

# Adult Congenital Heart Disease Evidence Update



**October 2017**  
**(Quarterly)**

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## Lunchtime Drop-in Sessions

*All sessions last one hour*

### November (13.00-14.00)

Thu 2nd	Literature searching
Fri 10th	Critical Appraisal
Mon 13th	Statistics
Tue 21st	Literature searching
29 <sup>th</sup> Wed	Critical Appraisal

### December (12.00-13:00)

7 <sup>th</sup> Thu	Statistics
15 <sup>th</sup> Fri	Literature Searching

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

**NICE** National Institute for  
Health and Care Excellence

[Durable mechanical circulatory support in teenagers and adults with congenital heart disease: A systematic review](#)

Source: [PubMed](#) - 02 August 2017 - Publisher: International Journal Of Cardiology



No relevant updates

**UpToDate®**

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

### [Medical management of cyanotic congenital heart disease in adults](#)

Author: [Heidi M Connolly, MD, FASE](#)

All topics are updated as new evidence becomes available and our [peer review process](#) is complete.

**Literature review current through:** Sep 2017. | **This topic last updated:** Aug 18, 2017.

### [Pulmonary hypertension in adults with congenital heart disease](#)

Authors: [Heidi M Connolly, MD, FASE](#); [Robert P Frantz, MD](#)

All topics are updated as new evidence becomes available and our [peer review process](#) is complete.

**Literature review current through:** Sep 2017. | **This topic last updated:** Aug 24, 2017.

### [Atrial arrhythmias \(including AV block\) in congenital heart disease](#)

Authors: [Samuel Asirvatham, MD](#); [Heidi M Connolly, MD, FASE](#); [Christopher J McLeod, MB, ChB, PhD](#)

All topics are updated as new evidence becomes available and our [peer review process](#) is complete.

**Literature review current through:** Sep 2017. | **This topic last updated:** Sep 05, 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](mailto:library@uhbristol.nhs.uk)

### 1. Perceptions of Disease-Related Stress: A Key to Better Understanding Patient-Reported Outcomes Among Survivors of Congenital Heart Disease.

**Author(s):** Jackson, Jamie L; Gerardo, Gina M; Daniels, Curt J; Vannatta, Kathryn

**Source:** The Journal of cardiovascular nursing; ; vol. 32 (no. 6); p. 587-593

**Publication Type(s):** Journal Article

**PubMedID:** 27685861

**Abstract:** BACKGROUND Disease-related stressors for survivors of congenital heart disease (CHD) have been qualitatively described but not quantified nor examined in relationship to important patient-reported outcomes (PROs). OBJECTIVE The aims of this study are to (1) identify the types and degree of disease-related stress experienced by CHD survivors based on age, functional status, and sex, (2) examine differences in stress and PROs by age, functional status, and sex, and (3) determine the unique contribution of perceived stress to variability in PROs. METHODS A cross-sectional study of 173 adolescents and emerging and young adults who were recruited from both pediatric and adult CHD clinics was conducted. Participants rated the degree to which they found various aspects of CHD stressful and completed PROs of health-related quality of life and emotional distress. Differences in perceptions of stress across predictors were determined using analyses of variance and  $\chi^2$  analyses. The relative contribution of perceived stress predicting PROs was examined using stepwise linear regression. RESULTS Two items emerged as being stressful for almost half of the sample, including concerns about future health and having scars or other signs of medical procedures. Adolescents reported less perceived stress than emerging or young adults, and survivors with even mild functional limitations reported higher perceived stress than did those without any symptoms. Perceptions of stress significantly contributed to variability in PROs above and beyond other predictors and was the only variable to explain unique variance in emotional distress. CONCLUSIONS Having even mild functional impairment may have significant deleterious consequences on PROs via increased perceptions of stress. Stress may be modifiable using cognitive behavioral therapy.

**Database:** Medline

### 2. Process of Transition for Congenital Heart Patients: Preventing Loss to Follow-up.

**Author(s):** Habibi, Hajar; Emmanuel, Yaso; Chung, Natali

**Source:** Clinical nurse specialist CNS; ; vol. 31 (no. 6); p. 329-334

**Publication Type(s):** Journal Article

**PubMedID:** 28991016

**Abstract:** **PURPOSE** The aim of this article is to provide an overview of our nurse-led transition clinic provided to congenital heart disease patients moving from pediatric into adult care setting. **DESCRIPTION OF THE SERVICE** Nurse-led transition clinic was analyzed at various stages of young adult care from an early stage of 12 to 14 years to entering adult setting at 16 years or older. **METHODS** Overview of current transition service for young adults being transferred from pediatric into adult services highlights the integral role of clinical nurse specialist as a coordinator of care. **RESULTS** The result of the service overview indicates that nurse-led transition service enables patients to build on their knowledge. This is achieved by providing them time and the opportunities to develop an understanding of their condition and the attitudes required to engage with the adult care setting as indicated in the psychology questionnaire from transition day. **CONCLUSION** A nurse-led transition clinic enhances long-term care of patients by supporting the young adults and their family/carer through the transition and transfer of the care to promote the young adult's understanding of their condition and to prevent any lost to follow-up.

**Database:** Medline

### 3. Mortality in adults with congenital heart disease.

**Author(s):** Naidu, Pavithra; Grigg, Leeanne; Zentner, Dominica

**Source:** International journal of cardiology; Oct 2017; vol. 245 ; p. 125-130

**Publication Date:** Oct 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28874283

**Abstract:** **AIM** Retrospective ascertainment of the causes of mortality in the adult congenital heart disease (ACHD) cohort of the Royal Melbourne Hospital (RMH). **METHODS** Deceased patients (n=73) of the 2519 ACHD patients in the Royal Melbourne Hospital registry (commenced in 1991) were identified. Retrospective analysis was undertaken. Age, gender of deceased individuals, and frequency and cause of death in different congenital diagnosis groups was explored. **RESULTS** Between 1991 and 2015, death occurred in 3.3% of the ACHD cohort. Median age at death was 32 years (IQR 26-41.5) and 51% were male. The most frequent underlying cardiac conditions were Eisenmenger's syndrome (22%), pulmonary atresia and ventricular septal defect +/- major aorto-pulmonary collateral arteries (12%), Tetralogy of Fallot (10%), transposition of great arteries (TGA) with intact ventricular septum (8%), single ventricle (8%) and congenitally corrected TGA (5%). The cause of death was available from medical records in 60 (82%) of the 73 patients. The majority of deaths were due to cardiac causes (67%) including sudden death (40%), heart failure (13%), and documented ventricular arrhythmias (8%). The most common non-cardiac cause of death was sepsis (10%). **CONCLUSION** The majority of deaths in this group were due to cardiac causes with sudden death and heart failure being the most common. Identification of risk factors for sudden death might assist identification of patients who may benefit from preventative therapies including implantable cardiac defibrillator.

**Database:** Medline

### 4. Adult congenital heart disease in Greece: Preliminary data from the CHALLENGE registry.

**Author(s):** Giannakoulas, G; Vasiliadis, K; Frogoudaki, A; Ntellos, C; Tzifa, A; Brili, S; Manginas, A; Papaphylactou, M; Parcharidou, D; Kampouridis, N; Pitsis, A; Chamaidi, A; Kolios, M; Papadopoulos, G; Douras, A; Davluros, P; Ntiloudi, D; Karvounis, H; Kalangos, A; Tsioufis, C; Rammos, S; CHALLENGE investigators

**Source:** International journal of cardiology; Oct 2017; vol. 245 ; p. 109-113

**Publication Date:** Oct 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28743482

**Abstract:**BACKGROUNDThe majority of patients with congenital heart disease (CHD), nowadays, survives into adulthood and is faced with long-term complications. We aimed to study the basic demographic and clinical characteristics of adult patients with congenital heart disease (ACHD) in Greece.METHODSA registry named CHALLENGE (Adult Congenital Heart Disease Registry. A registry from Hellenic Cardiology Society) was initiated in January 2012. Patients with structural CHD older than 16years old were enrolled by 16 specialized centers nationwide.RESULTSOut of a population of 2115 patients with ACHD, who have been registered, (mean age 38years (SD 16), 52% women), 47% were classified as suffering from mild, 37% from moderate and 15% from severe ACHD. Atrial septal defect (ASD) was the most prevalent diagnosis (33%). The vast majority of ACHD patients (92%) was asymptomatic or mildly symptomatic (NYHA class I/II). The most symptomatic patients were suffering from an ASD, most often the elderly or those under targeted therapy for pulmonary arterial hypertension. Elderly patients (>60years old) accounted for 12% of the ACHD population. Half of patients had undergone at least one open-heart surgery, while 39% were under cardiac medications (15% under antiarrhythmic drugs, 16% under anticoagulants, 16% under medications for heart failure and 4% under targeted therapy for pulmonary arterial hypertension).CONCLUSIONSACHD patients are an emerging patient population and national prospective registries such as CHALLENGE are of unique importance in order to identify the ongoing needs of these patients and match them with the appropriate resource allocation.

**Database:** Medline

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

**Author(s):** Talluto, Christopher

**Source:** Current opinion in cardiology; Oct 2017

**Publication Date:** Oct 2017

**Publication Type(s):** Journal Article

**PubMedID:** 29028634

**Abstract:**PURPOSE OF REVIEWIn 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 FINDINGSWe 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.SUMMARYWith the development of a successful transition program patients will hopefully receive high-quality, continuous life-long care in the appropriate setting.

**Database:** Medline

## 6. Diagnosis and Management of Noncardiac Complications in Adults With Congenital Heart Disease: A Scientific Statement From the American Heart Association.

**Author(s):** Lui, George K; Saidi, Arwa; Bhatt, Ami B; Burchill, Luke J; Deen, Jason F; Earing, Michael G; Gewitz, Michael; Ginns, Jonathan; Kay, Joseph D; Kim, Yuli Y; Kovacs, Adrienne H; Krieger, Eric V; Wu, Fred M; Yoo, Shi-Joon; American Heart Association Adult Congenital Heart Disease Committee of the



Council on Clinical Cardiology and Council on Cardiovascular Disease in the Young; Council on Cardiovascular Radiology and Intervention; and Council on Quality of Care and Outcomes Research

**Source:** Circulation; Oct 2017

**Publication Date:** Oct 2017

**Publication Type(s):** Journal Article Review

**PubMedID:** 28993401

**Abstract:**Life expectancy and quality of life for those born with congenital heart disease (CHD) have greatly improved over the past 3 decades. While representing a great advance for these patients, who have been able to move from childhood to successful adult lives in increasing numbers, this development has resulted in an epidemiological shift and a generation of patients who are at risk of developing chronic multisystem disease in adulthood. Noncardiac complications significantly contribute to the morbidity and mortality of adults with CHD. Reduced survival has been documented in patients with CHD with renal dysfunction, restrictive lung disease, anemia, and cirrhosis. Furthermore, as this population ages, atherosclerotic cardiovascular disease and its risk factors are becoming increasingly prevalent. Disorders of psychosocial and cognitive development are key factors affecting the quality of life of these individuals. It is incumbent on physicians who care for patients with CHD to be mindful of the effects that disease of organs other than the heart may have on the well-being of adults with CHD. Further research is needed to understand how these noncardiac complications may affect the long-term outcome in these patients and what modifiable factors can be targeted for preventive intervention.

**Database:** Medline

### **7. 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; Oct 2017

**Publication Date:** Oct 2017

**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. METHOD 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. RESULT 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

### **8. Challenges of congenital heart disease in grown-up patients.**

**Author(s):** Schwerzmann, Markus; Schwitz, Fabienne; Thomet, Corina; Kadner, Alexander; Pfammatter, Jean-Pierre; Wustmann, Kerstin

**Source:** Swiss medical weekly; Oct 2017; vol. 147 ; p. w14495

**Publication Date:** Oct 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28975959

**Abstract:** Nowadays, more than 90% of all children born with congenital heart disease (CHD) reach adult life. Although initially considered to be cured, the majority of them continue to need specialised follow-up because they require re-do interventions or are at increased risk of cardiovascular complications and premature death. Arrhythmias are the most common cause of unscheduled hospital visits for grown-up CHD (GUCH) patients, accounting for one third of emergency admissions in these patients. Some GUCH patients are also at increased risk for sudden cardiac death. The principles of arrhythmia management and the prevention of sudden cardiac death in GUCH patients are similar to those used in adults with acquired heart disease, but are not evidence based. Decompensated heart failure is the other leading cause of death. Conventional medical heart-failure therapy for left ventricular dysfunction is not effective in GUCH patients at highest risk of heart failure, i.e., those with right or single ventricular failure. Careful haemodynamic assessment and structural interventions are the first step to consider in GUCH patients presenting with heart failure symptoms. Adults with moderate or complex CHD and regular follow-up in specialised GUCH centres have a survival benefit compared with patients without such follow-up. Cardiac surgery in GUCH patients should be performed by surgeons trained in treatment of CHD, i.e., surgeons also operating on paediatric patients. A structured transition programme with a defined transfer of care from the paediatric to the adult care environment is important to avoid lapses of care in today's adolescents with CHD. For GUCH patients with an intervention performed decades ago and no specific cardiac follow-up in later life, referral to a specialised GUCH centre is recommended and may save lives.

**Database:** Medline

### **9. Illness perceptions in adult congenital heart disease: A multi-center international study.**

**Author(s):** Rassart, Jessica; Apers, Silke; Kovacs, Adrienne H; Moons, Philip; Thomet, Corina; Budts, Werner; Enomoto, Junko; Sluman, Maayke A; Wang, Jou-Kou; Jackson, Jamie L; Khairy, Paul; Cook, Stephen C; Subramanian, Raghavan; Alday, Luis; Eriksen, Katrine; Dellborg, Mikael; Berghammer, Malin; Johansson, Bengt; Rempel, Gwen R; Menahem, Samuel; Caruana, Maryanne; Veldtman, Gruschen; Soufi, Alexandra; Fernandes, Susan M; White, Kamila S; Callus, Edward; Kutty, Shelby; Luyckx, Koen; APPROACH-IS consortium and the International Society for Adult Congenital Heart Disease (ISACHD)

**Source:** International journal of cardiology; Oct 2017; vol. 244 ; p. 130-138

**Publication Date:** Oct 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28669511

**Abstract:**BACKGROUND Illness perceptions are cognitive frameworks that patients construct to make sense of their illness. Although the importance of these perceptions has been demonstrated in other chronic illness populations, few studies have focused on the illness perceptions of adults with congenital heart disease (CHD). This study examined (1) inter-country variation in illness perceptions, (2) associations between patient characteristics and illness perceptions, and (3) associations between illness perceptions and patient-reported outcomes. METHODS Our sample, taken from APPROACH-IS, consisted of 3258 adults with CHD from 15 different countries. Patients completed questionnaires on illness perceptions and patient-reported outcomes (i.e., quality of life, perceived health status, and symptoms of depression and anxiety). Patient characteristics included sex, age, marital status, educational level, employment status, CHD complexity, functional class, and ethnicity. Linear mixed models were applied. RESULTS The inter-country variation in illness perceptions was generally small, yet patients from different countries differed in the extent to which they perceived their illness as chronic and worried about their illness. Patient characteristics that were linked to illness perceptions were sex, age, employment status, CHD complexity, functional class, and ethnicity. Higher scores on consequences, identity, and emotional representation, as well as lower scores on illness coherence and personal and treatment control, were associated with poorer patient-reported outcomes. CONCLUSION This study emphasizes that, in order to gain a deeper understanding of patients' functioning, health-care providers should focus not only on objective indicators of illness severity such as the complexity of the heart defect, but also on subjective illness experiences.

**Database:** Medline

#### 10. 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); Sep 2017

**Publication Date:** Sep 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28954835

Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals

Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals - NHS

**Abstract:**BACKGROUND The 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. METHODS A 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. RESULTS 50 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. **CONCLUSION** Women 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

### 11. 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); Sep 2017

**Publication Date:** Sep 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28942393

Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals

Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals - NHS

**Abstract:** **OBJECTIVE** Galectin-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). **METHODS** In 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. **RESULTS** Galectin-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

### 12. Mortality in pulmonary arterial hypertension due to congenital heart disease: Serial changes improve prognostication.

**Author(s):** Schuijt, M T U; Blok, I M; Zwinderman, A H; van Riel, A C M J; Schuurings, M J; de Winter, R J; Duijnhouwer, A L; van Dijk, A P J; Mulder, B J M; Bouma, B J

**Source:** International journal of cardiology; Sep 2017; vol. 243 ; p. 449-453

**Publication Date:** Sep 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28606655

**Abstract:**BACKGROUND Adult patients with pulmonary arterial hypertension due to congenital heart disease (PAH-CHD) suffer from high mortality. This underlines the importance of adequate risk stratification to guide treatment decisions. Several baseline parameters are associated with mortality, however, their prognostic value may weaken after years of follow-up. Therefore we investigated the prognostic value of serial changes in standard clinical parameters in PAH-CHD. METHODS In this prospective observational cohort study we included consecutive PAH-CHD adults, between 2005 and 2016. Control visits to the outpatient clinic were standardized, including functional, biochemical and echocardiographic tests, according to the guidelines. The prognostic value of serial changes was determined with time-dependent Cox regression. RESULTS Ninety-two patients with PAH-CHD were included (age  $43 \pm 15$  years, 34% male, 38% Down, 73% Eisenmenger). During a median follow-up of 6.0 (IQR 3.7-9.3) years, 35 (38%) patients died. Serial changes in World Health Organization functional classification (WHO-FC, HR 18.34 for onset class IV), six-minute walk distance (6-MWD, HR 0.65 per 50m), oxygen saturation at peak exercise (peak SaO<sub>2</sub>, HR 0.74 per 5%), NTproBNP (HR 2.25 per 1000ng/l) and echocardiographic right ventricular function (TAPSE, HR 0.80 per 0.5cm) significantly predicted mortality. Moreover, serial changes in these parameters were more potent predictors compared to baseline parameters, based on reduction in -2 log likelihood. CONCLUSION Serial changes in standard clinical parameters have more prognostic value compared to baseline parameters in PAH-CHD. Our results emphasize the importance of screening for serial changes since periodical assessment could guide treatment decisions to delay disease progression.

**Database:** Medline

### **13. Modeling Major Adverse Outcomes of Pediatric and Adult Patients with Congenital Heart Disease Undergoing Cardiac Catheterization: Observations from the NCDR IMPACT Registry.**

**Author(s):** Jayaram, Natalie; Spertus, John A; Kennedy, Kevin F; Vincent, Robert; Martin, Gerard R; Curtis, Jephtha P; Nykanen, David G; Moore, Phillip M; Bergersen, Lisa

**Source:** Circulation; Sep 2017

**Publication Date:** Sep 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28882885

**Abstract:**Background -Risk-standardization for adverse events following congenital cardiac catheterization is needed to equitably compare patient outcomes among different hospitals as a foundation for quality improvement. The goal of this project was to develop a risk-standardization methodology to adjust for patient characteristics when comparing major adverse outcomes in the NCDR® IMPACT™ (Improving Pediatric and Adult Congenital Treatment) Registry. Methods -39,725 consecutive patients within IMPACT undergoing cardiac catheterization between January 2011 and March 2014 were identified. Given the heterogeneity of interventional procedures for congenital heart disease, new procedure-type risk categories were derived with empiric data and expert opinion, as were markers of hemodynamic vulnerability. A multivariable hierarchical logistic regression model to identify patient and procedural characteristics predictive of a major adverse event (MAE) or death following cardiac catheterization was derived in 70% of the cohort and validated in the remaining 30%. Results -The rate of MAE or death was 7.1% and 7.2% in the derivation and validation cohorts, respectively. Six procedure-type risk categories and six independent indicators of hemodynamic vulnerability were identified. The final risk adjustment model included procedure-type risk category, number of hemodynamic vulnerability indicators,

renal insufficiency, single-ventricle physiology, and coagulation disorder. The model had good discrimination with a C-statistic of 0.76 and 0.75 in the derivation and validation cohorts, respectively. Model calibration in the validation cohort was excellent with a slope of 0.97 (standard error [SE] 0.04; p-value [for difference from 1]= 0.53) and an intercept of 0.007 (SE 0.12; p-value [for difference from 0]= 0.95). Conclusions -The creation of a validated risk-standardization model for adverse outcomes following congenital cardiac catheterization can support reporting of risk-adjusted outcomes in the IMPACT Registry as a foundation for quality improvement.

**Database:** Medline

#### **14. Pregnancy outcome in women with congenital heart disease: A single-center experience.**

**Author(s):** Ntiloudi, Despina; Zegkos, Thomas; Bazmpani, Maria Anna; Parcharidou, Despoina; Panagiotidis, Theofilos; Hadjimiltiades, Stavros; Karvounis, Haralambos; Giannakoulas, George

**Source:** Hellenic journal of cardiology : HJC = Hellenike kardiologike epitheorese; Sep 2017

**Publication Date:** Sep 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28873334

**Abstract:**OBJECTIVEPregnancies in patients with adult congenital heart disease (ACHD) are often complicated. We aimed to highlight the nature and the rate of these complications in a single-center patient population.METHODSWe retrospectively studied all the pregnancies of women who presented on an outpatient basis, and all pregnancies were reviewed for maternal and fetal outcomes.RESULTSO f 117 pregnancies from 52 ACHD patients (age at pregnancy  $28.3 \pm 6.6$  years), 10 were therapeutically aborted. A proportion of 41.1% of the remaining 107 pregnancies were complicated either with cardiac (3.7%), obstetric (15.0%), or fetal/neonatal (11.2%) adverse events or with spontaneous abortion (17.8%). Hospitalization during pregnancy was required in 10 patients. Elective cesarean sections were initially planned for 31% of the 87 pregnancies, which were finally completed, while 8% required an emergency cesarean section, mostly for obstetric reasons. NYHA class deterioration after pregnancy was detected in 9.3% of our cohort. Modified WHO class III/IV, prior medication use, and higher body mass index (BMI) were related to cardiac complications during pregnancy or NYHA deterioration.CONCLUSIONPregnancies in ACHD patients are at high risk for complications. Advanced modified WHO class, prior medication use, and high BMI were related to adverse cardiac events.

**Database:** Medline

#### **15. The World Database for Pediatric and Congenital Heart Surgery: The Dawn of a New Era of Global Communication and Quality Improvement in Congenital Heart Disease.**

**Author(s):** St Louis, James D; Kurosawa, Hiromi; Jonas, Richard A; Sandoval, Nestor; Cervantes, Jorge; Tchervenkov, Christo I; Jacobs, Jeffery P; Sakamoto, Kisaburo; Stellin, Giovanni; Kirklin, James K

**Source:** World journal for pediatric & congenital heart surgery; Sep 2017; vol. 8 (no. 5); p. 597-599

**Publication Date:** Sep 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28901228

**Abstract:**The World Society for Pediatric and Congenital Heart Surgery was founded with the mission to "promote the highest quality comprehensive cardiac care to all patients with congenital heart disease, from the fetus to the adult, regardless of the patient's economic means, with an emphasis

on excellence in teaching, research, and community service." Early on, the Society's members realized that a crucial step in meeting this goal was to establish a global database that would collect vital information, allowing cardiac surgical centers worldwide to benchmark their outcomes and improve the quality of congenital heart disease care. With tireless efforts from all corners of the globe and utilizing the vast experience and invaluable input of multiple international experts, such a platform of global information exchange was created: The World Database for Pediatric and Congenital Heart Disease went live on January 1, 2017. This database has been thoughtfully designed to produce meaningful performance and quality analyses of surgical outcomes extending beyond immediate hospital survival, allowing capture of important morbidities and mortalities for up to 1 year postoperatively. In order to advance the societal mission, this quality improvement program is available free of charge to WSPCHS members. In establishing the World Database, the Society has taken an essential step to further the process of global improvement in care for children with congenital heart disease.

**Database:** Medline

#### **16. Adult Congenital Heart Disease Intervention: The Canadian Landscape.**

**Author(s):** Frankfurter, Claudia; Asgar, Anita W; Webb, John G; Cantor, Warren J; Velianou, James L; Gobeil, François; Chan, Albert W; Welsh, Robert C; Love, Michael P; Wood, David A; McKenzie, Kevin; Horlick, Eric M

**Source:** The Canadian journal of cardiology; Sep 2017; vol. 33 (no. 9); p. 1201-1205

**Publication Date:** Sep 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28843330

**Abstract:** Once considered a childhood disease, the number of adults living with congenital heart disease (CHD) has now exceeded the number of pediatric patients. The landscape of percutaneous intervention for adult congenital heart disease (ACHD) has evolved over the past decade and has yet to be characterized in Canada. The aim of this study was to begin to understand the current infrastructure underlying ACHD interventions in Canada and to characterize the type and number of interventions being carried out across the country. A cross-sectional national survey was distributed by e-mail to all cardiac catheterization laboratory directors in 2015. All Canadian laboratories involved in ACHD interventions responded, encompassing 19 institutions spanning 69 cardiac catheterization laboratories. A total of 1451 percutaneous interventions were recorded. Nationwide, the most common simple ACHD interventions were for atrial septal defect and patent foramen ovale closures. The most common ACHD interventions of increased complexity were for coarctation stenting and transcatheter pulmonary valve implantation. There was a marked clustering of procedures in Ontario, Québec, British Columbia, and Alberta in keeping with Canada's population-density distribution. A total of 23 ACHD operators were identified, half of whom had ACHD-specific fellowship training. These data can be used as a starting point to inform the present state of affairs in the area and lay the groundwork for further work to assess resource allocation and human resource planning for the care of patients with ACHD in Canada.

**Database:** Medline

#### **17. Pulmonary Valve Procedures Late After Repair of Tetralogy of Fallot: Current Perspectives and Contemporary Approaches to Management.**

**Author(s):** Bhagra, Catriona J; Hickey, Edward J; Van De Bruaene, Alexander; Roche, S Lucy; Horlick, Eric M; Wald, Rachel M

**Source:** The Canadian journal of cardiology; Sep 2017; vol. 33 (no. 9); p. 1138-1149

**Publication Date:** Sep 2017

**Publication Type(s):** Journal Article Review

**PubMedID:** 28843325

**Abstract:** Few topics in adult congenital heart disease have approached the level of scrutiny bestowed on pulmonary valve replacement (PVR) strategies late after tetralogy of Fallot (TOF) repair. Despite the successes of primary surgery for TOF, there is a growing group of adults with residual right ventricular outflow tract and pulmonary valve dysfunction. Patients with residual chronic pulmonic regurgitation as a consequence of earlier surgery can later develop symptoms of exercise intolerance and complications including heart failure, tachyarrhythmias, and sudden cardiac death. Optimal timing of PVR has sparked debate, which has catalyzed increasing research efforts over the past decade. Although performance of PVR in the absence of symptoms is currently on the basis of the rationale that achievement of complete reverse remodelling is highly desirable, whether this approach results in improvement in patient outcomes in the long-term has yet to be shown. Surgical PVR and percutaneous pulmonary valve intervention are different techniques with specific advantages and disadvantages that require careful consideration for each individual patient, alongside the need for requisite reinterventions over the course of a patient's lifetime. Criteria pertaining to referral strategies are ever being refined as newer technologies for percutaneous therapies continue to evolve. In this article we review the literature surrounding the indications for, the optimal timing of, and the approaches to pulmonary valve procedures in adults with previously repaired TOF.

**Database:** Medline

### 18. Successful cardiac transplantation outcomes in patients with adult congenital heart disease.

**Author(s):** Menachem, Jonathan N; Golbus, Jessica R; Molina, Maria; Mazurek, Jeremy A; Hornsby, Nicole; Atluri, Pavan; Fuller, Stephanie; Birati, Edo Y; Kim, Yuli Y; Goldberg, Lee R; Wald, Joyce W

**Source:** Heart (British Cardiac Society); Sep 2017; vol. 103 (no. 18); p. 1449-1454

**Publication Date:** Sep 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28258242

Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals

Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals

Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals

Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals

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Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals - NHS

Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals - NHS

Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals - NHS

**Abstract:** OBJECTIVE The purpose of our study is (1) to characterise patients with congenital heart disease undergoing heart transplantation by adult cardiac surgeons in a large academic medical centre and (2) to describe successful outcomes associated with our multidisciplinary approach to the



evaluation and treatment of adults with congenital heart disease (ACHD) undergoing orthotopic heart transplantation (OHT). **BACKGROUND** Heart failure is the leading cause of death in patients with ACHD leading to increasing referrals for OHT. **METHODS** The Penn Congenital Transplant Database comprises a cohort of patients with ACHD who underwent OHT between March 2010 and April 2016. We performed a retrospective cohort study of the 20 consecutive patients. Original cardiac diagnoses include single ventricle palliated with Fontan (n=8), dextro-transposition of the great arteries after atrial switch (n=4), tetralogy of Fallot (n=4), pulmonary atresia (n=1), Ebstein anomaly (n=1), unrepaired ventricular septal defect (n=1) and Noonan syndrome with coarctation of the aorta (n=1). **RESULTS** Eight patients required pretransplant inotropes and two required pretransplant mechanical support. Nine patients underwent heart-liver transplant and three underwent heart-lung transplant. Three patients required postoperative mechanical circulatory support. Patients were followed for an average of 38 months as of April 2016, with 100% survival at 30 days and 1 year and 94% overall survival (19/20 patients). **CONCLUSIONS** ACHD-OHT patients require highly specialised, complex and multidisciplinary healthcare. The success of our programme is attributed to using team-based, patient-centred care including our multidisciplinary staff and specialists across programmes and departments.

**Database:** Medline

### **19. Surgery for supraventricular tachycardia and congenital heart defects: long-term efficacy of the combined approach in adult patients.**

**Author(s):** Giamberti, Alessandro; Pluchinotta, Francesca R; Chessa, Massimo; Varrica, Alessandro; Vitale, Raffaele; Frigiola, Alessandro; Pappone, Carlo; Ranucci, Marco

**Source:** Europace : European pacing, arrhythmias, and cardiac electrophysiology : journal of the working groups on cardiac pacing, arrhythmias, and cardiac cellular electrophysiology of the European Society of Cardiology; Sep 2017; vol. 19 (no. 9); p. 1542-1548

**Publication Date:** Sep 2017

**Publication Type(s):** Journal Article

**PubMedID:** 27738072

**Abstract:** Aims Supraventricular arrhythmias are a major cause of morbidity and mortality in adult patients with congenital heart disease (CHD). Intraoperative ablation offers an alternative for patients who failed ablation procedures or are requiring concomitant surgical intervention. We present our long-term results with the surgical treatment of arrhythmias in adults with CHD (ACHD) undergoing elective cardiac surgery and the clinical predictors for arrhythmia recurrence. **Methods and results** Between 2002 and 2013, 80 consecutive patients with CHD, mean age of 39 years, underwent intraoperative ablation with monopolar irrigated radiofrequency during cardiac surgery procedures. Significant clinical predictors of arrhythmia recurrence were determined by univariate analysis. We performed 47 right-sided Maze procedures, and 33 Cox-Maze III procedures. In 75 survivors, the ablation was effective immediately. Over an average follow-up period of 72 months (12-155 months), arrhythmias recurred in nine (20%) patients after right-sided Maze, and in six (19%) patients after Cox-Maze III. Eleven patients were controlled with medical therapy, three underwent catheter ablation of the arrhythmia, and one required a permanent pacemaker. Preoperative arrhythmia length  $\geq 3$  years ( $P \leq 0.001$ ), tetralogy of Fallot ( $P \leq 0.006$ ), and preoperative atrial fibrillation ( $P \leq 0.016$ ) were associated with recurrence arrhythmia. Conversely, NYHA class  $< 3$  ( $P \leq 0.047$ ) was associated with a lower risk of recurrence. **Conclusion** Surgical treatment of unresponsive supraventricular arrhythmia during concomitant cardiac surgery in ACHD is a safe and effective procedure. Freedom from arrhythmias recurrence is 75% after 6 years of follow-up. Long-

term recurrence of arrhythmia in these patients seems to be strongly correlated to preoperative arrhythmia duration.

**Database:** Medline

## **20. Adult congenital heart disease: magnitude of the problem.**

**Author(s):** Thakkar, Akanksha N; Chinnadurai, Ponraj; Lin, C Huie

**Source:** Current opinion in cardiology; Sep 2017; vol. 32 (no. 5); p. 467-474

**Publication Date:** Sep 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28617685

**Abstract:** PURPOSE OF REVIEW To define the magnitude of problems faced by patients with adult congenital heart disease (ACHD) and to identify unmet needs for this population. RECENT FINDINGS The ACHD population is estimated to include more than 1 million people in the United States and continues to grow at a steady rate. Owing to the decline in early mortality in this group, modern medicine is now faced by the long-term complications associated with congenital heart disease such as chronic heart failure, increased endocarditis risk, elevated burden of arrhythmias, pulmonary hypertension, valvular dysfunction, and pregnancy. SUMMARY Increasing access to ACHD care, evolution of imaging techniques and transcatheter technology and continued efforts at quality improvement will be key to successfully facing the challenges that are a product of the astounding success of pediatric cardiac surgery.

**Database:** Medline

## **21. Transcatheter interventions in adults with congenital heart disease: Surveys from the Society for Cardiovascular Angiography and Interventions to identify current patterns of care and perception on training requirements.**

**Author(s):** Wadia, Subeer K; Accavitti, Michael J; Morgan, Gareth J; Kenny, Damien; Hijazi, Ziyad M; Jones, Thomas K; Cabalka, Allison K; McElhinney, Doff B; Kavinsky, Clifford J

**Source:** Catheterization and cardiovascular interventions : official journal of the Society for Cardiac Angiography & Interventions; Sep 2017; vol. 90 (no. 3); p. 418-424

**Publication Date:** Sep 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28493591

**Abstract:** BACKGROUND Interventional catheterization is central to the care of Adults with Congenital Heart Disease (ACHD). Current standards for care provision and training in ACHD intervention are lacking. We sought to examine trends in current practice and training among interventionalists. METHODS We analyzed the results of two separate international surveys in June 2016. One was sent to all active members from the Society of Cardiovascular Angiography and Interventions (SCAI) who self-identified Structural Heart Disease or Congenital Heart Disease as a practice area. The second survey was conducted through the Pediatric Interventional Early Career Society (PICES) aimed at pediatric and adult congenital interventionalists in their first seven years after training. The total survey sample sizes were 1,535 and 112, respectively. RESULTS Response rates for the SCAI and PICES surveys were 15% (237/1,535) and 75% (84/112), respectively. Most respondents (74%) worked at institutions with pediatric and adult facilities in proximity (major medical centers). While 75% of SCAI respondents believed complex transcatheter procedures should

be performed by ACHD-trained interventionalists or multidisciplinary teams, only 32% reported such care is being provided at the present time. Most pediatric and adult cardiologists surveyed (73%) do not believe current interventional fellowships provide adequate training for proficiency in ACHD interventions. CONCLUSIONS ACHD management remains underdeveloped in relative terms, particularly in the United States. Significant gaps in interventional standards of practice and future training needs were recognized by this study. Our survey should serve as an impetus to establish training guidelines for physicians who seek to perform ACHD interventions.

**Database:** Medline

## **22. Attitudes and perceptions of pregnant women with CHD: results of a single-site survey.**

**Author(s):** Sabanayagam, Aarthi; Briston, David; Zaidi, Ali N

**Source:** Cardiology in the young; Sep 2017; vol. 27 (no. 7); p. 1257-1264

**Publication Date:** Sep 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28416028

**Abstract:** Introduction CHD occurs in about 1% of the United States population, and is now the most common cardiac condition affecting women during pregnancy. METHODS An anonymous, single-site, cross-sectional, 50-question survey was performed at a national Adult Congenital Heart Disease conference to assess the level of knowledge, attitudes, and perceptions regarding cardiac care during pregnancy in women with CHD. RESULTS A total of 77 women completed the survey. Among them, 50% (n=39) had moderate and 38% (n=29) had severely complex disease; 30% (n=23) of women were told that pregnancy was contraindicated given their underlying cardiac condition. Almost two-thirds (n=50) report being categorised as high risk for adverse cardiovascular events. During pregnancy, 84% (n=65) preferred their cardiologist to have trained in adult CHD, 44% (n=34) were satisfied with adult cardiologists, and 36% (n=28) with paediatric cardiologists. Only 48% (n=37) were aware that a fetal echocardiogram was indicated. Only 35% (n=27) discussed modes of delivery with their providers, and 70% (n=54) preferred their prenatal cardiology visits at an adult hospital. Up to 85% (n=64) of them had discussed contraception with their cardiologists, and 72% (n=56) felt they needed high-risk maternal-fetal medicine to be involved with their care. CONCLUSIONS Despite seeking medical care, these pregnant women did not have a full understanding of their condition and their cardiovascular risk during pregnancy. On the basis of these results, further efforts are needed to improve the knowledge, attitudes, and perceptions of women with CHD in relation to their cardiac and obstetric management during pregnancy.

**Database:** Medline

## **23. Imaging adult patients with Fontan circulation.**

**Author(s):** Ginde, Salil; Goot, Benjamin H.; Frommelt, Peter C.

**Source:** Current Opinion in Cardiology; Sep 2017; vol. 32 (no. 5); p. 521-528

**Publication Date:** Sep 2017

**Publication Type(s):** Academic Journal

**PubMedID:** 28548989

**Abstract:** Purpose Of Review: Survival after the Fontan procedure for palliation of single ventricle congenital heart disease has improved. However, adults with Fontan circulation are at risk for several complications including heart failure, thromboembolism, and protein-losing enteropathy.

This review discusses the role of noninvasive imaging for surveillance and early detection of anatomic and functional abnormalities of the Fontan circulation that can impact the risk for Fontan failure over time. Recent Findings: Echocardiography is the first-line imaging modality for the adult Fontan patient. Use of established techniques, such as tissue Doppler imaging, and newer techniques, such as myocardial deformation and three-dimensional imaging, has improved the ability of echocardiography to serially assess ventricular and valvular function in this population. Strain imaging, in particular, is effective for early detection of subclinical ventricular dysfunction, is reproducible and can be incorporated into a routine clinical echocardiography protocol. Cardiac magnetic resonance (CMR) imaging complements echocardiography and overcomes the limitation of poor acoustic windows in adult patients, especially with regards to visualizing the cavopulmonary anastomoses and pulmonary arteries. High resolution imaging with CMR provides reliable assessment of ventricular size and function. Novel techniques utilizing CMR, such as computational fluid dynamics, have provided important insights into Fontan fluid dynamics, and the impact of Fontan geometry on flow efficiency through the circulation. Summary: Recent advances in echocardiography and CMR have improved detection of structural and functional abnormalities in adults with Fontan circulation and are essential in monitoring for complications in this growing population.

**Database:** CINAHL

#### **24. Factors associated with coronary artery disease and stroke in adults with congenital heart disease.**

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

**Source:** Heart (British Cardiac Society); Aug 2017

**Publication Date:** Aug 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28847851

Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals

Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals - NHS

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

mechanical valves) suggesting a cardioembolic aetiology. These findings may inform surveillance and prevention strategies.

**Database:** Medline

## **25. Pregnancy in Women with Congenital Heart Disease.**

**Author(s):** Yucel, Evin; DeFaria Yeh, Doreen

**Source:** Current treatment options in cardiovascular medicine; Aug 2017; vol. 19 (no. 9); p. 73

**Publication Date:** Aug 2017

**Publication Type(s):** Journal Article Review

**PubMedID:** 28828743

**Abstract:**OPINION STATEMENTAdvances in cardiac surgical interventions in infancy and childhood have led to an increased number of women with congenital heart disease of childbearing age. For these women, individualized preconception counseling and pregnancy planning should be a vital component of their medical management, and presentation for obstetric care may even be an opportunity to re-establish cardiovascular care for patients who have been lost to follow-up. These patients have unique cardiovascular anatomy and physiology, which is dependent upon the surgical intervention they may have undergone during childhood or adolescence. These factors are associated with a variety of long-term complications, and the normal hemodynamic changes of pregnancy may unmask cardiac dysfunction and pose significant risk. Among three published risk assessment algorithms, the World Health Organization classification is the most sensitive in predicting maternal cardiovascular events in this population. Women with simple congenital heart defects generally tolerate pregnancy well and can be cared for in the community with careful monitoring. Conversely, women with complex congenital defects, with or without surgical repair and/or residual defects, should be managed in tertiary care centers under a multidisciplinary team of physicians experienced in adult congenital heart disease and high-risk obstetrics, who collaboratively participate in pregnancy planning, management, and care through childbirth and postpartum. Women who are cyanotic with oxygen saturation less than 85%, have significant pulmonary arterial hypertension of any cause, or have systemic ventricular dysfunction should be counseled to avoid pregnancy due to a very high risk of maternal and fetal mortality.

**Database:** Medline

## **26. Pacing in congenital heart disease - A four-decade experience in a single tertiary centre.**

**Author(s):** Midha, Disha; Chen, Zhong; Jones, David G; Williams, Howell J; Lascelles, Karen; Jarman, Julian; Clague, Jonathan; Till, Janice; Dimopoulos, Konstantinos; Babu-Narayan, Sonya V; Markides, Vias; Gatzoulis, Michael A; Wong, Tom

**Source:** International journal of cardiology; Aug 2017; vol. 241 ; p. 177-181

**Publication Date:** Aug 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28291620

**Abstract:**BACKGROUNDThe increased risk of brady- and tachy-arrhythmias in the congenital heart disease (CHD) population means that cardiac rhythm management devices are often required at an early age and expose patients to device-related complications. The present study drew upon four decades of experience at a tertiary adult congenital heart disease ACHD center and aimed to investigate the indication for cardiac implantable electronic devices (CIEDs) and predictors of late

device-related complication requiring re-intervention. **METHODS** A retrospective review of pacing records of ACHD patients over forty years was carried out. The primary outcome measure was device related complication requiring re-intervention. **RESULTS** Between 1970 and 2009, 238 structural CHD patients who received CIEDs with follow-up data were identified (structural group). As a comparator group, 98 patients with congenital conduction disease or long QT syndrome with a structurally normal heart (electrical group) were included in the study. During a mean follow-up of  $9.6 \pm 8.5$  years, 72 (21%) patients (44 structural group, 28 electrical group) required  $\geq 1$  re-intervention due to device related complications. Multivariate analysis showed that age at the time of device implant was an independent predictor of late device-related complications (HR 0.77, 95% CI 0.60-0.98,  $p=0.04$ ). Sub-analysis of the structural group showed that ACHD complexity (Bethesda guideline) was the only predictor late device-related complication in the structural group (HR 2.96, 95% CI: 1.67-5.26,  $p<0.01$ ). **CONCLUSION** Increasing age at device implant was inversely associated with late device-related complications. ACHD patients with complex anatomy are at increased risk of device-related complications at mid and long-term follow-up.

**Database:** Medline

## **27. Employment characteristics of a complex adult congenital heart disease cohort.**

**Author(s):** Pickup, L; Gaffey, T; Clift, P; Bowater, S; Thorne, S; Hudsmith, L

**Source:** Occupational medicine (Oxford, England); Aug 2017; vol. 67 (no. 6); p. 453-455

**Publication Date:** Aug 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28898966

**Abstract:** **Background** Due to advances in surgical techniques and subsequent management, there have been remarkable improvements in the survival of patients with congenital heart disease. In particular, larger numbers of patients with complex disease are now living into adulthood and are entering the workforce. **Aims** To establish the types of employment complex adult congenital heart disease (ACHD) patients are engaged in, based on the largest cohort of patients with a single-ventricle circulation in the UK. **Methods** Records of all patients with a univentricular (Fontan) circulation at the Queen Elizabeth Hospital were reviewed. Employment status was categorized according to the Standard Occupational Classification criteria (2010). **Results** A total of 210 patient records were reviewed. There was the same proportion of professionals in our cohort compared to the rest of the UK (20% versus 20%). There were greater proportions working in the caring, leisure and other service occupations (15% versus 9%), the elementary occupations (17% versus 11%), sales and customer service occupations (14% versus 8%) and administrative and secretarial occupations (12% versus 11%). The reverse trend was observed for associate professions and technical occupations (7% versus 14%), skilled trades (10% versus 11%), process, plant and machine operatives (3% versus 6%) and managers, directors and senior officials (2% versus 10%). **Conclusions** The data show that ACHD patients with a single ventricle are engaged in a diverse range of occupations. It is essential that early education and employment advice are given to this cohort to maximize future employment potential.

**Database:** Medline

## **28. Acquired coronary artery disease in adult patients with congenital heart disease: a true or a false problem?**

**Author(s):** Giamberti, Alessandro; Lo Rito, Mauro; Conforti, Erika; Varrica, Alessandro; Carminati, Mario; Frigiola, Alessandro; Menicanti, Lorenzo; Chessa, Massimo

**Source:** Journal of cardiovascular medicine (Hagerstown, Md.); Aug 2017; vol. 18 (no. 8); p. 605-609

**Publication Date:** Aug 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28072626

**Abstract:**BACKGROUNDThe population of adults with congenital heart disease (ACHD) is increasing and aging, and a large percentage of this population is now over 65 years of age. For this reason, it is probable that acquired coronary artery disease (CAD) will become an important issue that needs to be addressed also in these patients. We retrospectively analyzed all ACHD patients who underwent surgery in our Institution with the aim to investigate the incidence of associated CAD and the results of surgical treatment.METHODSFrom January 2000 to December 2015, a total of 1154 ACHD underwent surgery in our center. Fifty patients (4.3%) were diagnosed with acquired CAD and required coronary artery bypass grafting. The mean age at surgery was 66 years (range 41-78 years). The primary diagnosis were atrial septal defect (n=40 patients), Tetralogy of Fallot (n=4 patients), ventricular septal defect (n=2 patients), partial AV canal (n=1), partial anomalous pulmonary venous drainage (n=1), Ebstein's anomaly (n=1), and subaortic stenosis (n=1).RESULTSHospital mortality was 2% (one patient). During a mean follow-up of 9 years (maximum follow-up: 15 years), seven patients died (14%). The actuarial survival was 83% at 5 years and 77% at 10 years. Freedom from reoperation for coronary artery bypass grafting or percutaneous coronary intervention was 88% at 5 years and 82% at 10 years.CONCLUSIONAcquired CAD may coexist with congenital heart defects but the association is quite rare. It typically occurs later during adulthood and it is usually associated with atrial septal defect. Acquired CAD and congenital heart defects can be treated contemporarily with good early and late results.

**Database:** Medline

## 29. Clinical predictors of length of stay in adults with congenital heart disease.

**Author(s):** Cedars, Ari; Benjamin, Lawrence; Burns, Sara V; Novak, Eric; Amin, Amit

**Source:** Heart (British Cardiac Society); Aug 2017; vol. 103 (no. 16); p. 1258-1263

**Publication Date:** Aug 2017

**Publication Type(s):** Multicenter Study Journal Article

**PubMedID:** 28237970

Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals

Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals

Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals

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Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals - NHS

Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals - NHS

**Abstract:**OBJECTIVELength of stay (LOS) is a major driver of inpatient care costs. To date, few studies have investigated risk factors associated with increased LOS in patients with adult congenital heart disease (ACHD). In the present work, we sought to address this knowledge gap.METHODSWe

conducted an analysis of the State Inpatient Databases from Arkansas, California, Florida, Hawaii, Nebraska and New York. We analysed data on admissions in patients with ACHD and constructed a series of hierarchical regression models to identify the clinical factors having the greatest effects on LOS. RESULTS We identified 99 103 inpatient hospitalisations meeting criteria for inclusion. Diagnoses associated with the longest LOS were septicaemia (LOS=14.2 days in patients atrial septal defect, and 11.7 days among all other ACHD) and pericarditis, endocarditis and myocarditis (LOS=13.6 days and 10.0 days, respectively). When separated by underlying anatomy, the variables most consistently associated with longer LOS were bacterial infection, complications of surgeries or medical care, acute renal disease and anaemia. CONCLUSIONS In the present study, we identified risk factors associated with longer LOS in ACHD. These data may be used to identify at-risk patients for targeted intervention to decrease LOS and thereby cost.

**Database:** Medline

### 30. Body mass index in adult congenital heart disease.

**Author(s):** Brida, Margarita; Dimopoulos, Konstantinos; Kempny, Alexander; Liodakis, Emmanouil; Alonso-Gonzalez, Rafael; Swan, Lorna; Uebing, Anselm; Baumgartner, Helmut; Gatzoulis, Michael A; Diller, Gerhard-Paul

**Source:** Heart (British Cardiac Society); Aug 2017; vol. 103 (no. 16); p. 1250-1257

**Publication Date:** Aug 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28237971

Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals

Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals

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**Abstract:** OBJECTIVE Abnormal body mass index (BMI) is associated with higher mortality in various cardiovascular cohorts. The prognostic implications of BMI in adults with congenital heart disease (ACHD) are unknown. We aim to assess the distribution of BMI and its association with symptoms and survival in the ACHD population. METHODS We included 3069 ACHD patients (median age 32.6 years) under follow-up at our institution between 2001 and 2015. Patients were classified based on BMI as underweight (30), and symptoms, exercise capacity and mortality were assessed. RESULTS Overall, 6.2% of patients were underweight, 51.1% had normal weight, 28.2% were overweight and 14.6% were obese. Higher BMI values were associated with lower all-cause and cardiac mortality on univariable Cox analysis, and this effect persisted after adjustment for age, defect complexity, cyanosis and objective exercise capacity. Higher BMI was especially associated with better prognosis in symptomatic ACHD patients (HR 0.94 (95% CI 0.90 to 0.98), p=0.002) and those with complex underlying cardiac defects (HR 0.96 (95% CI 0.91 to 0.997), p=0.048) In patients with a complex cardiac defect who had repeated weight measurements, weight loss was also associated with a worse survival (HR 1.82 (95% CI 1.02 to 3.24), p=0.04). CONCLUSIONS ACHD patients with a higher BMI had a lower mortality. The association between BMI and mortality was especially pronounced in symptomatic patients with complex underlying cardiac defects, suggesting that



cardiac cachexia may play a role. Indeed, weight loss in complex ACHD patients was linked to an even higher mortality.

**Database:** Medline

### 31. Contemporary cardiac surgery for adults with congenital heart disease.

**Author(s):** Beurtheret, Sylvain; Tutarel, Oktay; Diller, Gerhard Paul; West, Cathy; Ntalarizou, Evangelia; Resseguier, Noémie; Papaioannou, Vasileios; Jabbour, Richard; Simpkin, Victoria; Bastin, Anthony J; Babu-Narayan, Sonya V; Bonello, Beatrice; Li, Wei; Sethia, Babulal; Uemura, Hideki; Gatzoulis, Michael A; Shore, Darryl

**Source:** Heart (British Cardiac Society); Aug 2017; vol. 103 (no. 15); p. 1194-1202

**Publication Date:** Aug 2017

**Publication Type(s):** Journal Article Observational Study

**PubMedID:** 28270427

Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals

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Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals

Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals

Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals - NHS

Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals - NHS

Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals - NHS

Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals - NHS

**Abstract:**OBJECTIVEAdvances in early management of congenital heart disease (CHD) have led to an exponential growth in adults with CHD (ACHD). Many of these patients require cardiac surgery. This study sought to examine outcome and its predictors for ACHD cardiac surgery.METHODSThis is an observational cohort study of prospectively collected data on 1090 consecutive adult patients with CHD, undergoing 1130 cardiac operations for CHD at the Royal Brompton Hospital between 2002 and 2011. Early mortality was the primary outcome measure. Midterm to longer-term survival, cumulative incidence of reoperation, other interventions and/or new-onset arrhythmia were secondary outcome measures. Predictors of early/total mortality were identified.RESULTSAge at surgery was 35±15 years, 53% male, 52.3% were in New York Heart Association (NYHA) class I, 37.2% in class II and 10.4% in class III/IV. Early mortality was 1.77% with independent predictors NYHA class ≥ III, tricuspid annular plane systolic excursion (TAPSE) <15 mm and female gender. Over a mean follow-up of 2.8±2.6 years, 46 patients died. Baseline predictors of total mortality were NYHA class ≥ III, TAPSE <15 mm and non-elective surgery. The number of sternotomies was not independently associated with neither early nor total mortality. At 10 years, probability of survival was 94%. NYHA class among survivors was significantly improved, compared with baseline.CONCLUSIONSContemporary cardiac surgery for ACHD performed at a single, tertiary reference centre with a multidisciplinary approach is associated with low mortality and improved functional status. Also, our findings emphasise the point that surgery should not be delayed because of reluctance to reoperate only.

**Database:** Medline

### 32. Chance of surgery in adult congenital heart disease.

**Author(s):** Verheugt, Carianne L; Uiterwaal, Cuno Spm; Vaartjes, Ilonca; van der Velde, Enno T; Zomer, A C; Meijboom, Folkert J; Pieper, Petronella G; Post, Marco C; Vliegen, Hubert W; Hazekamp, Mark G; Grobbee, Diederick E; Mulder, Barbara Jm

**Source:** European journal of preventive cardiology; Aug 2017; vol. 24 (no. 12); p. 1319-1327

**Publication Date:** Aug 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28541122

**Abstract:**Background Young patients with congenital heart disease reaching adulthood face mandatory transition to adult cardiology. Their new cardiologist needs to assess the chances of major future events such as surgery. Using a large national registry, we assessed if patient characteristics at the age of 18 years could predict the chance of congenital heart surgery in adulthood. Design and methods Of 10,300 patients from the CONCOR national registry, we used general patient characteristics at age 18 years, underlying congenital heart defect, history of complications, and interventions in childhood as potential predictors of congenital heart surgery occurring from age 18 years up to age 40 and 60 years. Cox regression was used to calculate hazard ratios with 95% confidence intervals. Analyses were performed separately for all congenital heart surgery and for valvular surgery alone. Results Altogether 2427 patients underwent congenital heart surgery after age 18 years, 1389 of whom underwent valvular surgery. Underlying heart defect, male sex, multiple defects, childhood endocarditis, supraventricular arrhythmia, aortic complications and paediatric cardiovascular surgery, independently predicted adult congenital heart surgery. The mean chance of congenital heart surgery was 22% up to age 40 and 43% up to age 60 years; individual chances spanned from 9-68% up to age 40 and from 19-93% up to age 60 years. Conclusion At the time of transition from paediatric to adult cardiology, an easily obtainable set of characteristics of patients with congenital heart disease can meaningfully inform cardiologists about the patient's individual chance of surgery in adulthood. Our findings warrant validation in other cohorts.

**Database:** Medline

### **33. Incidence, risk factors, and outcomes of acute kidney injury in adults undergoing surgery for congenital heart disease.**

**Author(s):** Kwiatkowski, David M; Price, Elizabeth; Axelrod, David M; Romfh, Anitra W; Han, Brian S; Sutherland, Scott M; Krawczeski, Catherine D

**Source:** Cardiology in the young; Aug 2017; vol. 27 (no. 6); p. 1068-1075

**Publication Date:** Aug 2017

**Publication Type(s):** Journal Article

**PubMedID:** 27869053

**Abstract:**BACKGROUND Acute kidney injury after cardiac surgery is a frequent and serious complication among children with congenital heart disease (CHD) and adults with acquired heart disease; however, the significance of kidney injury in adults after congenital heart surgery is unknown. The primary objective of this study was to determine the incidence of acute kidney injury after surgery for adult CHD. Secondary objectives included determination of risk factors and associations with clinical outcomes. METHODSThis single-centre, retrospective cohort study was performed in a quaternary cardiovascular ICU in a paediatric hospital including all consecutive patients  $\geq 18$  years between 2010 and 2013. RESULTSData from 118 patients with a median age of 29 years undergoing cardiac surgery were analysed. Using Kidney Disease: Improving Global Outcome creatinine criteria, 36% of patients developed kidney injury, with 5% being moderate to severe (stage 2/3). Among higher-complexity surgeries, incidence was 59%. Age  $\geq 35$  years, preoperative left

ventricular dysfunction, preoperative arrhythmia, longer bypass time, higher Risk Adjustment for Congenital Heart Surgery-1 category, and perioperative vancomycin use were significant risk factors for kidney injury development. In multivariable analysis, age  $\geq 35$  years and vancomycin use were significant predictors. Those with kidney injury were more likely to have prolonged duration of mechanical ventilation and cardiovascular ICU stay in the univariable regression analysis. **CONCLUSIONS** We demonstrated that acute kidney injury is a frequent complication in adults after surgery for CHD and is associated with poor outcomes. Risk factors for development were identified but largely not modifiable. Further investigation within this cohort is necessary to better understand the problem of kidney injury.

**Database:** Medline

### **34. Effect of Obesity and Underweight Status on Perioperative Outcomes of Congenital Heart Operations in Children, Adolescents, and Young Adults: An Analysis of Data From the Society of Thoracic Surgeons Database.**

**Author(s):** O'Byrne, Michael L.; Kim, Sunghee; Yerokun, Babatunde A.; Hornik, Christoph P.; Matsouaka, Roland A.; Jacobs, Jeffrey P.; Jacobs, Marshall L.; Jonas, Richard A.

**Source:** Circulation; Aug 2017; vol. 136 (no. 8); p. 704-718

**Publication Date:** Aug 2017

**Publication Type(s):** Academic Journal

**PubMedID:** 28626087

**Abstract:**Background: Extreme body mass index (BMI; either very high or very low) has been associated with increased risk of adverse perioperative outcome in adults undergoing cardiac surgery. The effect of BMI on perioperative outcomes in congenital heart disease patients has not been evaluated.Methods: A multicenter retrospective cohort study was performed studying patients 10 to 35 years of age undergoing a congenital heart disease operation in the Society of Thoracic Surgeons Congenital Heart Surgery Database between January 1, 2010, and December 31, 2015. The primary outcomes were operative mortality and a composite outcome (1 or more of operative mortality, major adverse event, prolonged hospital length of stay, and wound infection/dehiscence). The associations between age- and sex-adjusted BMI percentiles and these outcomes were assessed, with adjustment for patient-level risk factors, with multivariate logistic regression.Results: Of 18 337 patients (118 centers), 16% were obese, 15% were overweight, 53% were normal weight, 7% were underweight, and 9% were severely underweight. Observed risks of operative mortality ( $P=0.04$ ) and composite outcome ( $P<0.0001$ ) were higher in severely underweight and obese subjects. Severely underweight BMI was associated with increased unplanned cardiac operation and reoperation for bleeding. Obesity was associated with increased risk of wound infection. In multivariable analysis, the association between BMI and operative mortality was no longer significant. Obese (odds ratio, 1.28;  $P=0.008$ ), severely underweight (odds ratio, 1.29;  $P<0.0001$ ), and underweight (odds ratio, 1.39;  $P=0.002$ ) subjects were associated with increased risk of composite outcome.Conclusions: Obesity and underweight BMI were associated with increased risk of composite adverse outcome independently of other risk factors. Further research is necessary to determine whether BMI represents a modifiable risk factor for perioperative outcome.

**Database:** CINAHL

### **35. 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); Jul 2017

**Publication Date:** Jul 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28754810

Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals

Available at [Heart \(British Cardiac Society\)](#) - from BMJ Journals - NHS

**Abstract:**OBJECTIVE Infective 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. METHODS All 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. RESULTS There 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. CONCLUSIONS IE-associated morbidity and mortality in a contemporary cohort of ACHD patients is still high in the current era.

**Database:** Medline

### 36. 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; Jul 2017

**Publication Date:** Jul 2017

**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

### **37. Strengths, Limitations, and Geographical Discrepancies in the Eligibility Criteria for Sport Participation in Young Patients With Congenital Heart Disease.**

**Author(s):** Cantinotti, Massimiliano; Giordano, Raffaele; Assanta, Nadia; Murzi, Bruno; Melo, Manuel; Franchi, Eliana; Crocetti, Maura; Iervasi, Giorgio; Kutty, Shelby

**Source:** Clinical journal of sport medicine : official journal of the Canadian Academy of Sport Medicine; Jul 2017

**Publication Date:** Jul 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28742603

**Abstract:** **OBJECTIVE** Benefits of physical activity has been shown in children with congenital heart disease (CHD). In several forms of CHD, the risk of sudden death remains a major concern both for parents and clinicians, who in turn will have to consider the risk-benefit ratio of sport participation versus restriction. **DATA SOURCE** A literature search was performed within the National Library of Medicine using the keywords: Sport, CHD, and Eligibility. The search was further refined by adding the keywords: Children, Adult, and Criteria. **MAIN RESULTS** Fifteen published studies evaluating sport eligibility criteria in CHD were included. Seven documents from various scientific societies have been published in the past decade but which of them should be adopted remains unclear. Our research highlighted accuracy and consistency of the latest documents; however, differences have emerged between the US and European recommendations. Eligibility criteria were consistent between countries for simple congenital heart defects, whereas there are discrepancies for borderline conditions including moderate valvular lesions and mild or moderate residual defects after CHD repair. Furthermore, some of the more severe defects were not evaluated. Multiple recommendations have been made for the same CHD, and cut-off values used to define disease severity have varied. Published eligibility criteria have mainly focused on competitive sports. Little attention was paid to recreational activities, and the psychosocial consequences of activity restriction were seldom evaluated. **CONCLUSIONS** Comprehensive consensus recommendations for sport eligibility evaluating all CHD types and stages of repair are needed. These should include competitive and recreational activities, use standardized classifications to grade disease severity, and address the consequences of restriction.

**Database:** Medline

### **38. 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; Jul 2017

**Publication Date:** Jul 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.

**Database:** Medline

### **39. Prediction of adverse events after catheter-based procedures in adolescents and adults with congenital heart disease in the IMPACT registry.**

**Author(s):** Stefanescu Schmidt, Ada C; Armstrong, Aimee; Kennedy, Kevin F; Nykanen, David; Aboulhosn, Jamil; Bhatt, Ami B

**Source:** European heart journal; Jul 2017; vol. 38 (no. 26); p. 2070-2077

**Publication Date:** Jul 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28430913

**Abstract:**AimsWe sought to identify factors associated with major adverse events (MAE) after cardiac catheterization in adolescents and adults with congenital heart disease (CHD), and create the first model to individualize risk discussions in this growing population.Methods and resultsImproving Pediatric and Adult Congenital Treatment (IMPACT), a National Cardiovascular Data Registry, contains congenital catheterization data from over 87 hospitals in the United States. Demographics, pre-procedure, and procedural variables were collected for patients over age 10. Multivariable logistic regression was used to identify significant predictors of MAE, a composite of death, urgent surgery or procedure due to a catheterization complication, transfusion, embolic stroke, tamponade, extracorporeal membrane oxygenation or ventricular assist device placement, and device

embolization, malposition or thrombosis requiring surgical intervention. A risk score was built based on the effect sizes of each predictor and validated in a split sample. A MAE occurred in 686 (2.5%) of the 27 293 index procedures meeting inclusion criteria. The independent multivariate predictors of MAE were older age, pre-procedural anticoagulation use, renal disease, lower haemoglobin, lower oxygen saturation, non-elective procedure, higher index procedure risk and having had no prior cardiac procedures. Being underweight or overweight had borderline significance and was added to the model. The C-statistic for the model was robust at 0.787 in the derivation and 0.773 in the validation cohort. Conclusion The factors predicting adverse events after cardiac catheterization in adolescents and adults with CHD are different than in the general population. Validation of this model in other national or multi-institutional datasets is the next step.

**Database:** Medline

#### **40. Incidence, risk factors, and predictors of infective endocarditis in adult congenital heart disease: focus on the use of prosthetic material.**

**Author(s):** Kuijpers, Joey M; Koolbergen, Dave R; Groenink, Maarten; Peels, Kathinka C H; Reichert, Constant L A; Post, Marco C; Bosker, Hans A; Wajon, Elly M C J; Zwinderman, Aeilko H; Mulder, Barbara J M; Bouma, Berto J

**Source:** European heart journal; Jul 2017; vol. 38 (no. 26); p. 2048-2056

**Publication Date:** Jul 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28065906

**Abstract:** Aims Adult congenital heart disease (ACHD) predisposes to infective endocarditis (IE). Surgical advancements have changed the ACHD population, whereas associated prosthetic material may constitute additional IE targets. We aimed to prospectively determine contemporary incidence, risk factors, and predictors of IE in a nationwide ACHD cohort, focusing on the presence of prosthetics. Methods and results We identified 14 224 patients prospectively followed in the CONCOR ACHD registry (50.5% female, median age 33.6 years). IE incidence was determined using Poisson regression, risk factors and predictors using Cox regression. Overall incidence was 1.33 cases/1000 person-years (124 cases in 93 562 person-years). For risk-factor analysis, presence of prosthetics was forced-as separate time-updated variables for specific prosthetics-into a model with baseline characteristics univariably associated with IE. Valve-containing prosthetics were independently associated with greater risk both short- and long term after implantation [0-6 months: hazard ratio (HR) = 17.29; 7.34-40.70, 6-12 months: HR = 15.91; 6.76-37.45, beyond 12 months: HR = 5.26; 3.52-7.86], non-valve-containing prosthetics, including valve repair, only in the first 6 months after implantation (HR = 3.34; 1.33-8.41), not thereafter. A prediction model was derived and validated using bootstrapping techniques. Independent predictors of IE were baseline valve-containing prosthetics, main congenital heart defect, multiple defects, previous IE, and sex. The model had fair discriminative ability and provided accurate predictions up to 10 years. Conclusions This study provides IE incidence estimates, and determinants of IE risk in a nationwide ACHD cohort. Our findings, essentially informing IE prevention guidelines, indicate valve-containing prosthetics as a main determinant of IE risk whereas other prosthetics, including valve-repair, are not associated with increased risk long term after implantation.

**Database:** Medline

#### **41. Adult congenital heart disease: the challenges of a lifetime.**

**Author(s):** Warnes, Carole A

**Source:** European heart journal; Jul 2017; vol. 38 (no. 26); p. 2041-2047

**Publication Date:** Jul 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28011704

**Abstract:**The growing population of adults with congenital heart disease (CHD) poses challenges for cardiac surgeons, general cardiologists and CHD sub specialists. The patients themselves, often believing themselves to be 'totally corrected' following operative repair, also face challenges with re-operations and lifelong cardiac problems. This review examines the challenges of the past, present and future for both medical providers and patients.

**Database:** Medline

#### **42. 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; Jul 2017

**Publication Date:** Jul 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,  $\leq 0.22$ ; 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  $\leq 0.25$  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

#### **43. Three-dimensional echocardiography in adult congenital heart disease.**

**Author(s):** Yang, Hyun Suk

**Source:** The Korean journal of internal medicine; Jul 2017; vol. 32 (no. 4); p. 577-588

**Publication Date:** Jul 2017

**Publication Type(s):** Journal Article Review



**PubMedID:** 28704916

Available at [The Korean journal of internal medicine](#) - from Europe PubMed Central - Open Access

**Abstract:** Congenital heart disease (CHD) is now more common in adults than in children due to improvements in fetal echo, neonatal and pediatric care, and surgical techniques leading to dramatically increased survivability into adulthood. Adult patients with CHD, regardless of prior cardiac surgery, experience further cardiac problems or therapeutic challenges; therefore, a non-invasive, easily accessible echocardiographic examination is an essential follow-up tool. Among echocardiographic modalities, three-dimensional (3D) echocardiography provides better delineation of spatial relationships in complex cardiac geometries and more accurate volumetric information without geometric assumptions. For atrial septal defects, an en face view of the tissue defect allows better decisions on device closure. For tricuspid valve malformations, an en face view provides diagnostic information that is difficult to obtain from routine 2D tomography. In repaired tetralogy of fallot with pulmonary regurgitation, preoperative 3D echocardiography-based right ventricular volume may be used to determine the timing of a pulmonary valve replacement in conjunction with cardiovascular magnetic imaging. For optimal adult CHD care, 3D echocardiography is an important complement to routine 2D echocardiography.

**Database:** Medline

#### **44. Outcomes and Costs of Cardiac Surgery in Adults with Congenital Heart Disease.**

**Author(s):** Nasr, Viviane G; Faraoni, David; Valente, Anne Marie; DiNardo, James A

**Source:** Pediatric cardiology; Jul 2017

**Publication Date:** Jul 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28669107

**Abstract:** Advances in pediatric cardiac surgical and medical care have led to increased survival of patients with congenital heart disease (CHD). Consequently, many CHD patients survive long enough to require cardiac surgery as adults. Using the 2013 Nationwide Inpatient Sample (NIS) database, we compared costs and outcomes for adult patients undergoing surgery for treatment of CHD to a reference population of adults undergoing CABG. Patients were identified using International Classification of Diseases, Ninth Revision, Clinical Modification (ICD-9 CM) procedure codes. We recorded the demographic characteristics, gender, ethnicity, hospital bed size, hospital length of stay, in-hospital mortality, and comorbidities. Patients with ACHD have higher incidences of in-hospital mortality (2.6 vs. 1.8%), and complication rates including neurologic complications (2.6 vs. 0.9%), thromboembolic complications (3.9 vs. 1.4%), arrhythmias (51.6 vs. 29.8%), hepatic failure (4.44 vs. 2.03%), and sepsis (7.24 vs. 4.61%) (all p < 6 days (Coefficient = 0.703, 95% CI, 0.697-0.710; p < 0.001). This study shows that ACHD patients undergoing cardiac surgery experience higher hospital costs and poorer outcomes than a reference population of adult CABG patients. Recognition and treatment of comorbidities in ACHD patients undergoing cardiac surgery may provide an opportunity to improve perioperative outcomes in this growing patient population.

**Database:** Medline

#### **45. An Overview of Cardiac Computed Tomography in Adults With Congenital Heart Disease.**

**Author(s):** Suranyi, Pal; Varga-Szemes, Akos; Hlavacek, Anthony M

**Source:** Journal of thoracic imaging; Jul 2017; vol. 32 (no. 4); p. 258-273

**Publication Date:** Jul 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28632654

**Abstract:** Familiarity with congenital heart disease (CHD) and its manifestations in adults is becoming increasingly important for the practicing cardiothoracic imager. The use of computed tomographic angiography is becoming commonplace not only in adults with a history and subsequent interventions for CHD as a child but also in de novo detection of typically-milder, survivable forms of CHD, which are clinically suspected because of declining cardiac performance, cardiac events, or murmurs. Occasionally, adult CHD (ACHD) is found incidentally on scans performed for other indications (eg, trauma or neoplasm staging) because of improvements in computed tomographic technology and advanced visualization. A growing number of patients with CHD survive into adulthood and lead essentially normal lives all over the world and may present in community hospitals and emergency rooms to receive care. Advancements in computed tomographic angiography technology have further allowed us to individually tailor cardiovascular scans according to the patient's anatomy and specifically for the clinical question raised, aiming at reduced exposure to iodinated contrast and radiation dose, while obtaining diagnostic-quality images. This task, however, is not simple. There is no one-size-fits-all cardiac scan that serves as a one-stop shop for ACHD patients. Careful planning and patient preparation, close supervision of the scan, sophisticated postprocessing, and interdisciplinary interpretation of the findings are all required ingredients for managing patients with ACHD. In this review we highlight the technical principles that should be observed to optimize imaging in ACHD. We also provide practical insight into how specific clinical questions can be answered utilizing this technology in the most frequently encountered forms of ACHD.

**Database:** Medline

#### **46. Utility of Cardiac Magnetic Resonance Imaging in the Management of Adult Congenital Heart Disease.**

**Author(s):** Muscogiuri, Giuseppe; Secinaro, Aurelio; Ciliberti, Paolo; Fuqua, Megan; Nutting, Arni

**Source:** Journal of thoracic imaging; Jul 2017; vol. 32 (no. 4); p. 233-244

**Publication Date:** Jul 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28632652

**Abstract:** The increasing number of patients with adult congenital heart disease (ACHD) calls for the development of noninvasive imaging techniques that allow a long-term evaluation of native and postsurgical anatomy and function. Echocardiography remains the imaging modality of choice for congenital heart disease, but it is affected by limited acoustic windows and poor tissue characterization. Cardiac computed tomography and cardiac catheter angiography are 2 valid alternatives for the anatomic and functional assessment of ACHD; however, both use ionizing radiation, and cardiac catheter angiography requires an invasive approach. Cardiac magnetic resonance (CMR), noninvasively and in the absence of ionizing radiation, has the ability to evaluate the biventricular function, quantify flows, characterize tissue, and provide information on cardiac anatomy. Despite the long acquisition time and lower spatial resolution compared with cardiac computed tomography, CMR represents the ideal technique for long-term follow-up of ACHD. CMR is now widely utilized and is well described in the literature with regard to diagnosis, identification of complications, timing of surgery, and postoperative prognosis in ACHD. CMR represents a fundamental technique for the evaluation of patients with ACHD.

**Database:** Medline

#### **47. Pictorial Review of Surgical Anatomy in Adult Congenital Heart Disease.**

**Author(s):** De Cecco, Carlo N; Muscogiuri, Giuseppe; Madrid Pérez, José M; Eid, Marwen; Suranyi, Pal; Lesslie, Virginia W; Bastarrika, Gorka

**Source:** Journal of thoracic imaging; Jul 2017; vol. 32 (no. 4); p. 217-232

**Publication Date:** Jul 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28549022

**Abstract:**The survival rate of patients with congenital heart disease (CHD) has dramatically improved over the last 2 decades because of technological and surgical advances in diagnosis and treatment, respectively. The vast majority of CHD patients are, in fact, amenable to treatment by either device closure or surgery. Considering the wide spectrum of surgical procedures and complex native and derived anatomy, continuous and detailed follow-up is of paramount importance. Cardiac magnetic resonance and cardiac computed tomography angiography are the cornerstones of diagnosis and follow-up of CHD, allowing for comprehensive noninvasive assessment of the heart, coronary tree, and intrathoracic great vessels, along with both morphological and functional evaluation. The aim of this pictorial review is to provide an overview of the most common CHDs and their related surgical procedures as familiarity with the radiological findings of grown-up congenital heart disease patients is crucial for proper diagnostic and follow-up pathways.

**Database:** Medline

#### **48. Imaging in Adult Congenital Heart Disease.**

**Author(s):** Gaydos, Stephanie S; Varga-Szemes, Akos; Judd, Rochelle N; Suranyi, Pal; Gregg, David

**Source:** Journal of thoracic imaging; Jul 2017; vol. 32 (no. 4); p. 205-216

**Publication Date:** Jul 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28632651

**Abstract:**There has been tremendous growth in the population of adults with congenital heart disease (CHD) over the last few decades because of advances in medical care. Whereas some cases are cured during childhood, most patients instead undergo palliation, which leaves them at risk for late complications. Lifelong clinical follow-up involving serial multimodality imaging is helpful to monitor and guide the treatment of late complications. Imaging of these individuals is challenging because of their unique anatomy and therefore requires careful consideration on a case-by-case basis. Knowledge of late complications associated with various diseases and surgical palliations provides a basis for selection and interpretation of an appropriate imaging study. This review highlights the most common conditions in adult CHD and the clinical questions that imaging hopes to answer, as well as the advantages and disadvantages of available imaging modalities. We illustrate clinical scenarios and the use of noninvasive imaging modalities including echocardiography, computed tomography, and magnetic resonance imaging, as well as applications in the following CHDs: tetralogy of Fallot, transposition of the great arteries, coarctation of the aorta, shunts, functional single ventricle, and coronary anomalies.

**Database:** Medline

#### **49. Association Between Depressive Symptoms and Exercise Capacity in Patients With Heart Disease: A META-ANALYSIS.**

**Author(s):** Papasavvas, Theodoros; Alhashemi, Mohammad; Micklewright, Dominic

**Source:** Journal of cardiopulmonary rehabilitation and prevention; Jul 2017; vol. 37 (no. 4); p. 239-249

**Publication Date:** Jul 2017

**Publication Type(s):** Journal Article

**PubMedID:** 27428818

**Abstract:** PURPOSE Depression and reduced exercise capacity are risk factors for poor prognosis in patients with heart disease, but the relationship between the 2 is unclear. We assessed the relationship between depressive symptoms and exercise capacity in patients with heart disease. METHOD PubMed, Cochrane Library, Google Scholar, and ProQuest databases were browsed for English-language studies published from January 2000 to September 2013. Studies including adult patients with coronary artery disease, heart failure, congenital heart disease, and implantable cardioverter defibrillator, reporting correlation between a depression scale and exercise capacity ((Equation is included in full-text article.)  $O_2$  peak, peak watts, estimated metabolic equivalents, and incremental shuttle walk test distance), as well as studies from which such a correlation could be calculated and provided by the authors, were included. Correlation coefficients (CCs) were converted to Fischer z values, and the analysis was performed using a random-effects model. Then, summary effects and 95% CIs were converted back to CCs. RESULTS Fifty-nine studies (25 733 participants) were included. Depressive symptoms were inversely correlated to exercise capacity (CC = -0.15; 95% CI, -0.17 to -0.12). Heterogeneity was significant (I = 64%; P < .001). There was no evidence of publication bias (Fail-safe N = 4681; Egger test: P = .06; Kendall test: P = .29). CONCLUSION Patients with heart disease and elevated depressive symptoms may tend to have reduced exercise capacity, and vice versa. This finding has clinical and prognostic implications. It also encourages research on the effects of improving depression on exercise capacity, and vice versa. The effects of potential moderators need to be explored.

**Database:** Medline

#### **50. Arterial stiffness and arterial function in adult cyanotic patients with congenital heart disease.**

**Author(s):** Trojnariska, Olga; Szczepaniak-Chicheł, Ludwina; Gabriel, Marcin; Bartczak-Rutkowska, Agnieszka; Rupa-Matysek, Joanna; Tykarski, Andrzej; Grajek, Stefan

**Source:** Journal of cardiology; Jul 2017; vol. 70 (no. 1); p. 62-67

**Publication Date:** Jul 2017

**Publication Type(s):** Journal Article

**PubMedID:** 27756510

**Abstract:** BACKGROUND Mortality in cyanotic patients with congenital heart diseases (CHD) is high, mainly due to cardiovascular complications. It is known that endothelial dysfunction, increased arterial stiffness, and impaired vascular function have negative influence on cardiovascular prognosis. The aim of the study was to assess parameters of arterial stiffness and vascular dysfunction in cyanotic patients with CHD as well as their potential relation to impaired blood oxygen saturation and polycythemia parameters typical for cyanosis. METHOD SA total of 36 CHD cyanotic patients (17 males) (42.3±16.3 years) and 35 healthy individuals (16 males) (39.6±10.4 years) were enrolled. Assessed parameters were intima media thickness and flow-mediated dilatation (FMD). Assessed parameters using applanation tonometry methods were aortic systolic

pressure, aortic pulse pressure (AoPP), augmentation pressure (AP), augmentation index (AI), pulse pressure amplification (PPampl), and pulse wave velocity (PWV). RESULTS AoPP ( $37.3 \pm 11.1$  mmHg vs.  $29 \pm 6.5$  mmHg;  $p=0.002$ ), AP ( $10.1 \pm 7.3$  mmHg vs.  $1.1 \pm 3.9$  mmHg;  $p=0.00001$ ), AI ( $24.7 \pm 13.5\%$  vs.  $3.0 \pm 13.6\%$ ;  $p=0.00001$ ), and PWV ( $7.4 \pm 2.1$  m/s vs.  $6.3 \pm 0.7$  m/s;  $p=0.003$ ) were higher, and PPampl was lower ( $135.3 \pm 16.1\%$  vs.  $160.4 \pm 12.8\%$ ;  $p=0.00001$ ) in the studied group compared to controls and proved the presence of the increased stiffness of arteries. Impairment of FMD was observed ( $9.0 \pm 5.6$  vs.  $10.9 \pm 4.7$ ;  $p=0.04$ ). No significant correlations were found between analyzed arterial parameters and biochemical ones characterizing cyanotic patients depicting rheological properties of blood. CONCLUSION Cyanotic patients with CHD are characterized by increased arterial stiffness estimated with pulse wave analysis parameters and by deteriorated arterial function expressed with worse vasodilative response in comparison with healthy population. It may confirm relevance of those mechanisms in development of increased rate of cardiovascular events in this population. Association between oxygen saturation or polycythemia and arterial stiffening or vascular dysfunction was not found in these patients.

**Database:** Medline

### **51. Invasive Hemodynamics of Adult Congenital Heart Disease: From Shunts to Coarctation.**

**Author(s):** Veeram Reddy, Surendranath R; Nugent, Alan W; Zellers, Thomas M; Dimas, V Vivian

**Source:** Interventional cardiology clinics; Jul 2017; vol. 6 (no. 3); p. 345-358

**Publication Date:** Jul 2017

**Publication Type(s):** Journal Article Review

**PubMedID:** 28600089

**Abstract:** Adults with congenital heart disease are a growing population with increasingly more complex disease, in large part due to improvements in delivery of care to the pediatric population. Cardiac catheterization is an integral component of diagnosis and management in these patients. Careful attention to detail and a thorough understanding of intracardiac hemodynamics are critical to performing complete diagnostic evaluations. This article outlines the most commonly encountered lesions with guidelines for invasive assessment to help guide further therapy.

**Database:** Medline

### **52. Adolescents with congenital heart disease: their opinions about the preparation for transfer to adult care.**

**Author(s):** Burström, Åsa; Bratt, Ewa-Lena; Frenckner, Björn; Nisell, Margret; Hanséus, Katarina; Rydberg, Annika; Öjmyr-Joelsson, Maria

**Source:** European journal of pediatrics; Jul 2017; vol. 176 (no. 7); p. 881-889

**Publication Date:** Jul 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28508990

**Abstract:** The aim of the study was to explore what adolescents with congenital heart disease (CHD) view as important in the preparation for the transfer to adult care. We performed interviews in four focus groups with adolescents (14-18 years old) at four university hospitals in Sweden. Data was analysed using qualitative content analysis. The analysis revealed one main category; Becoming a manager of the condition and four subcategories; Sufficient knowledge about the health, Be a participant in the care, Parental support, and Communicate with others about the health. The

adolescents' ages differentiated the discussion in the groups. The older adolescents seemed to have more interest in transition planning, information and transfer. The younger described more frustrations about communication and handling the disease. **CONCLUSION** To become a manager of the CHD in daily life, the adolescents want disease specific knowledge, which should be communicated in a developmentally appropriate way. Adolescents want to participate and be involved in the transition process. They need support and guidance in how to communicate their CHD. Parental support is fundamental but it change over time. Moreover, peer-support is becoming more significant during the transition process. **What is Known:** • Transition during adolescence and transfer to adult care for adolescents with CHD is complex, and there is a shift in roles. • Adolescents often have poor knowledge and understanding about their heart condition and the consequences. **What is New:** • Adolescents call for disease specific information regarding health issues of importance for them in daily life. • Communicating the disease with other is a challenge- peer support from other adolescents with CHD could be a facilitator.

**Database:** Medline

### **53. Preparing adolescents with heart problems for transition to adult care, 2009-2010 National Survey of Children with Special Health Care Needs.**

**Author(s):** Downing, Karrie F; Oster, Matthew E; Farr, Sherry L

**Source:** Congenital heart disease; Jul 2017; vol. 12 (no. 4); p. 497-506

**Publication Date:** Jul 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28523852

**Abstract:** **OBJECTIVE** A substantial percentage of children with congenital heart disease (CHD) fail to transfer to adult care, resulting in increased risk of morbidity and mortality. Transition planning discussions with a provider may increase rates of transfer, yet little is known about frequency and content of these discussions. We assessed prevalence and predictors of transition-related discussions between providers and parents of children with special healthcare needs (CSHCN) and heart problems, including CHD. **DESIGN** Using parent-reported data on 12- to 17-year-olds from the 2009-2010 National Survey of CSHCN, we calculated adjusted prevalence ratios (aPR) for associations between demographic factors and provider discussions on shift to adult care, future insurance, and adult healthcare needs, weighted to generate population-based estimates. **RESULTS** Of the 5.3% of adolescents with heart problems in our sample (n = 724), 52.8% were female, 65.3% white, 62.2% privately insured, and 37.1% had medical homes. Less than 50% had parents who discussed with providers their child's future health insurance (26.4%), shift to adult care (22.9%), and adult healthcare needs (49.0%). Transition planning did not differ between children with and without heart problems (aPR range: 1.0-1.1). Among parents of CSHCN with heart problems who did not have discussions, up to 66% desired one. Compared to 1-/13-year-olds, a larger percentage of 16-/17-year-olds had parents who discussed their shift to adult care (aPR 2.1, 95% confidence interval (CI) [1.1, 3.9]), and future insurance (aPR 1.8, 95% CI [1.1, 2.9]). Having a medical home was associated with discussing adult healthcare needs (aPR 1.5, 95% CI [1.2, 1.8]) and future insurance (aPR 1.8, 95% CI [1.3, 2.6]). **CONCLUSIONS** Nationally, less than half of adolescents with heart problems had parents who discussed their child's transition with providers, which could be contributing to the large percentage of CHD patients who do not successfully transfer to adult care.

**Database:** Medline

#### 54. Prevention of Sudden Cardiac Death in Adults With Congenital Heart Disease: Do the Guidelines Fall Short?

**Author(s):** Vehmeijer, Jim T; Koyak, Zeliha; Budts, Werner; Harris, Louise; Silversides, Candice K; Oechslin, Erwin N; Bouma, Berto J; Zwinderman, Aeilko H; Mulder, Barbara J M; de Groot, Joris R

**Source:** Circulation. Arrhythmia and electrophysiology; Jul 2017; vol. 10 (no. 7)

**Publication Date:** Jul 2017

**Publication Type(s):** Multicenter Study Journal Article

**PubMedID:** 28696220

**Abstract:**BACKGROUND Sudden cardiac death (SCD) is a major cause of mortality in adult congenital heart disease (ACHD) patients. SCD may be prevented by implantable cardioverter-defibrillator (ICD) implantation, but patient stratification remains troublesome. The 2014 Consensus Statement on Arrhythmias in ACHD patients and the 2015 European Society of Cardiology Guidelines specified recommendations for ICD implantation in ACHD patients for the first time. We assess the discriminative ability of these ICD recommendations for SCD in ACHD patients. METHODS AND RESULTS Of 25 790 ACHD patients in an international multicenter registry, we identified all SCD cases, matched to living controls by age, sex, congenital defect, and surgical repair. We assessed all primary prevention ICD recommendations listed in both documents. We used conditional logistic regression models to calculate odds ratios and receiver operating characteristic curves with area under the curve. Consensus Statement: One hundred twenty-four cases (median age at death, 33 years [26-44]; 67% men) and 230 controls were studied. In total, 41% of SCD cases and 17% of controls had an ICD recommendation (odds ratio, 5.9;  $P < 0.001$ ). European Society of Cardiology Guidelines: Of one hundred fifty-seven cases (median age at death, 33 years [26-48]; 64% men) and 292 controls, 35% and 14% had an ICD recommendation, respectively (odds ratio, 4.8;  $P < 0.001$ ). CONCLUSION A minority of SCD cases had an ICD recommendation according to these guidelines, whereas the majority of SCD victims remained unrecognized. With an area under the curve of 0.6 to 0.7, the discriminative ability of both guidelines was mediocre. Critical clinical reasoning when deciding on ICD implantation in ACHD patients, therefore, remains vital.

**Database:** Medline

#### 55. Cognitive dysfunction in adult CHD with different structural complexity.

**Author(s):** Tyagi, Manavi; Fteropoulli, Theodora; Hurt, Catherine S; Hirani, Shashivadan P; Rixon, Lorna; Davies, Anna; Picaut, Nathalie; Kennedy, Fiona; Deanfield, John; Cullen, Shay; Newman, Stanton P

**Source:** Cardiology in the young; Jul 2017; vol. 27 (no. 5); p. 851-859

**Publication Date:** Jul 2017

**Publication Type(s):** Journal Article

**PubMedID:** 27751192

Available at [Cardiology in the young](#) - from EBSCO (MEDLINE Complete)

Available at [Cardiology in the young](#) - from EBSCO (MEDLINE Complete)

Available at [Cardiology in the young](#) - from EBSCO (MEDLINE Complete)

Available at [Cardiology in the young](#) - from ProQuest (Hospital Premium Collection) - NHS Version

**Abstract:**OBJECTIVE We carried out a cross-sectional study to assess cognitive function in a sample of adult CHD patients, within the Functioning in Adult Congenital Heart Disease study London. The association between cognitive functioning and disease complexity was examined. METHODS A total of

310 patients participated in this study. Patients were classified into four structural complexity groups - tetralogy of Fallot, transposition of the great arteries, single ventricle, and simple conditions. Each patient underwent neuropsychological assessment to evaluate cognitive function, including memory and executive function, and completed questionnaires to assess depression and anxiety. RESULTS Among all, 41% of the sample showed impaired performance (>1.5 SD below the normative mean) on at least three tests of cognitive function compared with established normative data. This was higher than the 8% that was expected in a normal population. The sample exhibited significant deficits in divided attention, motor function, and executive functioning. There was a significant group difference in divided attention ( $F=5.01$ ,  $p=0.002$ ) and the mean total composite score ( $F=5.19$ ,  $p=0.002$ ) between different structural complexity groups, with the simple group displaying better cognitive function. CONCLUSION The results indicate that many adult CHD patients display impaired cognitive function relative to a healthy population, which differs in relation to disease complexity. These findings may have implications for clinical decision making in this group of patients during childhood. Possible mechanisms underlying these deficits and how they may be reduced or prevented are discussed; however, further work is needed to draw conclusive judgements.

**Database:** Medline

#### **56. Cardiopulmonary Exercise Testing in Adult Congenital Heart Disease.**

**Author(s):** Mantegazza, Valentina; Apostolo, Anna; Hager, Alfred

**Source:** Annals of the American Thoracic Society; Jul 2017; vol. 14 ; p. S93

**Publication Date:** Jul 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28375677

Available at [Annals of the American Thoracic Society](#) - from EBSCO (MEDLINE Complete)

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Available at [Annals of the American Thoracic Society](#) - from EBSCO (MEDLINE Complete)

Available at [Annals of the American Thoracic Society](#) - from ProQuest (Hospital Premium Collection)  
- NHS Version

**Abstract:** Recently, the number of patients with congenital heart diseases reaching adulthood has been progressively increasing in developed countries, and new issues are emerging: the evaluation of their capacity to cope with physical activity and whether this knowledge can be used to optimize medical management. A symptom-limited cardiopulmonary exercise test has proven to be an essential tool, because it can objectively evaluate the functional cardiovascular capacity of these patients, identify the pathological mechanisms of the defect (circulatory failure, shunts, and/or pulmonary hypertension), and help prescribe an individualized rehabilitation program when needed. The common findings on cardiopulmonary exercise testing in patients with congenital heart diseases are a reduced peak  $\dot{V}O_2$ , an early anaerobic threshold, a blunted heart rate response, a reduced increase of  $\dot{V}_t$ , and an increased  $\dot{V}_E/\dot{V}_t$ . All these measures suggest common pathophysiological abnormalities: (1) a compromised exercise capacity from anomalies affecting the heart, vessels, lungs, or muscles; (2) chronotropic incompetence secondary to cardiac autonomic dysfunction or  $\beta$ -blockers and antiarrhythmic therapy; and (3) ventilatory inefficiency caused by left-heart failure with pulmonary congestion, pulmonary hypertension, pulmonary obstructive vascular disease, or cachexia. Most of these variables also have prognostic significance. For these patients, cardiopulmonary exercise testing allows evaluation and decisions affecting lifestyle and therapeutic interventions.



**Database:** Medline

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

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

**Source:** Heart, lung & circulation; Jun 2017

**Publication Date:** Jun 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28709919

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

**Database:** Medline

**58. Atrial tachyarrhythmia in adult congenital heart disease.**

**Author(s):** Karbassi, Arsha; Nair, Krishnakumar; Harris, Louise; Wald, Rachel M; Roche, S Lucy

**Source:** World journal of cardiology; Jun 2017; vol. 9 (no. 6); p. 496-507

**Publication Date:** Jun 2017

**Publication Type(s):** Journal Article Review

**PubMedID:** 28706585

Available at [World journal of cardiology](#) - from Europe PubMed Central - Open Access

**Abstract:** The adult congenital heart disease (ACHD) population continues to grow and most cardiologists, emergency room physicians and family doctors will intermittently come into contact with these patients. Oftentimes this may be in the setting of a presentation with atrial tachyarrhythmia; one of the commonest late complications of ACHD and problem with potentially serious implications. Providing appropriate initial care and ongoing management of atrial tachyarrhythmia in ACHD patients requires a degree of specialist knowledge and an awareness of

certain key issues. In ACHD, atrial tachyarrhythmia is usually related to the abnormal anatomy of the underlying heart defect and often occurs as a result of surgical scar or a consequence of residual hemodynamic or electrical disturbances. Arrhythmias significantly increase mortality and morbidity in ACHD and are the most frequent reason for ACHD hospitalization. Intra-atrial reentrant tachycardia and atrial fibrillation are the most prevalent type of arrhythmia in this patient group. In hemodynamically unstable patients, urgent cardioversion is required. Acute management of the stable patient includes anticoagulation, rate control, and electrical or pharmacological cardioversion. In ACHD, rhythm control is the preferred management strategy and can often be achieved. However, in the long-term, medication side-effects can prove problematic. Electrophysiology studies and catheter ablation are important treatment modalities and in certain cases, surgical or percutaneous treatment of the underlying cardiac defect has a role. ACHD patients, especially those with complex CHD, are at increased risk of thromboembolic events and anticoagulation is usually required. Female ACHD patients of child bearing age may wish to pursue pregnancies. The risk of atrial arrhythmias is increased during pregnancy and management of atrial tachyarrhythmia during pregnancy needs specific consideration.

**Database:** Medline

#### **59. Impact of growing cohorts of adults with con-genital heart disease on clinical workload: a 20-year experience at a tertiary care centre.**

**Author(s):** Padrutt, Maria; Bracher, Isabelle; Bonassin, Francesca; Santos Lopes, Bruno; Gruner, Christiane; Stämpfli, Simon F; Wolber, Thomas; Kretschmar, Oliver; Oxenius, Angela; De Pasquale, Gabriella; Seeliger, Theresa; Lüscher, Thomas F; Attenhofer Jost, Christine; Greutmann, Matthias

**Source:** Swiss medical weekly; Jun 2017; vol. 147 ; p. w14443

**Publication Date:** Jun 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28634971

**Abstract:**BACKGROUND Population based studies show a steady increase in adult patients with congenital heart defects. The aim of this study was to assess the evolution of such a patient cohort and its burden on clinical care at a dedicated tertiary care centre. METHODS All patients with congenital heart disease followed up by a dedicated multidisciplinary team at our institution were identified (n = 1725). Disease characteristics, the increase in patient numbers and interventions and the increase in selected complications were analysed and compared between the first (1996-2005) and second (2006-2015) decades of the study period. RESULTS Between the two decades of the study period, the number of patients in follow-up increased by 109%, the number of patients who died or underwent transplantation more than doubled and the number of outpatient visits increased by 195%. One fourth of all patients underwent at least one surgical procedure and 14% had at least one percutaneous intervention. The increase in surgical procedures between the two decades was 27% and the increase in percutaneous interventions 159%. Between the two decades the number of patients requiring direct current cardioversion increased from 32 to 95 (+197%), the number of patients requiring admission for infective endocarditis increased from 7 to 29 (+314%) and the number of women followed up during pregnancy increased from 18 to 115 (+539%). CONCLUSION As a result of the increasing number and complexity of adult survivors with congenital heart disease more resources will be needed to cope with the demands of this novel cohort of complex patients in adult cardiology.

**Database:** Medline

### 60. Cost of Congenital Heart Disease Hospitalizations in Canada: A Population-Based Study.

**Author(s):** Mackie, Andrew S; Tran, Dat T; Marelli, Ariane J; Kaul, Padma

**Source:** The Canadian journal of cardiology; Jun 2017; vol. 33 (no. 6); p. 792-798

**Publication Date:** Jun 2017

**Publication Type(s):** Journal Article Observational Study

**PubMedID:** 28457736

**Abstract:**BACKGROUNDThe prevalence of congenital heart disease (CHD) is rising, and late complications are common. The impact of these factors on health-care costs is not well understood. We sought to describe inpatient CHD costs in Canada.METHODSWe conducted an observational retrospective cohort study. The Canadian Institute for Health Information (CIHI) Discharge Abstract Database was used for all Canadian provinces, except Quebec, between April 2004 and March 2014. We included hospitalizations with a main diagnosis of CHD (International Classification of Diseases, 10th revision, codes Q20.0-26.9) and hospitalizations having CHD as a secondary diagnosis if the main diagnosis was a comorbid condition related to CHD. CIHI patient cost estimates were used to provide dollar values. Costs were inflated to 2016 Canadian dollars.RESULTSAmong 59,917 hospitalizations, annual CHD costs increased by 21.6% from CAD\$99.7 million (95% confidence interval [CI], \$89.4-\$110.1 million) in 2004 to \$121.2 million (95% CI, \$112.8-\$129.6 million) in 2013 ( $P < 0.001$ ). Costs were higher for children compared with adults. However, the cost increase was greater in adults (4.5%/y;  $P < 0.001$ ) than in children (0.7%/y;  $P = 0.006$ ). Adults accounted for 38.2% of costs in 2004 vs 45.8% in 2013 ( $P = 0.002$ ). Costs increased most among adults with complex CHD (7.2%/y;  $P = 0.001$ ). Adult men accounted for greater increases in costs relative to women ( $P < 0.001$ ). Length of stay was unchanged over time.CONCLUSIONSInpatient CHD costs are increasing independent of inflation, particularly among adults with complex lesions. Although children still account for greater inpatient CHD costs, a larger increase was observed among adults. These data are important in allocating inpatient resources for adults with CHD.

**Database:** Medline

### 61. Social independence of adult congenital heart disease patients in Japan.

**Author(s):** Ochiai, Ryota; Ikeda, Yukitaka; Kato, Hitoshi; Shiraishi, Isao; Parents' Association of Heart Disease Children

**Source:** Pediatrics international : official journal of the Japan Pediatric Society; Jun 2017; vol. 59 (no. 6); p. 675-681

**Publication Date:** Jun 2017

**Publication Type(s):** Journal Article

**PubMedID:** 28199782

**Abstract:**BACKGROUNDAs treatment outcomes for congenital heart disease (CHD) have improved, the social independence of adult CHD patients has become a key goal. The aims of this study were therefore to (i) determine the relationship between social independence and psychological profile, and (ii) identify patient anxieties, difficulties, and demands related to life in society.METHODSA total of 143 patients aged  $\geq 15$  years with physical disability certificates were selected using a questionnaire distributed by a patients' association. Each participant was asked about employment status, income, and receipt of disability pension as a social independence index, and about financial and psychological distress as a psychological status index. Furthermore, each participant was asked to freely describe his or her difficulties, anxieties, and needs pertaining to life in society.RESULTSThe subjects were 15-73 years old. Seventy-one (50%) were female, and 94 (66%) had a grade 1 physical

disability certificate. Fifty-nine subjects (41%) were employed, 37 (26%) were unemployed, and 45 (31%) were students. Of those employed, 34 subjects (58%) reported annual individual income  $\leq$  2 million yen. Frequent hospital visits, low total household income, low individual annual income, work dissatisfaction, and receipt of a disability pension were associated with poorer psychological profile. In an open description section, subjects expressed desires for better pension systems, support for medical fees, and employment support. **CONCLUSIONS** Because financial issues can adversely affect the psychological profiles of adult CHD patients, enhancement of social welfare and employment support may improve their social independence.

**Database:** Medline

## **62. Red Flags for Maltese Adults with Congenital Heart Disease: Poorer Dental Care and Less Sports Participation Compared to Other European Patients-An APPROACH-IS Substudy.**

**Author(s):** Caruana, Maryanne; Apers, Silke; Kovacs, Adrienne H; Luyckx, Koen; Thomet, Corina; Budts, Werner; Sluman, Maayke; Eriksen, Katrine; Dellborg, Mikael; Berghammer, Malin; Johansson, Bengt; Soufi, Alexandra; Callus, Edward; Moons, Philip; Grech, Victor; APPROACH-IS consortium and the International Society for Adult Congenital Heart Disease (ISACHD)

**Source:** Pediatric cardiology; Jun 2017; vol. 38 (no. 5); p. 965-973

**Publication Date:** Jun 2017

**Publication Type(s):** Comparative Study Journal Article

**PubMedID:** 28341902

**Abstract:** Studies in recent years have explored lifestyle habits and health-risk behaviours in adult congenital heart disease (ACHD) patients when compared to controls. The aim of this study was to investigate differences in lifestyle habits between Maltese and other European ACHD patients. Data on alcohol consumption, cigarette smoking, substance misuse, dental care and physical activity collected in 2013-2015 during "Assessment of Patterns of Patient-Reported Outcomes in Adults with Congenital Heart disease-International Study" (APPROACH-IS) were analysed. Responses from 119 Maltese participants were compared to those of 1616 participants from Belgium, France, Italy, Norway, Sweden, Switzerland and the Netherlands. Significantly fewer Maltese patients with simple (Maltese 84.1% vs. European 97.5%,  $p < 0.001$ ) and moderately complex CHD (Maltese 83.6% vs. European 97.4%,  $p < 0.001$ ) brushed their teeth daily. Only 67.2% of Maltese with moderately complex disease had dental reviews in the previous year compared to 80.3% of Europeans ( $p = 0.02$ ). Maltese patients with simple (Maltese 31.8% vs. European 56.1%,  $p = 0.002$ ) and moderately complex lesions (Maltese 30.0% vs. European 59.2%,  $p < 0.001$ ) performed less regular sport activities. Comparison by country showed Maltese patients to have significantly poorer tooth brushing and sports participation than patients from any other participating country. Alcohol consumption, cigarette smoking and substance misuse were not significantly different. This study highlights lifestyle aspects that Maltese ACHD patients need to improve on, which might not be evident upon comparing patients to non-CHD controls. These findings should also caution researchers against considering behaviours among patients in one country as necessarily representative of patients on the larger scale.

**Database:** Medline

## **63. Heart transplantation for adults with congenital heart disease: current status and future prospects.**

**Author(s):** Matsuda, Hikaru; Ichikawa, Hajime; Ueno, Takayoshi; Sawa, Yoshiki

**Source:** General thoracic and cardiovascular surgery; Jun 2017; vol. 65 (no. 6); p. 309-320

**Publication Date:** Jun 2017

**Publication Type(s):** Journal Article Review

**PubMedID:** 28439697

**Abstract:** Increased survival rates after corrective or palliative surgery for complex congenital heart disease (CHD) in infancy and childhood are now being coupled with increased numbers of patients who survive to adulthood with various residual lesions or sequelae. These patients are likely to deteriorate in cardiac function or end-organ function, eventually requiring lifesaving treatment including heart transplantation. Although early and late outcomes of heart transplantation have been improving for adult survivors of CHD, outcomes and pretransplant management could still be improved. Survivors of Fontan procedures are a vulnerable cohort, particularly when single ventricle physiology fails, mostly with protein-losing enteropathy and hepatic dysfunction. Therefore, we reviewed single-institution and larger database analyses of adults who underwent heart transplantation for CHD, to enable risk stratification by identifying the indications and outcomes. As the results, despite relatively high early mortality, long-term results were encouraging after heart transplantation. However, further investigations are needed to improve the indication criteria for complex CHD, especially for failed Fontan. In addition, the current system of status criteria and donor heart allocation system in heart transplantation should be arranged as suitable for adults with complex CHD. Furthermore, there is a strong need to develop ventricular assist devices as a bridge to transplantation or destination therapy, especially where right-sided circulatory support is needed.

**Database:** Medline

#### **64. How Pregnancy Impacts Adult Cyanotic Congenital Heart Disease: A Multicenter Observational Study.**

**Author(s):** Ladouceur, Magalie; Benoit, Louise; Basquin, Adeline; Radojevic, Jelena; Hauet, Quentin; Hascoet, Sébastien; Mocerri, Pamela; Le Gloan, Laurianne; Amedro, Pascal; Lucron, Hugues; Richard, Adelaide; Gouton, Marielle; Nizard, Jacky

**Source:** Circulation; Jun 2017; vol. 135 (no. 24); p. 2444-2447

**Publication Date:** Jun 2017

**Publication Type(s):** Academic Journal

**PubMedID:** 28606952

**Abstract:** The article presents a study which examines the impacts of pregnancy of adult cyanotic congenital heart disease (CHD). The multicenter observational study conducted a review of the charts of all pregnant women with CHD. Results showed the need for larger prospective studies on pregnancy related complications in the unique patient population with cyanotic CHDs.

**Database:** CINAHL

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### [Journal of the American College of Cardiology](#)

October 24 2017, Volume 70, Issue 17

### [Circulation](#)

October 17 2017, Volume 136, Issue 16

### [European Heart Journal](#)

October 7 2017, Volume 38, Issue 38

### [Heart BMJ](#)

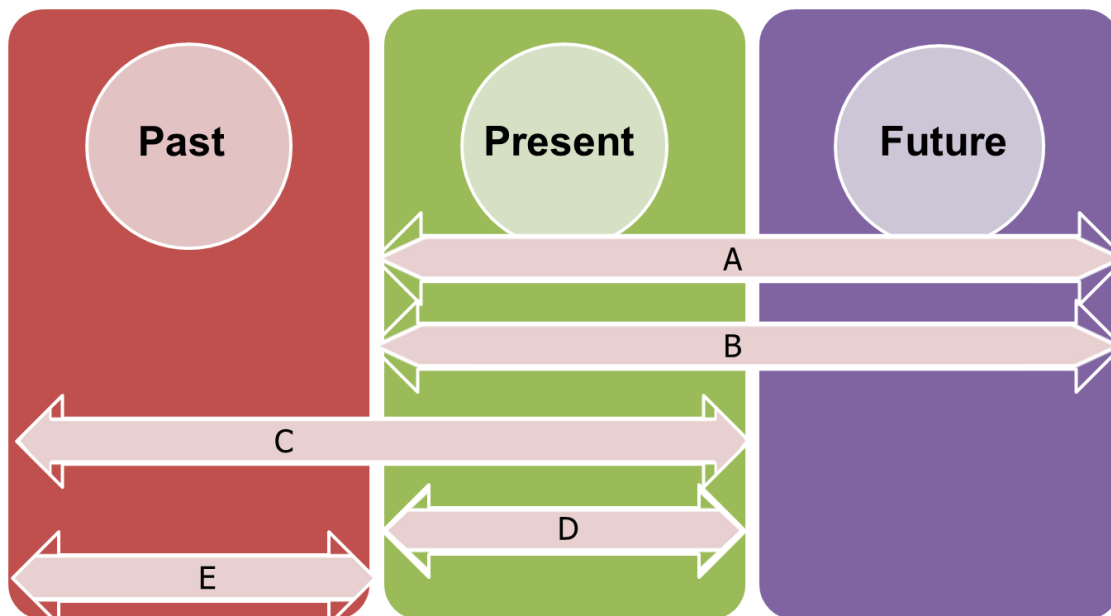
November 2017, Volume 103, Issue 21

### [Pediatric Cardiology](#)

October 2017, Volume 38, Issue 7

## Exercise: Study Design Timeframes

Match the study design with the timeframe it covers



1. Randomised Controlled Trial
2. Cross-Sectional Study
3. Case-control Study
4. Cohort Study
5. Case Report

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Answers: 1A/B; 2D; 3C; 4A/B; 5E



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