Background: The possibility that congenital heart defects signal a familial predisposition to cardiovascular disease has not been investigated. We aimed to determine whether the risk of cardiovascular disorders later in life was higher for women who have had newborns with congenital heart defects.

Methods: We studied a cohort of 1,084,251 women who had delivered infants between 1989 and 2013 in Quebec, Canada. We identified women whose infants had critical, noncritical, or no heart defects, and tracked the women over time for future hospitalizations for cardiovascular disease, with follow-up extending up to 25 years past pregnancy. We calculated the incidence of cardiovascular hospitalization per 1000 person-years, and used Cox proportional hazards regression to estimate hazard ratios and 95% confidence intervals (CIs) for the association between infant heart defects and risk of maternal cardiovascular hospitalization. Models were adjusted for age, parity, preeclampsia, comorbidity, material deprivation, and time period.

Results: Women whose infants had heart defects had a higher overall incidence of cardiovascular hospitalization. There were 3.38 cardiovascular hospitalizations per 1000 person-years for those with critical defects (95% CI, 2.67–4.27), 3.19 for noncritical defects (95% CI, 2.96–3.45), and 2.42 for no heart defects (95% CI, 2.39–2.44). In comparison with no heart defects, women whose infants had critical defects had a hazard ratio of 1.43 (95% CI, 1.13–1.82) for any cardiovascular hospitalization, and women whose infants had noncritical defects had a hazard ratio of 1.24 (95% CI, 1.15–1.34), in adjusted models. Risks of specific causes of cardiovascular hospitalization, including myocardial infarction, heart failure, and other atherosclerotic disorders, were also greater for mothers of infants with congenital heart defects than with no defects.

Conclusions: Women whose infants have congenital heart defects have a greater risk of cardiovascular hospitalization later in life. Congenital heart defects in offspring may be an early marker of predisposition to cardiovascular disease.

2) Pregnancy Outcomes in Women With Heart Disease

The CARPREG II Study Candice K. Silversides, MD, MS, Jasmine Grewal, MD, Jennifer Mason, RN, Mathew Sermer, MD, Marla Kiess, MD, Valerie Rychel, MD, Rachel M. Wald, MD, Jack M. Colman, MD, Samuel C. Siu, MD, SM,

BACKGROUND: Identifying women at high risk is an important aspect of care for women with heart disease.

OBJECTIVES: This study sought to: 1) examine cardiac complications during pregnancy and their temporal trends; and 2) derive a risk stratification index.

METHODS: We prospectively enrolled consecutive pregnant women with heart disease and determined their cardiac outcomes during pregnancy. Temporal trends in complications were
examined. A multivariate analysis was performed to identify predictors of cardiac complications and these were incorporated into a new risk index.

**RESULTS:** In total, 1,938 pregnancies were included. Cardiac complications occurred in 16% of pregnancies and were primarily related to arrhythmias and heart failure. Although the overall rates of cardiac complications during pregnancy did not change over the years, the frequency of pulmonary edema decreased (8% from 1994 to 2001 vs. 4% from 2001 to 2014; p value \( \approx 0.012 \)). Ten predictors of maternal cardiac complications were identified: 5 general predictors (prior cardiac events or arrhythmias, poor functional class or cyanosis, high-risk valve disease/left ventricular outflow tract obstruction, systemic ventricular dysfunction, no prior cardiac interventions); 4 lesion-specific predictors (mechanical valves, high-risk aortopathies, pulmonary hypertension, coronary artery disease); and 1 delivery of care predictor (late pregnancy assessment). These 10 predictors were incorporated into a new risk index (CARPREG II [Cardiac Disease in Pregnancy Study]).

**CONCLUSIONS:** Pregnancy in women with heart disease continues to be associated with significant morbidity, although mortality is rare. Prediction of maternal cardiac complications in women with heart disease is enhanced by integration of general, lesion-specific, and delivery of care variable

3)  **Evaluation and Management of Right-Sided Heart Failure: A Scientific Statement From the American Heart Association**


**Background and Purpose:** The diverse causes of right-sided heart failure (RHF) include, among others, primary cardiomyopathies with right ventricular (RV) involvement, RV ischemia and infarction, volume loading caused by cardiac lesions associated with congenital heart disease and valvular pathologies, and pressure loading resulting from pulmonic stenosis or pulmonary hypertension from a variety of causes, including left-sided heart disease. Progressive RV dysfunction in these disease states is associated with increased morbidity and mortality. The purpose of this scientific statement is to provide guidance on the assessment and management of RHF.

**Methods:** The writing group used systematic literature reviews, published translational and clinical studies, clinical practice guidelines, and expert opinion/statements to summarize existing evidence and to identify areas of inadequacy requiring future research. The panel reviewed the most relevant adult medical literature excluding routine laboratory tests using MEDLINE, EMBASE, and Web of Science through September 2017. The document is organized and classified according to the American Heart Association to provide specific suggestions, considerations, or reference to contemporary clinical practice recommendations.

**Results:** Chronic RHF is associated with decreased exercise tolerance, poor functional capacity, decreased cardiac output and progressive end-organ damage (caused by a combination of end-organ venous congestion and underperfusion), and cachexia resulting from poor absorption of nutrients, as well as a systemic proinflammatory state. It is the principal cause of death in patients with pulmonary arterial hypertension. Similarly, acute RHF is associated with hemodynamic instability and is the primary cause of death in patients presenting with massive pulmonary embolism, RV myocardial infarction, and postcardiotomy shock associated with cardiac surgery. Functional assessment of the right side of the heart can be hindered by its complex geometry. Multiple hemodynamic and
biochemical markers are associated with worsening RHF and can serve to guide clinical assessment and therapeutic decision making. Pharmacological and mechanical interventions targeting isolated acute and chronic RHF have not been well investigated. Specific therapies promoting stabilization and recovery of RV function are lacking.

Conclusions: RHF is a complex syndrome including diverse causes, pathways, and pathological processes. In this scientific statement, we review the causes and epidemiology of RV dysfunction and the pathophysiology of acute and chronic RHF and provide guidance for the management of the associated conditions leading to and caused by RHF.


BACKGROUND: The Fontan operation has provided life-saving palliation and adult survival for individuals born with single ventricle physiology. Many now seek advice about safe pregnancy. Little data are, however, available, consisting mainly of anecdotal experience and small series. This article seeks to review the published literature and identify lessons learnt from this collective experience.

METHODS AND RESULTS: We conducted a systematic review to evaluate maternal and fetal outcomes of pregnancy in women with a Fontan circulation. Among 1150 studies that were screened, 6 studies had sufficient longitudinal data points to qualify for meaningful inclusion, yielding 255 pregnancies in 133 women after Fontan procedure resulting in 115 live births (45%; including reports from 1986 to 2015). There was a total of 137 pregnancy losses (69%), with 115 miscarriages (45%), 19 elective terminations of pregnancy (7%), 2 stillbirths (1%), and 1 ectopic pregnancy (1%). The most common cardiovascular adverse events were supraventricular arrhythmia affecting 8.4% (range, 3%-37%) and heart failure affecting 3.9% (range, 3%-11%) of pregnancies. These complications were successfully managed with conventional approaches. No maternal deaths were reported. Postpartum hemorrhage was the predominant obstetric complication affecting 14% of the patients. Most patients were on antiplatelet agents (27%) or anticoagulants (50%) whereas only a minority (11%) were on neither. Among the 115 live births, 68 were premature (59%), 17 were small for gestational age (20%), and neonatal death occurred in 6 patients (5%).

CONCLUSIONS: The most commonly reported cardiovascular complications in patients with Fontan physiology-associated pregnancy were arrhythmia and heart failure. Miscarriages were highly prevalent as was prematurity and intrauterine growth restriction. Postpartum hemorrhage seems to be the most common obstetric complication. Large-scale data sets are needed to confirm these early observations and address the late sequelae of pregnancy in women with a Fontan circulation.


BACKGROUND: Current guidelines on oral anticoagulation (OAC) in adults with congenital heart disease (ACHD) and atrial arrhythmias (AA) consist of heterogeneous and divergent recommendations with limited level of evidence, possibly leading to diverse OAC management and different outcomes.
Therefore, we aimed to evaluate real-world implementation and outcome of three guidelines on OAC management in ACHD patients with AA.

**METHODS:** The ESC GUCH 2010, PACES/HRS 2014 and ESC atrial fibrillation (AF) 2016 guidelines were assessed for implementation. ACHD patients with recurrent or sustained non-valvular AA from 5 tertiary centers were identified using a national ACHD registry. After two years of prospective follow-up, thromboembolism, major bleeding and death were assessed.

**RESULTS:** In total, 225 adults (mean age 54±15 years, 55% male) with various defects (simple 43%; moderate 37%; complex 20%) and AA were included. Following the most strict indication (OAC is recommended in all three guidelines), one should treat a mere 37% of ACHD patients with AA, whereas following the least strict indication (OAC is recommended in any one of the three guidelines), one should treat 98% of patients. The various guidelines were implemented in 54–80% of patients.

From all recommendations, Fontan circulation, CHA2DS2-VASc≥1 and AF were independently associated with OAC prescription. Superiority of any guideline in identifying outcome (n=15) could not be demonstrated.

**CONCLUSIONS:** The implementation of current guidelines on OAC management in ACHD patients with AA is low, probably due to substantial heterogeneity among guidelines. OAC prescription in daily practice was most consistent in patients with AF and CHA2DS2-VASc≥1 or Fontan circulation.

6) Scimitar Syndrome in Children and Adults: Natural History, Outcomes, and Risk Analysis.

**Wang H, Kalfa D, Rosenbaum MS, Ginns JN, Lewis MJ, Glickstein JS, Bacha EA, Chai PJ.**


**BACKGROUND:** Scimitar syndrome involves congenital anomalous pulmonary venous return to the inferior vena cava. Optimal management remains controversial. We describe the natural history of disease, nonsurgical and surgical outcomes, and risk factors for poor outcomes at our institution.

**METHODS:** Patients with anomalous pulmonary venous return to the inferior vena cava documented on echocardiography at our institution between January 1994 and January 2015 were reviewed retrospectively. The study protocol IRB-AAAO1805 was approved.

**RESULTS:** Forty-seven patients were identified, including 20 infants with significant associated congenital heart defects (42.6%, including 7 with single ventricle physiology), and 10 infants (21.3%) and 16 noninfants (34.0%) with isolated scimitar syndrome. Median follow-up was 3.55 years. Noninfants exhibited lower incidences of right pulmonary artery hypoplasia (p < 0.001), aortopulmonary collaterals (p = 0.004), and scimitar vein obstruction at the caval confluence (p = 0.032). Eighteen patients (38.3%) underwent surgical repair for scimitar syndrome. Overall mortality after baffle repair or scimitar vein reimplantation was 37.5% (3 of 8) for infants and 0% (0 of 6) for noninfants (p = 0.209). Overall mortality for medically managed infants was 46.7% (7 of 15) compared with 0% (0 of 8) for noninfants (p = 0.052). Multivariable analyses identified infantile onset as an independent risk factor for stenosis or obstruction after repair (hazard ratio 9.34, p = 0.048), and single ventricle physiology as an independent risk factor for mortality among unrepaired patients (hazard ratio 29.8, p = 0.004).

**CONCLUSIONS:** The severity of scimitar syndrome depends on presenting age and associated congenital heart disease. Nonsurgical and surgical outcomes are suboptimal for infantile disease,
which is a risk factor for stenosis after repair. Single ventricle physiology is associated with poor prognosis.