

# Adult Congenital Heart Disease Current Awareness Bulletin



**June 2017**

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## Lunchtime Drop-in Sessions

*All sessions last one hour*

### June (12.00-13.00)

29th (Thurs) Literature Searching

### July (13.00-14.00)

3rd (Mon) Interpreting Statistics

12th (Wed) Critical Appraisal

21st (Fri) Literature Searching

26th (Wed) Interpreting Statistics

### August (12.00-13.00)

4th (Fri) Critical Appraisal

9th (Wed) Literature Searching

15th (Tues) Interpreting Statistics

24th (Thurs) Critical Appraisal

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

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



**UpToDate®**

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

## **Pregnancy in women with congenital heart disease: General principles**

Authors: [Carol A Waksmonski, MD](#); [Michael R Foley, MD](#); Section Editors: [Charles J Lockwood, MD, MHCM](#); [Heidi M Connolly, MD, FASE](#); Deputy Editor: [Susan B Yeon, MD, JD, FACC](#)

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

**Literature review current through:** May 2017. | **This topic last updated:** Nov 14, 2016.

[https://www.uptodate.com/contents/pregnancy-in-women-with-congenital-heart-disease-general-principles?source=search\\_result&search=congenital%20heart%20disease%20and%20pregnancy&selectedTitle=2~150](https://www.uptodate.com/contents/pregnancy-in-women-with-congenital-heart-disease-general-principles?source=search_result&search=congenital%20heart%20disease%20and%20pregnancy&selectedTitle=2~150)

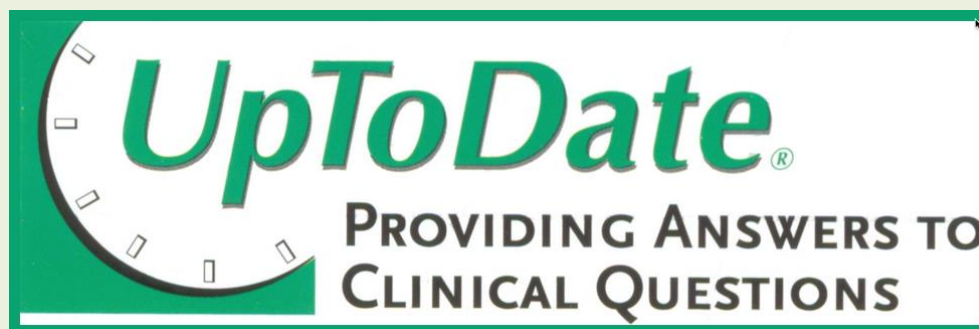
## **Pregnancy in women with congenital heart disease: Specific lesions**

Authors: [Carol A Waksmonski, MD](#); [Michael R Foley, MD](#); Section Editors: [Charles J Lockwood, MD, MHCM](#); [Heidi M Connolly, MD, FASE](#); Deputy Editor: [Susan B Yeon, MD, JD, FACC](#)

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

**Literature review current through:** May 2017. | **This topic last updated:** Mar 02, 2016.

[https://www.uptodate.com/contents/pregnancy-in-women-with-congenital-heart-disease-specific-lesions?source=related\\_link](https://www.uptodate.com/contents/pregnancy-in-women-with-congenital-heart-disease-specific-lesions?source=related_link)



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## Current Awareness Database

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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)

### Adult Congenital Heart Disease

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1. Cognitive dysfunction in adult CHD with different structural complexity.
2. Transition of Adolescents and Young Adults with Congenital Heart Disease: Challenges, Progress, and Future Improvements.
3. Transition of Care in Congenital Heart Disease: Ensuring the Proper Handoff.
4. Current Role of Blood and Urine Biomarkers in the Clinical Care of Adults with Congenital Heart Disease.
5. Drug Therapy in Adult Congenital Heart Disease.
6. Sudden Cardiac Death in Adult Congenital Heart Disease.
7. Atrial fibrillation in pregnancy: a growing challenge.
8. Preparing adolescents with heart problems for transition to adult care, 2009-2010 National Survey of Children with Special Health Care Needs.
9. Adolescents with congenital heart disease: their opinions about the preparation for transfer to adult care.
10. ACR Appropriateness Criteria® Known or Suspected Congenital Heart Disease in the Adult.
11. Increased risk of thromboembolic events in adult congenital heart disease patients with atrial tachyarrhythmias.
12. Improving the quality of transition and transfer of care in young adults with congenital heart disease.
13. Validity of the Montreal Cognitive Assessment Screener in Adolescents and Young Adults With and Without Congenital Heart Disease.

14. Maternal Cardiovascular Mortality in Illinois, 2002-2011.
15. Inferior Vena Cava Thrombus in a Postpartum Patient With Fontan Physiology: A Case Report.
16. [Adult congenital heart disease: Medical and psychosocial issues].
17. Effect of maternal age and cardiac disease severity on outcome of pregnancy in women with congenital heart disease.
18. Cardiovascular causes of maternal sudden death. Sudden arrhythmic death syndrome is leading cause in UK.
19. Attitudes and perceptions of pregnant women with CHD: results of a single-site survey.
20. Person-centred transition programme to empower adolescents with congenital heart disease in the transition to adulthood: a study protocol for a hybrid randomised controlled trial (STEPSTONES project).
21. Pregnancy in Adult Congenital Heart Disease: Special Delivery.
22. Sudden cardiac death in adults with congenital heart disease: does QRS-complex fragmentation discriminate in structurally abnormal hearts?
23. Cardiopulmonary Exercise Testing in Adult Congenital Heart Disease.
24. Readiness for Transition to Adult Health Care for Young Adolescents with Congenital Heart Disease.
25. Arrhythmias in Adults with Congenital Heart Disease: What Are Risk Factors for Specific Arrhythmias?
26. Maternal and Fetal Outcomes of Admission for Delivery in Women With Congenital Heart Disease.
27. Incidence and predictors of obstetric and fetal complications in women with structural heart disease.
28. National In-Hospital Outcomes of Pregnancy in Women With Single Ventricle Congenital Heart Disease.
29. Educational needs of adolescents with congenital heart disease: Impact of a transition intervention programme.
30. Advance Care Planning in Adults with Congenital Heart Disease: A Patient Priority.
31. Characteristics and outcomes of heart failure-related hospitalization in adults with congenital heart disease.
32. Adult Congenital Cardiac Care.

33. Pregnancy in women with complete transposition of the great arteries following the atrial switch procedure. A study from three of the largest Adult Congenital Heart Disease centers in Poland.
34. Hopelessness among adults with congenital heart disease: Cause for despair or hope?
35. Sudden cardiac death in adult congenital heart disease: can the unpredictable be foreseen?
36. Neurocognitive functioning in adults with congenital heart disease.
37. NEUROCOGNITIVE IMPAIRMENT IS COMMON IN THE ADULT WITH CONGENITAL HEART DISEASE: IDENTIFICATION USING A NOVEL CLINICAL QUESTIONNAIRE.
38. MORTALITY IN PREGNANT WOMEN WITH CONGENITAL HEART DISEASE: A SUBANALYSIS OF THE CALIFORNIA PREGNANCY-ASSOCIATED MORTALITY REVIEW.
39. The role of palliative care in critical congenital heart disease.
40. Changing Landscape of Congenital Heart Disease.
41. Contraceptive Practices of Women With Complex Congenital Heart Disease.
42. Body mass index in adult congenital heart disease.
43. Social independence of adult congenital heart disease patients in Japan.
44. Adult congenital heart disease nurse coordination: Essential skills and role in optimizing team-based care a position statement from the International Society for Adult Congenital Heart Disease (ISACHD).
45. Healthcare needs of adolescents with congenital heart disease transitioning into adulthood: a Delphi survey of patients, parents, and healthcare providers.
46. Survivorship in Children and Young Adults With Congenital Heart Disease in Sweden.
47. Management of Pregnancy in Patients With Complex Congenital Heart Disease: A Scientific Statement for Healthcare Professionals From the American Heart Association.
48. Pregnancy outcomes in patients with pulmonary arterial hypertension associated with congenital heart disease.
49. Use of 3D models of congenital heart disease as an education tool for cardiac nurses.
50. Neurocognitive and executive functioning in adult survivors of congenital heart disease.
51. Health-related quality of life of young people with long-term illnesses before and after transfer from child to adult healthcare.
52. Multiple pregnancy in a primigravida with uncorrected Pentalogy of Fallot.



### 1. 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 Type(s):** Journal Article

**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.

### 2. Transition of Adolescents and Young Adults with Congenital Heart Disease: Challenges, Progress, and Future Improvements.

**Author(s):** Nicolarsen, Jeremy

**Source:** Pediatric annals; Jun 2017; vol. 46 (no. 6); p. e224

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

Available in full text at [Pediatric Annals](#) - from ProQuest

**Abstract:** Congenital heart disease (CHD) is the most common and perhaps most widely variable birth defect. Decades of improved CHD care has resulted in a steady growth in the number and complexity of adults with CHD, and many of these patients require lifelong, specialized follow-up care. This begins with successful transition from pediatric-based to adult-based care. Despite the remarkable advances in this field, many adults with CHD still experience lapses in care that have significant health consequences. This review outlines some of the challenges, progress, and areas for improvement in CHD transition medicine. [Pediatr Ann. 2017;46(6):e224-e228.].

### 3. Transition of Care in Congenital Heart Disease: Ensuring the Proper Handoff.

**Author(s):** Lee, Angela; Bailey, Barbara; Cullen-Dean, Geraldine; Aiello, Sandra; Morin, Joanne; Oechslin, Erwin

**Source:** Current cardiology reports; Jun 2017; vol. 19 (no. 6); p. 55

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

**Abstract:** BACKGROUND With great advances in medical and surgical care, most congenital heart disease patients are living in to adulthood and require lifelong surveillance and expert care for adult onset complications. Care lapse and lack of successful transfer from pediatric to adult care put young adults at risk for increased morbidity and premature death. Hence, transition and transfer from pediatric to adult care is a crucial and critical process to provide access to specialized care and lifelong surveillance. PURPOSE OF REVIEW The aim of this article is to describe barriers to successful transition and transfer and to share practical strategies and concepts to overcome these barriers in order to successfully implement a transition program. RECENT FINDINGS There are patient-specific, local, and institutional specific barriers to establish a successful transition program which involves many stakeholders. Collaboration of the Pediatric and Adult Congenital Heart Disease programs is paramount; the understanding of the benefit and the need of a structured transition program, dedication, and a proactive approach are essential. Youth- and family-centered education improves healthcare knowledge, self-management, self-advocacy, and appropriate interdependence and helps young adults to take ownership of their health. Nurses play an integral role within the multidisciplinary team in supporting seamless, successful transition and transfer of CHD patients from pediatric to adult care thereby reducing loss to follow-up and lapses in care. Most experiences and recommendations are based on retrospective studies and expert consensus. It is imperative to evaluate the impact of structured and planned transition/transfer programs on the outcomes. Hence, prospective, randomized trials are required to document if implementation of structured intervention transition programs improve knowledge, patient experiences, and outcomes of congenital heart defect survivors.

**4. Current Role of Blood and Urine Biomarkers in the Clinical Care of Adults with Congenital Heart Disease.**

**Author(s):** Rajpal, Saurabh; Alshawabkeh, Laith; Opotowsky, Alexander R

**Source:** Current cardiology reports; Jun 2017; vol. 19 (no. 6); p. 50

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

**Abstract:** PURPOSE OF REVIEW There is an increasing number of adult patients with congenital heart disease (CHD). While several biomarkers have been validated and integrated into general cardiology clinical practice, these tests are often applied to adults with CHD in the absence of disease-specific validation. Although these patients are often grouped into a single population, there is heterogeneous pathophysiology, variable disease chronicity, extensive multisystem involvement, and a low event rate relative to acquired heart disease. These stand as challenges to systematic investigation and clinical application of biomarkers for adults with CHD. This paper reviews recent studies investigating the use of biomarkers in this population, with emphasis on biomarkers applied in clinical adult CHD care. RECENT FINDINGS A handful of biomarkers have been integrated into adult CHD practice, such as iron studies in cyanotic heart disease and stool alpha-1 antitrypsin for diagnosis of protein losing enteropathy in the Fontan circulation. Use of kidney and liver tests has been studied in prognostication of adult CHD patients. A few other biomarkers like natriuretic peptides and troponins seem likely to provide useful information in other ACHD situations based on limited disease-specific data and extrapolation from acquired heart disease. More research is needed to support the robust validity of most existing clinical biomarkers in adult congenital cardiology practice. Until data from larger, prospectively enrolled cohorts are available, clinical use of biomarkers in these patients will require careful interpretation with attention to underlying pathophysiology, as well as detailed understanding of potential pitfalls of specific assays and clinical contexts.

### 5. Drug Therapy in Adult Congenital Heart Disease.

**Author(s):** Contractor, Tahmeed; Levin, Vadim; Mandapati, Ravi

**Source:** Cardiac electrophysiology clinics; Jun 2017; vol. 9 (no. 2); p. 295-309

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

**Abstract:** Adults with congenital heart disease are at risk for atrial and ventricular arrhythmias that can lead to an increased morbidity as well as mortality. When catheter ablation is not an option or unsuccessful, antiarrhythmic drugs are the mainstay of treatment. There is limited data on the use of antiarrhythmics in this population. The purpose of this article is to discuss the practical aspects of the use of antiarrhythmics in adults with congenital heart disease. Several tables have been provided to provide clinicians a reference for daily use.

### 6. Sudden Cardiac Death in Adult Congenital Heart Disease.

**Author(s):** Ávila, Pablo; Chaix, Marie-A; Mondésert, Blandine; Khairy, Paul

**Source:** Cardiac electrophysiology clinics; Jun 2017; vol. 9 (no. 2); p. 225-234

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

**Abstract:** Sudden death of presumed arrhythmic etiology is a leading cause of mortality in adults with congenital heart disease. Anticipated benefits of the implantable cardioverter-defibrillator (ICD) must be weighed against high complication rates. Without robust evidence from randomized trials, caregivers face difficult decisions in selecting appropriate candidates. Although secondary prevention indications are often clear-cut, risk stratification for primary prevention ICDs is more challenging. Factors associated with sudden death in patients with tetralogy of Fallot are reasonably consistent across studies. In contrast, identification of high-risk patients with systemic right ventricles or univentricular hearts remains controversial.

### 7. Atrial fibrillation in pregnancy: a growing challenge.

**Author(s):** Katsi, Vasiliki; Georgiopoulos, Georgios; Marketou, Maria; Oikonomou, Dimitrios; Parthenakis, Fragiskos; Makris, Thomas; Nihoyannopoulos, Petros; Vardas, P; Tousoulis, Dimitris

**Source:** Current medical research and opinion; Jun 2017 ; p. 1-8

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

**Abstract:** BACKGROUND Atrial fibrillation (AF) constitutes a relatively infrequent pregnancy complication, which may be a therapeutic Gordian knot. Indeed, sparse data exist regarding the prevalence, prognosis, and management of AF during pregnancy. In general, AF occurs as a benign, self-limited arrhythmia, but occasionally may have severe hemodynamic consequences in pregnant patients suffering from heart failure, congenital heart disease, or other comorbidities. Extra-cardiac causes of AF should always be meticulously excluded. REVIEW Treatment decisions are difficult, since medications may cross the placental barrier and potentially affect fetal growth and organogenesis, or even result in fetal bradyarrhythmias. Treatment goals are not differentiated in comparison to those regarding AF occurring in the general population. Still, while maternal treatment is prioritized, issues regarding fetal health must deliberately be considered. Consequently, hemodynamic instability is to be promptly treated with synchronized electrical cardioversion. In contrast, in stable patients, pharmacologic cardioversion, under appropriate antithrombotic regimen, should be attempted. Selection of appropriate antithrombotic therapy, including novel oral anticoagulants, imposes further difficulties on therapeutic decision-making. Further clinical trials are

warranted in order to assess the pathophysiology and prognosis of AF in pregnancy and ameliorate the evidence-based therapeutic strategy in this specific group of the population.

### **8. 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; May 2017

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

**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.

### **9. 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; May 2017

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

**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.

#### **10. ACR Appropriateness Criteria® Known or Suspected Congenital Heart Disease in the Adult.**

**Author(s):** Expert Panel on Cardiac Imaging;; Woodard, Pamela K; Ho, Vincent B; Akers, Scott R; Beache, Garth; Brown, Richard K J; Cummings, Kristopher W; Greenberg, S Bruce; Min, James K; Stillman, Arthur E; Stojanovska, Jadranka; Jacobs, Jill E

**Source:** Journal of the American College of Radiology : JACR; May 2017; vol. 14 (no. 5S); p. S166

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

**Abstract:** The incidence of congenital heart disease (CHD) has been increasing in the adult patient population in part as a result of better patient survival. Patients with more severe CHD are living longer. Nearly all adults with known CHD require periodic imaging as a means of monitoring their disease process. Furthermore, adult patients with suspected CHD require imaging as a means of definitive diagnosis. As a result, it is important for both the referring clinician and the imager to be aware of the most appropriate imaging modality needed to obtain the data most needed to direct the next steps in patient care. Imaging procedures for the diagnosis of known or suspected CHD in the adult include chest radiography, fluoroscopy, echocardiography, nuclear scintigraphy, cardiac-gated CT, MRI, and cardiac catheterization/angiography. The physician trying to diagnose these often complex conditions needs complete and reliable information that includes details about intracardiac and vascular anatomy, hemodynamics, and function. The American College of Radiology Appropriateness Criteria are evidence-based guidelines for specific clinical conditions that are reviewed annually by a multidisciplinary expert panel. The guideline development and revision include an extensive analysis of current medical literature from peer-reviewed journals and the application of well-established methodologies (RAND/UCLA Appropriateness Method and Grading of Recommendations Assessment, Development, and Evaluation or GRADE) to rate the appropriateness of imaging and treatment procedures for specific clinical scenarios. In those instances where evidence is lacking or equivocal, expert opinion may supplement the available evidence to recommend imaging or treatment.

#### **11. Increased risk of thromboembolic events in adult congenital heart disease patients with atrial tachyarrhythmias.**

**Author(s):** Masuda, Keita; Ishizu, Tomoko; Niwa, Koichiro; Takechi, Fumie; Tateno, Shigeru; Horigome, Hitoshi; Aonuma, Kazutaka

**Source:** International journal of cardiology; May 2017; vol. 234 ; p. 69-75

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

**Abstract:** BACKGROUND Atrial tachyarrhythmias are a major morbidity in patients with adult congenital heart disease (ACHD). However, few studies have investigated risk stratification of thromboembolic events in ACHD patients with atrial tachyarrhythmias. METHODS AND RESULTS This retrospective cohort study reviewed the clinical records of 2314 ACHD patients from 1977 to 2014.

We found 242 (10.4%) patients with atrial tachyarrhythmias and excluded 84 patients already being treated with anticoagulant therapy. The remaining 158 patients without anticoagulant therapy were retrospectively followed up from the onset of atrial tachyarrhythmia to the incidence of thromboembolic events. Fourteen thromboembolic events and 5 hemorrhagic events occurred. All patients with thromboembolic events had atrial fibrillation (AF). Thromboembolic events occurred even in the patients with low or intermediate risk as indicated by CHADS2 or CHA2DS2-VASc score. Event rates were higher than those in data from the general adult population in previous studies. Univariate analysis revealed that age $\geq$ 60years (OR 4.54, 95% CI 1.47-14.06, P=0.009), vascular disease (OR 7.83, 95% CI 1.19-51.53, P=0.032), and persistent AF (OR 5.60, 95% CI 1.73-18.11, P=0.004) were the independent risk factors of thromboembolic events. CONCLUSIONSACHD patients with atrial tachyarrhythmias and even those with low or intermediate risk as indicated by the CHADS2 or CHA2DS2-VASc score had a higher risk of thromboembolic events. Therefore, anticoagulation should be considered earlier than in the general population in patients with risk factors of age $\geq$ 60years, vascular disease, or persistent AF.

## **12. Improving the quality of transition and transfer of care in young adults with congenital heart disease.**

**Author(s):** Everitt, Ian K; Gerardin, Jennifer F; Rodriguez, Fred H; Book, Wendy M

**Source:** Congenital heart disease; May 2017; vol. 12 (no. 3); p. 242-250

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

**Abstract:** The transition and transfer from pediatric to adult care is becoming increasingly important as improvements in the diagnosis and management of congenital heart disease allow patients to live longer. Transition is a complex and continuous process that requires careful planning. Inadequate transition has adverse effects on patients, their families and healthcare delivery systems. Currently, significant gaps exist in patient care as adolescents transfer to adult care and there are little data to drive the informed management of transition and transfer of care in adolescent congenital heart disease patients. Appropriate congenital heart disease care has been shown to decrease mortality in the adult population. This paper reviews the transition and transfer of care processes and outlines current congenital heart disease specific guidelines in the United States and compares these recommendations to Canadian and European guidelines. It then reviews perceived and real barriers to successful transition and identifies predictors of success during transfer to adult congenital heart disease care. Lastly, it explores how disease-specific markers of outcomes and quality indicators are being utilized to guide transition and transfer of care in other chronic childhood illnesses, and identifies existing knowledge gaps and structural impediments to improving the management of transition and transfer among congenital heart disease patients.

## **13. Validity of the Montreal Cognitive Assessment Screener in Adolescents and Young Adults With and Without Congenital Heart Disease.**

**Author(s):** Pike, Nancy A.; Poulsen, Marie K.; Woo, Mary A.

**Source:** Nursing Research; May 2017; vol. 66 (no. 3); p. 222-230

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

Available in full text at [Nursing Research](#) - from Ovid

**Abstract:** Background: Cognitive deficits are common, long-term sequelae in children and adolescents with congenital heart disease (CHD) who have undergone surgical palliation. However, there is a lack of a validated brief cognitive screening tool appropriate for the outpatient setting for

adolescents with CHD. One candidate instrument is the Montreal Cognitive Assessment (MoCA) questionnaire. Objective: The purpose of the research was to validate scores from the MoCA against the General Memory Index (GMI) of the Wide Range Assessment of Memory and Learning, 2nd Edition (WRAML2), a widely accepted measure of cognition/memory, in adolescents and young adults with CHD. Methods: We administered the MoCA and the WRAML2 to 156 adolescents and young adults ages 14-21 (80 youth with CHD and 76 healthy controls who were gender and age matched). Spearman's rank order correlations were used to assess concurrent validity. To assess construct validity, the Mann-Whitney Utest was used to compare differences in scores in youth with CHD and the healthy control group. Receiver operating characteristic curves were created and area under the curve, sensitivity, specificity, positive predictive value, and negative predictive value were also calculated. Results: The MoCA median scores in the CHD versus healthy controls were (23, range 15-29 vs. 28, range 22-30;  $p < .001$ ), respectively. With the screening cutoff scores at  $<26$  points for the MoCA and 85 for GMI ( $<1$  SD,  $M = 100$ ,  $SD = 15$ ), the CHD versus healthy control groups showed sensitivity of .96 and specificity of .67 versus sensitivity of .75 and specificity of .90, respectively, in the detection of cognitive deficits. A cutoff score of 26 on the MoCA was optimal in the CHD group; a cutoff of 25 had similar properties except for a lower negative predictive value. The area under the receiver operating characteristic curve (95% CI) for the MoCA was 0.84 (95% CI [0.75, 0.93],  $p < .001$ ) and 0.84 (95% CI [0.62, 1.00],  $p = .02$ ) for the CHD and controls, respectively. Discussion: Scores on the MoCA were valid for screening to detect cognitive deficits in adolescents and young adults aged 14-21 with CHD when a cutoff score of 26 is used to differentiate youth with and without significant cognitive impairment. Future studies are needed in other adolescent disease groups with known cognitive deficits and healthy populations to explore the generalizability of validity of MoCA scores in adolescents and young adults.

#### **14. Maternal Cardiovascular Mortality in Illinois, 2002-2011.**

**Author(s):** Briller, Joan; Koch, Abigail R.; Geller, Stacie E.

**Source:** *Obstetrics & Gynecology*; May 2017; vol. 129 (no. 5); p. 819-826

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

Available in full text at [Obstetrics and Gynecology](#) - from Ovid

**Abstract:** Objective: To describe the demographic characteristics of women in Illinois who died from cardiovascular disease during pregnancy or up until 1 year postpartum, addressing specific etiologies, timing of death, proportion of potentially preventable mortality, and factors associated with preventability. Methods: This is a retrospective analysis from the Illinois Department of Public Health Maternal Mortality Review process using International Classification of Diseases, 9th Revision codes that attributed cardiovascular disease as the immediate or underlying cause of maternal death in Illinois from 2002 to 2011. We categorized the etiology of cardiovascular mortality, analyzed demographic factors associated with cardiovascular mortality in comparison with noncardiovascular causes, defined the relationship to pregnancy, and identified factors associated with preventability. Results: There were 636 deaths in Illinois from 2002 to 2011 of pregnant women or within 1 year postpartum. One hundred forty women (22.2%) died of cardiovascular causes, for a cardiovascular mortality rate of 8.2 (95% confidence interval 6.9-9.6) per 100,000 live births. Women with cardiovascular mortality were likely to be older and die postpartum. The most common etiologies were related to acquired cardiovascular disease (97.1%) as compared with congenital heart disease (2.9%). Cardiomyopathy was the most common etiology ( $n=39$  [27.9%]), followed by stroke ( $n=32$  [22.9%]), hypertensive disorders ( $n=18$  [12.9%]), arrhythmias ( $n=15$  [10.7%]), and coronary disease ( $n=13$  [9.3%]). Nearly 75% of cardiac deaths were related to pregnancy as compared with 35.3% of noncardiac deaths. More than one fourth of cardiac deaths (28.1%) were potentially preventable, attributable primarily to health care provider and patient factors. Conclusion: From 2002 to 2011,

more than one fifth of maternal deaths in Illinois were attributed to cardiovascular disease such as cardiomyopathy. More than one fourth of these deaths were potentially preventable. Health care provider and patient factors were identified, which may be modifiable through education and intensive postpartum monitoring, which may diminish mortality. State maternal mortality reviews can identify opportunities for reducing maternal deaths.

#### **15. Inferior Vena Cava Thrombus in a Postpartum Patient With Fontan Physiology: A Case Report.**

**Author(s):** Tashjian, Jessica A; Fraint, Hannah; DiNardo, James; Rouine-Rapp, Kathryn

**Source:** A & A case reports; May 2017

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

Available in full text at [A&A Case Reports](#) - from Ovid

**Abstract:** Women with complex congenital heart disease, such as those with single-ventricle physiology, are surviving into adulthood and becoming pregnant. Because of their complex physiology, common peripartum complications pose unique risks. We describe a patient with a single ventricle who underwent an external vascular conduit, nonfenestrated Fontan procedure in childhood and then presented during the postpartum period with extensive thrombosis in her lower extremity deep venous system and inferior vena cava. In this article, we will discuss single-ventricle physiology and the implications of pregnancy, anesthetic considerations, and data for maternal and fetal outcomes in this population.

#### **16. [Adult congenital heart disease: Medical and psychosocial issues].**

**Author(s):** Ladouceur, Magalie; Pontnau, Florence; Iserin, Laurence

**Source:** Presse medicale (Paris, France : 1983); May 2017; vol. 46 (no. 5); p. 523-529

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

**Abstract:** The population of adults with congenital heart disease (ACHD) is continuously increasing with now a higher prevalence than that of the pediatric population. This concerns above all complex congenital heart diseases. Heart failure is the primary cause of death followed by arrhythmia, which is very common in ACHD. A specialized follow-up by dedicated centers is significantly associated with an improvement of survival of ACHD patients compared to non-expert follow-up. Extracardiac disorders (liver, kidney, respiratory) are frequent and require an accurate and specific management. The psychosocial impact, particularly the professional difficulties, is common and may require implementation of appropriate measures to improve the patient social life. Unplanned pregnancy and/or a lack of information about contraception may induce severe cardiovascular complications in ACHD women. Education about contraceptive methods at adolescence and pre-conceptual counseling are requested in this population.

#### **17. Effect of maternal age and cardiac disease severity on outcome of pregnancy in women with congenital heart disease.**

**Author(s):** Furenäs, Eva; Eriksson, Peter; Wennerholm, Ulla-Britt; Dellborg, Mikael

**Source:** International journal of cardiology; May 2017

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

**Abstract:** BACKGROUND There is an increasing prevalence of women with congenital heart defects reaching childbearing age. In western countries women tend to give birth at a higher age compared



to some decades ago. We evaluated the CARDiac disease in PREGnancy (CARPREG) and modified World Health Organization (mWHO) risk classifications for cardiac complications during pregnancies in women with congenital heart defects and analyzed the impact of age on risk of obstetric and fetal outcome. METHODS A single-center observational study of cardiac, obstetric, and neonatal complications with data from cardiac and obstetric records of pregnancies in women with congenital heart disease. Outcomes of 496 pregnancies in 232 women, including induced abortion, miscarriage, stillbirth, and live birth were analyzed regarding complications, maternal age, mode of delivery, and two risk classifications: CARPREG and mWHO. RESULTSThere were 28 induced abortions, 59 fetal loss, 409 deliveries with 412 neonates. Cardiac (14%), obstetric (14%), and neonatal (15%) complications were noted, including one maternal death and five stillbirths. The rate of cesarean section was 19%. Age above 35years was of borderline importance for cardiac complications ( $p=0.054$ ) and was not a significant additional risk factor for obstetric or neonatal complications. Both risk classifications had moderate clinical utility, with area under the curve (AUC) 0.71 for CARPREG and 0.65 for mWHO on cardiac complications. CONCLUSIONS Pregnancy complications in women with congenital heart disease are common but severe complications are rare. Advanced maternal age does not seem to affect complication rate. Existing risk classification systems are insufficient in predicting complications.

#### **18. Cardiovascular causes of maternal sudden death. Sudden arrhythmic death syndrome is leading cause in UK.**

**Author(s):** Krexii, Dimitra; Sheppard, Mary N

**Source:** European journal of obstetrics, gynecology, and reproductive biology; May 2017; vol. 212 ; p. 155-159

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

**Abstract:** OBJECTIVEThis study aims to determine the causes of sudden cardiac death during pregnancy and in the postpartum period and patients' characteristics. There are few studies in the literature. METHODS Eighty cases of sudden unexpected death due to cardiac causes in relation to pregnancy and postpartum period in a database of 4678 patients were found and examined macroscopically and microscopically. RESULTS The mean age was  $30\pm 7$  years with a range from 16 to 43 years. About 30% were 35 years old or older; 50% of deaths occurred during pregnancy and 50% during the postpartum period. About 59.18% were obese or overweight where body mass index data were available. The leading causes of death were sudden arrhythmic death syndrome (SADS) (53.75%) and cardiomyopathies (13.80%). Other causes include dissection of aorta or its branches (8.75%), congenital heart disease (2.50%) and valvular disease (3.75%).CONCLUSION This study highlights sudden cardiac death in pregnancy or in the postpartum period, which is mainly due to SADS with underlying channelopathies and cardiomyopathy. We wish to raise awareness of these frequently under-recognised entities in maternal deaths and the need of cardiological screening of the family as a result of the diagnosis.

#### **19. 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; Apr 2017 ; p. 1-8

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

**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.

**20. Person-centred transition programme to empower adolescents with congenital heart disease in the transition to adulthood: a study protocol for a hybrid randomised controlled trial (STEPSTONES project).**

**Author(s):** Acuña Mora, Mariela; Sparud-Lundin, Carina; Bratt, Ewa-Lena; Moons, Philip

**Source:** BMJ open; Apr 2017; vol. 7 (no. 4); p. e014593

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

**Abstract:** INTRODUCTION When a young person grows up, they evolve from an independent child to an empowered adult. If an individual has a chronic condition, this additional burden may hamper adequate development and independence. Transition programmes for young persons with chronic disorders aim to provide the necessary skills for self-management and participation in care. However, strong evidence on the effects of these interventions is lacking. Therefore, as part of the STEPSTONES project (Swedish Transition Effects Project Supporting Teenagers with chrONic mEdical conditionS), we propose a trial to assess the effectiveness of a structured, person-centred transition programme to empower adolescents with congenital heart disease in the transition to adulthood. METHODS/DESIGN STEPSTONES will use a hybrid experimental design in which a randomised controlled trial is embedded in a longitudinal, observational study. It will be conducted in 4 paediatric cardiology centres in Sweden. 2 centres will be allocated to the randomised controlled trial group, assigning patients randomly to the intervention group (n=63) or the comparison group (n=63). The other 2 centres will form the intervention-naïve control group (n=63). The primary outcome is the level of patient empowerment, as measured by the Gothenburg Young Persons Empowerment Scale (GYPES). ETHICS AND DISSEMINATION The study has been approved by the Regional Ethical Board of Gothenburg, Sweden. Findings will be reported following the CONSORT statement and disseminated at international conferences and as published papers in peer-reviewed journals. TRIAL REGISTRATION NUMBER NCT02675361; pre-results.

**21. Pregnancy in Adult Congenital Heart Disease: Special Delivery.**

**Author(s):** Davidson, William R

**Source:** JAMA cardiology; Apr 2017

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

## 22. Sudden cardiac death in adults with congenital heart disease: does QRS-complex fragmentation discriminate in structurally abnormal hearts?

**Author(s):** Vehmeijer, Jim T; Koyak, Zeliha; Bokma, Jouke P; Budts, Werner; Harris, Louise; Mulder, Barbara J M; de Groot, Joris R

**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; Apr 2017

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

Available in full text at [Europace](#) - from Highwire Press

**Abstract:** Aims Sudden cardiac death (SCD) causes a large portion of all mortality in adult congenital heart disease (ACHD) patients. However, identification of high-risk patients remains challenging. Fragmented QRS-complexes (fQRS) are a marker for SCD in patients with acquired heart disease but data in ACHD patients are lacking. We therefore aim to evaluate the prognostic value of fQRS for SCD in ACHD patients. Methods and results From a multicentre cohort of 25 790 ACHD patients, we included tachyarrhythmic SCD cases (n = 147), and controls (n = 266) matched by age, gender, congenital defect and (surgical) intervention. fQRS was defined as  $\geq 1$  discontinuous deflection in narrow QRS-complexes, and  $\geq 2$  in wide QRS-complexes ( $>120$  ms), in two contiguous ECG leads. We calculated odds ratios (OR) using univariable and multivariable conditional logistic regression models correcting for impaired systemic ventricular function, heart failure and QRS duration  $>120$  ms. ECGs of 147 SCD cases (65% male, median age of death 34 years) and of 266 controls were assessed. fQRS was present in 51% of cases and 34% of controls (OR 2.0,  $P = 0.003$ ). In multivariable analysis, fQRS was independently associated with SCD (OR 1.9,  $P = 0.01$ ). The most common diagnose of SCD cases was tetralogy of Fallot (ToF, 34 cases). In ToF, fQRS was present in 71% of cases vs. 43% of controls (OR for SCD 2.8,  $P = 0.03$ ). Conclusions fQRS was independently associated with SCD in ACHD patients in a cohort of SCD patients and matched controls. fQRS may therefore contribute to the decision when evaluating ACHD patients for primary prevention of SCD.

## 23. Cardiopulmonary Exercise Testing in Adult Congenital Heart Disease.

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

**Source:** Annals of the American Thoracic Society; Apr 2017

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

Available in full text at [Annals of the American Thoracic Society](#) - from EBSCOhost

Available in full text at [Annals of the American Thoracic Society](#) - from ProQuest

**Abstract:** Recently, the number of patients with congenital heart diseases reaching adulthood has been progressively increasing in developed countries and new issues are emerging regarding them: 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, 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 oxygen uptake, an early anaerobic threshold, a blunted heart rate response, a reduced rise of tidal volume, and an increased ratio of minute ventilation to carbon dioxide production. 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 have also prognostic significance. For these patients, cardiopulmonary exercise testing allows evaluation and decisions affecting life-style and therapeutic interventions.

#### **24. Readiness for Transition to Adult Health Care for Young Adolescents with Congenital Heart Disease.**

**Author(s):** Stewart, Kimberly T; Chahal, Nita; Kovacs, Adrienne H; Manlihot, Cedric; Jelen, Ahlexxi; Collins, Tanveer; McCrindle, Brian W

**Source:** Pediatric cardiology; Apr 2017; vol. 38 (no. 4); p. 778-786

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

**Abstract:** This study evaluates transition readiness, medical condition knowledge, self-efficacy, and illness uncertainty in young adolescents (ages 12 to 15 years) with congenital heart disease (CHD), and medical, patient, and parental factors associated with transition readiness. We enrolled 82 patients with moderate or complex CHD (n = 36, 44% male; mean age 13.6 ± 1.3 years), and their parents. Patients completed standardized self-report measures: Transition Readiness Assessment Questionnaire (TRAQ), MyHeart scale, General Self-Efficacy scale, and Children's Uncertainty in Illness Scale. Parents completed the MyHeart scale and demographic information. Many young adolescents had not discussed transfer with a health care provider (n = 20, 24%) or parent (n = 34, 41%). Transition readiness was higher among patients who were older, more knowledgeable about their condition, had a history of primary cardiac repair and greater self-efficacy, and was lower for boys and patients on cardiac medications. Transition readiness was unrelated to CHD diagnosis and patients' illness uncertainty. Patients' self-advocacy skills were superior to their chronic disease self-management skills. Increased parental medical condition knowledge was positively correlated with patient knowledge, and patient-parent discussion of transfer was associated with increased patient's self-management skills. Transition is not uniformly discussed with young adolescent CHD patients. Parental involvement is correlated with increased transition readiness and patient disease self-management skills. Young adolescent transition programs should focus on education around improving patient medical condition knowledge, promote chronic disease self-management skills development, and include parental involvement.

#### **25. Arrhythmias in Adults with Congenital Heart Disease: What Are Risk Factors for Specific Arrhythmias?**

**Author(s):** Loomba, Rohit S; Buelow, Matthew W; Aggarwal, Saurabh; Arora, Rohit R; Kovach, Joshua; Ginde, Salil

**Source:** Pacing and clinical electrophysiology : PACE; Apr 2017; vol. 40 (no. 4); p. 353-361

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

**Abstract:** INTRODUCTION An increasing number of patients with congenital heart disease are now surviving into adulthood. This has also led to the emergence of complications from the underlying congenital heart disease, related surgical interventions, and associated comorbidities. While the prevalence of particular arrhythmias with specific congenital heart disease has been previously described, a detailed analysis of all lesions and a large number of comorbidities has not been previously published. METHODS Admissions with congenital heart disease were identified in the National Inpatient Sample. Associated comorbidities were also identified for these patients. Univariate analysis was done to compare those risk factors associated with specific arrhythmias in the setting of congenital heart disease. Next, regression analysis was done to identify what patient

characteristics and comorbidities were associated with increased risk of specific arrhythmias. RESULTS A total of 52,725,227 admissions were included in the analysis. Of these, 109,168 (0.21%) had congenital heart disease. Of those with congenital heart disease, 27,088 (25%) had an arrhythmia at some point. The most common arrhythmia in those with congenital heart disease was atrial fibrillation, which was noted in 86% of those with arrhythmia followed by atrial flutter which was noted in 20% of those with congenital heart disease. The largest burden of arrhythmia was found to be in those with tricuspid atresia with a 51% prevalence of arrhythmia in this group followed by Ebstein anomaly which had an arrhythmia prevalence of 39%. Increasing age, male gender, double outlet right ventricle, atrioventricular septal defect, heart failure, obstructive sleep apnea, transposition of the great arteries, congenitally corrected transposition, and tetralogy of Fallot were frequently noted to be independent risk factors of specific arrhythmias. CONCLUSION Approximately, 25% of adult admissions with congenital heart disease are associated with arrhythmia. The burden of arrhythmia varies by the specific lesion and other risk factors as well. Understanding of these can help in risk stratification and can help devise strategies to lower this risk.

## **26. Maternal and Fetal Outcomes of Admission for Delivery in Women With Congenital Heart Disease.**

**Author(s):** Hayward, Robert M; Foster, Elyse; Tseng, Zian H

**Source:** JAMA cardiology; Apr 2017

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

**Abstract:** Background Women with congenital heart disease (CHD) may be at increased risk for adverse events during pregnancy and delivery. Objective To compare delivery outcomes between women with and without CHD. Design, Setting, and Participants This retrospective study of inpatient delivery admissions in the Healthcare Cost and Utilization Project's California State Inpatient Database compared maternal and fetal outcomes between women with and without CHD by using multivariate logistic regression. Female patients with codes for delivery from the International Classification of Diseases, Ninth Revision, from January 1, 2005, through December 31, 2011, were included. The association of CHD with readmission was assessed to 7 years after delivery. Cardiovascular morbidity and mortality were hypothesized to be higher among women with CHD. Data were analyzed from April 4, 2014, through January 23, 2017. Exposures Noncomplex and complex CHD. Main Outcomes and Measures Maternal outcomes included in-hospital arrhythmias, eclampsia or preeclampsia, congestive heart failure (CHF), length of stay, preterm labor, anemia complicating pregnancy, placental abnormalities, infection during labor, maternal readmission at 1 year, and in-hospital mortality. Fetal outcomes included growth restriction, distress, and death. Results Among 3 642 041 identified delivery admissions, 3189 women had noncomplex CHD (mean [SD] age, 28.6 [7.6] years) and 262 had complex CHD (mean [SD] age, 26.5 [6.8] years). Women with CHD were more likely to undergo cesarean delivery (1357 [39.3%] vs 1 164 509 women without CHD [32.0%];  $P < .001$ ). Incident CHF, atrial arrhythmias, ventricular arrhythmias, and maternal mortality were uncommon during hospitalization, with each occurring in fewer than 10 women with noncomplex or complex CHD (<0.5% each). After multivariate adjustment, noncomplex CHD (odds ratio [OR], 9.7; 95% CI, 4.7-20.0) and complex CHD (OR, 56.6; 95% CI, 17.6-182.5) were associated with greater odds of incident CHF. Similar odds were found for atrial arrhythmias in noncomplex (OR, 8.2; 95% CI, 3.0-22.7) and complex (OR, 31.8; 95% CI, 4.3-236.3) CHD, for fetal growth restriction in noncomplex (OR, 1.6; 95% CI, 1.3-2.0) and complex (OR, 3.5; 95% CI, 2.1-6.1) CHD, and for hospital readmission in both CHD groups combined (OR, 3.6; 95% CI, 3.3-4.0). Complex CHD was associated with greater adjusted odds of serious ventricular arrhythmias (OR, 31.8; 95% CI, 4.3-236.3) and maternal in-hospital mortality (OR, 79.1; 95% CI, 23.9-261.8). Conclusions and Relevance In this study of hospital admissions for delivery in California, CHD was associated with incident CHF,

atrial arrhythmias, and fetal growth restriction and complex CHD was associated with ventricular arrhythmias and maternal in-hospital mortality, although these outcomes were rare, even in women with complex CHD. These findings may guide monitoring decisions and risk assessment for pregnant women with CHD at the time of delivery.

### **27. Incidence and predictors of obstetric and fetal complications in women with structural heart disease.**

**Author(s):** van Hagen, Iris M; Roos-Hesselink, Jolien W; Donvito, Valentina; Liptai, Csilla; Morissens, Marielle; Murphy, Daniel J; Galian, Laura; Bazargani, Nooshin Mohd; Cornette, Jérôme; Hall, Roger; Johnson, Mark R

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

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

Available in full text at [Heart](#) - from Highwire Press

**Abstract:** OBJECTIVE Women with cardiac disease becoming pregnant have an increased risk of obstetric and fetal events. The aim of this study was to study the incidence of events, to validate the modified WHO (mWHO) risk classification and to search for event-specific predictors. METHODS The Registry Of Pregnancy And Cardiac disease is a worldwide ongoing prospective registry that has enrolled 2742 pregnancies in women with known cardiac disease (mainly congenital and valvular disease) before pregnancy, from January 2008 up to April 2014. RESULTS Mean age was 28.2±5.5 years, 45% were nulliparous and 33.3% came from emerging countries. Obstetric events occurred in 231 pregnancies (8.4%). Fetal events occurred in 651 pregnancies (23.7%). The mWHO classification performed poorly in predicting obstetric (c-statistic=0.601) and fetal events (c-statistic=0.561). In multivariable analysis, aortic valve disease was associated with pre-eclampsia (OR=2.6, 95%CI=1.3 to 5.5). Congenital heart disease (CHD) was associated with spontaneous preterm birth (OR=1.8, 95%CI=1.2 to 2.7). Complex CHD was associated with small-for-gestational-age neonates (OR=2.3, 95%CI=1.5 to 3.5). Multiple gestation was the strongest predictor of fetal events: fetal/neonatal death (OR=6.4, 95%CI=2.5 to 16), spontaneous preterm birth (OR=5.3, 95%CI=2.5 to 11) and small-for-gestational age (OR=5.0, 95%CI=2.5 to 9.8). CONCLUSION The mWHO classification is not suitable for prediction of obstetric and fetal events in women with cardiac disease. Maternal complex CHD was independently associated with fetal growth restriction and aortic valve disease with pre-eclampsia, potentially offering an insight into the pathophysiology of these pregnancy complications. The increased rates of adverse obstetric and fetal outcomes in women with pre-existing heart disease should be highlighted during counselling.

### **28. National In-Hospital Outcomes of Pregnancy in Women With Single Ventricle Congenital Heart Disease.**

**Author(s):** Collins, R Thomas; Chang, Di; Sandlin, Adam; Goudie, Anthony; Robbins, James M

**Source:** The American journal of cardiology; Apr 2017; vol. 119 (no. 7); p. 1106-1110

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

Available in full text at [American Journal of Cardiology, The](#) - from ProQuest

**Abstract:** Most patients with single ventricle (SV) congenital heart disease are expected to survive to adulthood. Women with SV are often counseled against pregnancy; however, data on pregnancies in these women are lacking. We sought to evaluate in-hospital outcomes of pregnancy in women with SV. We used nationally representative data from the 1998 to 2012 National Inpatient Sample to identify women ≥18 years of age admitted to the hospital with International Classification of

Diseases-9th Revision codes for an intrauterine pregnancy and a diagnosis of hypoplastic left heart syndrome, tricuspid atresia, or common ventricle. A matched comparison group without a diagnosis of congenital heart disease or pulmonary hypertension was identified from the database. National estimates of hospitalizations were calculated. Length of stay, hospital charges, and complications were analyzed and compared between groups. Charge data were adjusted to 2012 dollars. There were 282 admissions of pregnant women with SV (69% with deliveries) and 1,405 admissions in the control group (88% with deliveries). Vaginal delivery was more common in SV (74% vs 71%,  $p < 0.001$ ). Length of stay ( $4.1 \pm 0.91$  vs  $2.8 \pm 0.18$  days,  $p < 0.001$ ) and charges ( $\$30,787 \pm 8,109$  vs  $\$15,536 \pm 1,006$ ,  $p < 0.0001$ ) were higher in the SV group. Complications occurred in most SV admissions and were more common in the SV group than in the control group. No deaths occurred. Cardiovascular complications occurred in 25% of pregnancy-related hospitalizations, although in-hospital pregnancy-related death is rare. Vaginal delivery is common in these patients. These data suggest that pregnancy and vaginal delivery can be tolerated in women with SV, although the risk for a cardiovascular event is significantly higher than in the general population.

### **29. Educational needs of adolescents with congenital heart disease: Impact of a transition intervention programme.**

**Author(s):** Ladouceur, Magalie; Calderon, Johanna; Traore, Maladon; Cheurfi, Radhia; Pagnon, Christine; Khraiche, Diala; Bajolle, Fanny; Bonnet, Damien

**Source:** Archives of cardiovascular diseases; Mar 2017

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

**Abstract:** BACKGROUND Adolescents and young adults with congenital heart disease (CHD) have complex health needs and require lifelong follow-up. Interventions to facilitate the paediatric-to-adult healthcare transition are recommended, but outcomes remain largely under-investigated. AIMS To identify the educational needs and the impact of a transition intervention on knowledge and self-management skills in adolescents and young adults with CHD. METHODS From September 2014 to May 2015, 115 adolescents and young adults with CHD (mean age  $17 \pm 2$  years; 47 girls) were consecutively enrolled. Among these, 22 had participated in a structured educational programme in the previous  $11 \pm 4$  months (education group) and 93 had not (comparison group). Knowledge about their health status was assessed using a targeted CHD questionnaire. RESULTS The mean overall health knowledge score (maximum of 20) in the education group was significantly higher than in the comparison group ( $11.7 \pm 3.5$  vs.  $8.6 \pm 3.2$ ;  $P < 0.001$ ). We observed significant gaps in knowledge in the comparison group: e.g. 61.3% vs. 90.0% knew their condition name ( $P = 0.01$ ), 21.5% vs. 63.6% were aware of recommended follow-up ( $P = 0.004$ ), and 12.8% vs. 75.0% of girls knew to check their heart condition before pregnancy ( $P < 0.01$ ). In multivariable analysis, after adjustment for age, structured CHD education and higher academic attainment were significant determinants of health-related knowledge ( $P < 0.01$ ). CONCLUSION Education during adolescent-to-adult transition has a significant impact on health knowledge. Structured CHD educational programmes could improve understanding and prevent potential future complications.

### **30. Advance Care Planning in Adults with Congenital Heart Disease: A Patient Priority.**

**Author(s):** Deng, Lisa X; Gleason, Lacey P; Khan, Abigail M; Drajpuch, David; Fuller, Stephanie; Goldberg, Leah A; Mascio, Christopher E; Partington, Sara L; Tobin, Lynda; Kim, Yuli Y; Kovacs, Adrienne H

**Source:** International journal of cardiology; Mar 2017; vol. 231 ; p. 105-109

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

**Abstract:** BACKGROUND Adult congenital heart disease (ACHD) patients with moderate or great defect complexity are at risk for premature death. Although early engagement in advance care planning (ACP) is recommended, previous research suggests that it seldom occurs. METHODS This study investigated ACHD patient preferences for ACP and factors that impact preferences. ACHD patients completed an ACP preferences questionnaire, the Hospital Anxiety and Depression Scale and a measure of attachment styles. RESULTS Of 152 ACHD patients (median age 33years, 50% female), 13% reported previous ACP discussions with providers and 21% had completed advance directives. On a 0-10 scale, the median rating for the importance of discussing ACP with providers was 7; 18years was identified as the most appropriate age to initiate this dialogue. Higher ratings for the importance of discussing ACP with providers was observed in patients who were female ( $p=0.03$ ), had lower disease complexity ( $p=0.03$ ), and had elevated anxiety symptoms ( $p=0.001$ ); elevated anxiety remained significant in a multivariable model. Interest in receiving information about life expectancy (61% overall) was greater among patients with lower disease complexity ( $p=0.04$ ) and a history of  $\geq 2$  cardiac surgeries ( $p=0.01$ ); disease complexity remained significant in a multivariable model. CONCLUSIONS As a group, ACHD patients value the opportunity for ACP discussions and prefer earlier communication. Although some clinicians might avoid ACP discussions in patients who are generally more anxious or have less complex CHD, such avoidance does not appear to be warranted.

### 31. Characteristics and outcomes of heart failure-related hospitalization in adults with congenital heart disease.

**Author(s):** Moussa, Nidhal Ben; Karsenty, Clement; Pontnau, Florence; Malekzadeh-Milani, Sophie; Boudjemline, Younes; Legendre, Antoine; Bonnet, Damien; Iserin, Laurence; Ladouceur, Magalie

**Source:** Archives of cardiovascular diseases; Mar 2017

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

**Abstract:** BACKGROUND Heart failure (HF) is the main cause of death in adult congenital heart disease (ACHD). AIMS We aimed to characterize HF-related hospitalization of patients with ACHD, and to determine HF risk factors and prognosis in this population. METHODS We prospectively included 471 patients with ACHD admitted to our unit over 24 months. Clinical and biological data and HF management were recorded. Major cardiovascular events were recorded for ACHD with HF. RESULTSHF was the main reason for hospitalization in 13% of cases (76/583 hospitalizations). Patients with HF were significantly older (median age  $44\pm 14$  years vs.  $37\pm 15$  years;  $P<0.01$ ), with more complex congenital heart disease ( $P=0.04$ ). In the multivariable analysis, pulmonary arterial hypertension (odds ratio [OR] 6.2, 95% confidence interval [CI] 3.5-10.7), history of HF (OR 9.8, 95% CI 5.7-16.8) and history of atrial arrhythmia (OR 3.6, 95% CI 2.2-5.9) were significant risk factors for HF-related admissions ( $P<0.001$ ). The mean hospital stay of patients with HF was longer (12.2 vs. 6.9 days;  $P<0.01$ ), and 25% of patients required intensive care. Overall, 11/55 (20%) patients with HF died, 10/55 (18%) were readmitted for HF, and 6/55 (11%) had heart transplantation during the median follow-up of 18 months (95% CI 14-20 months). The risk of cardiovascular events was 19-fold higher after HF-related hospitalization. CONCLUSIONS HF is emerging as a leading cause of morbidity and mortality in the ACHD population. Earlier diagnosis and more active management are required to improve outcomes of HF in ACHD.

### 32. Adult Congenital Cardiac Care.

**Author(s):** Kogon, Brian E; Miller, Kati; Miller, Paula; Alsoufi, Bahaaldin; Rosenblum, Joshua M

**Source:** World journal for pediatric & congenital heart surgery; Mar 2017; vol. 8 (no. 2); p. 242-247



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

**Abstract:** BACKGROUND The Adult Congenital Heart Association (ACHA) is dedicated to supporting patients with congenital heart disease. To guide patients to qualified providers and programs, it maintains a publicly accessible directory of dedicated adult congenital cardiac programs. We analyzed the directory in 2006 and 2015, aiming to evaluate the growth of the directory as a whole and to evaluate the growth of individual programs within the directory. We also hope this raises awareness of the growing opportunities that exist in adult congenital cardiology and cardiac surgery. METHODS Data in the directory are self-reported. Only data from US programs were collected and analyzed. RESULTS By the end of 2015, compared to 2006, there were more programs reporting to the directory in more states (107 programs across 42 states vs 57 programs across 33 states), with higher overall clinical volume (591 vs 164 half-day clinics per week, 96,611 vs 34,446 patient visits). On average, each program was busier (5 vs 2 half-day clinics per week per program). Over the time period, the number of reported annual operations performed nearly doubled (4,346 operations by 210 surgeons vs 2,461 operations by 125 surgeons). Access to ancillary services including specific clinical diagnostic and therapeutic services also expanded. CONCLUSION Between 2006 and 2015, the clinical directory and the individual programs have grown. Current directory data may provide benchmarks for staffing and services for newly emerging and existing programs. Verifying the accuracy of the information and inclusion of all programs will be important in the future.

**33. Pregnancy in women with complete transposition of the great arteries following the atrial switch procedure. A study from three of the largest Adult Congenital Heart Disease centers in Poland.**

**Author(s):** Lipczyńska, Magdalena; Szymański, Piotr; Trojnarowska, Olga; Tomkiewicz-Pająk, Lidia; Pietrzak, Bronisława; Klisiewicz, Anna; Kumor, Magdalena; Podolec, Piotr; Hoffman, Piotr

**Source:** The journal of maternal-fetal & neonatal medicine : the official journal of the European Association of Perinatal Medicine, the Federation of Asia and Oceania Perinatal Societies, the International Society of Perinatal Obstetricians; Mar 2017; vol. 30 (no. 5); p. 563-567

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

**Abstract:** OBJECTIVE We sought to identify maternal/neonatal and cardiovascular complications in pregnant women with complete transposition of great arteries (D-TGA) following atrial switch. METHODS Clinical records of all women with D-TGA after the Mustard/Senning (M/S) operation who were followed at the three largest Adult Congenital Heart Disease (ACHD) centers in Poland were reviewed. RESULTS Fifteen of the fifty-nine women followed had a total of 24 pregnancies, including two spontaneous miscarriages. Twenty-two pregnancies (92%) resulted in a live birth, 91% were by cesarean section. During 5 (23%) of the 22 completed pregnancies obstetric complications were observed (one gestational diabetes, one hypertension in pregnancy, one placenta increta and two preterm labors). The mean pregnancy duration was 37.2 weeks (range: 26-41 weeks). We observed one neonatal death due to extreme prematurity. Six (25%) children had a birth weight of  $\leq 2500$  g. None of women had severe cardiac complications during pregnancy nor in the postpartum period. CONCLUSIONS In our study, we demonstrated a large number of obstetric complications and low birth weight in the presence of a systemic right ventricle. However, from a cardiologist's point of view pregnancy after the M/S operation was well-tolerated and relatively safe.

**34. Hopelessness among adults with congenital heart disease: Cause for despair or hope?**

**Author(s):** Eslami, Bahareh; Kovacs, Adrienne H; Moons, Philip; Abbasi, Kyomars; Jackson, Jamie L

**Source:** International journal of cardiology; Mar 2017; vol. 230; p. 64-69

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

**Abstract:** BACKGROUND Adults with congenital heart disease (CHD) face unique life courses and challenges that may negatively influence their psychological functioning. The aims of this study were to (1) examine the level of hopelessness among adults with CHD in comparison with non-CHD participants and (2) identify correlates of elevated hopelessness among adults with CHD. METHODS We enrolled 347 patients with CHD (18-64years, 52.2% female) and 353 matched (by sex/age) non-CHD persons in this cross-sectional study. Hopelessness was assessed by Beck Hopelessness Scale. Hierarchical multiple logistic regression analyses were performed to explore correlates of elevated hopelessness. RESULTS The mean total hopelessness score did not significantly differ between the CHD and non-CHD groups. Twenty-eight percent of CHD patients had elevated hopelessness scores. Within the CHD patient sample, regression analyses revealed that being male (odds ratio=2.62), not having children (odds ratio=3.57), being unemployed (odds ratio=2.27), and elevated depressive symptoms (odds ratio=1.21) were significantly associated with hopelessness. Regular physical activity (odds ratio=0.36) emerged as a protective factor and all CHD disease parameters were unrelated to hopelessness. The final model explained 43% of the variance in hopelessness. CONCLUSIONS Adult CHD teams are encouraged to continue to explore strategies to support patients to live as rich and full as lives as possible by pursuing relationships, employment and physical activity, as well as managing depression and hopelessness.

**35. Sudden cardiac death in adult congenital heart disease: can the unpredictable be foreseen?**

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

**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; Mar 2017; vol. 19 (no. 3); p. 401-406

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

Available in full text at [Europace](#) - from Highwire Press

**Abstract:** Aims Sudden cardiac death (SCD) is a major cause of mortality in adults with congenital heart disease (CHD). Several risk factors for SCD including conduction disturbances and ventricular dysfunction have been described previously. However, electrocardiogram (ECG) and echocardiographic parameters may change over time, and the predictive value of such temporal changes, rather than their point estimates, for SCD remains unknown. Methods and results This was a retrospective case-control study in adults with CHD and proven or presumed SCD and matched controls. Data were obtained from three databases including 25 000 adults with CHD. Sequential measurements were performed on electrocardiograms and echocardiograms. Ventricular function was assessed by echocardiography and graded on a four-point ordinal scale: 1, normal [ejection fraction (EF)  $\geq 50\%$ ]; 2, mildly impaired (EF 40-49%); 3, moderately impaired (EF 30-39%); and 4, severely impaired (EF  $< 30\%$ ). Overall, 131 SCDs (mean age  $36 \pm 14$  years, 67% male) and 260 controls (mean age  $37 \pm 13$  years, 63% male) were included. At baseline, median QRS duration was 108 ms (range 58-168 ms) in SCDs and 97 ms (range 50-168 ms) in controls and increased over time at a rate of  $1.6 \pm 0.5$  vs.  $0.5 \pm 0.2$  ms/year in SCDs and controls, respectively ( $P = 0.011$ ). QT dispersion at baseline was 61 ms (range 31-168 ms) in SCDs and 50 ms (range 21-129 ms) in controls. QT dispersion increased at a rate of  $1.1 \pm 0.4$  ms/year in SCD victims and decreased at a rate of  $0.2 \pm 0.2$  ms/year in controls ( $P = 0.004$ ). Increase of QRS duration  $\geq 5$  ms/year was associated with an increased risk of SCD [OR 1.9, 95% confidence interval (CI) 1.1-3.3,  $P = 0.013$ ]. Change from any baseline systemic ventricular function (normal, mild, or moderately impaired) to severe ventricular

dysfunction over time was associated with the highest risk of SCD (OR 16.9, 95% CI 1.8-120.1, P = 0.008). Conclusion In adults with CHD, QRS duration and ventricular dysfunction progress over time. Progression of QRS duration and the rate of impairment of ventricular function served to identify those at increased risk of SCD.

### **36. Neurocognitive functioning in adults with congenital heart disease.**

**Author(s):** Ilardi, Dawn; Ono, Kim E; McCartney, Rebecca; Book, Wendy; Stringer, Anthony Y

**Source:** Congenital heart disease; Mar 2017; vol. 12 (no. 2); p. 166-173

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

**Abstract:** OBJECTIVE Adults with congenital heart disease (CHD) are at increased risk of psychological disorders and cognitive deficiencies due to structural/acquired neurological abnormalities and neurodevelopmental disorders as children. However, limited information is known about the neuropsychological functioning of adults with CHD. This study screened neuropsychological abilities and explored group differences related to cardiac disease severity and neurological risk factors in adults with CHD. DESIGN Participants completed brief neuropsychological testing. Information about neurobehavioral and psychological symptoms, employment, education, and disability were also collected from the patient and a family member. RESULTS Forty-eight participants with adult CHD completed neuropsychological testing. Visuospatial skills and working memory were worse than expected compared to the typical population. Frequency of neurological comorbidities (e.g., stroke, seizures) was higher in those with more severe heart disease (e.g., single ventricle or cyanotic disease), and executive functioning was weaker in those with neurological comorbidities. Those with more severe heart disease were more likely to be unemployed and to receive disability benefits, but educational attainment did not differ. Those who received disability performed worse on tasks of executive functioning. CONCLUSIONS Findings suggest concerns about neuropsychological functioning that need to be more comprehensively assessed in adults with CHD. Understanding the cognitive limitations of this aging population can help guide access to resources, transition of care, and medical care engagement, thus improving quality of care and quality of life.

### **37. NEUROCOGNITIVE IMPAIRMENT IS COMMON IN THE ADULT WITH CONGENITAL HEART DISEASE: IDENTIFICATION USING A NOVEL CLINICAL QUESTIONNAIRE.**

**Author(s):** Cohen, Scott; Leverenz, Ashley; Reis, Michael; Umfleet, Laura; Ginde, Salil; Bartz, Peter; Earing, Michael

**Source:** Journal of the American College of Cardiology (JACC); Mar 2017; vol. 69 ; p. 565-565

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

Available in full text at [Journal of the American College of Cardiology](#) - from ProQuest

### **38. MORTALITY IN PREGNANT WOMEN WITH CONGENITAL HEART DISEASE: A SUBANALYSIS OF THE CALIFORNIA PREGNANCY-ASSOCIATED MORTALITY REVIEW.**

**Author(s):** Sabanayagam, Aarthi; Agarwal, Anushree; MacCain, Christy; Lawton, Elizabeth; Main, Elliot; Hameed, Afshan; Harris, Ian; Foster, Elyse

**Source:** Journal of the American College of Cardiology (JACC); Mar 2017; vol. 69 ; p. 606-606

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

Available in full text at [Journal of the American College of Cardiology](#) - from ProQuest

### 39. The role of palliative care in critical congenital heart disease.

**Author(s):** Mazwi, Mjaye L; Henner, Natalia; Kirsch, Roxanne

**Source:** Seminars in perinatology; Mar 2017; vol. 41 (no. 2); p. 128-132

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

**Abstract:** Patients with critical congenital heart disease are exposed to significant lifetime morbidity and mortality. Prenatal diagnosis can provide opportunities for anticipatory co-management of patients between palliative subspecialists and the cardiac care team. The benefits of palliative care include support for longitudinal decision-making and avoidance of interventions not consistent with family goals. Effectively counseling families requires an up-to-date understanding of outcomes and knowledge of provider biases. Patient-proxy reported quality of life (QOL) is highly variable in this population and healthcare providers need to be aware of limitations in their own subjective assessment of QOL.

### 40. Changing Landscape of Congenital Heart Disease.

**Author(s):** Bouma, Berto J; Mulder, Barbara J M

**Source:** Circulation research; Mar 2017; vol. 120 (no. 6); p. 908-922

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

Available in full text at [Circulation Research](#) - from Highwire Press

**Abstract:** Congenital heart disease is the most frequently occurring congenital disorder affecting ≈0.8% of live births. Thanks to great efforts and technical improvements, including the development of cardiopulmonary bypass in the 1950s, large-scale repair in these patients became possible, with subsequent dramatic reduction in morbidity and mortality. The ongoing search for progress and the growing understanding of the cardiovascular system and its pathophysiology refined all aspects of care for these patients. As a consequence, survival further increased over the past decades, and a new group of patients, those who survived congenital heart disease into adulthood, emerged. However, a large range of complications raised at the horizon as arrhythmias, endocarditis, pulmonary hypertension, and heart failure, and the need for additional treatment became clear. Technical solutions were sought in perfection and creation of new surgical techniques by developing catheter-based interventions, with elimination of open heart surgery and new electronic devices enabling, for example, multisite pacing and implantation of internal cardiac defibrillators to prevent sudden death. Over time, many pharmaceutical studies were conducted, changing clinical treatment slowly toward evidence-based care, although results were often limited by low numbers and clinical heterogeneity. More attention has been given to secondary issues like sports participation, pregnancy, work, and social-related difficulties. The relevance of these issues was already recognized in the 1970s when the need for specialized centers with multidisciplinary teams was proclaimed. Finally, research has become incorporated in care. Results of intervention studies and registries increased the knowledge on epidemiology of adults with congenital heart disease and their complications during life, and at the end, several guidelines became easily accessible, guiding physicians to deliver care appropriately. Over the past decades, the landscape of adult congenital heart disease has changed dramatically, which has to be continued in the future.

#### 41. Contraceptive Practices of Women With Complex Congenital Heart Disease.

**Author(s):** Miner, Pamela D; Canobbio, Mary M; Pearson, Dorothy D; Schlater, Mary; Balon, Yvonne; Junge, Kathryn J; Bhatt, Ami; Barber, Deena; Nickolaus, Michelle J; Kovacs, Adrienne H; Moons, Philip; Shaw, Kate; Fernandes, Susan M

**Source:** The American journal of cardiology; Mar 2017; vol. 119 (no. 6); p. 911-915

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

Available in full text at [American Journal of Cardiology, The](#) - from ProQuest

**Abstract:** Understanding the contraceptive practices of women with complex congenital heart disease (CHD) and providing them individualized contraception counseling may prevent adverse events and unplanned high-risk pregnancies. Given this, we sought to examine the contraceptive practices in women with CHD, describe adverse events associated with contraceptive use, and describe the provision of contraception counseling. Women >18 years were recruited from 2011 to 2014 from 9 adult CHD (ACHD) centers throughout North America. Subjects completed a 48-item questionnaire regarding contraceptive use and perceptions of contraception counseling, and a medical record review was performed. Of 505 subjects, median age was 33 (interquartile range 26 to 44) and 81% had CHD of moderate or great complexity. The majority (86%, 435 of 505) of the cohort had used contraception. The types included barrier methods (87%), oral contraception (OC) 84%, intrauterine device (18%), Depo-Provera (15%), vaginal ring (7%), patch (6%), hormonal implant (2%), Plan B (19%), and sterilization (16%). Overall OC use was not significantly different by CHD complexity. Women with CHD of great complexity were more likely to report a thrombotic event while taking OC than those with less complex CHD (9% vs 1%,  $p = 0.003$ ). Contraception counseling by the ACHD team was noted by 43% of subjects. Unplanned pregnancy was reported by 25% with no statistical difference by CHD complexity. In conclusion, contraceptive practices of women with complex CHD are highly variable, and the prevalence of blood clots while taking OC is not insignificant while provision of contraception counseling by ACHD providers appears lacking.

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

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

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

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

Available in full text at [Heart](#) - from Highwire Press

**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.

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

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

**Source:** Pediatrics international : official journal of the Japan Pediatric Society; Feb 2017

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

**Abstract:** BACKGROUND As 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. METHODS A 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. RESULTS The 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.

#### **44. Adult congenital heart disease nurse coordination: Essential skills and role in optimizing team-based care a position statement from the International Society for Adult Congenital Heart Disease (ISACHD).**

**Author(s):** Sillman, Christina; Morin, Joanne; Thomet, Corina; Barber, Deena; Mizuno, Yoshiko; Yang, Hsiao-Ling; Malpas, Theresa; Flocco, Serena Francesca; Finlay, Clare; Chen, Chi-Wen; Balon, Yvonne; Fernandes, Susan M

**Source:** International journal of cardiology; Feb 2017; vol. 229 ; p. 125-131

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

**Abstract:** BACKGROUND Founded in 1992, the International Society for Adult Congenital Heart Disease (ISACHD) is the leading global organization of professionals dedicated to pursuing excellence in the care of adults with congenital heart disease (CHD) worldwide. Among ISACHD's objectives is to "promote a holistic team-based approach to the care of the adult with CHD that is comprehensive, patient-centered, and interdisciplinary" (<http://www.isachd.org>). This emphasis on team-based care reflects the fact that adults with CHD constitute a heterogeneous population with a wide spectrum of disease complexity, frequent association with other organ involvement, and varied co-morbidities and psychosocial issues. METHODS Recognizing the vital role of the adult CHD (ACHD) nurse

coordinator (ACHD-NC) in optimizing team-based care, ISACHD established a task force to elucidate and provide guidance on the roles and responsibilities of the ACHD-NC. Acknowledging that nursing roles can vary widely from region to region based on factors such as credentials, scopes of practice, regulations, and local culture and tradition, an international panel was assembled with experts from North America, Europe, East Asia, and Oceania. The writing committee was tasked with reviewing key aspects of the ACHD-NC's role in team-based ACHD care. **RESULTS/CONCLUSION** The resulting ISACHD position statement addresses the ACHD-NC's role and skills required in organizing, coordinating, and facilitating the care of adults with CHD, holistic assessment of the ACHD patient, patient education and counseling, and support for self-care management and self-advocacy.

#### **45. Healthcare needs of adolescents with congenital heart disease transitioning into adulthood: a Delphi survey of patients, parents, and healthcare providers.**

**Author(s):** Chi-Wen Chen; Wen-Jen Su; Yueh-Tao Chiang; Ying-Mei Shu; Philip Moons

**Source:** European Journal of Cardiovascular Nursing; Feb 2017; vol. 16 (no. 2); p. 125-135

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

**Abstract:**Background: The increasing survival of children with congenital heart disease (CHD) challenges healthcare systems regarding how to manage the many health needs of patients undergoing transitional care. A comprehensive understanding of the perspectives of patients, parents, and healthcare providers is required. Objective: This study systematically identified the healthcare needs of adolescents with CHD transitioning into young adults by collecting the perspectives of patients, parents, and healthcare providers. Methods: A sample of CHD patients (n = 29), parents of children with CHD (n = 29), and healthcare providers (n = 16) completed the two-round Delphi study, and 64 healthcare needs were identified. The central tendency and the level of dispersion were computed in order to establish a consensus. Results: A consensus was reached on 25 healthcare needs including health, family, individual, interpersonal interaction, and policy dimensions, which were classified as important with a moderate to high level of agreement by all three groups. The three groups were strongly agreed that "encouraging the patient to learn health self-management" and "cultivating a positive attitude toward the illness" were very important. The opinions of the three groups differed significantly on 12 needs ( $p < 0.05$ ) related to health, family, and policy dimensions. Conclusion: A consensus was reached on the needs that were identified as being potentially valid measures of the healthcare needs of adolescents with CHD transitioning into young adults. The identified needs can serve as the basis for establishing a transitional health passport and developing a clinical intervention for adolescents with CHD transitioning into young adults.

#### **46. Survivorship in Children and Young Adults With Congenital Heart Disease in Sweden.**

**Author(s):** Mandalenakis, Zacharias; Rosengren, Annika; Skoglund, Kristofer; Lappas, Georgios; Eriksson, Peter; Dellborg, Mikael

**Source:** JAMA Internal Medicine; Feb 2017; vol. 177 (no. 2); p. 224-230

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

Available in full text at [JAMA Internal Medicine](#) - from EBSCOhost

**Abstract:** Importance: Mortality in patients with congenital heart disease (CHD) has markedly decreased during recent decades because of advancement in pediatric care. However, there are limited data on survival trends in children and young adults with CHD compared with the general population. Objective: To determine survivorship in children and young adults with CHD compared

with matched controls. Design, Setting, and Participants: A registry-based, prospective, matched-cohort study was conducted in Sweden. Data from the national patient and cause of death registers were linked to identify individuals with CHD born between January 1, 1970, and December 31, 1993, who were registered at or after birth. Follow-up and comorbidity data were collected until December 31, 2011. Survival analyses were performed with the Cox proportional hazards model; these analyses were performed from January 1, 1970, to December 31, 2011. A total of 21 982 patients with CHD in Sweden were identified. The mean (SD) follow-up time was 27.0 (8.86) years. Children serving as controls (n = 219 816) (10 for each patient), matched for birth year, sex, and county, were randomly selected from the general population. Main Outcomes and Measures: Survivorship in young patients with CHD and controls. Results: Of the 21 982 patients who were born between 1970 and 1993 and were registered with the diagnosis of CHD, 10 650 were female (48.4%). Median age at index registration was 4.22 years (interquartile range, 17.07 years). Survivorship among children younger than 5 years was increased from 96% in those born in 1970-1979 to 98% in those born in 1990-1993. Hazard ratios (HRs) of death in relation to that in control individuals decreased from 225.84 (95% CI, 136.84-372.70) to 33.47 (95% CI, 22.54-49.70). A substantial, but less pronounced, absolute and relative increase in survivorship was found in older patients (HRs ranged from 24.52; 95% CI, 11.72-51.26, at 5-9 years to 4.27; 95% CI, 2.29-7.95, at 18-29 years). According to a hierarchical CHD classification, the group of patients with the most severe complex defects (ie, common arterial trunk, transposition of the great vessels, double inlet ventricle, hypoplastic left heart syndrome, tetralogy of Fallot, and atrioventricular septal defect) had the highest risk for death (HR, 64.07; 95% CI, 53.39-76.89). Conclusions and Relevance: Despite substantially increasing absolute and relative survivorship in children and young adults with CHD, the mortality risk remains high compared with the risk in matched controls. Further research on reducing the death rate in this vulnerable group is required.

#### **47. Management of Pregnancy in Patients With Complex Congenital Heart Disease: A Scientific Statement for Healthcare Professionals From the American Heart Association.**

**Author(s):** Canobbio, Mary M; Warnes, Carole A; Aboulhosn, Jamil; Connolly, Heidi M; Khanna, Amber; Koos, Brian J; Mital, Seema; Rose, Carl; Silversides, Candice; Stout, Karen; American Heart Association Council on Cardiovascular and Stroke Nursing; Council on Clinical Cardiology; Council on Cardiovascular Disease in the Young; Council on Functional Genomics and Translational Biology; and Council on Quality of Care and Outcomes Research

**Source:** *Circulation*; Feb 2017; vol. 135 (no. 8); p. e50

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

Available in full text at [Circulation](#) - from Highwire Press

**Abstract:** Today, most female children born with congenital heart disease will reach childbearing age. For many women with complex congenital heart disease, carrying a pregnancy carries a moderate to high risk for both the mother and her fetus. Many such women, however, do not have access to adult congenital heart disease tertiary centers with experienced reproductive programs. Therefore, it is important that all practitioners who will be managing these women have current information not only on preconception counseling and diagnostic evaluation to determine maternal and fetal risk but also on how to manage them once they are pregnant and when to refer them to a regional center with expertise in pregnancy management.



#### 48. Pregnancy outcomes in patients with pulmonary arterial hypertension associated with congenital heart disease.

**Author(s):** Ladouceur, Magalie; Benoit, Louise; Radojevic, Jelena; Basquin, Adeline; Dauphin, Claire; Hascoet, Sébastien; Mocerri, Pamela; Bredy, Charlene; Iserin, Laurence; Gouton, Marielle; Nizard, Jacky

**Source:** Heart (British Cardiac Society); Feb 2017; vol. 103 (no. 4); p. 287-292

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

Available in full text at [Heart](#) - from Highwire Press

**Abstract:** OBJECTIVE There is growing evidence that maternal mortality in pregnant women with pulmonary arterial hypertension associated with congenital heart disease (PAH-CHD) is lower than that in available data. In order to evaluate this hypothesis, we collected data of pregnancies in women with PAH-CHD. METHODS Women with PAH-CHD followed in seven French referral centres were retrospectively included from 1997 to 2015. All pregnancies were recorded. We collected data on maternal, obstetrical and neonatal outcomes. RESULTS 28 pregnancies in 20 women (26±6 years old) with PAH-CHD were managed during this period. There were 18 complete pregnancies (≥20 weeks' gestation (WG)), 8 abortions and 2 miscarriages. Six (33%, 95% CI (11.9 to 54.3)) patients experienced severe cardiac events. The concerned women had lower resting oxygen saturation (79.6±4.1% vs 89.3±3.8%, p<0.01). The most common cardiac complications during the complete pregnancies were heart failure (n=4) and severe hypoxaemia (n=5). Heart failure was overall severe, requiring inotropic treatment in three patients, mechanical circulatory support in one and led to one maternal death (mortality=5.0% 95% CI (0.1 to 24.9)). Obstetrical complications occurred in 25% of pregnancies. Small for gestational age was diagnosed in 39% (7/18) of fetuses. 12/18 (67%) pregnancies were delivered by caesarean section, of which 10 in emergency for obstetrical reason. Prematurity was frequent (78%), but no neonatal death occurred. CONCLUSIONS Outcome of pregnancy in women with PAH-CHD is better than previously reported, with only 5% maternal mortality in our cohort. However, because of the severity of heart failure and the high rate of neonatal complications, patients should still be advised against pregnancy.

#### 49. Use of 3D models of congenital heart disease as an education tool for cardiac nurses.

**Author(s):** Biglino, Giovanni; Capelli, Claudio; Koniordou, Despina; Robertshaw, Di; Leaver, Lindsay-Kay; Schievano, Silvia; Taylor, Andrew M; Wray, Jo

**Source:** Congenital heart disease; Jan 2017; vol. 12 (no. 1); p. 113-118

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

**Abstract:** BACKGROUND Nurse education and training are key to providing congenital heart disease (CHD) patients with consistent high standards of care as well as enabling career progression. One approach for improving educational experience is the use of 3D patient-specific models. OBJECTIVES To gather pilot data to assess the feasibility of using 3D models of CHD during a training course for cardiac nurses; to evaluate the potential of 3D models in this context, from the nurses' perspective; and to identify possible improvements to optimise their use for teaching. DESIGN A cross-sectional survey. SETTING A national training week for cardiac nurses. PARTICIPANTS One hundred cardiac nurses (of which 65 paediatric and 35 adult). METHODS Nurses were shown nine CHD models within the context of a specialized course, following a lecture on the process of making the models themselves, starting from medical imaging. Participants were asked about their general learning experience, if models were more/less informative than diagrams/drawings and lesion-specific/generic models, and their overall reaction to the models. Possible differences between adult and paediatric nurses were investigated. Written feedback was subjected to content analysis and quantitative data were analyzed using nonparametric statistics. RESULTS Generally models were well

liked and nurses considered them more informative than diagrams. Nurses found that 3D models helped in the appreciation of overall anatomy (86%), spatial orientation (70%), and anatomical complexity after treatment (66%). There was no statistically significant difference between adult and pediatric nurses' responses. Thematic analysis highlighted the need for further explanation, use of labels and use of colors to highlight the lesion of interest amongst improvements for optimizing 3D models for teaching/training purposes. **CONCLUSION** 3D patient-specific models are useful tools for training adult and pediatric cardiac nurses and are particularly helpful for understanding CHD anatomy after repair.

#### **50. Neurocognitive and executive functioning in adult survivors of congenital heart disease.**

**Author(s):** Klouda, Leda; Franklin, Wayne J; Saraf, Anita; Parekh, Dhaval R; Schwartz, David D

**Source:** Congenital heart disease; Jan 2017; vol. 12 (no. 1); p. 91-98

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

**Abstract:** **OBJECTIVE** Congenital heart disease (CHD) can affect the developing central nervous system, resulting in neurocognitive and behavioral deficits. Preoperative neurological abnormalities as well as sequelae of the open heart operations required to correct structural abnormalities of the heart contribute to these deficits. There are few studies examining the neurocognitive functioning of adults with CHD. This study sought to investigate multiple domains of neurocognitive functioning in adult survivors of CHD who had childhood cardiac surgery with either moderate or severe disease complexity. **DESIGN** A total of 48 adults (18-49 years of age) who had undergone cardiac surgery for CHD prior to five years of age participated in the study. CHD severity was classified as moderate or severe according to the 32nd Bethesda Guidelines. A computerized battery of standardized neurocognitive tests (CNS-Vital Signs), a validated rating scale of executive functioning, and demographic questionnaires were administered. **RESULTS** There were no significant differences between the moderate CHD group and normative data on any cognitive measure. In contrast, the severe CHD group differed from norms in multiple domains: psychomotor speed, processing speed, complex attention, reaction time, and on the overall neurocognitive index. Number of surgeries was strongly related to worse executive functioning. There was no association between age at first surgery or time since last surgery and neuropsychological functioning. Number of surgeries was also unrelated to neurocognitive test performance. **CONCLUSIONS** Patients with severe CHD performed significantly worse on measures of processing speed, attention, and executive functioning. These findings may be useful in the long-term care of adults with congenital heart disease.

#### **51. Health-related quality of life of young people with long-term illnesses before and after transfer from child to adult healthcare.**

**Author(s):** While, A E; Heery, E; Sheehan, A M; Coyne, I

**Source:** Child: care, health and development; Jan 2017; vol. 43 (no. 1); p. 144-151

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

**Abstract:** **BACKGROUND** The numbers of children with long-term illnesses surviving into adulthood and transferring from child to adult services has increased dramatically in the last 30 years. This study aimed to examine health-related quality of life pre- and post-transfer from child to adult healthcare for young people with three long-term illnesses. **METHODS** A total of 217 young people with cystic fibrosis, congenital heart defects or diabetes attending child and adult hospital services in Dublin, Ireland completed a questionnaire survey. Multiple linear regression was used to identify predictors of five dimensions of health-related quality of life pre- and post-transfer. **RESULTS** Post-transfer young people with congenital heart disease and diabetes reported significantly lower

physical well-being than their pre-transfer counterparts. Pre-transfer young people with cystic fibrosis reported significantly lower physical well-being than those with diabetes, but there was no significant difference post-transfer. Pre-transfer females reported lower scores than males on the Psychological Well-being and Autonomy and Parent Relation dimensions; however, these differences disappeared post-transfer. Higher maternal overprotection scores were associated with significantly lower scores on the Psychological Well-being, Autonomy and Parent Relation, and Social Support and Peers dimensions, regardless of transfer status. **CONCLUSIONS** Disease group, gender and maternal overprotection were predictors of health-related quality of life pre- and post-transfer from child to adult healthcare. Transition programmes should promote self-management and discourage parental overprotection.

## **52. Multiple pregnancy in a primigravida with uncorrected Pentalogy of Fallot.**

**Author(s):** Partana, Pamela; Tan, Jarrod Kah Hwee; Tan, Ju Le; Tan, Lay Kok

**Source:** BMJ case reports; Jan 2017; vol. 2017

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

**Abstract:** Pentalogy of Fallot is a cyanotic congenital heart disease that has guarded prognosis without surgical intervention in infancy. Women with uncorrected defects rarely survive into childbearing age and pregnancy in this group is associated with a high rate of perinatal loss. Physiological cardiovascular changes in pregnancy can lead to maternal haemodynamic instability with subsequent adverse cardiac sequelae with or without fetal decompensation. Optimum management and pregnancy outcomes in mother with uncorrected Pentalogy of Fallot and twin pregnancy have not been described in the literature. We describe a successful case of monochorionic diamniotic twin pregnancy in an affected woman who has not undergone surgical repair. Her pregnancy progressed without any adverse cardiopulmonary complications. Her caesarean delivery and postpartum recovery were favourable, with successful birth of two healthy babies at 35.7 weeks. This case emphasises the importance of a multidisciplinary team, especially of obstetricians with expertise in high-risk pregnancies, adult congenital heart disease cardiologists and anaesthesiologist.

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## Exercise: Creating a Search Strategy

**Scenario:** A 64 year old obese male who has tried many ways to lose weight presents with a newspaper article about 'fat-blazer' (chitosan). He asks for your advice.

### 1. What would your PICO format be?

Population/problem	
Intervention/indicator	
Comparator	
Outcome	

### 2. What would your research question be?

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**PICO:** P = obese patients; I = chitosan; C = placebo; O = decrease weight  
**Research question:** In obese patients, does chitosan, compared to a placebo, decrease weight?



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