Mortality in First 5 Years in Infants With Functional Single Ventricle Born in Texas, 1996 to 2003

David E. Fixler, MD, MSc; Wendy N. Nembhard, PhD; Jason L. Salemi, MPH; Mary K. Ethen, MPH; Mark A. Canfield, PhD

Background—Infants with functional single ventricle have a high risk of death during the early years of life. Studies have reported improvement in postoperative survival, but they do not include preoperative deaths or those occurring before transfer. The purpose of this population-based study was to estimate 5-year survival in infants with functional single ventricle, to define factors associated with survival, and to estimate improvement in outcome.

Methods and Results—Patients with hypoplastic left heart syndrome, pulmonary atresia intact ventricular septum, single ventricle, and tricuspid atresia born in 1996 to 2003 were identified from the Texas Birth Defects Registry and linked to state and national birth and death vital records. We examined the effects of defect type, birth era, birth weight, gestational age, maternal race/ethnicity, extracardiac anomalies, sex, and maternal age and education on survival. Five-year survival varied by defect type: hypoplastic left heart syndrome, 38.0% (95% confidence interval, 32.6 to 43.5); single ventricle, 56.1% (95% confidence interval, 49.9 to 61.7); pulmonary atresia intact ventricular septum, 55.7% (95% confidence interval, 45.8 to 64.4); and tricuspid atresia, 74.6% (95% confidence interval, 62.4 to 83.4). The presence of extracardiac defects increased the adjusted risk of death by 84%. Non-Hispanic blacks had an adjusted risk of death that was 41% higher than that for non-Hispanic whites, and Hispanics had a 26% higher risk. Patients born in 2001 to 2003 had a 47% lower risk than those born in 1996 to 2000.

Conclusions—This population-based study demonstrates significant improvement in overall 5-year survival, particularly in cases of hypoplastic left heart syndrome and single ventricle. Additional studies are needed to determine the factors causing racial/ethnic and regional differences in outcome. (Circulation. 2010;121:644-650.)

Key Words: epidemiology | heart defects, congenital | survival | single ventricle

Advances in clinical care by pediatric cardiologists and cardiac surgeons have improved the outcome of most infants with congenital heart defects.1–3 Several tertiary cardiac centers have reported, from their own surgical experience, improvement in survival for specific single ventricle defects.4–7 However, such center-specific data may not be applicable to all centers and do not include patients born in regional hospitals who died before transfer to the tertiary center. In 2001, Boneva et al8 examined population-based data of mortality from congenital heart defects for the United States from 1979 to 1997 using the National Center for Health Statistics multiple-cause mortality files. Looking at temporal trends, they found that for most anomalies the median age of death increased considerably during this 19-year time period. Nevertheless, patients with hypoplastic left heart syndrome (HLHS) and single ventricle did not show evidence for improvement in outcome. In 2001, Nembhard et al8 reported first-year survival among infants with congenital anomalies in Texas born between 1995 and 1997. This population-based registry study showed that 1-year survival was lower for HLHS (20.9%) than for all other birth defects studied except anencephaly. In 2003, Cleves et al9 reported first-year survival of infants born with congenital heart defects in Arkansas using a data registry of births between 1993 and 1998. They found that first-year survival was low for HLHS (22.6%) and for pulmonary atresia with intact ventricular septum (PA-IVS; 50%), a form of hypoplastic right ventricle. However, no population-based study has been reported on 5-year survival for infants born after 2001 with various forms of functional single ventricle.

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The purpose of this study was to describe survival during the first 5 years of life in infants born with functional single ventricles, namely HLHS, PA-IVS, single ventricle complex (SV), and tricuspid atresia (TA). During the neonatal period, such patients are considered potential candidates for single ventricle surgical palliation or heart transplantation. The outcome from these surgical approaches can best be demonstrated

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by survival beyond the first year of life. The specific aims of this study were to estimate temporal trends in survival that may reflect improvement in medical and surgical care, to estimate hazard ratios (HRs) for these 4 lesions compared with simple d-transposition (d-TGA) case survival, and to estimate the effect of extracardiac defects, gestational age, birth weight, and maternal race/ethnicity, age, and education on survival in these high-risk infants.

Methods

Infants with congenital heart disease were identified by the Texas Birth Defects Registry, which is maintained by the Birth Defects Epidemiology and Surveillance Branch of the Texas Department of State Health Services. This population-based registry collects information on major structural and chromosomal anomalies. During the period of study, the registry covered 35% of Texas resident live births in 1997, 85% in 1998, and 100% in 1999 to 2003. The staff of this active surveillance system visit all delivery units, pediatric hospitals, birthing centers, and midwife personnel in the state to review medical records of all pregnancy outcomes. If an infant or fetus has a birth defect that was monitored by the registry, the mother is a Texas resident at the time of delivery, and the birth defect is diagnosed prenatally or within 1 year after delivery, registry staff abstract detailed demographic and diagnostic information and enter that information into the registry. In cases when records had discrepant diagnoses, priority for reliability was as follows: noncardiologist note < cardiologist note < echocardiography report < surgery report. Data were not collected relative to timing or types of surgical procedures. To provide additional information, registry cases are linked to their birth or fetal death certificates through the use of the distribution of cases by maternal race/ethnicity showed a significant difference in mortality. An HR was considered statistically significantly different from 1 if its 95% confidence interval (CI) did not include 1. For each factor, a proportional-hazards model was used to estimate the adjusted effect of each factor on survival. Only variables with statistically significant findings (P < 0.05) in the unadjusted (crude) analyses were included in the multivariable analysis. To determine whether the increase in survival was significant for each functional single ventricle subtype and for each race/ethnic group, a crude proportional-hazards model was used to compare survival in the other diagnostic groups. Although the distribution of cases by maternal race/ethnicity showed a relatively low number of patients born to NH black mothers (only 9.0%), the distribution was similar to their frequency of 9.6% among 50 268 infants in the Texas Birth Defects Registry born during the same time period. In patients having a functional

Table 1. Distribution of Cases of Selected Congenital Heart Defects by Maternal Race/Ethnicity, Texas Birth Defects Registry, 1996 to 2003

<table>
<thead>
<tr>
<th>Defect</th>
<th>Total, n</th>
<th>NH White, n (%)</th>
<th>NH Black, n (%)</th>
<th>Hispanic, n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>HLHS</td>
<td>311</td>
<td>148 (47.6)</td>
<td>29 (9.3)</td>
<td>134 (43.1)</td>
</tr>
<tr>
<td>SV</td>
<td>286</td>
<td>99 (34.6)</td>
<td>29 (10.1)</td>
<td>158 (55.2)</td>
</tr>
<tr>
<td>PA-IVS</td>
<td>118</td>
<td>45 (38.1)</td>
<td>13 (11.0)</td>
<td>60 (50.9)</td>
</tr>
<tr>
<td>TA</td>
<td>67</td>
<td>17 (25.4)</td>
<td>11 (16.5)</td>
<td>39 (58.2)</td>
</tr>
<tr>
<td>d-TGA</td>
<td>225</td>
<td>111 (49.3)</td>
<td>9 (4.0)</td>
<td>105 (46.7)</td>
</tr>
<tr>
<td>Total</td>
<td>1007</td>
<td>420 (41.7)</td>
<td>91 (9.0)</td>
<td>496 (49.3)</td>
</tr>
</tbody>
</table>

Results

Table 1 shows the race/ethnic characteristics of the study group by specific diagnoses. The total number of cases was 1007, of which 782 had functional single ventricle. The 225 cases of d-TGA were included as a referent group to compare their survival with survival in the other diagnostic groups. Although the distribution of cases by maternal race/ethnicity showed a relatively low number of patients born to NH black mothers (only 9.0%), the distribution was similar to their frequency of 9.6% among 50 268 infants in the Texas Birth Defects Registry born during the same time period. In patients having a functional...
single ventricle, major extracardiac defects were reported in 17.1%. Of these extracardiac defects, obstructive uropathy accounted for 23.7%, microcephaly for 8.7%, renal agenesis/dysgenesis for 7.3%, diaphragmatic hernia for 5.5%, pulmonary hypoplasia/agenesis for 4.6%, cleft lip/palate for 4.6%, and tracheoesophageal fistula for 4.1%. Table 2 shows the impact of major extracardiac defects on 1-, 3-, and 4-year survival. Crude 5-year survival among isolated cases was lowest for HLHS (40.9%). When associated with a major extracardiac defect, survival was only 20.8%. In cases of SV, the presence of an extracardiac defect reduced crude 5-year survival from 63.5% to 28.1%. Figure 1 shows the estimated survival over 5 years for all patients according to diagnosis.

Table 3 displays the results of the univariate and multivariable analyses of factors associated with 5-year mortality. Infant gestational age, infant sex, maternal age, and maternal

Table 2. Kaplan–Meier Estimates of Survival for Infants With Selected Congenital Heart Defects With and Without Major Extracardiac Defects, Texas Birth Defects Registry, 1996 to 2003

<table>
<thead>
<tr>
<th>Defect</th>
<th>n</th>
<th>Died, n</th>
<th>1-Year Survival, % (95% CI)</th>
<th>3-Year Survival, % (95% CI)</th>
<th>5-Year Survival, % (95% CI)</th>
</tr>
</thead>
<tbody>
<tr>
<td>HLHS</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No extracardiac defects</td>
<td>267</td>
<td>157</td>
<td>44.2 (38.2–50.0)</td>
<td>41.5 (35.5–47.3)</td>
<td>40.9 (34.9–46.8)</td>
</tr>
<tr>
<td>Extracardiac defects</td>
<td>44</td>
<td>34</td>
<td>27.3 (15.2–40.8)</td>
<td>25.0 (13.5–38.4)</td>
<td>20.8 (9.7–34.9)</td>
</tr>
<tr>
<td>Overall</td>
<td>311</td>
<td>191</td>
<td>41.8 (36.3–46.9)</td>
<td>39.1 (33.7–44.5)</td>
<td>38.0 (32.6–43.5)</td>
</tr>
<tr>
<td>SV</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No extracardiac defects</td>
<td>226</td>
<td>83</td>
<td>72.1 (65.8–77.5)</td>
<td>66.7 (59.0–71.5)</td>
<td>63.5 (56.6–69.6)</td>
</tr>
<tr>
<td>Extracardiac defects</td>
<td>60</td>
<td>43</td>
<td>36.7 (24.7–48.7)</td>
<td>28.1 (17.4–39.8)</td>
<td>28.1 (17.4–39.8)</td>
</tr>
<tr>
<td>Overall</td>
<td>286</td>
<td>126</td>
<td>64.7 (58.8–69.9)</td>
<td>57.8 (51.8–63.3)</td>
<td>56.1 (49.9–61.7)</td>
</tr>
<tr>
<td>PA-IVS</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No extracardiac defects</td>
<td>100</td>
<td>42</td>
<td>61.0 (50.7–89.8)</td>
<td>59.8 (49.4–68.7)</td>
<td>56.6 (45.8–66.0)</td>
</tr>
<tr>
<td>Extracardiac defects</td>
<td>18</td>
<td>9</td>
<td>50.0 (25.9–70.1)</td>
<td>50.0 (25.9–70.1)</td>
<td>50.0 (25.9–70.1)</td>
</tr>
<tr>
<td>Overall</td>
<td>118</td>
<td>51</td>
<td>59.3 (49.9–67.6)</td>
<td>58.3 (49.8–66.6)</td>
<td>55.7 (45.8–64.4)</td>
</tr>
<tr>
<td>TA</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No extracardiac defects</td>
<td>55</td>
<td>12</td>
<td>80.0 (66.8–88.4)</td>
<td>78.1 (64.7–86.9)</td>
<td>78.1 (64.7–86.9)</td>
</tr>
<tr>
<td>Extracardiac defects</td>
<td>12</td>
<td>5</td>
<td>58.3 (27.0–80.1)</td>
<td>58.3 (27.0–80.1)</td>
<td>58.3 (27.0–80.1)</td>
</tr>
<tr>
<td>Overall</td>
<td>67</td>
<td>17</td>
<td>76.1 (64.0–84.6)</td>
<td>74.6 (62.4–83.9)</td>
<td>74.6 (62.4–83.4)</td>
</tr>
<tr>
<td>d-TGA</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No extracardiac defects</td>
<td>204</td>
<td>15</td>
<td>93.6 (89.3–96.2)</td>
<td>92.6 (88.0–95.5)</td>
<td>92.6 (88.0–95.5)</td>
</tr>
<tr>
<td>Extracardiac defects</td>
<td>21</td>
<td>8</td>
<td>61.9 (38.1–78.8)</td>
<td>61.9 (38.1–78.8)</td>
<td>61.9 (38.1–78.8)</td>
</tr>
<tr>
<td>Overall</td>
<td>225</td>
<td>23</td>
<td>90.7 (86.0–93.8)</td>
<td>89.7 (85.0–93.1)</td>
<td>89.7 (85.0–93.1)</td>
</tr>
</tbody>
</table>

Figure 1. Five-year survival for cases by diagnostic categories. On abscissa, time 0 is birth. Numbers of nondeceased/non-censored cases at each year are shown below.
Gestational age, wk
Birth weight, g
Residence in
Birth era
Maternal race/ethnicity
Defect type
Maternal age, y
Maternal education, y
Infant sex

Similarity, infants with SV and PA-IVS had an adjusted risk of death 4.1- and 3.9-fold higher, respectively, than the referent cases of d-TGA. The subgroup TA had an adjusted HR of 1.6, but this was not statistically significant, possibly because of small sample size.

In evaluating the effect of maternal race/ethnicity, NH white was selected as the referent group. After adjustment for defect type, presence of extracardiac defects, birth era, maternal residence in border county, and birth weight, infants born to NH black mothers had a 41% greater risk for death than infants of NH white mothers (adjusted HR, 1.41; 95% CI, 1.01 to 1.97). Infants of Hispanic mothers tended to have a greater risk of death (adjusted HR, 1.26; 95% CI, 1.00 to 1.58), but the difference was of borderline significance. No significant race/ethnic differences were found in the age at first postnatal echocardiogram; the median age for each race/ethnic group was <3 days. After adjustment for defect type, presence of extracardiac defects, maternal race/ethnicity, birth era, and birth weight, infants born to mothers residing in Texas counties bordering Mexico had a 36% greater risk of death than those born to mothers residing in nonborder counties (adjusted HR, 1.36; 95% CI, 1.04 to 1.79). Although female patients had a 27% greater risk of death than male patients on univariate analysis, infant sex did not remain significant after adjustment for defect type, presence of extracardiac defects, maternal race/ethnicity, birth era, residence in nonborder county, and birth weight. Gestational age and maternal age and education were not associated with mortality after adjustment and thus were not included in the multivariable model.

Infants born in the more recent era, 2001 to 2003, had a 47% lower risk of death than infants born in 1996 to 2000 (adjusted HR, 0.53; 95% CI, 0.43 to 0.66). To determine whether the smaller sampling of Texas resident live births in 1996 (35%) and 1997 (56%) affected the magnitude of the decreased mortality, we reran the multivariable analysis, excluding the 1996 and 1997 cases. Using 1998 to 2000 as the early era only slightly reduced the magnitude of the improvement in survival from 47% to 42%; however, it remained statistically significant.

Mortality was significantly lower in the later era for HLHS and SV; the corresponding crude HRs were 0.49 (95% CI, 0.36 to 0.66) and 0.47 (95% CI, 0.32 to 0.69; Figure 2). Although the magnitude of the era effect was similar in the TA subgroup (crude HR, 0.45; 95% CI, 0.15 to 1.39), it did not achieve statistical significance because of its small sample size. No significant change was noted for the PA-IVS and d-TGA subgroup (crude HR, 0.92; 95% CI, 0.53 to 1.60) or the SV subgroup (crude HR, 0.82; 95% CI, 0.35 to 1.90). On further subgroup analysis, mortality for SV cases significantly decreased in NH whites (P<0.05), NH blacks (P<0.05), and Hispanics (P<0.01). Mortality for HLHS cases significantly decreased in NH whites (P<0.01) and in Hispanics (P<0.001) but not in NH blacks because of the small magnitude of the change and the small number of cases.

Discussion

The rationale for studying survival of functional single ventricle cases is that they are the most lethal forms of congenital heart disease and require a large expenditure of resources for intensive medical and surgical care. In the first months of life, these infants are selected for either single
ventricle surgical palliation or heart transplantation. This population-based study presents 5-year survival outcomes for functional single ventricle cases from the Texas Birth Defects Registry. Most clinical outcome studies have the limitation of being based on center referrals and reporting postsurgical survival.\(^{4-7}\) Furthermore, prior published population-based studies have the limitation of using broadly defined heart defect groups and reporting survival of cases born \(>10\) years ago.\(^{1-3}\) The strengths of our study rest on it being population based, having carefully edited specific diagnostic categories, having births from a more recent era (1996 to 2003), and having follow-up to 5 years of age.

Survival of these infants with functional single ventricle was significantly less than that of infants with d-transposition requiring neonatal arterial switch surgery. One-year survival differed between the various categories of functional single ventricle. The lowest 1-year survival was seen with HLHS (41.8%); the highest survival was seen with TA (76.1%). Survival was similar for PA-IVS and SV at 59.3% and 64.7%, respectively. These differences in survival are probably related to the frequency of aortic arch hypoplasia requiring Norwood type of aortic arch reconstruction in cases of HLHS and in some cases of SV, whereas such repair is not necessary in cases of PA-IVS or TA. An earlier study, covering 1986 to 1999, of first-year mortality rates for selected birth defects using data from the Hawaii Birth Defects Program reported 1-year survival for HLHS of 29% and for SV of 65%.\(^{2}\) Cleves et al\(^{9}\) reported 1-year survival for infants born with congenital heart defects in Arkansas in 1993 to 1998 based on a state registry in which cardiac cases were edited by a pediatric cardiologist. They reported 1-year survival for HLHS of 22.6%; for TA, 76.7%; and for PA-IVS, 63.3%. In a large multicenter study, Li and coworkers\(^{14}\) reported 1-year postoperative survival for HLHS of 64% and for TA of 84%. More recently, Tham et al\(^{6}\) reported the outcomes of infants with single ventricle of the double-inlet left ventricle type who were diagnosed from 1990 to 2004 at 3 major referral centers, namely Boston Children’s Hospital, the Toronto Hospital for Sick Children, and the University of California San Francisco. Their 1-year survival was 82%, which is higher than our findings for SV cases. The lower survival in our study compared with these postoperative clinical reports may be explained in part by our inclusion of deaths that occurred before referral to a tertiary care center and those that occurred before any surgical intervention.

The majority of deaths occur in the first year of life, after which survival is relatively stable over the next 4 years (Figure 1). This stability in survival cannot be explained by subjects lost to follow-up as a result of their moving out of state because out-of-state deaths were ascertained from the National Death Index. The greater risk in the first year of life is due to these fragile young infants undergoing surgical procedures that are associated with significant mortality, as well as 17.1% having a life-threatening extracardiac birth defect. The presence of such noncardiac congenital anomalies in our patients with functional single ventricle increased the adjusted risk by 84%. In their study of infants with congenital heart disease, Cleves and coworkers\(^{9}\) reported that survival was highest in infants with no other organ malformations and declined as the number of extracardiac defects increased. This effect of multiple congenital anomalies on survival has been described in other population-based studies of birth defects.\(^{2,8,15,16}\)

Racial/ethnic disparities in survival were found in the present study, as reported by others. After multivariable adjustment, NH blacks with functional single ventricle had a 41% greater risk of death compared with NH whites. Hispanics tended to have a 26% greater risk than NH whites. From their study of the national patterns of mortality from congenital heart disease, Boneva et al\(^{1}\) also reported that blacks died at an earlier age than whites. Looking at 4-state (Massachusetts, Pennsylvania, New York, California) in-hospital mortality after congenital heart surgery, Gonzalez et al\(^{17}\) reported that Hispanics had 71% greater risk of dying than NH whites. The risk for blacks varied among the states and overall did not differ from that of NH whites. Benavides and coworkers,\(^{18}\) studying racial disparities in mortality after open heart surgery for congenital heart disease from a 19-state database, found that blacks had a 65% and Hispanics had a 24% greater risk of mortality after adjustment for Risk Adjusted Classification for Congenital Heart Surgery (RACHS-1) categories of risk, a method that accounts for differences in case severity mix among racial groups.\(^{19}\) They reported that the racial differences were increased further after adjustment for median family income. It has been proposed that racial disparities in mortality may be due to differences in access to medical care. In a population-based study of congenital heart disease patients born in Dallas County, Texas, in 1971 to 1984, Fixler et al\(^{20}\) reported that the timing of referral for pediatric cardiac care was not related to ethnicity, median family income, or household education. Chang et al,\(^{21}\) using
data from hospital discharges for 1995 to 1996 in California, found no difference between whites, blacks, and Asians in age at operation for congenital heart disease. However, Milazzo and coworkers22 reported finding significant racial differences in the timing of Glenn and Fontan procedures in their single-center study from Duke University Medical Center in North Carolina. At the time of either procedure, black children had a median age more than twice that of white children. No significant differences were found in median family income or type of insurance coverage. In our study, we looked at the age at the first postnatal echocardiographic diagnosis as a surrogate measure of access to pediatric cardiac consultant care. No significant differences were found among ethnic groups, with the median age for each ethnic group being <3 days.

We found that the offspring of mothers living in counties bordering Mexico had a 36% greater risk of death after adjustment for maternal race/ethnicity and other variables listed in Table 3. The higher mortality during the first 5 years may have been due to factors affecting access to subspecialty care. Families in these counties are more likely to live in rural areas of Texas. Chang et al21 found a direct correlation between population density of the county of residence in California and earlier age at heart surgery. Many of the border counties are at substantial distances from centers having the capability to care for complex congenital heart disease, which may affect postoperative care after discharge. In addition, the higher frequency of non–English-speaking mothers in the border counties may be an impediment to postdischarge care and follow-up. Further studies seem warranted to identify the causes of the regional differences in mortality of children with life-threatening birth defects.

Several limitations of this study should be considered. The use of a restrictive case definition of HLHS, which excluded patients with ventricular septal defect, may limit the ability to generalize to other study populations. However, this was done to include only patients with severe left ventricular hypoplasia resulting from mitral stenosis/atresia and aortic stenosis/atresia. A second limitation of this study is the racial/ethnic distribution, which differs from that of other regions and the United States in general. The large percentage of Hispanic births (49%) in Texas results in a lower prevalence of HLHS because the prevalence in this group is reported to be 23% lower than in NH whites.23 The relatively small number of NH black cases limits the power to show statistically significant improvement in survival of HLHS in this group. A third, and most important, limitation of this study is that no clinical data were available for review other than those recorded by registry abstractors. In addition, the lack of clinical follow-up information in the registry such as subsequent medical and surgical treatment did not allow us to determine whether families chose nonintervention, which patients underwent palliative surgery or heart transplantation, or which patients died before surgery. Unfortunately, we could not address 5-year survival in the most recent cohort of cases born in 2004 to 2008 because of the short period of follow-up and the delay in updating registry records and state vital records of births and deaths.

Despite these limitations, this population-based study demonstrated significant improvement in overall survival in infants with functional single ventricle born in the later era, 2001 to 2003. The risk of death for those born after 2000 was 47% lower than for infants born in 1996 to 2000. An important improvement in survival occurred in infants with HLHS and SV. In a US population-based study, Boneva et al1 reported that mortality from congenital heart disease declined by 40% from 1979 through 1997 but first-year mortality for HLHS declined by only 7.5%. Scott and Niebuhr,24 in their study of survival in HLHS cases in US military families, reported a marked increase in survival comparing cases from the 1980s and the 1990s, but no change was observed thereafter. In a single-center study of HLHS, Mahle et al5 reported progressive improvement in hospital survival after each stage of palliation (ie, Norwood, bidirectional Glenn, and Fontan procedures). For example, survival after Norwood procedures improved from 56.2% in 1984 to 1988 to 71.3% in 1995 to 1998. Such data indicate that improvement in 5-year survival can be attributed to a large extent on better surgical outcomes in the first year of life.

Conclusions

Although all functional single ventricle patients may undergo complex surgical procedures, 5-year survival varies significantly among diagnostic categories, probably because of the need for aortic arch reconstruction. After the first year of life, the risk of death is relatively low through 5 years of age for all subtypes. This study demonstrates that significant improvement in 5-year survival occurred in infants born after 2000 who had functional single ventricle. Important factors affecting survival include birth era, birth weight, presence of extracardiac defects, maternal race/ethnicity, and maternal residence location. Additional studies are needed to determine whether race/ethnic and regional differences in outcome are due to factors affecting referral to cardiac specialists and the subsequent communication between cardiologists, primary care staff, and families.

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Disclosures

None.

References


**CLINICAL PERSPECTIVE**

Although several postoperative outcome studies have indicated improvement in survival of functional single ventricles, none have included deaths before referral or surgery or patients lost to follow-up. This population-based study of 782 Texas liveborn infants with functional single ventricle includes deaths occurring before referral or surgery during their first 5 years of life anywhere in the United States. The overall 5-year survival for hypoplastic left heart syndrome was 38%; for pulmonary atresia with intact ventricular septum, 56%; for single ventricle, 56%; and for tricuspid atresia, 75%. Five-year survival in patients with significant extracardiac defects was 21% for hypoplastic left heart syndrome, 28% for single ventricle, 50% for pulmonary atresia with intact ventricular septum, and 58% for tricuspid atresia. For infants weighing <1500 g, mortality was 6-fold greater than for those with birth weights of ≥2500 g. The adjusted 5-year mortality for those born in 2001 to 2003 was 47% lower than for those born in 1996 to 2000, indicating a significant improvement in outcome. This study provides important data on specific risk factors affecting mortality in the first 5 years of life. Such information should enable physicians to estimate more realistically the outlook for an infant with functional single ventricle during discussions with family.
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In the article by Fixler et al, “Mortality in First 5 Years in Infants With Functional Single Ventricle Born in Texas, 1996 to 2003,” which appeared in the February 9, 2010 issue of journal (Circulation. 2010;121:644–650), the authors stated that Duke University is located in South Carolina rather than in North Carolina. The third line of the first paragraph on page 649 should read: “However, Milazzo and coworkers22 reported finding significant racial differences in the timing of Glenn and Fontan procedures in their single-center study from Duke University Medical Center in North Carolina.”

The correction has been made to the current online version of the article. The authors regret the error.

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