving a team of heart specialists, the physiological changes that may occur during pregnancy, the types of defects present in patients with congenital heart disease, and the importance of health education about the risk of CVD in pregnancy.

Database: CINAHL

45. Maternal congenital cardiac disease: Evaluating prognostic scores and the location of the lesion

Author(s): Phippen J.L.; Thompson J.L.; Markham L.; Frischhertz B.

Source: American Journal of Obstetrics and Gynecology; Jan 2018; vol. 218 (no. 1)

Publication Date: Jan 2018

Publication Type(s): Conference Abstract

Abstract:OBJECTIVE: We sought to describe both maternal and neonatal outcomes in women with congenital heart disease delivering at a single academic medical center and compare three prognostic scoring systems - mWHO, ZAHARA, CARPREG, to determine the best predictor of cardiac event in pregnancy. STUDY DESIGN: We conducted a retrospective cohort study of pregnancies occurring in women with congenital heart disease who received prenatal care and delivered at Vanderbilt University Medical Center between 2009 - 2016. We evaluated the association of each cardiac risk system with postpartum cardiac event and calculated the sensitivity, specificity, PPV and NPV of each system. We also sought to compare the frequency of postpartum cardiac event and other maternal and neonatal morbidity markers with the type of predominant cardiac lesion (left/complex versus right). Student's t-test and chi-square tests were used for analysis. RESULTS: A total of 81 pregnancies were included. Most patients were Caucasian (56, 69.1%), utilized public insurance (55, 67.9%) and had surgically repaired lesions (61, 75%). Most cardiac lesions were mWHO I/II (63, 77.8%). Seven patients had a postpartum cardiac event. Women with a mWHO ≥ 3 were more likely to have a postpartum cardiac event (RR 4.67, $p = 0.03$). Specificity was greatest with the ZAHARA system, 95%. Sensitivity was greatest for the CARPREG, 71%. PPV was low for all three scoring systems (mWHO 22%, CARPREG 16%, ZAHARA 33%). NPV was similar among the three systems (mWHO 95%, CARPREG 96%, ZAHARA 93%). Postpartum ICU admission (57% vs 42.9%) and overall cardiac events (57 vs 42.9%) occurred more frequently in women with right sided cardiac lesions ($p = 0.40$). Preterm birth (68.5% vs 31.5%, $p = 0.16$) and IUGR (68.5% vs 31.5%, $p = 0.13$) were more common in women with right sided cardiac lesions. CONCLUSION: A mWHO ≥ 3 increases risk for significant postpartum cardiac event. The mWHO, CARPREG and ZAHARA systems demonstrate the ability to rule out the possibility of a postpartum cardiac event with higher specificity and NPV. There is no evidence of association of location of cardiac lesion with maternal or neonatal morbidity; however, results are limited due to sample size. Additional research is needed to further evaluate the maternal and obstetrical risks in pregnancy in women with congenital heart disease.

Database: EMBASE

46. Heart failure hospitalization in adults with congenital heart diseases: What predicts it and how does it affect mortality?

Author(s): Cohen S.; Liu A.; Wang F.; Guo L.; Therrien J.; Marelli A.

Source: Archives of Cardiovascular Diseases Supplements; Jan 2018; vol. 10 (no. 1); p. 137

Publication Date: Jan 2018

Publication Type(s): Conference Abstract

Abstract:Background Adults with congenital heart disease (ACHD) are not cured and residual abnormalities predispose them to heart failure (HF). Aim To calculate the cumulative incidence of HF and assess the impact of HF on mortality. To identify predictors of one-year risk of HF. Methods This population-based retrospective cohort of 27,975 ACHD aged 18-65 in 1995-2010 was based on the Quebec CHD database. We calculated the cumulative probability of HF hospitalization using the practical incidence estimator macro to adjust for the competing risk of death. To assess the impact of HF on mortality, we first used propensity score matching to select random controls for each HF

hospitalized patient. We then compared the mortality rates between cases and their matched controls. Finally, we applied nested-case control study and conducted logistic regression analyses to identify the predictors of one-year risk of HF hospitalization. We further used the regression model to construct a risk scoring system (RAAID-HF) for HF hospitalization to identify patients at high-risk of 1-year HF hospitalization. Results The lifetime cumulative risk of HF hospitalization by age 65 was 33.2%. HF hospitalization was associated with a 5-fold increase in mortality risk (HR = 5.4, 95% CI: 3.5, 8.3). Age, sex, CHD severity, HF hospitalization history and comorbidities (arrhythmia, pulmonary hypertension, coronary heart disease, diabetes, hypertension) were significant predictors of one-year HF hospitalization. The RAAID-HF had excellent predictive performance for HF hospitalization (C-statistics = 0.92). Conclusion HF is a common comorbid condition in ACHD patients and is strongly associated with an risk of death in ACHD population. We developed a convenient clinical risk score for predicting the risk of HF hospitalization within a year. These data enable targeting patients at high risk for 1-year HF hospitalization for accelerated referral to CHD centers.

Database: EMBASE

47. Maternal and fetal outcome in patients with cyanotic congenital heart disease: A multicenter observational study

Author(s): Ladouceur M.; Basquin A.; Radojevic J.; Hauet Q.; Hascoet S.; Mocerri P.; Le Gloan L.; Amedro P.; Lucron H.; Richard A.; Gouton M.; Benoit L.; Nizard J.

Source: Archives of Cardiovascular Diseases Supplements; Jan 2018; vol. 10 (no. 1); p. 134

Publication Date: Jan 2018

Publication Type(s): Conference Abstract

Abstract:Background Maternal cyanotic congenital heart disease (CHD) is considered a great maternal and fetal risks during pregnancy, but information on management of these pregnancies are lacking. The purpose of this study was to assess maternal and fetal outcome in patients with cyanotic CHD in a large cohort of patients. Methods This multicenter retrospective study included pregnant women with cyanotic CHD followed in 11 French specialized centers from 1997 to 2015. Patients with pulmonary hypertension were excluded. We recorded maternal, obstetrical and neonatal outcome. Results Thirty-one patients (mean age 27+/-6 years) had 71 pregnancies. There were 17 (26%) miscarriages and 48 (73%) complete pregnancies (≥ 20 week gestation (WG)). All pregnancies were singleton. Severe cardiac events occurred in 7 patients (23%, 95% CI [10-41]) and 6 complete pregnancies (8%, 95% CI [3-17]). Heart failure (n = 3) and arrhythmia (n = 2) were the main cardiovascular complications. There was no maternal death. No thromboembolism event occurred, and one patient experienced an infective endocarditis during postpartum. Obstetrical complications included mainly hemorrhages (n = 9, 13% of pregnancies). Small for gestational age (SGA) was diagnosed in 28%. The mean birth weight was 1897+/-607 g at a mean gestational age of 33+/-3WG, and 85% of newborns were premature. These two comorbidities were associated with an 11% neonatal mortality. Pre-pregnancy maternal oxygen saturation $\leq 85\%$ was related to miscarriages and SGA ($P \leq 0.04$). Conclusion Women with cyanotic CHD can go through pregnancy with a low risk for themselves. However, cyanotic CHD is associated with a high incidence of fetal and neonatal complications.

Database: EMBASE

48. Exposure to low-dose ionizing radiation from cardiac procedures and risk of cancer in adults with congenital heart disease

Author(s): Cohen S.; Liu A.; Guo L.; Therrien J.; Marelli A.; Gurvitz M.; Laprise C.; Kaufman J.; Abrahamowicz M.

Source: Archives of Cardiovascular Diseases Supplements; Jan 2018; vol. 10 (no. 1); p. 133-134

Publication Date: Jan 2018

Publication Type(s): Conference Abstract

Abstract:Background The increasing exposure to low-dose ionizing radiation (LDIR) from cardiac imaging in adults with congenital heart disease (ACHD) has raised concerns about the risk of malignancy. Aim To estimate the association between LDIR exposure from cardiac procedures and incident cancer in ACHD. Methods This retrospective study of 24,833 ACHD aged 18-64 years from 1995-2009 was based on the Quebec CHD Database. Cumulative numbers and cumulative effective dose of LDIR-related cardiac procedures were measured for each patient. We assessed if high LDIR-exposure (≥ 6 procedures) was associated with an increased risk of cancer than low LDIR-exposure (≤ 1). Propensity score and inverse probability weighting were used to adjust for potential confounders. Further, we conducted a nested case-control study to investigate if LDIR-exposure was predictive of cancer using a multivariable logistic regression model. Each case was matched on sex, CHD severity, age and calendar time with 4 randomly selected controls. Results In over 250,791 person-years of follow-up, 602 cancer cases were observed. High LDIR-exposure was associated with a 2.6-fold increase risk of cancer (HR = 2.61, 95% CI: 2.30-2.96). In the nested case-control study, cumulative LDIR-exposure was independently associated with cancer (OR = 1.08 per procedure; 95% CI, 1.04-1.13). Similar results were obtained using dose estimates (OR = 1.10 per 10 milliSieverts; 95% CI: 1.05-1.15) with an apparent dose-response relationship. The effect measure was in the same direction after excluding smoking-related cancer cases (OR = 1.10 per procedure; 95% CI: 1.05-1.16) and after applying a three-year lag time (OR = 1.09 per procedure; 95% CI: 1.03-1.14). Conclusion To our knowledge, this is the first population-based study to document the association between LDIR from cardiac procedures and cancer in ACHD. This finding supports policy recommendations for radiation surveillance in CHD patients where no regulation currently exists.

Database: EMBASE

49. Surgical closure of ventricular septal defect in adults: A multicenter study

Author(s): Han Yee Yu M.; Iriart X.; Jalal Z.; Thambo J.B.; Seguela P.E.; Roubertie F.; Henaine R.

Source: Archives of Cardiovascular Diseases Supplements; Jan 2018; vol. 10 (no. 1); p. 133

Publication Date: Jan 2018

Publication Type(s): Conference Abstract

Abstract:Background Although ventricular septal defect (VSD) is one of the most common congenital heart disease, this defect is not so frequent in adults because of early surgery and spontaneous closure. Complications such as aortic insufficiency, infectious endocarditis or supraventricular arrhythmia are classically described. Purpose The main objective of this study was to determine the outcome of adult patients who were operated for VSD after childhood. Methods We conducted an observational, retrospective and multicenter study including patients operated for VSD from the age of 18 years, over a 15-year period (2000-2015). Overall survival and occurrence of adverse events were analyzed. Results Seventy-three patients were included. Mean age was 37.47 \pm 2.1 years with a male predominance (62%). Type 2 VSDs were the most common type encountered. After surgery, overall survival was 92% with a mean follow-up of 6 years. Excluding events occurring during the early postoperative period, overall survival was significantly better in patients operated for isolated VSD than in patients operated for VSD associated with other cardiac injuries (95% versus 92%, P = 0.02). Early postoperative complications were found in 14% of patients (principally atrioventricular

block requiring early postoperative permanent pacing). Aortic cross clamp and cardiopulmonary bypass time were significantly lower in patients operated for isolated VSD than in patients with associated lesion. Conclusion As morbi-mortality is low, surgical closure of VSD in adults is not a high-risk intervention. Survival of patients with isolated VSD is better than that of patients with associated cardiac lesions. This last finding advocates for early surgery before the apparition of complications.

Database: EMBASE

50. Increasing prevalence of atrial fibrillation and permanent atrial tachyarrhythmias in the aging population with congenital heart disease: A multicenter study

Author(s): Labombarda F.; Dore A.; Macle L.; Mondesert B.; Mongeon F.P.; Proietti A.; Rivard L.; Khairy P.; Hamilton R.; Shohoudi A.; Aboulhosn J.; Broberg C.; Cohen S.; Cook S.; Fernandes S.; Fournier A.; Kay J.; Opatowsky A.R.; Ting J.G.; Zaidi A.

Source: Archives of Cardiovascular Diseases Supplements; Jan 2018; vol. 10 (no. 1); p. 132

Publication Date: Jan 2018

Publication Type(s): Conference Abstract

Abstract:Background Atrial arrhythmias are the most common complication encountered in the growing and aging population with congenital heart disease (CHD). We assessed the types and patterns of atrial arrhythmias, associated factors, and age-related trends. Methods A multicenter cohort study enrolled 482 patients with CHD and atrial arrhythmias, age 32.0+/-18.0 years, 45.2% female, from 12 North American centers. Qualifying arrhythmias were classified by a blinded adjudicating committee. Results The most common presenting arrhythmia was intra-atrial reentrant tachycardia (IART; 61.6%), followed by atrial fibrillation (AF; 28.8%), and focal atrial tachycardia (FAT; 9.5%). Patients with FAT were the youngest at presentation (23.6+/-18.5 years) followed by IART (28.8+/-16.8 years) and AF (41.0+/-17.2 years), $P < 0.0001$. The proportion of arrhythmias due to IART increased with CHD complexity from 47.2% to 62.1% to 67.0% in patients with simple, moderate, and complex defects, respectively ($P = 0.0013$). AF comprised 9.3% of arrhythmias in patients < 20 years and increased with age to surpass IART as the most common arrhythmia in those ≥ 50 years (51.2% versus 44.2%, $P < 0.0001$). Older age [odd ratio (OR) 1.024 per year, 95% confidence interval (CI) 1.010, 1.039], $P = 0.001$ and hypertension [OR 2.00, 95% CI (1.08, 3.71), $P = 0.029$] were independently associated with AF. During a mean followup of 11.3+/-9.4 years, the predominant arrhythmia pattern was paroxysmal in 62.3%, persistent in 28.2%, and permanent in 9.5%. Permanent atrial arrhythmias increased with age from 3.1% in patients < 20 years to 5.5%, 12.9%, and 22.6% in ages 20-35, 35-50, and ≥ 50 years, respectively ($P < 0.0001$). Conclusion IART is the most common presenting atrial arrhythmia in patients with CHD, with a predominantly paroxysmal pattern. However, as the population ages, atrial arrhythmias become increasingly permanent. AF surpasses IART as the most prevalent type of atrial arrhythmia over the age of 50 years.

Database: EMBASE

51. Administrative Health Databases for addressing emerging issues in adults with congenital heart diseases

Author(s): Cohen S.; Iserin L.; Gilutz H.; Marelli A.; Bonnet D.; Burgun A.

Source: Archives of Cardiovascular Diseases Supplements; Jan 2018; vol. 10 (no. 1); p. 142-143

Publication Date: Jan 2018

Publication Type(s): Conference Abstract

Abstract:Background As congenital heart disease (CHD) patients get older, they become at an increased risk of life-long disease burden. Given the lack of epidemiological and longitudinal data on CHD patients across the life-span, secondary use of administrative health databases (AHD) provides opportunities to address emerging issues in adults with CHD (ACHD) population and to study specific outcomes. Aim To systematically review all studies using AHD for ACHD research purpose. Methods and results We systematically searched PubMed and Embase from January 1, 2006 to December 31, 2015 for studies based on secondary use of AHD and providing new knowledge on ACHD. Of the 2217 identified abstracts, 59 studies were included in this review. They derived from 12 different AHD from 6 different countries. The majority of them were performed in North America [n = 32, (55%) from the US; n = 17, (28%) from Canada]. Only 4 (7%) were conducted in Europe and 6 (10%) in Asia. No qualified study was published before 2007. From 2006 to 2015, the number of articles grew exponentially. Study designs were various with a majority of cross-sectional and cohort studies. Since the data routinely derived from health insurance claims and cover long period of follow-up, assessing resource utilization and temporal trends were the most reported objectives. Due to their large size, they allow investigating relatively rare topics on a large scale, such as ACHD long-term complications or associated comorbidities. Furthermore, health care systems and thus, AHD differ substantially from one another. Conclusion Although not designed for research purposes, AHD are particularly powerful for studying ACHD population because they record all diagnoses on large groups of patients from large areas and for a long period of follow-up. Since CHD is associated with lifelong comorbidities and requires lifelong specialized care, longitudinal studies across the lifespan are the cornerstone in assessing ACHD issues.

Database: EMBASE

52. The burden of breast cancer in adult women with congenital heart disease

Author(s): Cohen S.; Liu A.; Guo L.; Marelli A.

Source: Archives of Cardiovascular Diseases Supplements; Jan 2018; vol. 10 (no. 1); p. 142

Publication Date: Jan 2018

Publication Type(s): Conference Abstract

Abstract:Background Breast cancer (BC) is the most common cancer among women in Quebec. Screening plays an important role in reducing its burden. BC incidence in adult women with congenital heart disease (ACHD) has not been studied. Aim To compare ACHD women BC incidence rates to general population. To assess if ACHD women have comparable BC screening rates to non-ACHD women. Methods The Quebec CHD database which includes 84,498 CHD patients was used to calculate the incidence rate of BC among ACHD women in 1992-2009. Age-specific and cumulative incidence of BC were calculated. The age-standardized incidence rate ratio (ASIR) was also calculated using direct standardization to compare rate to the general population and for 4 periods of time. Yearly BC screening participation rate was measured from 1989-2009, as the percentage of screen-eligible women, aged 50-69, who completed at least one mammogram within the 24 months. We compared it to the rate in Quebec general population. Results The incidence rate of BC progressively increased with advancing age with 21% of cases < 50 y, 45% in 50-69 y. In our cohort, 1 out of 10 women (9.8%; 95% CI, 8.6-11.0) developed BC over the course of adulthood. The ASIR was less < 1 before 2006 and 1.25 (95% CI, 1.23-1.26) in 2006-2009 indicating an increased risk of BC among ACHD women than general population in the recent era. Compared to the general population, ACHD women had lower yearly BC screening participation rates from 1989 (18.3% vs. 22.1%) to 2009 (64.6% vs. 67.9%) and still inferior to the official target participation rate (70%). Conclusion The incidence of BC in ACHD women has increased and is now higher than the general population.

Primary care physicians and cardiologists should collaborate to ensure appropriate cancer screening for ACHD women among whom attention to cancer screening may be overlooked. Further analyses will assess the impact of radiation from cardiac imaging and BC risk in this high exposed population.

Database: EMBASE

53. New York Heart Association (NYHA) classification in adults with congenital heart disease: Relation to objective measures of exercise and outcome

Author(s): Bredy C.; Ministeri M.; Kempny A.; Alonso-Gonzalez R.; Swan L.; Uebing A.; Diller G.-P.; Gatzoulis M.A.; Dimopoulos K.

Source: European Heart Journal - Quality of Care and Clinical Outcomes; Jan 2018; vol. 4 (no. 1); p. 51-58

Publication Date: Jan 2018

Publication Type(s): Article

Abstract: Aims The New York Heart Association functional classification (NYHA class) is often used to describe the functional capacity of adults with congenital heart disease (ACHD), albeit with limited evidence on its validity in this heterogeneous population. We aimed to validate the NYHA functional classification in ACHD by examining its relation to objective measures of limitation using cardiopulmonary exercise testing (CPET) and mortality. Methods and results This study included all ACHD patients who underwent a CPET between 2005 and 2015 at the Royal Brompton, in whom functional capacity was graded according to the NYHA classification. Congenital heart diagnoses were classified according to the Bethesda score. Time to all-cause mortality from CPET was recorded in all 2781 ACHD patients (mean age 33.8 +/- 14.2 years) enrolled in the study. There was a strong relation between NYHA class and peak oxygen consumption (peak VO₂), ventilation per unit in carbon dioxide production (VE/VCO₂) slope and the Bethesda classification (P < 0.0001). Although a large number of 'asymptomatic' (NYHA class 1) patients did not achieve a 'normal' peak VO₂, the NYHA class was a strong predictor of mortality, with an 8.7-fold increased mortality risk in class 3 compared with class 1 (hazard ratio 8.68, 95% confidence interval: 5.26-14.35, P < 0.0001). Conclusion Despite underestimating the degree of limitation in some ACHD patients, NYHA classification remains a valuable clinical tool. It correlates with objective measures of exercise and the severity of underlying cardiac disease, as well as mid- to long-term mortality and should, thus, be incorporated into the routine assessment and risk stratification of these patients. Copyright © Published on behalf of the European Society of Cardiology. All rights reserved. © The Author 2017.

Database: EMBASE

54. Health condition and familial factors associated with health-related quality of life in adolescents with congenital heart disease: A cross sectional study

Author(s): Im Y.-M.; Yun T.-J.; Lee S.

Source: Health and Quality of Life Outcomes; Jan 2018; vol. 16 (no. 1)

Publication Date: Jan 2018

Publication Type(s): Article

Available at [Health and quality of life outcomes](#) - from BioMed Central

Available at [Health and quality of life outcomes](#) - from Europe PubMed Central - Open Access

Available at [Health and quality of life outcomes](#) - from EBSCO (MEDLINE Complete)

Available at [Health and quality of life outcomes](#) - from PubMed Central

Abstract:Background: The focus of clinical care after the repair of congenital heart disease has shifted from saving life of the patient to the patient's quality of life. The purpose of this study was to examine the health condition and familial factors associated with the health related quality of life of adolescents with congenital heart disease. Methods: Ninety-eight adolescents aged 13-19 years were collected from a congenital heart clinic from July 22 to August 23, 2013. Perceptions of parental rearing behaviors, health related quality of life of adolescent with congenital heart disease, and general characteristics were measured. We used multiple linear regression analysis to explore factors that are associated with the health related quality of life of adolescents with congenital heart disease. Results: New York heart association class (Adj R² =.186, p =.000), presence of siblings (Adj R² =.240, p =.010), and mother's emotional warmth (Adj R² =.265, p =.043) were significantly associated with the health related quality of life of adolescents with congenital heart disease. Conclusions: Emotionally warm parental rearing behaviors and the presence of siblings were important familial factors that were positively associated with HRQOL in adolescents with CHD. Therefore, it is important for healthcare providers to develop a greater sensitivity to, and awareness of, the familial influences that may be impacting a subject's HRQOL, as well as the exigencies of the CHD, itself.

Database: EMBASE

55. Educational level and employment status in adults with congenital heart disease

Author(s): Pfitzer C.; Rosenthal L.-M.; Walker C.; Ferentzi H.; Berger F.; Schmitt K.R.L.; Helm P.C.; Bauer U.M.M.

Source: Cardiology in the Young; Jan 2018; vol. 28 (no. 1); p. 32-38

Publication Date: Jan 2018

Publication Type(s): Article

Abstract:Purpose Through this study we aimed to assess the educational level and employment status of adults with CHD in Germany. Methods Data were acquired from an online survey carried out in 2015 by the German National Register for Congenital Heart Defects. A total of 1458 adults with CHD participated in the survey (response rate: 37.6%). For 1198 participants, detailed medical information, such as main cardiac diagnosis and information from medical reports, was available. Results Of the participants surveyed (n=1198), 54.5% (n=653) were female, and the mean age was 30 years. The majority of respondents (59.4%) stated that they had high education levels and that they were currently employed (51.1%). Patients with simple CHD had significantly higher levels of education (p<0.001) and were more likely to be employed (p=0.01) than were patients with complex CHD. Conclusions More than half of the participants had high education levels and the majority were employed. The association between CHD and its severity and individuals' educational attainment should be investigated more closely in future studies. Copyright © Cambridge University Press 2017.

Database: EMBASE

56. Patient-reported outcomes in adults with congenital heart disease: Inter-country variation, standard of living and healthcare system factors

Author(s): Moons P.; Van Bulck L.; Apers S.; Dellborg M.; Berghammer M.; Kovacs A.H.; Luyckx K.; Thomet C.; Budts W.; Enomoto J.; Sluman M.A.; Yang H.-L.; Jackson J.L.; Khairy P.; Cook S.C.; Subramanyan R.; Alday L.; Eriksen K.; Johansson B.; Mackie A.S.; Menahem S.; Caruana M.; Veldtman G.; Soufi A.; Fernandes S.M.; White K.; Callus E.; Kutty S.

Source: International Journal of Cardiology; Jan 2018; vol. 251 ; p. 34-41

Publication Date: Jan 2018

Publication Type(s): Article

Abstract: Aims Geographical differences in patient-reported outcomes (PROs) of adults with congenital heart disease (ConHD) have been observed, but are poorly understood. We aimed to: (1) investigate inter-country variation in PROs in adults with ConHD; (2) identify patient-related predictors of PROs; and (3) explore standard of living and healthcare system characteristics as predictors of PROs. Methods and results Assessment of Patterns of Patient-Reported Outcomes in Adults with Congenital Heart disease - International Study (APPROACH-IS) was a cross-sectional, observational study, in which 4028 patients from 15 countries in 5 continents were enrolled. Self-report questionnaires were administered: patient-reported health (12-item Short Form Health Survey; EuroQOL-5D Visual Analog Scale); psychological functioning (Hospital Anxiety and Depression Scale); health behaviors (Health Behavior Scale-Congenital Heart Disease) and quality of life (Linear Analog Scale for quality of life; Satisfaction With Life Scale). A composite PRO score was calculated. Standard of living was expressed as Gross Domestic Product per capita and Human Development Index. Healthcare systems were operationalized as the total health expenditure per capita and the overall health system performance. Substantial inter-country variation in PROs was observed, with Switzerland having the highest composite PRO score (81.0) and India the lowest (71.3). Functional class, age, and unemployment status were patient-related factors that independently and consistently predicted PROs. Standard of living and healthcare system characteristics predicted PROs above and beyond patient characteristics. Conclusions This international collaboration allowed us to determine that PROs in ConHD vary as a function of patient-related factors as well as the countries in which patients live.

Database: EMBASE

57. Incidence, Predictors, and Mortality of Infective Endocarditis in Adults With Congenital Heart Disease Without Prosthetic Valves.

Author(s): Mylotte, Darren; Rushani, Dinela; Therrien, Judith; Guo, Liming; Liu, Aihua; Guo, Kenneth; Martucci, Giuseppe; Mackie, Andrew S; Kaufman, Jay S; Marelli, Ariane

Source: The American journal of cardiology; Dec 2017; vol. 120 (no. 12); p. 2278-2283

Publication Date: Dec 2017

Publication Type(s): Multicenter Study Journal Article

PubMedID: 29103604

Available at [The American journal of cardiology](#) - from ProQuest (Hospital Premium Collection) - NHS Version

Abstract: Congenital heart disease (CHD) increases the risk of infective endocarditis (IE), though the lesion-specific risk and mortality are poorly defined. Using the population-based Quebec CHD database, we sought to describe the predictors of IE and to evaluate if IE was associated with mortality among adult CHD (ACHD) patients without prior valve replacement surgery. We extracted data on ACHD patients with IE and assessed the lesion-specific incidence of IE, risk factors for IE acquisition, and all-cause 1-year mortality. Among 29,866 ACHD patients, 285 (0.95%) developed IE during follow-up period of 378,901 patient-years, from 1988-2010. The highest and lowest lesion-specific incidences of IE were observed with left-sided lesions (1.61/1000 patient-years) and patent ductus arteriosus (0.24/1000 patient-years), respectively. The following predicted the risk of IE acquisition (odds ratio (OR), 95% confidence interval [CI]): cardiac surgery in the previous 6 months (9.07, 3.98-20.67), endocardial cushion defects (6.65, 3.84-11.53), left-sided lesions (5.11, 3.60-7.25), cyanosis at birth (4.82, 3.12-7.46), ventricular septal defect (2.81, 1.87-4.21), diabetes mellitus (1.65,

1.10-2.48), and recent medical interventions (12.52, 5.23-29.97). Twenty-five (8.77%) patients died within 1-year of IE diagnosis, a substantially elevated rate compared to patients without IE (OR 31.07, 95%CI 16.23-59.49). The risk of death following IE diagnosis was similarly elevated among patients with left-sided, cyanotic and other CHD lesions. In conclusion, the risk of IE in ACHD patients is lesion-specific and is greatest in the context of recent medical interventions. IE is associated with increased 1-year mortality, irrespective of broad CHD lesion grouping.

Database: Medline

58. Incidence, predictors and outcomes of infective endocarditis in a contemporary adult congenital heart disease population.

Author(s): Moore, Benjamin; Cao, Jacob; Kotchetkova, Irina; Celermajer, David S

Source: International journal of cardiology; Dec 2017; vol. 249 ; p. 161-165

Publication Date: Dec 2017

Publication Type(s): Journal Article

PubMedID: 29121720

Abstract:BACKGROUNDThe prevalence of congenital heart disease (CHD) in the adult population is steadily increasing. A substrate of prosthetic material and residual lesions, constantly evolving as surgical techniques change over time, predispose these patients to the potentially devastating complication of infective endocarditis (IE).METHODSWe retrospectively reviewed 2935 patients in our adult CHD database for all cases of endocarditis between 1991 and 2016. Incidence, clinical course and predictors of outcomes were analysed.RESULTSWe document 74 episodes in 62 patients, with an incidence of 0.9 cases/1000 patient years (py). IE was more common in complex CHD (1.4 cases/1000py) and ventricular septal defects (VSDs) (1.9 cases/1000py). Prosthetic material was involved in 47% and left-sided infection predominated (66%). The incidence in bicuspid aortic valves post aortic valve replacement (AVR) was significantly higher than in unoperated valves, being 1.8 and 1.1 cases/1000 patient years respectively. Streptococcus was the most frequently implicated causative organism (37%). Emboli occurred in 34% of cases with a cerebral predilection. 46% of patients required surgery during the admission for IE, most frequently to replace a severely regurgitant bicuspid aortic valve. Early endocarditis-related mortality was 15%, associated with cerebral emboli and acute renal failure.CONCLUSIONSIn a contemporary adult CHD cohort, those with complex underlying lesions, VSDs or an AVR were at higher risk for IE. Mortality remains substantial and is more likely in patients with cerebral emboli and/or acute renal failure.

Database: Medline

59. Pregnancy in women with corrected aortic coarctation: Uteroplacental Doppler flow and pregnancy outcome.

Author(s): Siegmund, Anne S; Kampman, Marlies A M; Bilardo, Caterina M; Balci, Ali; van Dijk, Arie P J; Oudijk, Martijn A; Mulder, Barbara J M; Roos-Hesselink, Jolien W; Sieswerda, Gertjan Tj; Koenen, Steven V; Sollié-Szarynska, Krystyna M; Ebels, Tjark; van Veldhuisen, Dirk J; Pieper, Petronella G; ZAHARA investigators

Source: International journal of cardiology; Dec 2017; vol. 249 ; p. 145-150

Publication Date: Dec 2017

Publication Type(s): Journal Article

PubMedID: 28966042

Abstract:OBJECTIVE Women with repaired coarctation of the aorta (rCoA) are at risk of hypertensive disorders and other complications during pregnancy. Hypertensive disorders in pregnant women are associated with inadequate uteroplacental flow, which is related to adverse offspring outcome. The aim of this study was to investigate the relationship of maternal cardiac function, placental function and pregnancy complications in women with rCoA. METHODS We included 49 pregnant women with rCoA and 69 controls from the prospective ZAHARA-studies (Zwangerschap bij Aangeboren HARTafwijkingen, pregnancy in congenital heart disease). Clinical evaluation, echocardiography and uteroplacental Doppler flow (UDF) measurements were performed at 20 and 32 weeks gestation. Univariable regression analysis was performed. RESULTS Comparison of rCoA and healthy women. In women with rCoA, tricuspid annular plane systolic excursion (TAPSE) decreased during pregnancy (25.7mm to 22.8mm, $P=0.006$). UDF indices and pregnancy complication rates were similar in both groups. Offspring of rCoA women had lower birth weight (3233g versus 3578g, $P=0.001$), which was associated with β -blocker use during pregnancy ($\beta=-418.0$, $P=0.01$). Association of cardiac function and UDF. Right ventricular (RV) function before pregnancy (TAPSE) and at 20 weeks gestation (TAPSE and RV fractional area change) were associated with impaired UDF indices (umbilical artery pulsatility index at 20 weeks $\beta=-0.02$, $P=0.01$, resistance index at 20 and 32 weeks $\beta=-0.01$, $P=0.02$ and $\beta=-0.02$, $P=0.01$ and uterine artery pulsatility and resistance index at 20 weeks gestation $\beta=-0.02$, $P=0.05$ and $\beta=-0.01$, $P=0.02$). CONCLUSIONS Women with rCoA tolerate pregnancy well. However, RV function is altered and is associated with impaired placentation.

Database: Medline

60. Management errors in adults with congenital heart disease: prevalence, sources, and consequences.

Author(s): Cordina, Rachael; Nasir Ahmad, Subha; Kotchetkova, Irina; Eveborn, Gry; Pressley, Lynne; Ayer, Julian; Chard, Richard; Tanous, David; Robinson, Peter; Kilian, Jens; Deanfield, John E; Celermajer, David S

Source: European heart journal; Dec 2017

Publication Date: Dec 2017

Publication Type(s): Journal Article

PubMedID: 29236965

Abstract:Aims Improved survival has resulted in increasing numbers and complexity of adults with congenital heart disease (ACHD). International guidelines recommend specialized care but many patients are still not managed at dedicated ACHD centres. This study analysed referral sources and appropriateness of management for patients referred to our tertiary ACHD Centre over the past 3 years. Methods and results We compared differences in care between patients referred from paediatric/ACHD-trained vs. general adult cardiologists, according to Adherence (A) or Non-Adherence (NA) with published guidelines. Non-Adherent cases were graded according to the severity of adverse outcome or risk of adverse outcome. Of 309 consecutively referred patients (28 ± 14 years, 51% male), 134 (43%) were from general cardiologists (19% highly complex CHD) and 115 (37%) were from paediatric cardiology or ACHD specialists (33% highly complex CHD). Sixty referrals (20%) were from other medical teams and of those, 31 had been lost to follow-up. Guideline deviations were more common in referrals from general compared to CHD-trained cardiologists ($P < 0.001$). Of general cardiology referrals, 49 (37%) were NA; 18 had catastrophic or major complications ($n = 2, 16$ respectively). In contrast, only 12 (10%) of the paediatric/ACHD referrals were NA, but none of these were catastrophic and only 3 were major. Simple, moderate, and highly complex CHD patients were at increased risk of adverse outcome when not under specialized CHD cardiology care ($P = 0.04, 0.009$, and 0.002 , respectively). Conclusion Non-adherence

with guidelines was common in the ACHD population, and this frequently resulted in important adverse clinical consequences. These problems were more likely in patients who had not been receiving specialized CHD care. Configuring healthcare systems to optimize 'whole of life' care for this growing population is essential.

Database: Medline

61. Home-based interval training increases endurance capacity in adults with complex congenital heart disease.

Author(s): Sandberg, Camilla; Hedström, Magnus; Wadell, Karin; Dellborg, Mikael; Ahnfelt, Anders; Zetterström, Anna-Klara; Öhrn, Amanda; Johansson, Bengt

Source: Congenital heart disease; Dec 2017

Publication Date: Dec 2017

Publication Type(s): Journal Article

PubMedID: 29205923

Abstract:OBJECTIVEThe beneficial effects of exercise training in acquired heart failure and coronary artery disease are well known and have been implemented in current treatment guidelines. Knowledge on appropriate exercise training regimes for adults with congenital heart disease is limited, thus further studies are needed. The aim of this study was to examine the effect of home-based interval exercise training on maximal endurance capacity and peak exercise capacity.DESIGNRandomized controlled trial.METHODSTwenty-six adults with complex congenital heart disease were recruited from specialized units for adult congenital heart disease. Patients were randomized to either an intervention group-12 weeks of home-based interval exercise training on a cycle ergometer (n = 16), or a control group (n = 10). The latter was instructed to maintain their habitual physical activities. An incremental cardiopulmonary exercise test and a constant work rate cardiopulmonary exercise test at 75% of peak workload were performed preintervention and postintervention.RESULTSTwenty-three patients completed the protocol and were followed (intervention n = 13, control n = 10). Postintervention exercise time at constant work rate cardiopulmonary exercise test increased in the intervention group compared to controls (median[range] 12[-4 to 52]min vs 0[-4 to 5]min, P = .001). At incremental cardiopulmonary exercise test, peak VO₂ increased 15% within the intervention group (P = .019) compared to 2% within the control group (P = .8). However, in comparison between the groups no difference was found (285[-200 to 535] ml/min vs 17[-380 to 306] ml/min, P = .10). In addition, peak workload at incremental cardiopulmonary exercise test increased in the intervention group compared to controls (20[-10 to 70]W vs 0[-20 to 15]W, P = .003).CONCLUSIONHome-based interval exercise training increased endurance capacity and peak exercise capacity in adults with complex congenital heart disease. Aerobic endurance might be more relevant than peak oxygen uptake with regard to daily activities, and therefore a more clinically relevant measure to evaluate.

Database: Medline

62. 18F-FDG-PET/CT angiography in the diagnosis of infective endocarditis and cardiac device infection in adult patients with congenital heart disease and prosthetic material.

Author(s): Pizzi, María N; Dos-Subirà, L; Roque, Albert; Fernández-Hidalgo, Nuria; Cuéllar-Calabria, Hug; Pijuan Domènech, Antonia; González-Alujas, María T; Subirana-Domènech, M T; Miranda-Barrio, B; Ferreira-González, Ignacio; González-López, Juan J; Igual, Albert; Maisterra-Santos, Olga;

García-Dorado, David; Castell-Conesa, Joan; Almirante, Benito; Escobar Amores, Manuel; Tornos, Pilar; Aguadé-Bruix, Santiago

Source: International journal of cardiology; Dec 2017; vol. 248 ; p. 396-402

Publication Date: Dec 2017

Publication Type(s): Journal Article

PubMedID: 28807509

Abstract:OBJECTIVES Infective endocarditis (IE) and cardiac device infection (CDI) are a major complication in the growing number of patients with congenital heart disease (CHD) reaching adulthood. We aimed to evaluate the added value of 18F-FDG-PET/CT angiography (PET/CTA) in the diagnosis of IE-CDI in adults with CHD and intravascular or intracardiac prosthetic material, in whom echocardiography (ECHO) and modified Duke Criteria (DC) have limitations because of the patients' complex anatomy. METHODS A prospective study was conducted in a referral center with multidisciplinary IE and CHD Units. PET/CTA and ECHO findings were compared in consecutive adult (≥ 18 years) patients with CHD who have prosthetic material and suspected IE-CDI. The initial diagnosis using the DC and the diagnosis with the additional PET/CTA data (DC+PET/CTA) were compared with the final diagnostic consensus established by an expert team at three months. RESULTS Between November-2012 and April-2017, 25 patients (15 men; median age 40 years) were included. Cases were initially classified as definite in 8 (32%), possible in 14 (56%) and rejected in 3 (12%). DC+PET/CTA allowed reclassification of 12/14 (86%) cases initially identified as possible IE. The sensitivity, specificity, PPV, NPV, and accuracy of DC at IE suspicion were 39.1%/83.3%/90.4%/25.5%/61.2%, respectively. The diagnostic performance increased significantly with addition of PET/CTA data: 87%/83.3%/95.4%/61.5%/85.1%, respectively. PET/CTA also provided an alternative diagnosis in 3 patients with rejected IE, and detected pulmonary embolisms in 3 patients. CONCLUSIONS PET/CTA was a useful diagnostic tool in the complex group of adult patients with CHD who have cardiac or intravascular prosthetic material and suspected IE or CDI, providing added diagnostic value to the modified DC (increased sensitivity) and improving case classification.

Database: Medline

63. Clinical implications of eicosapentaenoic acid/arachidonic acid ratio (EPA/AA) in adult patients with congenital heart disease.

Author(s): Kanoh, Miki; Inai, Kei; Shinohara, Tokuko; Tomimatsu, Hirofumi; Nakanishi, Toshio

Source: Heart and vessels; Dec 2017; vol. 32 (no. 12); p. 1513-1522

Publication Date: Dec 2017

Publication Type(s): Journal Article

PubMedID: 28681101

Abstract:Recent studies showed that a low ratio between the levels of eicosapentaenoic acid and those of arachidonic acid (EPA/AA) is associated with higher incidence of coronary artery disease and poor prognosis of heart failure, arrhythmia, and cardiac sudden death. However, the clinical implications of EPA/AA in adult patients with congenital heart disease remain unclear. We aimed to assess the prognostic value of EPA/AA regarding cardiac events in adult patients with congenital heart disease. We measured the serum levels of eicosapentaenoic acid and arachidonic acid in 130 adult patients (median age, 31 years) stratified into two groups according to their EPA/AA (low, ≤ 0.22 ; high, > 0.22). We prospectively analyzed the association between EPA/AA and incidence of cardiac events during a mean observation period of 15 months, expressed in terms of hazard ratio (HR) with 95% confidence interval (95% CI). In the subgroup of patients with biventricular circulation (2VC) (n = 76), we analyzed the same clinical endpoints. In our study population, EPA/AA was not

associated with the incidence of arrhythmic events (HR, 1.52; 95% CI, 0.82-2.85; $p = 0.19$), but low EPA/AA was a predictor of heart failure hospitalization (HR, 2.83; 95% CI, 1.35-6.30; $p < 0.01$). Among patients with 2VC, an EPA/AA of ≤ 0.25 was associated with a significantly higher risk of arrhythmic events (HR, 2.55; 95% CI, 1.11-6.41; $p = 0.03$) and heart failure hospitalization (HR, 5.20; 95% CI, 1.78-18.1; $p < 0.01$). EPA/AA represents a useful predictor of cardiac events in adult patients with congenital heart disease.

Database: Medline

64. Organisation of care for pregnancy in patients with congenital heart disease.

Author(s): Roos-Hesselink, Jolien W; Budts, Werner; Walker, Fiona; De Backer, Julie F A; Swan, Lorna; Stones, William; Kranke, Peter; Sliwa-Hahnle, Karen; Johnson, Mark R

Source: Heart (British Cardiac Society); Dec 2017; vol. 103 (no. 23); p. 1854-1859

Publication Date: Dec 2017

Publication Type(s): Journal Article Review

PubMedID: 28739807

Available at [Heart](#) - from BMJ Journals - NHS

Available at [Heart](#) - from BMJ Journals

Abstract:Improvements in surgery have resulted in more women with repaired congenital heart disease (CHD) surviving to adulthood. Women with CHD, who wish to embark on pregnancy require pre-pregnancy counselling. This consultation should cover several issues such as the long-term prognosis of the mother, fertility and miscarriage rates, recurrence risk of CHD in the baby, drug therapy during pregnancy, estimated maternal risk and outcome, expected fetal outcomes and plans for pregnancy. Prenatal genetic testing is available for those patients with an identified genetic defect using pregestational diagnosis or prenatal diagnosis chorionic villus sampling or amniocentesis. Centralisation of care is needed for high-risk patients. Finally, currently there are no recommendations addressing the issue of the delivery. It is crucial that a dedicated plan for delivery should be available for all cardiac patients. The maternal mortality in low-income to middle-income countries is 14 times higher than in high-income countries and needs additional aspects and dedicated care.

Database: Medline

65. Employment after heart transplantation among adults with congenital heart disease.

Author(s): Tumin, Dmitry; Chou, Helen; Hayes, Don; Tobias, Joseph D; Galantowicz, Mark; McConnell, Patrick I

Source: Congenital heart disease; Dec 2017; vol. 12 (no. 6); p. 794-799

Publication Date: Dec 2017

Publication Type(s): Journal Article

PubMedID: 28703426

Abstract:OBJECTIVEAdults with congenital heart disease may require heart transplantation for end-stage heart failure. Whereas heart transplantation potentially allows adults with congenital heart disease to resume their usual activities, employment outcomes in this population are unknown. Therefore, we investigated the prevalence and predictors of work participation after heart transplantation for congenital heart disease.DESIGNRetrospective review of a prospective

registry.SETTINGUnited Network for Organ Sharing registry of transplant recipients in the United States.PATIENTSAdult recipients of first-time heart transplantation with a primary diagnosis of congenital heart disease, performed between 2004 and 2015.INTERVENTIONSNone.OUTCOME MEASURESEmployment status reported by transplant centers at required follow-up intervals up to 5 y posttransplant.RESULTSAmong 470 patients included in the analysis (mean follow-up: 5 ± 3 y), 127 (27%) worked after transplant, 69 (15%) died before beginning or returning to work, and 274 (58%) survived until censoring, but did not participate in paid work. Multivariable competing-risks regression analysis examined characteristics associated with posttransplant employment, accounting for mortality as a competing outcome. In descriptive and multivariable analysis, pretransplant work participation was associated with a greater likelihood of posttransplant employment, while the use of Medicaid insurance at the time of transplant was associated with a significantly lower likelihood of working after transplant (subhazard ratio compared to private insurance: 0.55; 95% confidence interval: 0.32, 0.95; $P = .032$).CONCLUSIONSEmployment was rare after heart transplantation for congenital heart disease, and was significantly less common than in the broader population of adults with congenital heart disease. Differences in return to work were primarily related to pretransplant employment and the use of public insurance, rather than clinical characteristics.

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March 7 2018, Volume 39, Issue 10

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