Survival Prospects and Circumstances of Death in Contemporary Adult Congenital Heart Disease Patients Under Follow-Up at a Large Tertiary Centre

Gerhard-Paul Diller, MD, MSc, PhD, MBA*; Aleksander Kempny, MD*; Rafael Alonso-Gonzalez, MD, MSc; Lorna Swan, MD, FRCP; Anselm Uebing, MD, PhD; Wei Li, MD, PhD; Sonya Babu-Narayan MB, BS, BSc, MRCP, PhD; Stephen J. Wort, PhD; Konstantinos Dimopoulos, MD, MSc, PhD; Michael A. Gatzoulis, MD, PhD

Background—Adult congenital heart disease (ACHD) patients have ongoing morbidity and reduced long-term survival. Recently, the importance of specialized follow-up at tertiary ACHD centers has been highlighted. We aimed to assess survival prospects and clarify causes of death in a large cohort of patients at a single, tertiary center.

Methods and Results—We included 6969 adult patients (age 29.9±15.4 years) under follow-up at our institution between 1991 and 2013. Causes of death were ascertained from official death certificates. Survival was compared with the expected survival in the general age- and sex-matched population, and standardized mortality rates were calculated. Over a median follow-up time of 9.1 years (interquartile range, 5.2–14.5), 524 patients died. Leading causes of death were chronic heart failure (42%), pneumonia (10%), sudden-cardiac death (7%), cancer (6%), and hemorrhage (5%), whereas perioperative mortality was comparatively low. Isolated simple defects exhibited mortality rates similar to those in the general population, whereas patients with Eisenmenger syndrome, complex congenital heart disease, and Fontan physiology had much poorer long-term survival (P<0.0001 for all). The probability of cardiac death decreased with increasing patient’s age, whereas the proportion of patients dying from noncardiac causes, such as cancer, increased.

Conclusions—ACHD patients continue to be afflicted by increased mortality in comparison with the general population as they grow older. Highest mortality rates were observed among patients with complex ACHD, Fontan physiology, and Eisenmenger syndrome. Our data provide an overview of causes of mortality and especially the spectrum of noncardiac causes of death in contemporary ACHD patients. (Circulation. 2015;132:2118-2125. DOI: 10.1161/CIRCULATIONAHA.115.017202.)

Key words: heart defects, congenital ■ heart failure ■ mortality ■ sudden cardiac death ■ survival

Life expectancy of patients born with congenital heart disease (CHD) has improved dramatically over the past few decades.1 In fact, >90% of these patients are now expected to survive to adulthood.2 This has led to the development of a large and growing population of adults with congenital heart disease (ACHD). Despite the surgical, interventional, and medical advancements, these patients are not cured and require life-long specialized health care. Beyond the obvious ongoing morbidity, including cardiac symptoms, reduced exercise capacity, and the need for electrophysiological, interventional, or surgical procedures, mortality is increased in this population of patients with chronic cardiac disease.3

Clinical Perspective on p 2125

Previous studies have investigated the long-term mortality of various ACHD cohorts and have delineated causes of death in this population.1-6 Because of ongoing improvement of care, survival prospects of adults with congenital heart disease are likely to have changed over recent decades. Khairy et al7 demonstrated that mortality in patients with congenital heart disease has shifted away from infants and toward adults, with a steady increase in age at death. The current study was designed to evaluate a contemporary ACHD cohort from a single tertiary center and attempt comparison with data from
Patients and Methods

We retrospectively reviewed data on all adult patients with congenital heart disease under active follow-up at the Royal Brompton Hospital, London between 1991 and 2014. For the scope of this study we defined the start of adulthood as age ≥16. Patients were divided into subgroups based on the major underlying heart defect. Patients were grouped by primary diagnosis into major ACHD groups. Those with combinations of lesions were assigned to the lesion of highest complexity. Patients with complex cardiac anatomy (comprising mainly patients with single-ventricle physiology) without Eisenmenger syndrome and without Fontan palliation were coded as ‘complex CHD.’ Patients with (generally isolated) congenital valvar lesions were coded as ‘valvar disease.’ Data on clinical status were obtained from medical records. Data on overall mortality were retrieved from the Office for National Statistics, which registers all United Kingdom deaths. The cause of death was established from medical records. Data on clinical status were obtained from medical records. Data on death certificates, available for all patients, by 1 investigator (G-P.D.). Where the likely immediate cause of mortality remained unclear, the case was discussed with 1 of the coprincipal investigators (A.K.) and consensus was reached. In addition, the records of the deceased patients were cross checked with data from the local surgical and interventional audit database to ascertain that no perioperative death was missed. Because this was a retrospective analysis based on data collected for routine clinical care and administrative purposes (UK National Research Ethics Service guidance), individual informed consent was not required. The study was locally registered and approved.

Statistical Analysis

Continuous variables are presented as mean±standard deviation or median and interquartile range, whereas categorical variables are presented as number (percentage). The association between various causes of mortality and age was assessed using a nonparametric competing risk survival model accounting for left truncation. To this end the cumulative incidence of death in the presence of competing risk was estimated as described by Coviello and Boggess using the stcompet package for Stata (Version IC 12.1, College Station, TX), and cumulative incidence plots with 95% confidence intervals were produced. This statistical approach models simultaneously the risk of multiple competing outcomes including heart failure, sudden cardiac death, acute myocardial infarction, cardiac surgery and interventions, cancer, or pneumonia-related mortality in our study. To estimate standardized mortality ratios (SMRs) compared with an age- and sex-matched sample of the general population the method reported by Finkelstein et al was used. Survival was compared with that predicted for an age- and sex-matched healthy cohort of UK residents using life table data (2007–2009 interim life tables) published by the Government Actuary’s Department (http://www.gad.gov.uk), as previously described. Equivalent age was defined as the age of UK population with the most similar 5-year mortality (ie, minimal sum of absolute differences). Statistical analyses were performed using R-package version 3.0.2. A 2-sided P-value of <0.05 was considered indicative of statistical significance.

Results

Demographics and Mortality

We included 6969 patients (49.9% females) under active follow-up at our institution as illustrated in Table 1. The mean age at baseline was 29.9±15.4 years. Overall, 69%, 26%, and 5% of patients were in the New York Health Association (NYHA) functional class I, II, and III/IV, respectively. According to the Bethesda disease complexity classification 52% of patients had simple defects, 33% moderate, and 15% great complexity defects.

During a median follow-up time of 9.1 years (interquartile range, 5.2–14.5; corresponding to a total of 70967 patient-years), 524 (7.7%) patients died yielding a mortality rate of 0.72%/patient-year.

The majority of patients (429; 81.9%) died outside hospital, whereas the remainder died in our institution or within 24 h from discharge. Death occurred after an elective or emergency procedure.
cardiac operation in 25 patients (Fontan-revision/conversion related surgery in 6, tricuspid valve surgery in 5, pulmonary valve replacement in 4, aortic surgery in 4, and other/complex surgery in 5). In addition, 1 Eisenmenger patient died early after heart-lung transplantation, whereas 4 patients succumbed to complications related to cardiac interventional procedures.

Table 2 provides an overview of the causes of death in this population. It illustrates that the leading cause of mortality in our cohort was chronic cardiac failure, followed by pneumonia and sudden cardiac death. Remarkably, cardiac surgery/cardiac intervention related mortality ranked only 5th in this statistic, after pneumonia and cancer. The same table also demonstrates the relatively high proportion of patients dying from noncardiac causes such as cancer, hemorrhage (56% cerebral, 19% pulmonary, 11% gastrointestinal), infection, or cerebrovascular events. In addition, we provide the percentage of patients dying because of aortic dissection or hepatic failure, both recognized causes of mortality in selected subgroups of patients with CHD.

With increasing patient age, the proportion of patients dying because of cardiac reasons (despite the inclusion of acute myocardial infarction) decreased and, by implication, proportionally more patients died because of competing noncardiac causes (Figure 1A). This association was especially evident in patients with simple defects (Figure 1B). In addition, Figure 2A shows the association between increasing age and cancer- or pneumonia-related deaths based on the results of a competing risk survival analysis. Regarding reasons for cardiac death, especially the risk of heart failure–related death and that of acute myocardial infarction–related mortality, increased with age (Figure 2B).

Survival in the entire ACHD cohort was significantly worse compared with the expected mortality for an age and gender matched sample from the general UK population (SMR, 2.29; 95% CI, 2.08–2.53; Logrank P<0.0001). There were, however, significant differences in mortality between subgroups of patients (Logrank P<0.0001; see Figure 3 and Figure I in the online-only Data Supplement). The SMR was highest in patients with Fontan circulation (SMR, 23.40; 95% CI, 15.97–34.29; P<0.0001), complex CHD (SMR, 14.13; 95% CI, 10.71–18.64; P<0.0001), and Eisenmenger syndrome (SMR, 12.79; 95% CI, 9.67–16.91; P<0.0001). In contrast, no significant difference in mortality was present in patients with ductus arteriosus and atrial or ventricular septal defects when compared with the general UK population (P>0.05, for all). The SMRs based on the Bethesda classification11 were 1.3 (95% CI, 1.1–1.5), 2.2 (95% CI, 1.8–2.3), and 10.9 (95% CI, 9.3–12.8) for patients with simple, moderate, and great complexity heart defects (P<0.001 for all). In addition, NYHA functional class was associated with prognosis for the overall cohort. The SMR increased from 1.6 (95% CI, 1.3–1.9; P<0.0001) for class 1, to 3.6 (95% CI, 3.0–4.2; P<0.0001) for class 2 and 4.6 (95% CI, 3.6–6.0; P<0.0001) for class 3 or 4.

Based on the fitted SMR models we calculated predicted 5-year risk of death for each diagnostic subgroup for hypothetical 40-year-old patients with CHD. These mortality risks were compared with the projected risk of the general population to obtain an 'equivalent age' with regards to mortality risk for each ACHD subgroup (Figure 4). For example, a 40-year-old average patient with Fontan physiology from our cohort had a 5-year risk of death (18.0%; 95% CI, 11.9–24.6%) comparable with that of a 75-year-old person without CHD. In addition, Figure 5 illustrates the equivalent age for the different diagnostic groups and

Table 2. Distribution of Causes of Death in Different Diagnostic Subgroups and Overall

<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</th>
<th>arterial switch</th>
<th>Valvular disease</th>
<th>VSD</th>
<th>All patients</th>
</tr>
</thead>
<tbody>
<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>
<td></td>
<td></td>
<td></td>
<td></td>
<td>40%</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>18%</td>
<td>—</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>17%</td>
</tr>
<tr>
<td>3</td>
<td>Sudden cardiac death</td>
<td>—</td>
<td>—</td>
<td>14%</td>
<td>11%</td>
<td>9%</td>
<td>13%</td>
<td>5%</td>
<td>13%</td>
<td>6%</td>
<td>33%</td>
<td>6%</td>
<td></td>
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<td></td>
<td></td>
<td>3%</td>
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<tr>
<td>4</td>
<td>Cancer</td>
<td>11%</td>
<td>14%</td>
<td>7%</td>
<td>—</td>
<td>13%</td>
<td>3%</td>
<td>25%</td>
<td>50%</td>
<td>—</td>
<td>4%</td>
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<td></td>
<td></td>
<td>2%</td>
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<tr>
<td>5</td>
<td>Cardiac Surgery/Intervention (peri-op)</td>
<td>11%</td>
<td>2%</td>
<td>7%</td>
<td>5%</td>
<td>19%</td>
<td>2%</td>
<td>19%</td>
<td>5%</td>
<td>9%</td>
<td>12%</td>
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<td></td>
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<td></td>
<td>1%</td>
</tr>
<tr>
<td>6</td>
<td>Sepsis/infection</td>
<td>8%</td>
<td>6%</td>
<td>—</td>
<td>8%</td>
<td>9%</td>
<td>3%</td>
<td>5%</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>7%</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>11%</td>
</tr>
<tr>
<td>7</td>
<td>Cerebrovascular</td>
<td>8%</td>
<td>8%</td>
<td>7%</td>
<td>6%</td>
<td>6%</td>
<td>3%</td>
<td>3%</td>
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<tr>
<td>8</td>
<td>Acute myocardial infarction</td>
<td>6%</td>
<td>5%</td>
<td>—</td>
<td>3%</td>
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<td>—</td>
<td>3%</td>
<td>8%</td>
<td>33%</td>
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</tr>
<tr>
<td>9</td>
<td>Endocarditis</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>2%</td>
<td>6%</td>
<td>2%</td>
<td>10%</td>
<td>4%</td>
<td>—</td>
<td>—</td>
<td>1%</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>1.6%</td>
</tr>
<tr>
<td>10</td>
<td>Aortic dissection</td>
<td>11%</td>
<td>2%</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>5%</td>
<td>—</td>
<td>1%</td>
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<td></td>
<td></td>
<td></td>
<td></td>
<td>1.4%</td>
</tr>
<tr>
<td></td>
<td>Hepatic failure</td>
<td>—</td>
<td>5%</td>
<td>—</td>
<td>6%</td>
<td>—</td>
<td>3%</td>
<td>—</td>
<td>3%</td>
<td>—</td>
<td>—</td>
<td>1%</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>1.4%</td>
</tr>
</tbody>
</table>

The ‘Cardiac-Surgery/peri-op’ row provides information on mortality related specifically to adult congenital heart surgery. ASD indicates atrial septal defect; AVSD, atrioventricular septal defect; CHD, congenital heart disease; PDA, patent ductus arteriosus; RV, right ventricle; TGA, transposition of the great arteries; and VSD, ventricular septal defect.
Discussion
Our data provide a contemporary overview over 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 in various ages in comparison to that observed in persons without congenital heart disease.
in the general UK population. The slightly elevated overall risk of death in patients with simple defects, as a group, is likely related to the inclusion of patients with valvar defects in this group. In contrast, patients with uncorrected, palliated, complex, or cyanotic underlying heart defects continue to be afflicted by substantial mortality. In addition, mortality rates in various diagnostic subgroups were compared with the mortality observed in the general population. To illustrate survival prospects, we introduce the concept of equivalent age. We contend that this may aid counseling of patients by projecting mortality risks for individual diagnostic subgroups compared to what is naturally expected at older age.

Previous studies have investigated primary causes of mortality in ACHD patients; these studies were different from the present report either because they referred to historical ACHD cohorts or because they represented registry studies including patients followed-up at numerous institutions. Oechslin and Connolly have described the circumstances of death in ACHD patients under follow-up at 2 large supraregional Canadian and US centers (Toronto and Mayo Clinic) in the 1980s and early 1990s, respectively. They reported a perioperative mortality of 18% and 37.7%, in what are now historic cohorts. These mortality rates were largely consistent with the proportion of patients dying perioperatively (26.3%) reported by Nieminen et al as part of a population-based Finnish study (albeit the latter study included also initial repair in children). In contrast, our data suggest that the focus of ACHD mortality has nowadays shifted to long-term cardiac and non-cardiac complications of the disease. Moreover, the perioperative ACHD mortality reported here is even lower compared with results from a recent Dutch national registry (7.1% perioperative deaths between 2002 and 2008), supporting the role of concentrating care at tertiary ACHD centres. The proportion of patients dying from heart failure in our study

![Figure 2. A, Cumulative incidence of pneumonia and cancer death with 95% confidence intervals based on the results of competing risk model. B, Cumulative incidence of various causes of cardiac mortality with 95% confidence intervals based on the results of competing risk model. AMI indicates acute myocardial infarction.](http://circ.ahajournals.org/).
is, however, similar to that seen in previous studies.\textsuperscript{4,5,13} It is likely that frequency of heart failure is increasing in ACHD patients,\textsuperscript{15} 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,\textsuperscript{16–18} 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\textsuperscript{19–21} and more liberal use of implantable cardiac defibrillators in the current era.\textsuperscript{7} 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,\textsuperscript{7} Afilalo et al,\textsuperscript{22} and our group.\textsuperscript{23} 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.\textsuperscript{24}

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

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**Figure 3.** Standardized mortality ratios (SMR) in various subgroups of patients. Points present the SMR, and horizontal lines the 95% confidence interval range. An SMR of 1 suggests that patients have comparable mortality as a sex- and age-matched sample from the general population. ASD indicates atrial septal defect; AVSD, atrioventricular septal defect; CHD, congenital heart disease; PDA, patent ductus arteriosus; RV, right ventricle; TGA, transposition of the great arteries; and VSD, ventricular septal defect.

**Figure 4.** Projected 5-year mortality rates for 40-year-old ACHD patients compared with that expected for the general UK population based on the results of the standardized mortality ratio (SMR) analysis. Points present the estimated mortality within 5 years (on the x axis) and also indicate the equivalent age—expressed as the age of subgroup of UK population with the most similar 5-years mortality (y axis). Red lines represent 95% confidence intervals for the 5-year mortality. The black curve presents 5-year mortality for the UK-population based on life table data. ASD indicates atrial septal defect; AVSD, atrioventricular septal defect; RV, right ventricle; and VSD, ventricular septal defect.
Figure 5. Mortality in subgroups of patients compared with mortality in age-matched UK population. Numbers on the colored surface present the equivalent age—expressed as the age of subgroup of UK population, having similar 5-year mortality rates. Colors reflect the difference between the relative age and the actual age of patients. The curved line corresponds to the 5-year risk of death in the general UK population according to life table data. AS indicates atrial septal defect; AVSD, atrioventricular septal defect; CHD, congenital heart disease; RV, right ventricle; and VSD, ventricular septal defect.

Table 5. Mortality in subgroups of patients compared with mortality in age-matched UK population. Numbers on the colored surface present the equivalent age, expressed as the age of subgroup of UK population, having similar 5-year mortality rates. Colors reflect the difference between the relative age and the actual age of patients. The curved line corresponds to the 5-year risk of death in the general UK population according to life table data.

<table>
<thead>
<tr>
<th>Patient’s age (years)</th>
<th>Age difference:</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>&gt;40</td>
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<tr>
<td></td>
<td>30-40</td>
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<td></td>
<td>20-30</td>
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<td></td>
<td>10-20</td>
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<td>5-10</td>
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<td></td>
<td>3-5</td>
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<td></td>
<td>&lt;2</td>
</tr>
</tbody>
</table>

Strength of the Current Report

To the best of our knowledge, the current report represents the largest single center study assessing the causes of mortality in contemporary patients (70967 patient-years versus 25,900 patient years in a previous nationwide registry study). This is explained by the relatively long history and the well-established nature of our center. Compared with a previously published national registry database (6933 patients, 197 deceased) and a pan-European registry study (4110 patients, 115 deceased), a group of 524 deceased patients formed the statistical basis of the current report. In addition, unlike registry data we had access to the entire medical/surgical database of the patients and could clarify equivocal information based on original medical records. This approach has been described to improve data quality and reliability of mortality data in the setting of ACHD. The mortality data presented here are based on official death certificates complimented by additional information available to us and should, therefore, provide robust estimates of the causes of mortality. A further theoretical advantage of this single center study is the consistent approach with a shared diagnostic and therapeutic strategy used over time.

Limitations

Because this represents a single-center retrospective study, the sample of patients included may not necessarily represent the pattern of ACHD patients present in the community. Studying long-term outcomes of community-based ACHD patients is, therefore, a recognized strength of registry-based studies. Like all similar studies, the distinction between the primary cause of death is not always unequivocal (e.g., pneumonia, which may be a consequence of cardiac pulmonary congestion). However, all causes of deaths were checked for plausibility through comparison with our clinical database and especially the data on surgical mortality is cross validated with information from our clinical/official surgical audit to improve data quality and minimize the number of patients with death due to unspecified reasons. The proportion of patients dying perioperatively is not equivalent to surgical mortality. Formally, the former is a function of surgical mortality, competing risks of deaths, and the number of operations performed. Therefore, this parameter cannot be compared directly with other studies reporting specifically surgical mortality rates. However, it can be compared with previous studies investigating circumstances of death in ACHD patients, in general, using the same metric. Estimates of mortality provided are for average patients stratified by diagnosis. Therefore, individual patients may exhibit different mortality, depending on additional factors specific to the patient. Furthermore, the average age and median age at death was different between the diagnostic groups. Especially for the young patients with arterial switch TGA it may be premature to extrapolate to mortality risks at older age and these data should, therefore, interpreted with caution.

Conclusions

The current report confirms that ACHD patients continue to be affected by increased mortality compared with general population as they grow older. Highest mortality rates were observed among patients with complex ACHD, Fontan physiology, and Eisenmenger syndrome. Our contemporary data show a clear shift from perioperative to chronic cardiac mortality and noncardiac death.

Sources of Funding

Dr Kempny was supported by the Deutsche Herzstiftung e.V. Prof 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. This project was supported by the National Institute of Health Research cardiovascular Biomedical Research Unit at the Royal Brompton and Harefield National Health Service Foundation Trust and Imperial College London. Sonya V. Babu-Narayan is supported by an Intermediate Clinical Research Fellowship from the British Heart Foundation (FS/11/38/28864).

Disclosures

None.
Adult congenital heart disease (ACHD) patients have ongoing morbidity and reduced long-term survival. Based on a large sample of patients under follow-up at an established supraregional ACHD center we provide data on contemporary survival of a sample to that of a standard population.

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Supplemental Figure 1

Kaplan Meier survival curves compared to expected mortality of an age and gender matched sample from the general UK population stratified by diagnostic group. SMR = standardized mortality rates.