Canadian Cardiovascular Society 2009 Consensus Conference on the management of adults with congenital heart disease: Executive summary

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With advances in pediatric cardiology and cardiac surgery, the population of adults with congenital heart disease (CHD) has increased. In the current era, there are more adults with CHD than children. This population has many unique issues and needs. They have distinctive forms of heart failure, and their cardiac disease can be associated with pulmonary hypertension, thromboemboli, complex arrhythmias and sudden death. Medical aspects that need to be considered relate to the long-term and multisystemic effects of single-ventricle physiology, cyanosis, systemic right ventricles, complex intracardiac baffles and failing subpulmonary right ventricles. Since the 2001 Canadian Cardiovascular Society Consensus Conference report on the management of adults with CHD, there have been significant advances in the understanding of the late outcomes, genetics, medical therapy and interventional approaches in the field of adult CHD. Therefore, new clinical guidelines have been written by Canadian adult CHD physicians in collaboration with an international panel of experts in the field. The present executive summary is a brief overview of the new guidelines and includes the recommendations for interventions. The complete document consists of four manuscripts that are published online in the present issue of *The Canadian Journal of Cardiology*, including sections on genetics, clinical outcomes, recommended diagnostic workup, surgical and interventional options, treatment of arrhythmias, assessment of pregnancy and contraception risks, and follow-up requirements. The complete document and references can also be found at www.ccs.ca or www.cachnet.org.

Key Words: *Adult congenital heart disease; Congenital heart disease; Consensus; Guidelines; Surgery*

La conférence consensuelle 2009 de la Société canadienne de cardiologie sur la prise en charge des adultes ayant une cardiopathie congénitale : Résumé

Étant donné les progrès de la cardiologie pédiatrique et de la chirurgie cardiaque, la population d'adultes ayant une cardiopathie congénitale (CPC) a augmenté. Il y a maintenant plus d'adultes que d'enfants ayant une CPC. Cette population a de nombreux problèmes et besoins uniques. Ils ont des formes particulières d'insuffisance cardiaque, et leur maladie cardiaque peut s'associer à une hypertension pulmonaire, à des thromboembolies, à des arythmies complexes et à une mort subite. Les aspects médicaux à envisager sont liés aux effets multisystémiques et à long terme de la physiologie monoventriculaire, de la cyanose, des ventricules droits systémiques, des cloisons intracardiaques complexes et de l'insuffisance du ventricule droit sous-pulmonaire. Depuis le rapport de la conférence consensuelle 2001 de la Société canadienne de cardiologie sur la prise en charge des adultes ayant une CPC, on constate d'importantes avancées dans la compréhension des issues tardives, de la génétique, de la thérapie médicale et des démarches d'intervention dans le domaine des CPC chez les adultes. Par conséquent, de nouvelles lignes directrices cliniques ont été rédigées par des médecins canadiens s'occupant des CPC chez les adultes, en collaboration avec un groupe d'experts internationaux dans le domaine. Le présent résumé donne un bref aperçu des nouvelles lignes directrices et contient les recommandations d'interventions. Le document complet se compose de quatre manuscrits publiés par voie électronique dans le présent numéro du *Journal canadien de cardiologie*, y compris des rubriques sur la génétique, les issues cliniques, les bilans diagnostiques recommandés, les possibilités chirurgicales et d'intervention, le traitement des arythmies, l'évaluation des risques de la grossesse et de la contraception et les recommandations de suivi. Le document complet et les références figurent également aux adresses www.ccs.ca et www.cachnet.org.

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The Canadian Cardiovascular Society published the original consensus conference report on the management of adult congenital heart disease (ACHD) patients in 1998 (1). This publication was followed by an update in 2001 (2). Due to advances in the field of adult congenital cardiology, including new information related to late outcomes, genetics, medical therapy and interventional techniques, the 2001 guidelines have now been updated. These recommendations were written by Canadian ACHD physicians in collaboration with an international panel of experts in the field. The format of the current update is similar to that used in the 2001 consensus statement and is divided into three parts, each of which provides recommendations for a number of congenital cardiac lesions. Table 1 outlines the classification definitions used for grading evidence. The present executive summary is an abbreviated version of the consensus conference, focusing on the recommendations pertaining to medical therapy and interventions. The complete document consists of four manuscripts, which are published online in the present issue of *The Canadian Journal of Cardiology* (3-6), and includes more detailed recommendations including sections on genetics, clinical outcomes, suggested diagnostic workup, surgical and interventional options and outcomes, treatment of arrhythmias, assessment of pregnancy and follow-up requirements. The complete document and references can be found at www.ccs.ca or www.cachnet.org.

EPIDEMIOLOGY AND SCOPE OF THE PROBLEM

Anomalies of the heart and circulation constitute one of the most common forms of congenital birth defects (7). Advances in pediatric cardiology and cardiac surgery have resulted in an increasing number of ACHD patients and a change in the epidemiology of congenital heart disease (CHD) (8-10). Although the overall prevalence of CHD has increased over time, population trends indicate proportionally different changes in children and adults. The prevalence of severe CHD increased by 85% in adults compared with 22% in children, consistent with the notion that the greatest survival benefit has occurred in those with more severe forms of CHD (10). Over the past two decades, the overall CHD population has aged, most notably in those with severe forms of CHD, where the mean age increased from 11 years of age in 1985 to 17 years of age in 2000. In 2000, the median age of the entire ACHD population was 40 years and was 29 years in the subset of adults with severe CHD (10).

Accurate determination of the numbers of adults with CHD, whether estimated or measured, is difficult (8,10). In a Quebec population-based study, the prevalence of CHD in the year 2000 was four per 1000 adults and 12 per 1000 children. Extrapolated to a Canadian population of 24 million adults, 96,000 adult patients in Canada were expected to have CHD in 2000. In the United States and Canada, there is one child for every three adults in the population. Therefore, although prevalence rates of CHD in children are higher than those in adults, the overall number of adults exceeds the number of children with CHD, and the number of adults and children with severe CHD was nearly equal by the year 2000 (10).

THE UNIQUE NEEDS OF ACHD PATIENTS

Unique issues specific to adults with CHD include long-term and multisystemic effects of single-ventricle physiology, cyanosis, systemic right ventricles, complex intracardiac baffles and failing subpulmonary right ventricles. Genetic counselling, birth control and high-risk pregnancy management have become integral components of care. Acquired comorbidities, such as diabetes, hypertension and coronary artery disease, may further impact the congenital substrate and potential for long-term adverse events. Complications include distinctive forms of heart failure, pulmonary hypertension, thromboemboli, complex arrhythmias and sudden death. Longer-term survival and quality-of-life issues, such as autonomy, employment, education, functional capacity and physical activities, have assumed increasing importance. To advance the care of adults with CHD, evidence-based approaches are increasingly sought.

THE CANADIAN ADULT CONGENITAL HEART NETWORK

Care for ACHD patients should be integrated from the primary care level to highly specialized subspecialty care in ACHD regional centres (2,11,12). Adult patients with CHD of great complexity should be followed in regional ACHD centres (8,12). Analysis of surgical trends in ACHD patients from 1990 to 2000 (13) revealed that the fastest growing segment of patients requiring interventions were those with disease of moderate complexity. The majority of new ACHD patients should be seen at least once by an ACHD specialist to determine the most appropriate venue of care. The Canadian Adult Congenital Heart Network, founded in 1991 by health care professionals, lists 15 self-identified ACHD care facilities of all kinds with varying size and services offered, a subset of which are regional ACHD centres (8,11,12).

ANTIBIOTIC PROPHYLAXIS

Infective endocarditis is a well-recognized complication of CHD (14). Although data on infective endocarditis in ACHD are limited, recent multi-institutional surveys suggest that morbidity and mortality rates remain elevated in this population (15,16). Guidelines from the American Heart Association (AHA) (17) have further defined the role of antibiotic prophylaxis in the prevention of infective endocarditis. Current recommendations reflect, in part, an increased emphasis on evidence, which has translated into a more restrictive use of antibiotic prophylaxis. The AHA guidelines also emphasize the notion that infective endocarditis in patients with certain high-risk cardiac conditions is associated with particularly poor clinical outcomes. Patients with these high-risk conditions are the ones who should receive antibiotic prophylaxis (17,18). The list of high-risk cardiac conditions is relevant to the ACHD population, and includes prosthetic cardiac valve or prosthetic material used for cardiac valve repair; previous infective endocarditis; CHD (specifically, unrepaired cyanotic CHD, including palliative shunts and conduits; completely repaired CHD with prosthetic material or device, whether placed during surgery or by catheter intervention, during the first six months after the procedure; and repaired CHD with residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic device); and cardiac transplantation recipients who develop cardiac valvulopathy. Finally, the AHA guidelines describe a more narrow list of procedures for which, in high-risk individuals, antibiotic prophylaxis is indicated (17). The members of this panel endorse the AHA antibiotic prophylaxis recommendations and their implementation to the ACHD population.

GENETIC EVALUATION

The genetic contribution to CHD has been significantly underestimated in the past. Clinically available genetic testing has increased over the years, as has the availability of newer technology that provides higher resolution to detect subtle genetic aberrations (deletions, duplications and mutations) causing disease. For the clinician caring for a patient with CHD, identifying a genetic etiology is important for several reasons: identification of a syndromic phenotype would help guide investigations for other potential medical problems involving other organ systems; risk stratification, because some syndromes are associated with poor prognosis; genetic and reproductive counselling for recurrence risk in future pregnancies; and screening family members to identify individuals at risk for the cardiac lesion. The vast majority of adults with CHD have not had genetic testing or family screening. The clinician is advised to consult the GeneTests Web site <http://www.genetests.org> for updates on what testing is currently available, as well as the AHA Congenital Cardiac Defects Committee's report on the genetic basis for congenital heart defects (19).

RECOMMENDATIONS – INDICATIONS FOR MEDICAL THERAPY, INTERVENTION AND REINTERVENTION

Atrial septal defects

Class I

- Surgical or percutaneous closure of an atrial septal defect (ASD) is indicated in the presence of a hemodynamically significant ASD with or without resulting symptoms. (Level B)
- In patients with large secundum ASDs (greater than 38 mm) not amenable to device closure, surgical closure should be undertaken. (Level B)
- Percutaneous ASD closure should be performed by individuals with expertise in the technique and its clinical evaluation. (Level C)
- A sinus venosus defect or ostium primum ASD cannot be closed by percutaneous devices and should be surgically repaired by congenital heart surgeons. (Level C)
- If atrial fibrillation/flutter occurs, anticoagulation is usually indicated in accordance with existing guidelines. (Level A)
- Atrial arrhythmias can be managed with either rate or rhythm control strategies, and the approach should be tailored to the individual patient. (Level B)

Class IIa

- Closure of an ASD may be indicated in patients with orthodeoxia-platypnea. (Level C)
- Closure of an ASD may be indicated in patients with paradoxical emboli. (Level C)
- Surgical closure of an ASD should be considered if patients are undergoing tricuspid valve repair or replacement. (Level C)
- Catheter ablation should be considered before device closure, while the surgical maze procedure would be performed concomitant with ASD closure. (Level B)
- Transvenous pacing should be avoided in patients with unrepaired ASDs because paradoxical emboli may occur. (Level B)
- Closure can be considered if pulmonary arterial hypertension (PAH) is present and there is a net left-to-right shunt greater than 1.5:1; or evidence of pulmonary artery reactivity when challenged with a pulmonary vasodilator (eg, oxygen, nitric oxide and/or prostaglandins). Such patients should receive care from a specialist with expertise in PAH. (Level C)

Class III

- If PAH is present (pulmonary artery pressure [PAP] greater than two-thirds the systemic arterial blood pressure [SABP], or pulmonary arteriolar resistance greater than two-thirds the systemic arteriolar resistance) and irreversible, the ASD should not be closed. Such patients should receive care from a specialist with expertise in PAH. (Level C)

Ventricular septal defects

Class I

- The following situations warrant closure:
  - The presence of a 'significant' ventricular septal defect (VSD) (symptomatic; left ventricular [LV] volume overload; deteriorating ventricular function due to volume [left ventricle] or pressure [right ventricle] overload, pulmonary-to-systemic flow ratio [Qp:Qs] of at least 2:1; pulmonary artery systolic pressure greater than 50 mmHg). (Level B)
  - Significant right ventricular outflow tract obstruction (RVOTO) (catheterization gradient or mean echocardiographic [echo] gradient greater than 50 mmHg). (Level B)
  - A perimembranous or subarterial VSD with more than mild aortic incompetence. (Level B)
  - In the presence of severe pulmonary hypertension (PAP greater than two-thirds the SABP or pulmonary arteriolar resistance greater than two-thirds the systemic arteriolar resistance), there must be a net left-to-right shunt of at least 1.5:1, or evidence of pulmonary artery reactivity when challenged with a pulmonary vasodilator (eg, oxygen, nitric oxide and/or prostaglandins). (Level B)
- Patients with an isolated VSD with or without associated lesions (RVOTO, aortic valve prolapse, subaortic stenosis or infective endocarditis) should be repaired by congenital heart surgeons. (Level C)

Class IIa

- The following situations may warrant closure:
  - A history of endocarditis (especially recurrent). (Level B)
  - If transvenous pacing is required, closure may be reasonable to prevent paradoxical emboli. (Level B)
- Device closure may be performed in the following settings:
  - Isolated trabecular muscular VSDs, especially if the VSD is remote from the tricuspid valve and the aorta. (Level B)
  - Perimembranous VSD if the defect is far enough from the aortic valve, although the risk of complete heart block is greater with device closure than with surgical closure. (Level B)
- Patients with an isolated VSD with or without associated lesions (RVOTO, aortic valve prolapse, subaortic stenosis or infective endocarditis) should be repaired by congenital heart surgeons. (Level C)

Class III

- If PAH is present (PAP greater than two-thirds the SABP, or pulmonary arteriolar resistance greater than two-thirds the systemic arteriolar resistance) and irreversible, the VSD should not be closed. Such patients should receive care from a specialist with expertise in PAH. (Level C)

Atrioventricular septal defects

Class I

- The following situations warrant intervention or reintervention:
  - An unoperated atrioventricular septal defect (AVSD) with the following:
    - Presumed paradoxical embolism. (Level B)
    - LV dysfunction. (Level B)
    - Right ventricular (RV) volume overload. (Level B)
    - Clinical heart failure. (Level B)
    - Reversible pulmonary hypertension. (Level B)

TABLE 1
Recommendation definitions for grading of evidence

Class	Definition
I	Conditions for which there is evidence for and/or general agreement that the procedure or treatment is useful and effective
II	Conditions for which there is conflicting evidence and/or a divergence of opinion about the usefulness/efficacy of a procedure or treatment
IIa	The weight of evidence or opinion is in favour of the procedure or treatment
IIb	Usefulness/efficacy is less well-established by evidence or opinion
III	Conditions for which there is evidence and/or general agreement that the procedure or treatment is not useful/effective and in some cases may be harmful
Level	**Definition**
A	When the data were derived from multiple randomized clinical trials involving a large number of individuals
B	When the data were derived from a limited number of randomized trials, nonrandomized studies or observational registries
C	When the primary basis for the recommendation was expert consensus

- An operated AVSD with the following:
  - Persisting or new hemodynamically significant defects arising after the original repair. (Level B)
  - Left atrioventricular (AV) ('mitral') valve regurgitation (or stenosis from previous repair) causing symptoms. (Level B)
  - Deterioration in ventricular function. (Level B)
- Significant subaortic obstruction (catheterization gradient or mean echo gradient greater than 50 mmHg at rest or on provocative testing with isoproterenol). (Level B)
- Transvenous pacing should be avoided if there are residual interatrial or interventricular communications because paradoxical emboli may occur. (Level B)
- A primum ASD (partial AVSD) cannot be closed using a percutaneous device and should be repaired by congenital heart surgeons. (Level C)

Class III

- If PAH is present (PAP greater than two-thirds the SABP, or pulmonary arteriolar resistance greater than two-thirds the systemic arteriolar resistance) and irreversible, the AVSD should not be closed. Such patients should receive care from a specialist with expertise in PAH. (Level C)

Patent ductus arteriosus

Class I

- No intervention is indicated if a small silent patent ductus arteriosus (PDA) is detected. (Level C)
- Surgical closure should be reserved for those in whom the PDA is too large for device closure. (Level B)
- Operative repair should be undertaken by congenital heart surgeons. (Level C)

Class IIa

- The following situations warrant intervention:
  - The presence of a PDA (except the silent duct at one extreme and the presence of severe, irreversible pulmonary vascular disease at the other extreme). (Level B)
  - Closure of a small but audible PDA is usually recommended, although this indication remains controversial given the low perceived risk of endarteritis. (Level B)
  - The occurrence of an episode of endarteritis on a clinically silent PDA. (Level B)
  - If pulmonary hypertension is present (PAP greater than two-thirds the SABP or pulmonary arteriolar resistance greater than two-thirds the systemic arteriolar resistance), there must be a net left-to-right shunt of at least 1.5:1, or evidence of pulmonary artery reactivity when challenged with a pulmonary vasodilator (eg, oxygen, nitric oxide and/or prostaglandin). (Level B)

Class III

- If PAH is present (PAP greater than two-thirds the SABP, or pulmonary arteriolar resistance greater than two-thirds the systemic arteriolar resistance) and irreversible, the PDA should not be closed. Such patients should receive care from a specialist with expertise in PAH. (Level C)

LV outflow tract obstruction and bicuspid aortic valve disease

Supravalvular LV outflow tract obstruction:

Class I

- Operative intervention is recommended for patients with supravalvular LV outflow tract obstruction (LVOTO) with symptoms, and/or a mean echo or catheter gradient of greater than 50 mmHg, or a peak instantaneous echo gradient of greater than 70 mmHg if the obstruction is discrete. (Level C)
- Patients who require operation for supravalvar LVOTO should be operated on by congenital heart surgeons. (Level C)

Valvular LVOTO:

Class I

- Valvular LVOTO requires intervention for symptoms (dyspnea, angina, presyncope or syncope) and significant left-sided outflow obstruction (mean echo gradient of greater than 40 mmHg or aortic valve area of less than 1.0 cm^2 or less than 0.6 cm^2/m^2). Gradients may be lower if there is significant LV systolic dysfunction. (Level C)
- Patients with bicuspid aortic valves require intervention for symptoms and severe regurgitation, or severe aortic regurgitation with LV end-systolic dimensions of greater than 55 mm, an end-diastolic diameter of greater than 75 mm or an LV ejection fraction of less than 50%. (Level B)
- Aortic root replacement is required for ascending aortic dissection and should be considered prophylactically for proximal aortic dilation (greater than 50 mm) or progressive dilation of greater than 5 mm/year. (Level B)
- Pulmonary autograft (Ross procedure) and balloon valvuloplasty for valvar LVOTO should be performed in centres and by physicians with substantial experience in these procedures. (Level C)

Class I – Reinterventions for valvular LVOTO

- Reoperation is indicated after valvotomy or after surgery for the following:
  - Recurrent LVOTO (same criteria as above). (Level C)
  - Severe aortic regurgitation. (Level C)
  - Combined restenosis with moderate or greater regurgitation, especially if symptoms or progressive LV dilation are present. (Level C)

Class IIb

- Intervention may be considered for asymptomatic patients with 'critical' aortic stenosis (valve area of less than 0.6 cm^2) and/or a mean Doppler gradient of greater than 60 mmHg. (Level C)
- Intervention may be indicated occasionally for other reasons (eg, a person with a lesser degree of obstruction who wishes to play vigorous sports or to become pregnant). (Level C)

Subvalvular LVOTO:

Class I

- Intervention is indicated for patients with subvalvular LVOTO with symptoms and a peak instantaneous echo gradient of greater than 50 mmHg or a mean echo gradient of greater than 30 mmHg, or if combined with progressive aortic regurgitation. (Level C)
- Patients who require operation for subvalvar LVOTO should be operated on by congenital heart surgeons. (Level C)

Coarctation of the aorta

Class I

- All patients with significant coarctation (native or recoarctation after repair) should be considered candidates for treatment. (Level C)
- For significant native aortic coarctation, a surgical or percutaneous approach (if the anatomy is suitable) is reasonable. The preferred approach should reflect centre expertise and patient preference. (Level B)
- For significant recoarctation after repair, a percutaneous approach (if the anatomy is suitable) is the preferred initial intervention. (Level B)
- Surgeries and percutaneous interventions should be performed in centres and by surgeons with expertise in the procedure. (Level C)

RVOTO

Class I

- In symptomatic patients with valvular RVOTO, a domed pulmonary valve, and a peak instantaneous Doppler gradient

greater than 50 mmHg or a mean echo gradient greater than 30 mmHg, balloon valvotomy is recommended. (Level C)
- In asymptomatic patients with valvular RVOTO, a domed pulmonary valve, and a peak instantaneous Doppler gradient greater than 60 mmHg or a mean gradient greater than 40 mmHg, balloon valvotomy should be considered. (Level C)
- The surgical approach is recommended for patients with significant RVOTO and dysplastic pulmonary valves, subvalvular or supravalvular pulmonary stenosis, associated pulmonary hypoplasia or severe pulmonary regurgitation. (Level C)
- Balloon valvuloplasty is the treatment of choice for valvar RVOTO. Occasionally, valve replacement may be necessary. (Level B)
- Balloon valvuloplasty for valvar RVOTO should still be performed only in centres and by teams with experience in this technique. (Level C)
- Patients who require operation for RVOTO should be operated on by congenital heart surgeons. (Level C)

Class I – Reinterventions for RVOTO
- Recurrent RVOTO after previous surgery or balloon valvotomy (same criteria as above). (Level C)
- Severe pulmonic regurgitation associated with reduced exercise capacity of cardiovascular cause or deteriorating RV function or substantial tricuspid regurgitation or sustained atrial flutter/fibrillation or sustained ventricular tachycardia. (Level C)

Class IIa
- In patients with valvular RVOTO, intervention is also probably indicated by the presence of the following:
  - Important arrhythmias (usually sustained atrial flutter). (Level C)
  - An associated ASD or VSD, especially if there is right-to-left shunting. (Level C)
  - Recurrent endocarditis. (Level C)
- In patients with a double-chambered right ventricle with significant midcavity obstruction (pullback gradient at catheterization of greater than 50 mmHg), surgery should be considered. (Level C)

Tetralogy of Fallot

Class I
- In patients with sustained ventricular tachyarrhythmia and/or resuscitated from sudden cardiac death with no clear identified reversible cause, implantable cardioverter defibrillators (ICDs) are indicated for secondary prevention. (Level B)
- Patients who require surgery for tetralogy of Fallot should be operated on by congenital heart surgeons. (Level C)

Class IIa
- Following palliative surgery, complete intracardiac repair should be considered in all patients, in the absence of severe irreversible pulmonary hypertension or unfavourable anatomy (inadequate pulmonary arteries). In palliated patients, the following situations particularly warrant complete repair:
  - Worsening symptoms. (Level C)
  - Cyanosis with erythrocytosis. (Level C)
  - Reduction or absence of the continuous shunt murmur (suspected shunt stenosis or occlusion). (Level C)
  - Aneurysm formation in the shunt. (Level C)
  - LV dilation due to aortic regurgitation or a residual shunt. (Level C)

Class IIa – Reinterventions for tetralogy of Fallot
- The following situations may warrant intervention after repair:
  - Free pulmonary regurgitation associated with progressive or moderate to severe RV enlargement (RV end-diastolic volume greater than 170 mL/m^2), moderate to severe RV dysfunction, important tricuspid regurgitation, atrial or ventricular arrhythmias, or symptoms such as deteriorating exercise performance. (Level C)
  - Residual VSD with a shunt greater than 1.5:1. (Level C)
  - Residual pulmonary stenosis with RV pressure at least two-thirds the systemic pressure (either the native RV outflow or valved conduit if one is present). (Level C)
  - Significant aortic regurgitation associated with symptoms and/or progressive LV systolic dysfunction. (Level C)
  - Aortic root enlargement of at least 55 mm in diameter. (Level C)
  - A large RV outflow tract aneurysm, or evidence of infection or false aneurysm. (Level of C)
  - Sustained clinical arrhythmias, most commonly either atrial flutter or fibrillation, or sustained monomorphic ventricular tachycardia. When any of these arrhythmias occur, the patient should also be evaluated for a treatable hemodynamic cause of the arrhythmia. (Level C)
  - The combination of residual VSD, and/or residual pulmonary stenosis and regurgitation, all mild-moderate but leading to substantial RV enlargement, reduced RV function or symptoms. (Level C)
- Patients deemed to be at particularly high risk for sudden cardiac death may benefit from ICDs for primary prevention. (Level B)
- Patients who require reoperation for tetralogy of Fallot should be operated on by congenital heart surgeons. (Level B)

Ebstein anomaly

Class I
- The following situations warrant intervention:
  - Limited exercise capacity (New York Heart Association class greater than II). (Level B)
  - Increasing heart size (cardiothoracic ratio greater than 65%). (Level B)
  - Important cyanosis (resting oxygen saturations of less than 90%). (Level B)
  - Severe tricuspid regurgitation with symptoms. (Level B)
  - Transient ischemic attack or stroke. (Level B)
- Patients who require operation for Ebstein anomaly should be operated on by congenital heart surgeons who have substantial specific experience and success with this operation. Every effort should be made to preserve the native tricuspid valve. (Level C)

Marfan's syndrome

Class I
- The following situations warrant surgical intervention:
  - A maximal aortic root/ascending aorta diameter greater than 50 mm. (Level B)
  - A maximal aortic root/ascending aorta diameter greater than 45 mm to 50 mm with rapid aortic root growth greater than 5 mm per year; progressive aortic regurgitation, especially if the surgeon believes the aortic valve can be spared and an aortic valve-sparing procedure is planned; family history of premature aortic dissection of less than 50 mm; and severe mitral valve regurgitation that requires surgery. (Level B)
  - A maximal aortic root/ascending aorta diameter of greater than 44 mm if pregnancy is desired. (Level B)
  - A maximal dimension of other parts of the aorta of 50 mm to 60 mm or progressive dilation. (Level B)
  - Severe mitral regurgitation with symptoms or progressive LV dilation/dysfunction as per the current guidelines on valvular heart disease. (Level B)
- Patients who require an operation for Marfan's syndrome should be operated on by surgeons with substantial experience performing these types of surgeries. (Level C)

Class IIa

- All patients with Marfan's syndrome should be advised to take beta-blockers and to remain on them unless side effects preclude their use. This is especially true – usually in association with other blood pressure-lowering agents – if dissection has occurred. (Level B)

Complete transposition of the great arteries

Class I

- Pacemaker insertion for symptomatic bradycardia or antitachycardia pacing for some atrial arrhythmias may be required. Before transvenous lead implantation, the superior baffle must be evaluated for stenosis and/or baffle leaks with appropriate intervention undertaken. (Level B)
- Given the association between rapidly conducting atrial arrhythmias and sudden death, an aggressive management strategy that includes catheter ablation is often recommended. (Level C)
- In patients with sustained ventricular tachyarrhythmia and/or resuscitated from sudden cardiac death with no clear identified reversible cause, ICDs are indicated for secondary prevention. (Level B)
- Ablation and device implantation should be undertaken by an electrophysiologist with appropriate training/experience in the ACHD population. (Level C)
- Patients who require intervention or reintervention should be treated by ACHD cardiologists and congenital heart surgeons with appropriate experience. (Level C)

Class IIa

- The following situations may warrant reintervention following the atrial switch procedures:
  - Significant systemic (tricuspid) AV valve regurgitation without significant ventricular dysfunction. (Level C)
  - Superior or inferior vena cava pathway obstruction. (Level C)
  - Pulmonary venous pathway obstruction. (Level C)
  - Baffle leak resulting in a significant left-to-right shunt (Qp:Qs greater than 1.5:1), symptoms, pulmonary hypertension or progressive ventricular enlargement/dysfunction. (Level C)
  - Baffle leak resulting in a significant right-to-left shunt and symptoms. (Level C)
  - Symptomatic bradyarrythmias or tachyarrhythmias. (Level C)
- The following situations may warrant reintervention following the arterial switch procedure:
  - Significant pulmonary artery stenosis (subvalvular, pulmonary trunk or branch pulmonary artery). (Level C)
  - Coronary arterial obstruction. (Level C)
  - Severe neoaortic valve regurgitation. (Level C)
  - Severe neoaortic root dilation. (Level C)
- The following situations may warrant reintervention following the Rastelli procedure:
  - Significant right ventricle to pulmonary artery conduit obstruction. (Level C)
  - Severe right ventricle to pulmonary artery conduit regurgitation with symptoms, progressive RV enlargement, and the occurrence of atrial or ventricular arrhythmia. (Level C)
  - Severe subaortic obstruction across the left ventricle to aorta tunnel (mean gradient greater than 50 mmHg). (Level C)
  - Significant branch pulmonary artery stenosis. (Level C)
  - Residual VSD resulting in a Qp:Qs greater than 1.5:1, pulmonary hypertension or progressive LV enlargement/dysfunction. (Level C)

Class IIb

- Patients deemed to be at particularly high risk for sudden cardiac death may benefit from ICDs for primary prevention. (Level C)

Congenitally corrected transposition of the great arteries

Class I

- Pacemakers are indicated in patients with spontaneous or postoperative third-degree and advanced second-degree AV block or documented periods of asystole (3.0 s or more). (Level C)
- Ablation and device implantation should be undertaken by an electrophysiologist with appropriate training/experience in the ACHD population. (Level C)
- Patients who require intervention should be treated by ACHD cardiologists and congenital heart surgeons with appropriate experience. (Level C)

Class IIa

- The following situations may warrant surgical intervention/reinterventions:
  - Presence of VSD or residual VSD. (Level C)
  - Moderate to severe systemic AV valve regurgitation. (Level B)
  - Hemodynamically significant pulmonary or subpulmonary obstruction. (Level B)
  - Significant stenosis across a left ventricle to pulmonary artery conduit. (Level C)
  - Deteriorating systemic (right) ventricular function. (Level C)

Fontan operation

Class I

- Reintervention after the Fontan procedure is warranted in the following situations:
  - Obstruction to systemic venous return in the Fontan circuit. (Level C)
  - Obstruction of pulmonary venous return. (Level C)
  - Significant (moderately severe or greater) systemic AV valve regurgitation. (Level C)
  - Development of venous collateral channels or pulmonary arteriovenous malformations resulting in symptomatic cyanosis. (Level C)
  - Residual ASD or fenestration resulting in significant right-to-left shunt. (Level C)
  - Residual shunt secondary to a previous palliative surgical shunt or residual ventricle-to-pulmonary artery connection causing a hemodynamically significant volume or pressure load. (Level C)
  - Subaortic obstruction with a peak-to-peak gradient of greater than 30 mmHg. (Level C)
  - Protein-losing enteropathy that is associated with high systemic venous pressures or Fontan abnormality. (Level C)
  - Recurrent or poorly tolerated atrial arrhythmias refractory to medical therapy. (Level C)
- Fontan patients with a history of atrial thrombus, thromboembolic event, interatrial communication or atrial arrhythmias should be therapeutically anticoagulated with warfarin. (Level C)
- When arrhythmias are present, an underlying hemodynamic cause should always be sought, and in particular, obstruction of the Fontan circuit, thrombus formation or ventricular dysfunction need to be excluded by comprehensive imaging. (Level C)
- Patients with arrhythmias should be referred for consultation with an electrophysiologist with expertise in CHD. (Level C)
- Electrophysiological studies in Fontan patients should be performed in centres with expertise in CHD. (Level C)
- Patients who require intervention or reintervention should be treated by ACHD cardiologists and congenital heart surgeons with appropriate experience. (Level C)

Class IIa

- Fontan patients with intracardiac pacemaker or defibrillator leads should be therapeutically anticoagulated with warfarin. (Level C)

- Anticoagulation may be considered in Fontan patients without atrial thrombus or arrhythmias. (Level C)
- Patients with serious refractory atrial arrhythmias may be considered for Fontan conversion to a total cavopulmonary connection with concomitant atrial maze procedure. (Level C)

Class IIb

- When clinical situations or hemodynamics warrant therapy, it may be reasonable to treat ventricular dysfunction in Fontan patients with diuretics, angiotensin-converting enzyme inhibitors and beta-blockers as tolerated. (Level C)

Eisenmenger's syndrome

Class I

- Advanced pulmonary vascular obstructive disease with a resistance that is fixed, in combination with the absence of left-to-right shunting, render a patient ineligible for cardiac repair. (Level C)
- The main interventions in patients with Eisenmenger's syndrome are directed toward preventing complications (eg, influenza and pneumococcal vaccination) or restoring physiological balance (eg, iron replacement for iron deficiency). (Level C)
- Phlebotomy with fluid replacement and iron supplementation should be performed only in patients who are symptomatic from secondary erythrocytosis. Prevention of iron deficiency is important. (Level C)
- Platelet transfusions, fresh frozen plasma, vitamin K, cryoprecipitate and desmopressin can be used to treat severe bleeding. (Level C)
- If iron deficiency anemia is confirmed, iron replacement should be prescribed. (Level C)
- Symptomatic hyperuricemia and gouty arthritis can be treated as necessary. (Level C)
- Sinus rhythm should be restored promptly and maintained whenever possible. (Level C)
- Symptomatic arrhythmias should be treated with individualized antiarrhythmic therapy. (Level C)
- Patients with atrial fibrillation/flutter should receive warfarin therapy with judicious monitoring of international normalized ratio levels (sodium citrate adjusted to hematocrit). (Level C)
- Insertion of an implantable defibrillator is a high-risk endeavour. It may be considered in patients with syncope and documented concurrent ventricular arrhythmia. Epicardial approaches should be used. (Level C)
- Transvenous pacing leads are not recommended and must be avoided in the presence of intracardiac shunts due to risk of paradoxical embolization. (Level B)
- Patients with Eisenmenger's syndrome should be treated by an ACHD cardiologist who understands and has experience in the management of Eisenmenger's syndrome. (Level C)
- Patients with Eisenmenger's syndrome benefit from the involvement of other specialists (nursing, respiratory, psychology/psychiatry, hematology, gynecology, anesthesia, intensive care and social work). (Level C)

Class IIa

- Cyanotic patients having surgery may undergo prophylactic phlebotomy to reduce the hematocrit level to less than 65%. (Level C)
- Pulmonary vasodilator therapy may help to improve quality of life in patients with Eisenmenger's syndrome. (Level B)

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