Adults with Congenital Heart Disease

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SJHMC
Disclosures & Disclaimer

- I have no lucrative financial relationships with industry to disclose.

- I do not intend to discuss unapproved, experimental, off-label or other magical therapies or devices.
Objectives:

- Describe the demographics of ACHD.
- Understand common lesions and surgical repair techniques in ACHD and their long-term sequelae.
- Recognize and implement recommendations for ACHD care based on national guidelines.
ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease

A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease)

Developed in Collaboration With the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons

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1.4. Epidemiology and Scope of the Problem

Remarkable improvement in survival of patients with congenital heart disease (CHD) has occurred over the past half century since reparative surgery has become commonplace. Since the advent of neonatal repair of complex lesions in the 1970s, an estimated 85% of patients survive into adult life. The 32nd Bethesda Conference report in 2000 estimated that there were approximately 800,000 adults with CHD in the United States (2,3). Given modern surgical mortality rates of less than 5%, one would expect that in the next decade, almost 1 in 150 young adults will have some form of CHD.
Pediatric to Adult Congenital Heart Disease

- Increased midterm survival
- Lower perioperative mortality
- Improved surgical techniques
- Fetal diagnosis
- Incidence of CHD
- Expanded population of adolescents and adults with CHD
- Increased early survival
- Early complete repair
- Advances in NICU care
Patients Reaching Adulthood With Congenital Heart Disease

20,000 new patients per year
4% increase per year

Adult Patients With CHD

1,500,000
1,250,000
1,000,000
750,000
500,000
250,000
0

1970
1980
1990
2000
2010

*Estimated numbers.
# Adult Congenital Heart Disease

<table>
<thead>
<tr>
<th></th>
<th>USA</th>
<th>Arizona</th>
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<td>Population</td>
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<td>Total ACHD</td>
<td>1,000,000</td>
<td>20,000</td>
</tr>
<tr>
<td>Simple</td>
<td>470,000</td>
<td>9400</td>
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<tr>
<td>Moderate Complexity</td>
<td>380,000</td>
<td>7600</td>
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<tr>
<td>Complex</td>
<td>150,000</td>
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The Usual Suspects

- Arrhythmias
  - Atrial
  - Ventricular
  - Sudden death

- Vascular Lesions

- Residual Shunts

- Valvular Disease

- Adults With Congenital Heart Disease

- Heart Failure
  - Right heart failure
  - Left heart failure
    - Systolic
    - Diastolic
  - Pulmonary hypertension
Congenital Heart Disease in the General Population
Changing Prevalence and Age Distribution

Ariane J. Marelli, MD; Andrew S. Mackie, MD, SM; Raluca Ionescu-Ittu, MSc;
Elham Rahme, PhD; Louise Pilote, MD, MPH, PhD

Background—Empirical data on the changing epidemiology of congenital heart disease (CHD) are scant. We determined the prevalence, age distribution, and proportion of adults and children with severe and other forms of CHD in the general population from 1985 to 2000.

Methods and Results—Where healthcare access is universal, we used administrative databases that systematically recorded all diagnoses and claims. Diagnostic codes conformed to the International Classification of Disease, ninth revision. Severe CHD was defined as tetralogy of Fallot, truncus arteriosus, transposition complexes, endocardial cushion defects, and univentricular heart. Prevalence of severe and other CHD lesions was determined in 1985, 1990, 1995, and 2000 using population numbers in Quebec. Children were subjects <18 years of age. The prevalence was 4.09 per 1000 adults in the year 2000 for all CHD and 0.38 per 1000 (9%) for those with severe lesions. Female subjects accounted for 57% of the adult CHD population. The median age of all patients with severe CHD was 11 years (interquartile range, 4 to 22 years) in 1985 and 17 years (interquartile range, 10 to 28 years) in 2000 (P<0.0001). The prevalence of severe CHD increased from 1985 to 2000, but the increase in adults was significantly higher than that observed in children. In the year 2000, 49% of those alive with severe CHD were adults.

Conclusions—The prevalence in adults and median age of patients with severe CHD increased in the general population from 1985 to 2000. In 2000, there were nearly equal numbers of adults and children with severe CHD. (Circulation. 2007;115:163-172.)
TABLE 3. Prevalence of Severe and Other CHD in a Population of 5 760 295 Adults and 1 596 734 Children in the Year 2000

<table>
<thead>
<tr>
<th></th>
<th>Adults Alive in 2000</th>
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<th>Children Alive in 2000</th>
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<tbody>
<tr>
<td></td>
<td>n (%)</td>
<td>Prevalence per 1000 Adults</td>
<td>n (%)</td>
<td>Prevalence per 1000 Children</td>
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<tr>
<td>All congenital heart lesions*</td>
<td>23 563 (100)</td>
<td>4.09</td>
<td>18 979 (100)</td>
<td>11.89</td>
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<tr>
<td>Severe lesions</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>TOF or truncus arteriosus</td>
<td>1001</td>
<td>0.17</td>
<td>778</td>
<td>0.49</td>
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<tr>
<td>AVCD</td>
<td>834</td>
<td>0.14</td>
<td>914</td>
<td>0.57</td>
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<tr>
<td>Transposition complex</td>
<td>235</td>
<td>0.04</td>
<td>424</td>
<td>0.27</td>
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<tr>
<td>Univentricular hearts</td>
<td>150</td>
<td>0.03</td>
<td>213</td>
<td>0.13</td>
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<tr>
<td>All severe lesions</td>
<td>2205 (9)</td>
<td>0.38</td>
<td>2316 (12)</td>
<td>1.45</td>
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<td>Other lesions</td>
<td></td>
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<td></td>
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<tr>
<td>ASD</td>
<td>5076</td>
<td>0.88</td>
<td>6205</td>
<td>3.89</td>
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<td>VSD</td>
<td>4486</td>
<td>0.78</td>
<td>6709</td>
<td>4.20</td>
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<td>PDA</td>
<td>103</td>
<td>0.02</td>
<td>493</td>
<td>0.31</td>
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<td>161</td>
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<td>1.67</td>
<td>1586</td>
<td>0.99</td>
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<tr>
<td>Congenital aortic stenosis or insufficiency‡</td>
<td>619</td>
<td>0.11</td>
<td>425</td>
<td>0.27</td>
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<tr>
<td>Anomalies of pulmonary artery or valve</td>
<td>698</td>
<td>0.12</td>
<td>798</td>
<td>0.50</td>
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<tr>
<td>Aortic coarctation</td>
<td>389</td>
<td>0.07</td>
<td>396</td>
<td>0.25</td>
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<tr>
<td>Congenital mitral or tricuspid valve disease</td>
<td>178</td>
<td>0.03</td>
<td>60</td>
<td>0.04</td>
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<tr>
<td>Ebstein’s anomaly</td>
<td>50</td>
<td>0.01</td>
<td>29</td>
<td>0.02</td>
</tr>
<tr>
<td>Unknown congenital heart lesions</td>
<td>91</td>
<td>0.02</td>
<td>35</td>
<td>0.02</td>
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<tr>
<td>Anomalies of the great veins</td>
<td>25</td>
<td>0.00</td>
<td>24</td>
<td>0.01</td>
</tr>
<tr>
<td>All other lesions</td>
<td>21 358 (91)</td>
<td>3.71</td>
<td>16 663 (88)</td>
<td>10.44</td>
</tr>
</tbody>
</table>
Atrial and Ventricular Septal Defects

- Non cyanotic forms of congenital heart disease.
- Characterized by left to right shunting at atrial or ventricular level.
- Results in pulmonary overcirculation.
  - Early CHF due to increased volume load
  - Late pulmonary hypertension
- Generally repaired in childhood in single procedure.
- Excellent long term hemodynamic prognosis.
- Late arrhythmias common.
Atrial Septal Defect

Figure 1. Atrial Septal Defect with Resultant Left-to-Right Shunting.
Blood from the pulmonary veins enters the left atrium, after which some of it crosses the atrial septal defect into the right atrium and ventricle (longer arrow).

- NEJM 1/27/2000
Atrial Septal Defect

- Murmur in pulmonary outflow area (LUSB)
- Wide split S2
- IRBBB on EKG
- Increased PVM on CXR
- Hole on echo…
Atrial Septal Defect

- Aortic cannula
- SVC cannula
- ASD
- Tendon of Todaro
- Right ventricle
- Septum primum
- Septum secundum
- Left atrium
- Right-to-left shunt
- Patent foramen ovale
- Tranverse sinus
- Right-sided disk
- Left-sided disk

St. Joseph's Hospital and Medical Center - CHW

Scott and Laura Eller Congenital Heart Center
Caring for Hearts for a Lifetime
Congenital Heart Disease for the Adult Cardiologist

Atrial Septal Defects in the Adult
Recent Progress and Overview

Gary Webb, MD; Michael A. Gatzoulis, MD, PhD

Management of Atrial Septal Defects in Adults

Indications for ASD closure

Right atrial and right ventricular dilation by echocardiography, MRI, or CT (in the presence of an ASD and in the absence of advanced pulmonary arterial hypertension) manifested with 1 or more of the following:

- ASD minimum diameter >10 mm on echocardiography
- Qp:Qs >1.5:1 by echocardiographic or cardiac MRI flow assessment, or from oxygen saturation runs, when cardiac catheterization is performed (for other reasons)

Anticipated benefits from ASD closure

- Improved functional class, dyspnea index, and exercise capacity (irrespective of age). Improvement occurs earlier after device closure than with surgical closure. Physical reconditioning is recommended.
- In addition, the following long-term prognostic benefits can be anticipated:
  - Improved survival after youthful repair
  - Improved quality of life
  - Prevention of right heart failure
  - Prevention of pulmonary arterial hypertension

(Circulation. 2006;114:1645-1653.)
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2.6. Recommendations for Postintervention Follow-Up

**CLASS I**

1. Early postoperative symptoms of undue fever, fatigue, vomiting, chest pain, or abdominal pain may represent postpericardiotomy syndrome with tamponade and should prompt immediate evaluation with echocardiography. *(Level of Evidence: C)*

2. Annual clinical follow-up is recommended for patients postoperatively if their ASD was repaired as an adult and the following conditions persist or develop:
   a. PAH. *(Level of Evidence: C)*
   b. Atrial arrhythmias. *(Level of Evidence: C)*
   c. RV or LV dysfunction. *(Level of Evidence: C)*
   d. Coexisting valvular or other cardiac lesions. *(Level of Evidence: C)*

3. Evaluation for possible device migration, erosion, or other complications is recommended for patients 3 months to 1 year after device closure and periodically thereafter. *(Level of Evidence: C)*

4. Device erosion, which may present with chest pain or syncope, should warrant urgent evaluation. *(Level of Evidence: C)*
Ventricular Septal Defect

Figure 2. Ventricular Septal Defect with Resultant Left-to-Right Shunting.
When the left ventricle contracts, it ejects some blood into the aorta and some across the ventricular septal defect into the right ventricle and pulmonary artery (arrow).

- NEJM 1/27/2000
Ventricular Septal Defect

- Holosystolic murmur at LLSB
  - Murmur intensity often exaggerates shunt (small holes make loud noises)
- Obscured S2
- LVH on EKG
- CM and increased PVM on CXR
- Hole on echo…
VSD Repair
VSD Follow Up

3.6. Key Issues to Evaluate and Follow-Up

3.6.1. Recommendations for Surgical and Catheter Intervention Follow-Up

**CLASS I**

1. Adults with VSD with residual heart failure, shunts, PAH, AR, or RVOT or LVOT obstruction should be seen at least annually at an ACHD regional center. *(Level of Evidence: C)*

2. Adults with a small residual VSD and no other lesions should be seen every 3 to 5 years at an ACHD regional center. *(Level of Evidence: C)*

3. Adults with device closure of a VSD should be followed up every 1 to 2 years at an ACHD center depending on the location of the VSD and other factors. *(Level of Evidence: C)*
<table>
<thead>
<tr>
<th>Lesion Type</th>
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<tr>
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<td>50 (0.00)</td>
<td>0.01</td>
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<tr>
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<td>91 (0.00)</td>
<td>0.02</td>
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<tr>
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<td>25 (0.00)</td>
<td>0.00</td>
</tr>
<tr>
<td>All other lesions</td>
<td>21358 (91)</td>
<td>3.71</td>
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</table>
Tetralogy of Fallot

- Complex cyanotic CHD lesion with:
  - Aortic override
  - Pulmonary stenosis
  - Ventricular septal defect
  - RV hypertrophy

- Generally repaired in infancy
- Multiple reoperations common
- “Complete repair <> cure!
  - Pulmonary stenosis
  - Pulmonary insufficiency
  - Atrial flutter
  - Ventricular tachycardia
ToF Repair

St. Joseph’s Hospital and Medical Center

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Pulmonary artery widened with a patch
Area below valve widened
VSD repaired with a patch
Late risk of outcomes for adults with repaired tetralogy of Fallot from an inception cohort spanning four decades☆

Edward J. Hickey a, Gruschen Veldtman b, Timothy J. Bradley b, Aungkana Gengsakul b, Cedric Manlhiot b, William G. Williams a, Gary D. Webb c, Brian W. McCrindle b, a

a Division of Cardiovascular Surgery, Department of Surgery, University of Toronto, The Hospital for Sick Children, Toronto, Canada
b Division of Cardiology, Department of Pediatrics, University of Toronto, The Hospital for Sick Children, Toronto, Canada
c Division of Cardiology, Department of Medicine, University of Toronto, Toronto General Hospital, Toronto, Canada

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Pulmonary Valve Replacement in Adults Late After Repair of Tetralogy of Fallot: Are We Operating Too Late?
Judith Therrien, MD, FRCP(C), Samuel C. Siu, MD, FRCP(C), Peter R. McLaughlin, MD, FRCP(C), Peter P. Liu, MD, FRCP(C), William G. Williams, MD, FRCS(C), Gary D. Webb, MD, FRCP(C)
Toronto, Canada

OBJECTIVES
The purpose of this study is to evaluate right ventricular (RV) volume and function after pulmonary valve replacement (PVR) and to address the issue of optimal surgical timing in these patients.

BACKGROUND
Chronic pulmonary regurgitation (PR) following repair of tetralogy of Fallot (TOF) leads to RV dilation and an increased incidence of sudden cardiac death in adult patients.

METHODS
We studied 25 consecutive adult patients who underwent PVR for significant PR late after repair of TOF. Radionuclide angiography was performed in all at a mean of 8.2 months (± 8 months) before PVR and repeated at a mean of 28.0 months (± 22.8 months) after the operation. Right ventricular (RV) end-systolic volume (RVESV), RV end-diastolic volume (RVEDV) and RV ejection fraction (RVEF) were measured.

RESULTS
Mean RVEDV, RVESV and RVEF remained unchanged after PVR (227.1 ml versus 214.9 ml, p = 0.74; 157.4 ml versus 155.4 ml, p = 0.94; 35.6% versus 34.7%, p = 0.78, respectively). Of the 10 patients with RVEF ≥ 0.40 before PVR, 5 patients (50%) maintained a RVEF ≥ 0.40 following PVR, whereas only 2 out of 15 patients (13%) with pre-operative values <0.40 reached an RVEF ≥ 0.40 postoperatively (p < 0.001).

CONCLUSIONS
Right ventricular recovery following PVR for chronic significant pulmonary regurgitation after repair of TOF may be compromised in the adult population. In order to maintain adequate RV contractility, pulmonary valve implant in these patients should be considered before RV function deteriorates. (J Am Coll Cardiol 2000;36:1670–5) © 2000 by the American College of Cardiology
Value of Programmed Ventricular Stimulation After Tetralogy of Fallot Repair
A Multicenter Study

Paul Khairy, MD, MSc; Michael J. Landzberg, MD; Michael A. Gatzoulis, MD, PhD;
Hugues Lucron, MD; Jean Lambert, PhD; François Marçon, MD;
Mark E. Alexander, MD; Edward P. Walsh, MD

Background—Studies have consistently shown that ventricular tachycardia (VT) and sudden cardiac death (SCD) complicate the long-term outcome after tetralogy of Fallot repair, yet the diagnostic and predictive value of electrophysiological testing in this population is uncertain.

Methods and Results—A multicenter cohort of 252 patients with repaired tetralogy of Fallot undergoing programmed ventricular stimulation was followed up for 18.5±9.6 and 6.5±4.5 years after corrective surgery and electrophysiological testing, respectively. Clinical VT and/or SCD occurred in 24.6%. Sustained monomorphic VT and polymorphic VT were induced in 30.2% and 4.4%. Including polymorphic VT in the definition of inducibility improved sensitivity (66.1±6.0% versus 77.4±5.3%, P=0.0082) with a marginal reduction in specificity (81.6±2.8% versus 79.5±2.9%, P=0.0433). Positive and negative predictive values were 55.2±5.3% and 91.5±2.2%. Independent risk factors for inducibility were age at study ≥18 years (OR, 3.3), palpitations (OR, 2.8), prior palliative surgery (OR, 3.1), modified Lown criteria ≥2 (OR, 5.6), and cardiothoracic ratio ≥0.6 (OR, 3.3). Event-free survival rates in noninducible and inducible patients at 1, 3, 10, and 15 years were 97.9%, 92.8%, 89.3%, and 89.3% versus 79.4%, 62.6%, 58.7%, and 50.3%, respectively (P<0.0001). Both inducible monomorphic VT [relative risk (RR), 5.0; P=0.0002] and polymorphic VT (RR, 12.9; P<0.0001) predicted future clinical VT and SCD. In a multivariate analysis, inducible sustained VT was an independent risk factor for subsequent events (RR, 4.7; 95% CI, 1.2 to 18.5; P=0.0268).

Conclusions—Programmed ventricular stimulation is of diagnostic and prognostic value in risk stratifying patients with repaired tetralogy of Fallot. In this patient population, inducible sustained polymorphic VT should not be disregarded as nonspecific. (Circulation. 2004;109:1994-2000.)
### TABLE 4. Predictors of Inducible Sustained VT

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<th>Variable</th>
<th>OR</th>
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<td><strong>Univariate analysis</strong></td>
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<td>1.04–1.11</td>
<td>&lt;0.0001</td>
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<td>Age ≥18 y</td>
<td>6.0</td>
<td>3.0–12.3</td>
<td>&lt;0.0001</td>
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<td>Age at corrective surgery</td>
<td>1.07</td>
<td>1.02–1.12</td>
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<td>Age ≥7 y</td>
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<td>1.9–5.6</td>
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<td>Palpitations</td>
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<td>Syncope</td>
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<td>Prior palliative surgery</td>
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<td>Documented AF/flutter</td>
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<td>At least moderate PR</td>
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<td>At least moderate TR</td>
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<td>Cardiothoracic ratio ≥0.60</td>
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<td>Age at EP study ≥18 y</td>
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<td>Palpitations</td>
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<td>Prior palliative surgery</td>
<td>3.1</td>
<td>1.2–7.6</td>
<td>0.0163</td>
</tr>
<tr>
<td>Modified Lown ≥2</td>
<td>5.6</td>
<td>1.0–30.9</td>
<td>0.0493</td>
</tr>
<tr>
<td>Cardiothoracic ratio ≥0.60</td>
<td>3.3</td>
<td>1.2–8.8</td>
<td>0.0200</td>
</tr>
</tbody>
</table>

**Figure 1.** A, Kaplan-Meier curve of survival free from clinical VT and SCD in entire cohort. B, Event-free survival according to results of programmed ventricular stimulation.
ToF Follow Up

10.4. Recommendations for Evaluation and Follow-Up of the Repaired Patient

CLASS I
1. Patients with repaired tetralogy of Fallot should have at least annual follow-up with a cardiologist who has expertise in ACHD. (Level of Evidence: C)
2. Patients with tetralogy of Fallot should have echocardiographic examinations and/or MRIs performed by staff with expertise in ACHD. (Level of Evidence: C)

3. Screening for heritable causes of their condition (eg, 22q11 deletion) should be offered to all patients with tetralogy of Fallot. (Level of Evidence: C)
4. Before pregnancy or if a genetic syndrome is identified, consultation with a geneticist should be arranged for patients with tetralogy of Fallot. (Level of Evidence: B)
5. Patients with unrepaired or palliated forms of tetralogy should have a formal evaluation at an ACHD center regarding suitability for repair. (Level of Evidence: B)
Transposition of the Great Arteries

- Complex cyanotic CHD due to parallel circulations.
- Extreme cyanosis at birth.
- Rapidly fatal at time of PDA closure if unoperated.

- Atrial baffle repair
  - Mustard & Senning
  - Palliative!
- Arterial switch repair
  - Jatene
  - Curative?
Atrial Baffle: Mustard & Senning

- Physiologic correction of cyanosis without coronary transfer.
  - (2 wrongs make a right?)
- Extensive atrial suture lines.
  - Sick sinus syndrome
  - Atrial flutter
- Baffle leaks.
- Baffle stenoses.
- Systemic RV failure.
- Systemic TV failure.
TGA Example

- 20 something man with varicose veins.
- Very SOB with minimal exertion.
- Ok at rest.
- Receiving laser sclerotherapy from adult cardiologist.

- Some kind of heart surgery as infant.
- Not much follow up.
- Also has a very old pacemaker.
TGA Example

- Atretic superior baffle limb.
- Stenostic inferior baffle limb.
- Sclerotherapy as first line treatment?
Transcatheter stenting & pacing
Arrhythmia and Mortality After the Mustard Procedure: A 30-Year Single-Center Experience

MARK GELATT, MD, ROBERT M. HAMILTON, MD, BRIAN W. McCRRINDLE, MD, MPH, FACC, MICHAEL CONNELLY, MB, ANDREW DAVIS, MB, BS, LOUISE HARRIS, MB, FACC, ROBERT M. GOW, MB, BS, WILLIAM G. WILLIAMS, MD, GEORGE A. TRUSLER, MD, ROBERT M. FREEDOM, MD, FACC

Toronto, Ontario, Canada

Objectives. Our purpose was to assess the risk factors for late mortality, loss of sinus rhythm and atrial flutter after the Mustard operation.

Background. The Mustard operation provides correction of cyanosis with low surgical risk in transposition of the great vessels. However, right ventricular failure, loss of sinus rhythm, atrial flutter and death are frequent long-term complications.

Methods. Records of 534 children who underwent the Mustard operation at a single center since 1962 were reviewed for demographic, anatomic, electrocardiographic and physiologic predictors and outcomes.

Results. There were 52 early deaths (9.7%). Survival analysis was undertaken for 478 early survivors with a mean follow-up interval of 11.6 ± 7.2 years. There were 77 late deaths (16.1%), with sudden death (n = 31) the most frequent cause. Survival estimates were 89% at 5 years and 76% at 20 years of age. Risk factors were an earlier date of operation, operative period arrhythmia and an associated ventricular septal defect. Risk (hazard) of late death declined in the first decade, with further peaks in the second decade. Sinus rhythm was present in 77% at 5 years and 40% at 20 years. Loss of sinus rhythm was associated with previous septectomy, postoperative bradycardia and late atrial flutter. Freedom from atrial flutter was 92% at 5 years and 73% at 20 years of age. Risk factors for atrial flutter were the occurrence of perioperative bradyarrhythmia, reoperation and loss of sinus rhythm during follow-up. Risk of atrial flutter demonstrates a late increase.

Conclusions. Ongoing loss of sinus rhythm and late peaks in the risk of atrial flutter and death necessitate continued follow-up.

(J Am Coll Cardiol 1997;29:194–201) ©1997 by the American College of Cardiology
Brady & Tachy Arrhythmias
Atrial Baffle Follow Up

11.4. Recommendation for Evaluation of the Operated Patient With Dextro-Transposition of the Great Arteries

CLASS I
1. Patients with repaired d-TGA should have annual follow-up with a cardiologist who has expertise in the management of ACHD patients. (Level of Evidence: C)

11.9. Recommendations for Electrophysiology Testing/Pacing Issues in Dextro-Transposition of the Great Arteries

CLASS I
1. Clinicians should be mindful of the risk of sudden arrhythmic death among adults after atrial baffle repair of d-TGA. These events usually relate to VT but may be caused in some cases by rapidly conducted IART or progressive AV block. (Level of Evidence: B)
2. Consultation with an electrophysiologist who is experienced with CHD is recommended to assist with treatment decisions. (Level of Evidence: B)
3. Pacemaker implantation is recommended for patients with d-TGA with either symptomatic sinus bradycardia or sick sinus syndrome. (Level of Evidence: B)
Arterial Switch

Coronary arteries are detached from the aortic valve and connected to the pulmonic valve.

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11.5.4. Recommendations for Imaging for Dextro-Transposition of the Great Arteries After Arterial Switch Operation

CLASS I
1. Comprehensive echocardiographic imaging to evaluate the anatomy and hemodynamics in patients with d-TGA and prior ASO repair should be performed at least every 2 years at a center with expertise in ACHD. *(Level of Evidence: C)*
2. After prior ASO repair for d-TGA, all adults should have at least 1 evaluation of coronary artery patency. Coronary angiography should be performed if this cannot be established noninvasively. *(Level of Evidence: C)*
### TABLE 3. Prevalence of Severe and Other CHD in a Population of 5,760,295 Adults and 1,596,734 Children in the Year 2000

<table>
<thead>
<tr>
<th></th>
<th>Adults Alive in 2000</th>
<th></th>
<th>Children Alive in 2000</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>n (%)</td>
<td>Prevalence per 1000 Adults</td>
<td>n (%)</td>
<td>Prevalence per 1000 Children</td>
</tr>
<tr>
<td><strong>All congenital heart lesions</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>TOF or truncus arteriosus</td>
<td>1001</td>
<td>0.17</td>
<td>778</td>
<td>0.49</td>
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<tr>
<td>AVCD</td>
<td>834</td>
<td>0.14</td>
<td>914</td>
<td>0.57</td>
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<tr>
<td>Transposition complex</td>
<td>235</td>
<td>0.04</td>
<td>424</td>
<td>0.27</td>
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<tr>
<td>Univentricular hearts</td>
<td>150</td>
<td>0.03</td>
<td>213</td>
<td>0.13</td>
</tr>
<tr>
<td><strong>All severe lesions</strong></td>
<td>2205 (9)</td>
<td>0.38</td>
<td>2316 (12)</td>
<td>1.45</td>
</tr>
<tr>
<td><strong>Other lesions</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>ASD</td>
<td>5076</td>
<td>0.88</td>
<td>6205</td>
<td>3.89</td>
</tr>
<tr>
<td>VSD</td>
<td>4486</td>
<td>0.78</td>
<td>6709</td>
<td>4.20</td>
</tr>
<tr>
<td>PDA</td>
<td>103</td>
<td>0.02</td>
<td>493</td>
<td>0.31</td>
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<tr>
<td>Unspecified defect of septal closure</td>
<td>161</td>
<td>0.03</td>
<td>9</td>
<td>0.01</td>
</tr>
<tr>
<td>Unspecified congenital anomalies†</td>
<td>9621</td>
<td>1.67</td>
<td>1586</td>
<td>0.99</td>
</tr>
<tr>
<td>Congenital aortic stenosis or insufficiency‡</td>
<td>619</td>
<td>0.11</td>
<td>425</td>
<td>0.27</td>
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<tr>
<td>Anomalies of pulmonary artery or valve</td>
<td>698</td>
<td>0.12</td>
<td>798</td>
<td>0.50</td>
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<tr>
<td>Aortic coarctation</td>
<td>389</td>
<td>0.07</td>
<td>396</td>
<td>0.25</td>
</tr>
<tr>
<td>Congenital mitral or tricuspid valve disease</td>
<td>178</td>
<td>0.03</td>
<td>60</td>
<td>0.04</td>
</tr>
<tr>
<td>Ebstein’s anomaly</td>
<td>50</td>
<td>0.01</td>
<td>29</td>
<td>0.02</td>
</tr>
<tr>
<td>Unknown congenital heart lesions</td>
<td>91</td>
<td>0.02</td>
<td>35</td>
<td>0.02</td>
</tr>
<tr>
<td>Anomalies of the great veins</td>
<td>25</td>
<td>0.00</td>
<td>24</td>
<td>0.01</td>
</tr>
<tr>
<td><strong>All other lesions</strong></td>
<td>21,358 (91)</td>
<td>3.71</td>
<td>16,663 (88)</td>
<td>10.44</td>
</tr>
</tbody>
</table>
Univentricular Hearts

- Most severe of all CHD types.
- Can be single LV, RV, or undefined common V.
- Always presents in infancy, however, presentation varies depending on great artery configuration:
  - Severe PS = cyanosis
  - No PS = CHF
  - Some PS = murmur alone
  - Systemic obstruction = shock!
- Staged palliative operations to Fontan circulation.
Fontan Circulation

- Separated but 1V circulation.
  - No desaturation / cyanosis
  - No paradoxical emboli
- Passive PBF
- Poor exercise capacity
- Venous hypertension
The Fontan Procedure
Contemporary Techniques Have Improved Long-Term Outcomes

Yves d’Udekein, MD, PhD; Ajay J. Iyengar, BmedSci; Andrew D. Cochrane, MD, FRACS; Leeanne E. Grigg, MBBS, FRACP; James M. Ramsay, MD, FRACP; Gavin R. Wheeldon, MD, FRACP; Dan J. Penny, MD, PhD, FRACP; Christian P. Brizard, MD

**Background**—To determine whether patients undergoing the lateral tunnel and extracardiac conduit modifications of the Fontan procedure have better outcomes than patients undergoing a classical atrio-pulmonary connection.

**Methods and Results**—Between 1980 and 2000, 305 consecutive patients underwent a Fontan procedure at our institution. There were 10 hospital deaths (mortality: 3%) with no death after 1990. Independent risk factors for mortality were preoperative elevated pulmonary artery pressures ($P=0.002$) and common atrioventricular valve ($P=0.04$). Fontan was taken down during hospital stay in 7 patients. A mean of 12±6 years of follow-up was obtained in the 257 non-foreign Fontan survivors. Completeness of concurrent follow-up was 96%. Twenty-year survival was 84% (95% CI: 79 to 89%). Recent techniques improved late survival. The 15-year survival after atrio-pulmonary connection was 81% (95% CI: 73% to 87%) versus 94% (95% CI: 79% to 98%) for lateral tunnel ($P=0.004$). Nine pts required heart transplantation (8 atrio-pulmonary connection, 1 lateral tunnel). Undergoing a Fontan modification independently predicted decreased occurrence of arrhythmia, and 15-year freedom from SVT was 61% (95% CI: 51% to 70%) for atrio-pulmonary connection versus 87% (95% CI: 76% to 93%) for lateral tunnel ($P=0.02$). Freedom from Fontan failure (death, take-down, transplantation, or NYHA class III-IV) was 70% (95% CI: 58% to 79%) at 20 years. After extra-cardiac conduits, no death, SVT, or failure was observed.

**Conclusions**—The Fontan procedure remains a palliation, but outcomes of patients have improved. Better patient selection minimizes hospital mortality. Patients with lateral tunnel and extracardiac conduit modifications experience less arrhythmia and are likely to have failure of their Fontan circulation postponed. (Circulation. 2007;116[suppl I]:I-157–I-164.)
Figure 1. Fontan surgical techniques: Classical atrio pulmonary connection (A), Lateral tunnel (B), and Extracardiac conduit (C).

Figure 2. Kaplan-Meier survival curves of hospital survivors by Fontan techniques.
14.8. Recommendations for Management Strategies for the Patient With Prior Fontan Repair

**CLASS I**

1. Management of patients with prior Fontan repair should be coordinated with a regional ACHD center. Local cardiologists, internists, and family care physicians should develop ongoing relationships with such a center with continuous availability of specialists. *(Level of Evidence: C)*

2. At least yearly follow-up is recommended for patients after Fontan repair. *(Level of Evidence: C)*

3. Arrhythmia management is frequently an issue, and consultation with an electrophysiologist is recommended as a vital part of care. *(Level of Evidence: C)*
Resource Documents

ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease: A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease) Developed in Collaboration With the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons

Best Practices in Managing Transition to Adulthood for Adolescents With Congenital Heart Disease: The Transition Process and Medical and Psychosocial Issues: A Scientific Statement From the American Heart Association
Craig Sable, Elsyse Foster, Karen Uzark, Katherine Bjorsem, Mary M. Canobbio, Heidi M. Connolly, Thomas P. Graham, Michelle Z. Gurvitz, Adrienne Kovacs, Alison K. Meadows, Graham J. Reid, John G. Reiss, Kenneth N. Rosenbaum, Paul J. Sagerman, Arwa Saidi, Rhonda Schonberg, Sangeeta Shah, Elizabeth Tong, Roberta G. Williams and on behalf of the American Heart Association Congenital Heart Defects Committee of the Council on Cardiovascular Disease in the Young, Council on Cardiovascular Nursing, Council on Clinical Cardiology, and Council on Peripheral Vascular Disease
Circulation published online Feb 28, 2011;
Delivery of Care and Ensuring Access

Healthcare for ACHD patients should be coordinated by regional ACHD centers of excellence that would serve as a resource for the surrounding medical community, affected individuals, and their families.

Every academic adult cardiology/cardiac surgery center should have access to a regional ACHD center for consultation and referral.

Each pediatric cardiology program should identify the ACHD center to which the transfer of patients can be made.

All emergency care facilities should have an affiliation with a regional ACHD center.
Delivery of Care and Ensuring Access

Ensuring and Improving Communication

Every ACHD patient should have a primary care physician. To ensure and improve communication, current clinical records should be on file with the primary care physician and a local cardiovascular specialist, as well as at a regional ACHD center; patients should also have copies of relevant records.

Referral Relationship

Every cardiovascular family caregiver should have a referral relationship with a regional ACHD center so that all patients have geographically accessible care.
Online Resources

http://www.achaheart.org/
Conclusions:

Adult Congenital Heart Disease is out there on your campus!

The spectrum of lesions runs from mild to extremely complex.

Each lesion has known long term problems based upon initial anatomy /physiology and surgical procedures performed.

Consensus documents exist to help you manage, refer, evaluate and treat this growing population of patients.
Guest Speakers

Beverly McCoy
Eva Gergely
J.R Trevas
Cindy Huie