

Contents

Preface: Adult Congenital Heart Disease: A Population that Has Come of Age xiii

Curt J. Daniels

Adult Congenital Heart Disease—Preparing for the Changing Work Force Demand 283

Michelle Gurvitz, George K. Lui, and Ariane Marelli

This volume is dedicated to advances in the care of adults with congenital heart disease (CHD). In this chapter the authors review the data cornerstone to the growing workforce needs. This first chapter serves as a backdrop to the second chapter that applies these observations to the planning of health care services delivery in the United States accounting for the definition and organization of multisystem expertise and centers for adults with CHD at a health systems level.

Access and Delivery of Adult Congenital Heart Disease Care in the United States: Quality-Driven Team-Based Care 295

Susan M. Fernandes, Ariane Marelli, Danielle M. Hile, and Curt J. Daniels

The landscape of congenital heart disease has changed rapidly over the past few decades. The shift from pediatric to adult congenital heart disease care has stretched resources and the ability to provide high-quality access and delivery of care for the more than 1.5 million adults with congenital heart disease in the United States. Meeting the demand for delivering high-quality care requires a team-based approach, with each member highly specialized. This review describes the deficits and deficiencies in providing care for adults with congenital heart disease in the United States and a team-based approach to improving access and delivery of care.


Psychological Needs, Assessment, and Treatment in the Care of Adults with Congenital Heart Disease 305

Jamie L. Jackson, Kristen R. Fox, and Adrienne H. Kovacs

Although the majority of congenital heart disease survivors are thriving, many are at risk for declining emotional well-being as they age. Emotional distress is a risk factor for poorer health outcomes and must be addressed. Primary care and cardiology teams may be the first line of defense in identifying and providing referral resources for symptoms of depression, anxiety, and medical trauma. The current review provides information about commonly used self-report measures of emotional distress to identify symptoms that warrant referral and describes multiple options for addressing these symptoms.

Atrial Septal Defect 317

Elisa A. Bradley and Ali N. Zaidi

 Video content accompanies this article at <http://www.cardiology.theclinics.com>.

Atrial septal defects are common congenital heart defects, characterized by insufficient/absent tissue at the interatrial septum. An unrepaired defect may be associated with right heart volume overload, atrial arrhythmia or pulmonary arterial hypertension.

The 3 major types of atrial septal defect are: ostium secundum defect, ostium primum defect, and sinus venosus. Characteristic physical findings include a midsystolic pulmonary flow or ejection murmur, accompanied by a fixed split-second heart sound. Small defects may spontaneously close; larger defects may persist and result in hemodynamic and clinical sequelae requiring percutaneous or surgical intervention. Severe pulmonary arterial hypertension is a contraindication to closure.

Aortopathy in Congenital Heart Disease

325

Timothy B. Cotts, Katherine B. Saliccioli, Sara K. Swanson, and Anji T. Yetman

Aortic dilatation is common in patients with congenital heart disease and is seen in patients with bicuspid aortic valve and those with conotruncal congenital heart defects. It is important to identify patients with bicuspid aortic valve at high risk for aortic dissection. High-risk patients include those with the aortic root phenotype and those with syndromic or familial aortopathies including Marfan syndrome, Loeys-Dietz syndrome, and Turner syndrome. Aortic dilatation is common in patients with conotruncal congenital heart defects and rarely results in aortic dissection.

Aortic Coarctation

337

Yuli Y. Kim, Lauren Andrade, and Stephen C. Cook

Aortic coarctation is a discrete narrowing of the thoracic aorta. In addition to anatomic obstruction, it can be considered an aortopathy with abnormal vascular properties characterized by stiffness and impaired relaxation. There are surgical and transcatheter techniques to address the obstruction but, despite relief, patients with aortic coarctation are at risk for hypertension, aortic complications, and abnormalities with left ventricular performance. This review covers the etiology, pathophysiology, diagnosis, and management of adults with aortic coarctation, with emphasis on multimodality imaging characteristics and lifelong surveillance to identify long-term complications.

Ebstein Anomaly in the Adult Patient

353

Margaret M. Fuchs and Heidi M. Connolly

Ebstein anomaly is a congenital malformation involving primarily the tricuspid valve, with failure of delamination from the underlying myocardium and right ventricular myopathy. Echocardiography is diagnostic in most patients and demonstrates apical displacement of the septal leaflet and variable tethering of leaflet tissue to the right ventricular myocardium. Operative intervention is considered for exertional symptoms, progressive right ventricular enlargement, or right ventricular dysfunction. Tricuspid valve cone repair is the preferred surgical approach. Tricuspid valve replacement and bidirectional cavopulmonary shunt also are considered in patients with advanced disease. Pregnancy generally is well tolerated. Patients with Ebstein anomaly require lifelong follow-up.

Tetralogy of Fallot

365

Eric V. Krieger and Anne Marie Valente



Video content accompanies this article at <http://www.cardiology.theclinics.com>.

Repaired tetralogy of Fallot is one of the most common conditions managed by adult congenital heart disease providers. Recent comprehensive review articles and book

chapters are devoted to this topic. The purpose of this article is to address several common clinical questions encountered in the management of patients with repaired tetralogy of Fallot. These answers are not intended to supplant Practice Guidelines.

The Adult Patient with a Fontan

379

Ahmed AlZahrani, Rahul Rathod, Ahmed Krimly, Yezan Salam, AlJuhara Thaar AlMarzoog, and Gruschen R. Veldtman

The authors summarize the most important anatomic and physiologic substrates of Fontan circulation. Common anatomic substrates include hypoplastic left heart syndrome, tricuspid atresia, double inlet left ventricle, and unbalanced atrioventricular septal defects. After the Fontan operation exercise capacity is limited and the key hemodynamic drivers is limited preload due to a relatively fixed pulmonary vascular resistance. The authors provide contemporary data on survival, morbidity, and need for reintervention. Operative morality is now expected to be less than 1% and 30 year survival approximately 89%. The authors delineate potential therapeutic approaches for the potential late complications.

Transcatheter Interventions in Adult Congenital Heart Disease

403

Jamil A. Aboulhosn and Ziyad M. Hijazi

This article provides a detailed review of the current practices and future directions of transcatheter interventions in adults with congenital heart disease. This includes indications for intervention, risks, and potential complications, as well as a review of available devices and their performance.

Adults with Congenital Heart Disease and Arrhythmia Management

417

Jeremy P. Moore and Paul Khairy



Video content accompanies this article at <http://www.cardiology.theclinics.com>.

Arrhythmia management in adult congenital heart disease (ACHD) encompasses a wide range of problems from bradyarrhythmia to tachyarrhythmia, sudden death, and heart failure-related electrical dyssynchrony. Major advances in the understanding of the pathophysiology and treatments of these problems over the past decade have resulted in improved therapeutic strategies and outcomes. This article attempts to define these problems and review contemporary management for the patient with ACHD presenting with cardiac arrhythmia.

Surgery for Adult Congenital Heart Disease

435

Tracy Geoffrion and Stephanie Fuller

Technical and medical improvements for congenital cardiac disease in children have contributed to an increasing population of patients who survive into adulthood. These patients may be prone to progression of their native palliated disease or suffer from sequelae of their childhood repair that requires repeat surgical intervention. Surgery for adult congenital cardiac disease poses unique challenges and risks.

Congenital Heart Disease and Pulmonary Hypertension**445**

Andrew Constantine, Konstantinos Dimopoulos, and Alexander R. Opatowsky

Pulmonary hypertension (PH) is common in adults with congenital heart disease and carries fundamental implications for management and prognosis. A high index of suspicion, combined with knowledge of the pathogenesis and pathophysiology of PH, is required to achieve a timely, accurate diagnosis, and appropriate classification and treatment. This article provides a guide on how to approach the adult with congenital heart disease and suspected PH of different types, including current management.

Heart Failure in Adult Congenital Heart Disease**457**

Luke J. Burchill, Melissa G.Y. Lee, Vidang P. Nguyen, and Karen K. Stout

As the population of adult congenital heart disease patients ages and grows, so too does the burden of heart failure in this population. Despite the advances in medical and surgical therapies over the last decades, heart failure in adult congenital heart disease remains a formidable complication with high morbidity and mortality. This review focuses on the challenges in determining the true burden and management of heart failure in adult congenital heart disease. There is a particular focus on the need for developing a common language for classifying and reporting heart failure in adult congenital heart disease, the clinical presentation and prognostication of heart failure in adult congenital heart disease, the application of hemodynamic evaluation, and advanced heart failure treatment. A common case study of heart failure in adult congenital heart disease is utilized to illustrate these key concepts.