A Growing Population: Outcomes and Expectations of Adult Patients with Congenital Heart Disease

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Harvard Medical School
Boston Children’s Hospital
No disclosures
Objectives

• Changing epidemiology
• Transition from pediatric to adult care
• Perioperative considerations
Congenital Heart Disease in the General Population: Changing Prevalence and Age Distribution
Ariane J. Marelli, Andrew S. Mackie, Raluca Ionescu-Ittu, Elham Rahme and Louise Pilote
Lifetime Prevalence of Congenital Heart Disease in the General Population From 2000 to 2010
Ariane J. Marelli, Raluca Ionescu-Ittu, Andrew S. Mackie, Liming Guo, Nandini Dendukuri and Mohammed Kaouache

Circulation. 2014;130:749-756; originally published online June 18, 2014;
The age of the most rapidly growing segment of the CHD population suggests that the next decades will witness an increasingly older population of patients with not only severe CHD but comorbidity that is expected to add to the disease burden.

Improved care, decreased mortality, and improved diagnosis over the life-span are likely contributors to the observation of an increasing prevalence of CHD. We and others have found a decrease in mortality of CHD patients, as documented in Canada, the United States, and other industrialized nations.

This decrease in mortality over time is likely an important contributing factor to an increasing pool of patients with prevalent CHD, especially for patients with severe CHD. For patients with mild forms of CHD, such as aortic and mitral valve disease, the rise in prevalence from childhood to adulthood is consistent with improvement in diagnostic techniques and presentation in adulthood, with an increased likelihood of being captured with longer observation periods. For example, bicuspid aortic valve is often not detected in childhood because the physical finding (a click) is subtle, and until stenosis or regurgitation develops, often not until adulthood, the diagnosis is not made. The reported rise in adults of mitral or tricuspid valve disease could be attributable to progressive atrioventricular valve regurgitation after endocardial cushion defect repair. Recently, we have observed a significant decrease in mortality associated with the delivery of specialized care for adults with severe and other forms of CHD.

These findings taken together provide the evidence base needed to improve quality of care for adults with CHD to improve outcomes.

In the Quebec CHD database, we observed a prevalence of CHD in infancy of 8.21 per 1000 from 1998 to 2005 and 8.12 per 1000 for the year 2010. Our results show that the 2010 CHD prevalence in infancy was lower than the prevalence in childhood. The most likely explanation for this is related to ascertainment of mild CHD, which is often diagnosed later in childhood. Therefore, it is not surprising that with up to 18 years of follow-up in children, we captured higher rates of total CHD prevalence, which included both adult and childhood cases.
Mortality Resulting From Congenital Heart Disease Among Children and Adults in the United States, 1999 to 2006

Suzanne M. Gilboa, PhD; Jason L. Salemi, MPH; Wendy N. Nembhard, PhD; David E. Fixler, MD; Adolfo Correa, MD, PhD
Specialized Adult Congenital Heart Disease Care: The Impact of Policy on Mortality
Darren Mylotte, Louise Pilote, Raluca Ionescu-Ittu, Michal Abrahamowicz, Paul Khairy, Judith Therrien, Andrew S. Mackie and Ariane Marelli
Geriatric Congenital Heart Disease

Burden of Disease and Predictors of Mortality

Jonathan Afilalo, MD, MSc,* Judith Therrien, MD,*‡ Louise Pilote, MD, MPH, PhD,† Raluca Ionescu-Ittu, MSc;‡ Giuseppe Martucci, MD;‡ Ariane J. Marelli, MD, MPH‡
Montreal, Quebec, Canada

- Dementia
- Gastrointestinal Bleed
- Chronic Kidney Disease
- Heart Failure
- Diabetes Mellitus
- Chronic Obstructive
- Pulmonary Disease
- Cancer
- Myocardial Infarction
Survival Prospects and Circumstances of Death in Contemporary Adult Congenital Heart Disease Patients Under Follow-Up at a Large Tertiary Centre

Gerhard-Paul Diller, Aleksander Kempny, Rafael Alonso-Gonzalez, Lorna Swan, Anselm Uebing, Wei Li, Sonya Babu-Narayan, Stephen J. Wort, Konstantinos Dimopoulos and Michael A. Gatzoulis

*Circulation.* 2015;132:2118-2125; originally published online September 14, 2015;

<table>
<thead>
<tr>
<th>Rank</th>
<th>Cause of death</th>
<th>Aortic coarctation</th>
<th>ASD</th>
<th>AVSD</th>
<th>Complex CHD</th>
<th>Ebstein</th>
<th>Eisenmenger</th>
<th>Fontan</th>
<th>Marfan syndrome</th>
<th>PDA</th>
<th>Systemic RV</th>
<th>Tetralogy of Fallot</th>
<th>TGA arterial switch</th>
<th>Valvular disease</th>
<th>VSD</th>
<th>All patients</th>
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<tr>
<td>1</td>
<td>Heart failure</td>
<td>31%</td>
<td>28%</td>
<td>57%</td>
<td>57%</td>
<td>38%</td>
<td>45%</td>
<td>52%</td>
<td>30%</td>
<td>50%</td>
<td>66%</td>
<td>40%</td>
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<td>40%</td>
<td>39%</td>
<td>42.5%</td>
</tr>
<tr>
<td>2</td>
<td>Pneumonia</td>
<td>8%</td>
<td>17%</td>
<td>7%</td>
<td>2%</td>
<td>—</td>
<td>16%</td>
<td>—</td>
<td>5%</td>
<td>—</td>
<td>—</td>
<td>18%</td>
<td>—</td>
<td>17%</td>
<td>6%</td>
<td>10.2%</td>
</tr>
<tr>
<td>3</td>
<td>Sudden cardiac death</td>
<td>—</td>
<td>—</td>
<td>14%</td>
<td>11%</td>
<td>—</td>
<td>9%</td>
<td>13%</td>
<td>5%</td>
<td>—</td>
<td>—</td>
<td>13%</td>
<td>6%</td>
<td>—</td>
<td>11%</td>
<td>7.0%</td>
</tr>
<tr>
<td>4</td>
<td>Cancer</td>
<td>11%</td>
<td>14%</td>
<td>7%</td>
<td>—</td>
<td>13%</td>
<td>—</td>
<td>3%</td>
<td>25%</td>
<td>50%</td>
<td>—</td>
<td>4%</td>
<td>—</td>
<td>2%</td>
<td>22%</td>
<td>6.3%</td>
</tr>
</tbody>
</table>
Discussion

Our data provide a contemporary overview of the causes of mortality in ACHD patients followed at a large, established supraregional center. In comparison with previous reports, a shift from perioperative death related to ACHD surgery to long-term cardiac and especially noncardiac mortality was evident. Moreover, long-term survival prospects of patients with simple, isolated congenital defects were found to be excellent and not statistically different from those expected.
is, however, similar to that seen in previous studies. It is likely that frequency of heart failure is increasing in ACHD patients, and—given the increasing complexity of disease as well as the growing incidence of comorbid conditions—more patients present with advanced forms of heart failure. On the other hand, progress in the management of advanced heart failure in ACHD has been slow and arguably unsatisfactory. The fact remains that standard heart failure therapy has still an unproven and possibly limited effect in this heterogeneous group of patients, whereas novel therapeutic options such as cardiac resynchronization therapy and assist systems have had a limited uptake so far. In contrast, sudden cardiac death rate was lower in the present study compared with previous reports, probably as a result of better risk stratification and more liberal use of implantable cardiac defibrillators in the current era.

The most remarkable finding, however, was the relatively large proportion of patients dying due to noncardiac complications, including cancer, cerebrovascular disease, infection, and pneumonia. This is consistent with previous data published by Khairy et al, Afilalo et al, and our group. Presumably as a consequence of the aging ACHD population the main causes of mortality are changing. Similar to these previous studies we could confirm that, with increasing age, the proportion of patients succumbing to myocardial infarction increases. However, we could not confirm that acute myocardial infarction was a leading cause of death, either in noncyanotic patients as a whole, or in a specific subgroup of patients. This is in contrast to a population-based US study, reporting acute myocardial infarction as the leading cause of death in elderly noncyanotic ACHD patients.

It is not surprising that survival prospects of ACHD patients are inferior to those observed in the general population. However, Figure 3 illustrates that especially Fontan, Eisenmenger syndrome, and complex CHD patients have greatly increased mortality rates. In contrast, simple defects were not found to fare significantly worse in terms of survival compared to the general population. We believe our findings are a testimony to the advance in the CHD field, represent the challenges ahead and may help to identify subgroups of patients where current and future research efforts need to be intensified.

Discussing life expectancy issues and short- to midterm risk of death with patients can be challenging. Beyond, obvious psychological barriers and anxiety associated with this.

![Standardized mortality ratio graph](image-url)

**Standardized mortality ratio**

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>SMR (95%CI)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>PDA</td>
<td>0.42 (0.10–1.78)</td>
<td>0.20</td>
</tr>
<tr>
<td>ASD</td>
<td>1.13 (0.86–1.48)</td>
<td>0.32</td>
</tr>
<tr>
<td>VSD</td>
<td>1.36 (0.82–2.27)</td>
<td>0.18</td>
</tr>
<tr>
<td>Valvar disease</td>
<td>1.39 (1.09–1.78)</td>
<td>0.002</td>
</tr>
<tr>
<td>Aortic Coarctation</td>
<td>1.73 (1.22–2.46)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>AVSD</td>
<td>1.86 (1.05–3.30)</td>
<td>0.014</td>
</tr>
<tr>
<td>Marfan syndrome</td>
<td>2.24 (1.41–3.57)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>2.34 (1.73–3.17)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>TGA arterial switch</td>
<td>2.61 (0.77–8.82)</td>
<td>0.08</td>
</tr>
<tr>
<td>Ebstein anomaly</td>
<td>3.30 (1.99–5.49)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Systemic RV</td>
<td>4.88 (3.33–7.16)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Eisenmenger syndrome</td>
<td>12.79 (9.67–16.91)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Complex CHD</td>
<td>14.13 (10.71–18.64)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Fontan–circulation</td>
<td>23.40 (15.97–34.29)</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>
The current report represents the first attempt to provide accurate estimates of long-term mortality in adults with congenital heart disease (ACHD) based on a large dataset of patients who were born with congenital heart disease.

### Strength of the Current Report

To the best of our knowledge, the current report represents the first attempt to provide accurate estimates of long-term mortality in adults with congenital heart disease (ACHD) based on a large dataset of patients who were born with congenital heart disease.

#### Sources of Funding

This project was supported by the National Institute of Health Research Cardiovascular Biomedical Research Unit at the Royal Brompton and Harefield NHS Foundation Trust and National Heart, Lung, and Blood Institute. Sonya V. Babu-Narayan is supported by an Intermediate Clinical Research Fellowship from the British Heart Foundation. Dr Kempny was supported by the Deutsche Herzstiftung e.V. Prof. Dr. M. Gatzoulis and the Adult Congenital Heart Center and National Center for Pulmonary Hypertension have received support from the Clinical Research Committee and the British Heart Foundation.

### Limitations

Because this represents a single-center retrospective study, the data on surgical mortality is cross validated with information from the United Kingdom Adult Congenital Heart Disease Society. Although the data is collected through comparison with our clinical database and especially for Pulmonary Hypertension patients, it may be a consequence of cardiac pulmonary congestion.

#### Estimating Mortality

The mortality data presented here are based on official death certificates complimented by published national registry database (6933 patients, 1979 deaths) and are therefore not subject to bias from ascertainment or misclassification. Compared with a previously published study, this approach may slightly reduce accuracy in reporting mortality for certain conditions.

#### Theoretical Advantage

This single-center study has a theoretical advantage of this single-center study is the consistency of the data collection and the ability to track patients over a long period of time.

#### Observations

Beyond, direct medical applications, there may also be a cognitive problem in understanding risks. One of the key deliverables of the current report is, therefore, the data on equivalent ages. The mortality in subgroups of patients compared with the general public, patients with complex, univentricular, or abnormal heart morphology, are afflicted by increased mortality compared with general population as they grow older. Highest mortality rates were observed for anterior aortic arch with subpulmonic stenosis, tetralogy of Fallot, and Eisenmenger syndrome. Our contemporary data, however, do not suggest a clear shift from perioperative to chronic cardiac mortality.

#### Conclusion

The current report confirms that ACHD patients continue to be at high risk of death and morbidity. Mortality estimates derived here are based on the current study and may be compared with previous studies investigating similar patient populations. The mortality rates among the patients stratified by diagnosis. Therefore, individual patients may exhibit different mortality, depending on additional factors and circumstances reporting specifically surgical mortality rates. However, it can be compared with previous studies investigating similar patient populations. This study provides the first robust estimate of mortality in ACHD patients that is compared with previous studies investigating similar patient populations. This study provides the first robust estimate of mortality in ACHD patients that is compared with previous studies investigating similar patient populations.

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### Table: Patient's age (years)

<table>
<thead>
<tr>
<th>Condition</th>
<th>20</th>
<th>25</th>
<th>30</th>
<th>35</th>
<th>40</th>
<th>45</th>
<th>50</th>
<th>55</th>
<th>60</th>
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<tbody>
<tr>
<td>ASD</td>
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<td>26</td>
<td>32</td>
<td>38</td>
<td>42</td>
<td>47</td>
<td>52</td>
<td>57</td>
<td>61</td>
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<tr>
<td>Valvar disease</td>
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<td>31</td>
<td>36</td>
<td>40</td>
<td>45</td>
<td>49</td>
<td>54</td>
<td>59</td>
<td>63</td>
</tr>
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<td>VSD</td>
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<td>30</td>
<td>36</td>
<td>40</td>
<td>44</td>
<td>49</td>
<td>53</td>
<td>59</td>
<td>63</td>
</tr>
<tr>
<td>Aortic Coarctation</td>
<td>32</td>
<td>33</td>
<td>38</td>
<td>43</td>
<td>47</td>
<td>52</td>
<td>56</td>
<td>62</td>
<td>66</td>
</tr>
<tr>
<td>AVSD</td>
<td>33</td>
<td>34</td>
<td>39</td>
<td>44</td>
<td>48</td>
<td>52</td>
<td>57</td>
<td>62</td>
<td>66</td>
</tr>
<tr>
<td>Marfan syndrome</td>
<td>37</td>
<td>38</td>
<td>42</td>
<td>46</td>
<td>50</td>
<td>54</td>
<td>59</td>
<td>64</td>
<td>68</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>37</td>
<td>38</td>
<td>42</td>
<td>47</td>
<td>50</td>
<td>54</td>
<td>60</td>
<td>65</td>
<td>69</td>
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<tr>
<td>Ebstein anomaly</td>
<td>42</td>
<td>43</td>
<td>47</td>
<td>51</td>
<td>54</td>
<td>59</td>
<td>63</td>
<td>68</td>
<td>72</td>
</tr>
<tr>
<td>Systemic RV</td>
<td>46</td>
<td>48</td>
<td>51</td>
<td>55</td>
<td>59</td>
<td>63</td>
<td>67</td>
<td>72</td>
<td>76</td>
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<tr>
<td>Eisenmenger syndrome</td>
<td>57</td>
<td>58</td>
<td>62</td>
<td>65</td>
<td>69</td>
<td>73</td>
<td>77</td>
<td>81</td>
<td>84</td>
</tr>
<tr>
<td>Complex CHD</td>
<td>58</td>
<td>59</td>
<td>63</td>
<td>67</td>
<td>70</td>
<td>74</td>
<td>78</td>
<td>82</td>
<td>85</td>
</tr>
<tr>
<td>Fontan</td>
<td>64</td>
<td>65</td>
<td>68</td>
<td>72</td>
<td>75</td>
<td>78</td>
<td>82</td>
<td>86</td>
<td>91</td>
</tr>
</tbody>
</table>

*Colors reflect the difference between the relative age and the actual age of patients. The curved line corresponds to the 5-year mortality in age-matched UK population. Numbers on the colored surface present the equivalent age—expressed as the age of the general UK population according to life table.*
Exercise Intolerance in Adult Congenital Heart Disease: Comparative Severity, Correlates, and Prognostic Implication

Gerhard-Paul Diller, Konstantinos Dimopoulos, Darlington Okonko, Wei Li, Sonya V. Babu-Narayan, Craig S. Broberg, Bengt Johansson, Beatriz Bouzas, Michael J. Mullen, Philip A. Poole-Wilson, Darrel P. Francis and Michael A. Gatzoulis
Oxygen consumption

Etiology of impaired exercise tolerance:
- Chronotropic Index
- Oxygen pulse VO2/HR

ACHD patients systematically severely overestimate their actual measured exercise capacity
Prognostic value of multiple biomarkers for cardiovascular mortality in adult congenital heart disease: comparisons of single-/two-ventricle physiology, and systemic morphologically right/left ventricles

Kenji Miyamoto¹ · Daiji Takeuchi¹ · Kei Inai¹ · Tokuko Shinohara¹ · Toshio Nakanishi¹

Heart Vessels. Feb 2016

Predictive biomarkers related to the NYHA classification and cardiothoracic ratio

<table>
<thead>
<tr>
<th>Overall group</th>
<th>Two ventricle</th>
<th>Single-Ventricle</th>
<th>Systemic left ventricle</th>
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<td>BNP</td>
<td>IL-6</td>
<td>BNP</td>
<td>BNP</td>
</tr>
<tr>
<td>ET-1</td>
<td>NE</td>
<td>ET-1</td>
<td>NE</td>
</tr>
<tr>
<td>sTNF-RI</td>
<td>hs-CRP</td>
<td></td>
<td>hs-CRP</td>
</tr>
<tr>
<td>NE</td>
<td>BNP</td>
<td></td>
<td>sTNF-RI</td>
</tr>
<tr>
<td>IL-6</td>
<td>ET-1</td>
<td></td>
<td>IL-6</td>
</tr>
</tbody>
</table>

K. Miyamoto and D. Takeuchi contributed equally to this work.

*Kenji Miyamoto
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1 Department of Pediatric Cardiology, Tokyo Women’s Medical University, 8-1, Kawada-cho, Shinjuku-ku, Tokyo 162-8666, Japan
Perioperative considerations

- Generalized Myopathy
- Cerebrovascular accidents
- Pregnancy
- Lung function
- Psychosocial Functionning
- Coronary Artery Disease
- Transcatheter Intervention
- Type of Cardiac Surgeon
- Heart transplant and Mechanical Circulatory Support
- Type of Anesthesiologist
- Cardiac Resynchronization Therapy
- Pediatric vs Adult Hospital

Adult Congenital Heart Disease

Risk factors for death in this study were "older" adults (25 to 49 years), male gender, higher surgical risk categories, and government-sponsored insurance. In a single-center report of ACHDs undergoing a more complex range of procedures performed by pediatric cardiothoracic surgeons, the overall mortality was 4.7% with risk factors that included chronic lung or liver dysfunction, prolonged CPB and aortic cross-clamp times, and higher postoperative inotrope scores.
Adult Congenital Heart Surgery: Adult or Pediatric Facility? Adult or Pediatric Surgeon?
Brian E. Kogon, MD, Courtney Plattner, BA, Traci Leong, PhD, Paul M. Kirshbom, MD, Kirk R. Kanter, MD, Mike McConnell, MD, and Wendy Book, MD
Divisions of Cardiothoracic Surgery and Cardiology, Emory University School of Medicine, Rollins School of Public Health, and Sibley Cardiology, Children’s Healthcare of Atlanta, Atlanta, Georgia

Adults or Big Kids: What Is the Ideal Clinical Environment for Management of Grown-Up Patients With Congenital Heart Disease?
Tara Karamlou, MD, Brian S. Diggs, PhD, Ross M. Ungerleider, MD, MBA, and Karl F. Welke, MD, MS
Divisions of Cardiothoracic Surgery and Surgery, Oregon Health and Science University, Portland, Oregon; Division of Pediatric Cardiothoracic Surgery, Department of Surgery, Case Western Reserve University, Cleveland, Ohio; and Mary Bridge/Swedish Pediatric Cardiothoracic Surgery Program, Mary Bridge Children’s Hospital and Health Center, Multicare Health System, Tacoma, Washington
Transition from Pediatric to Adults

- Financial and Insurance issues
- Insufficient Adult CHD-trained providers
- Pediatric vs. Adult Care Model

- Regional ACHD Centers
  - Complex and moderate lesions
  - Tertiary centers
  - Specialized & comprehensive services
Three parties, one direction: Research priorities in adults with congenital heart disease. What do professionals, patients and relatives want to know?☆

Paul C. Helm a,1,2, Marc-André Körten a,1,2, Hashim Abdul-Khaliq b,g,2, Boulos Asfour a,c,2, Helmut Baumgartner a,d,2, Günter Breithardt d,g,2, Deniz Kececioglu a,c,2, Christian Schlensak f,g,2, Gerhard-Paul Diller a,d,2, Ulrike M.M. Bauer a,g,⁎,2

Fontan circulation

![Fontan circulation chart](image-url)
everyday life) to be greater than their male counterparts. A similar prioritization is described by Goossens et al.\[10\]; their results are based on interviews primarily with female nursing staff members from the field of CHD. The main concern of these were issues such as patients’ disease related knowledge, the question of how to improve the care situation of CHD patients and, subsequently, quality of life of adult patients with CHD\[10\].

4.4. Leading research priorities

As to the question concerning which CHD related topics should become the particular focus of future research for the different CHD groups, those affected and physicians largely agree, although with different emphasis and, accordingly, ranking of single topics (Table 7).

The consensus of patients, relatives and physicians regarding the assessment of the need for research becomes especially apparent viewing the CHD group of “TGA after ASO”: Across all subgroups, the topics of “Vessel/valve related problems” and “Problems related to the coronary arteries” were listed among the top research topics. Both those affected and physicians concerned themselves with late complications associated with the arterial switch operation, as they are described in the literature\[11,12\]. The results suggest a great interest in studies on the long-term outcome of the arterial switch operation.

Regarding the CHD group of “Fontan circulation”, the topic of “Failing Fontan” is assigned an important role: this is a complication which to date has not been sufficiently investigated and which is associated with great difficulties in terms of treatment; due to lacking experience,

![Fig. 5.](image)

Overview of significant differences in perceived research needs between male and female physicians (higher mean levels indicates a higher importance). TGA/AS = transposition of the great arteries after atrial switch; TGA/ASO = transposition of the great arteries after arterial switch operation; TOF = tetralogy of Fallot.

<table>
<thead>
<tr>
<th>CHD group</th>
<th>Place</th>
<th>Patients</th>
<th>Relatives</th>
<th>Physicians</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fontan circulation</td>
<td>1</td>
<td>Catheter ablation rhythm disorders</td>
<td>Male: Sex, pregnancy Female: Diagnostic imaging</td>
<td>Male: Failing Fontan Female: Failing Fontan</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>Quality of life</td>
<td>Male: Quality of life Female: Cognitive capacity</td>
<td>Male: Quality of life Female: Quality of life</td>
</tr>
<tr>
<td></td>
<td>3</td>
<td>Diagnostic imaging</td>
<td>Male: Career choices, retirement Female: Sex, pregnancy</td>
<td>Male: Cognitive capacity Female: Career choices, retirement</td>
</tr>
<tr>
<td></td>
<td>4</td>
<td>Failing Fontan</td>
<td>Male: Diagnostic imaging Female: Heart failure/ intervention, reoperation</td>
<td>Male: Diagnostic imaging Female: Heart failure</td>
</tr>
<tr>
<td>TGA after AS</td>
<td>1</td>
<td>Career choices, retirement</td>
<td>Male: Catheter ablation, ICD, SCD Female: Diagnostic imaging</td>
<td>Male: Catheter ablation, ICD, SCD Female: Diagnostic imaging</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>Catheter ablation, ICD, SCD</td>
<td>Male: Catheter ablation, ICD, SCD Female: Heart failure/ intervention, reoperation</td>
<td>Male: Catheter ablation, ICD, SCD Female: Heart failure</td>
</tr>
<tr>
<td></td>
<td>3</td>
<td>Heart failure</td>
<td>Male: Cognitive capacity Female: Heart failure/ intervention, reoperation</td>
<td>Male: Diagnostic imaging Female: Heart failure</td>
</tr>
<tr>
<td>TGA after ASO</td>
<td>4</td>
<td>Cognitive capacity</td>
<td>Male: Heart failure Female: Diagnostic imaging</td>
<td>Male: Diagnostic imaging Female: Heart failure</td>
</tr>
<tr>
<td>Fontan circulation</td>
<td>1</td>
<td>Catheter ablation, ICD, SCD</td>
<td>Male: Heart failure Female: Vessel/valve related problems</td>
<td>Male: Vessel/valve related problems Female: Heart failure</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>Vessel/valve related problems</td>
<td>Male: Catheter ablation, ICD, SCD Female: Intervention, reoperation</td>
<td>Male: Problems related to coronary arteries Female: Intervention, reoperation</td>
</tr>
<tr>
<td></td>
<td>3</td>
<td>Problems related to coronary arteries</td>
<td>Male: Sex, pregnancy Female: Problems related to coronary arteries</td>
<td>Male: Cognitive capacity Female: Problems related to coronary arteries</td>
</tr>
<tr>
<td></td>
<td>4</td>
<td>Diagnostic imaging</td>
<td>Male: Problems related to coronary arteries Female: Cognitive capacity</td>
<td>Male: Problems related to coronary arteries Female: Cognitive capacity</td>
</tr>
<tr>
<td>TOF</td>
<td>1</td>
<td>Intervention, reoperation</td>
<td>Male: Catheter ablation, ICD, SCD Female: Intervention, reoperation</td>
<td>Male: Intervention, reoperation Female: Heart failure</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>Catheter ablation, ICD, SCD</td>
<td>Male: Catheter ablation, ICD, SCD Female: Intervention, reoperation</td>
<td>Male: Intervention, reoperation Female: Heart failure</td>
</tr>
<tr>
<td></td>
<td>3</td>
<td>Diagnostic imaging</td>
<td>Male: Diagnostic imaging Female: Heart failure</td>
<td>Male: Diagnostic imaging Female: Heart failure</td>
</tr>
<tr>
<td></td>
<td>4</td>
<td>Heart failure</td>
<td>Male: Heart failure Female: Quality of life</td>
<td>Male: Quality of life Female: Diagnostic imaging</td>
</tr>
</tbody>
</table>

\[a\] Shared place with equal ranking; ICD = implantable cardioverter defibrillator, SCD = sudden cardiac death; VA = ventriculo-arterial; TGA after AS = transposition of the great arteries after atrial switch; TGA after ASO = transposition of the great arteries after arterial switch operation; TOF = tetralogy of Fallot.
Physical activity counseling should be part of every patient interaction, whether or not the patient’s clinical status justifies activity restrictions.

### Table 5. Exercise Risk Factors Evaluated During a Cardiopulmonary Exercise Test

1. Abnormal hemodynamic response to exercise: Either a progressive drop in blood pressure (a normal response is an increase in systolic pressure and little or no change in diastolic pressure) or excessive rise in blood pressure (>250 mm Hg systolic), especially when it results in symptoms. Patients with coarctation of the aorta, repaired or unrepaired, may have normal resting right arm blood pressure but significantly elevated pressure during exercise.85
2. Abnormal heart rate profile: A blunted peak heart rate, lower heart rate reserve, and blunted heart rate recovery are associated with poorer overall prognosis in various forms of congenital heart disease.86
3. Arrhythmias: Supraventricular and ventricular tachycardia, ventricular fibrillation, and all forms of heart block can manifest during exercise testing.
4. Myocardial ischemia: On electrocardiography during exercise, significant ST depression of ≥2 mm can signify ischemia induced by the extra metabolic demands of exercise.
5. Reduced ventilatory efficiency: An increase in the $V_{E}/V_{CO_2}$ slope is associated with heart failure and an increased risk of sudden death in some congenital heart disease populations.87,88

$V_{CO_2}$ indicates carbon dioxide production; and $V_{E}$, minute ventilation.
Risks and Benefits of Exercise Training in Adults With Congenital Heart Disease

Marie-A Chaix, MD, MS, François Marcotte, MD, Annie Dore, MD, François-Pierre Mongeon, MD, MS, Blandine Mondésert, MD, Lise-Andrée Mercier, MD, and Paul Khairy, MD, PhD

Adult Congenital Heart Center, Montreal Heart Institute, Université de Montréal, Montreal, Quebec, Canada

Benefits:
• Improved health
• Improved exercise tolerance: VO2 max, peak oxygen consumption, peak oxygen pulse, heart rate recovery, walking distance and treadmill time.
• Quality of life
• Prevention of acquired coronary artery disease
• Reduction of depressive symptoms

Risks:
• Exercise induced ventricular tachyarrhythmia.
• Nature of the congenital malformation
• Underlying physiology
• Comorbidities
• Type and intensity of exercise

Eligibility and Disqualification
Recommendations for Competitive Athletes
With Cardiovascular Abnormalities:
Task Force 4: Congenital Heart Disease

A Scientific Statement From the American Heart Association and American College of Cardiology

- Consideration of both the training and the competitive aspects of the activity
- Patient’s functional status and history of surgery.
- Noninvasive testing (exercise testing, Holter monitoring, echocardiography, and cardiac magnetic resonance imaging)
Meeting the challenge: The evolving global landscape of adult congenital heart disease

Aleksander Kempny a,*, Rodrigo Fernández-Jiménez a, Oktay Tutarel a, Konstantinos Dimopoulos a,b, Anselm Uebing a, Yumi Shiina a, Rafael Alonso-Gonzalez a, Wei Li a, Lorna Swan a,b, Helmut Baumgartner c, Michael A Gatzoulis a,b, Gerhard-Paul Diller a,b,c

The Future

- Emergence of multicenter research networks and registries, particularly in North America and Europe-new opportunities for research coordination and grant funding.
- Developed countries devise and implement sustainable strategies beyond intermittent missions
- Expansion in training resources and development of multidisciplinary teams.
Thank you