Trends in Congenital Heart Disease in Dallas County Births 1971-1984

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Ellen Sigman, BS, and Clayton W. Eifler, PhD

To examine the changes in birth cohort prevalence rates and severity of congenital heart disease, we studied children with congenital heart disease born to blacks, whites, and Mexican-Americans in Dallas County from 1971 through 1984. Diagnoses were made by pediatric cardiologists’ clinical evaluations, echocardiography, catheterization, surgery, or autopsy. During this study period, 2,509 of 379,561 liveborn infants were diagnosed, a prevalence rate of 6.6/1000. The rates for whites was significantly higher than for blacks or Mexican-Americans — 7.2/1,000, 5.6/1,000, and 5.9/1,000, respectively. The rate for severe cases requiring cardiac catheterization or surgery or undergoing autopsy was 3.1/1,000 and did not differ among the three groups. The time trend for rates of congenital heart disease suggested an apparent increase in prevalence rate during the 1970s; however, the prevalence rate of severe forms remained relatively stable. This indicates that the apparent rise in prevalence could be accounted for by an increase in detection of mild cases. These findings were interpreted as reflecting a greater tendency for pediatricians to refer asymptomatic children with significant heart murmurs to a pediatric cardiologist. (Circulation 1990;81:137-142)

Congenital heart disease (CHD) is a leading cause of death during the first year of life. Malformations of the cardiovascular system are also associated with significant medical morbidity, which requires use of costly medical facilities. Children with CHD use 25-30% of the beds in most large pediatric intensive care units and, therefore, consume a large fraction of pediatric health care resources. To plan future health care needs, epidemiologic data are needed regarding changes in prevalence and severity of CHD. The surveillance systems for birth defects conducted by the Centers for Disease Control have indicated a more than twofold increase in the most common form of CHD, ventricular septal defect, between 1970 and 1977.1

To examine the changes in prevalence rates and severity of CHD, we gathered epidemiologic data on children with CHD who were born in a well-defined geographic area during a specific time period. The geographic area was restricted to ensure nearly complete ascertainment of cases and to use county birth frequencies to estimate population-based birth cohort prevalence rates. From these data, we were able to evaluate time trends of CHD prevalence from 1971 through 1984, ethnic prevalence rates, and frequency distribution of the anatomic types of heart malformations in the community.

Methods

Study Population

Cases were selected from the 379,561 children born between January 1, 1971, and December 31, 1984, to Dallas County residents. The county residence of cases was confirmed by searching the Texas State Birth Index and obtaining a copy of the birth certificate. Cases born in Dallas County to out-of-county residents were excluded from the study. In this region, 5% of Dallas County residents were born outside of the county; however, their birth certificates identify them as Dallas County resident births, and they are included in this study. The birth certificate data allowed determination of ethnicity (i.e., black, white, or Mexican-American) from the computer algorithm used by the Texas Department of Health.

Case Finding

In this county, the cardiology program at Children’s Medical Center of the University of Texas...
TABLE 1. Classification by Most Accurate Method of Diagnosis

<table>
<thead>
<tr>
<th>Method</th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Clinical only</td>
<td>1,043</td>
<td>41.6</td>
</tr>
<tr>
<td>Two-dimensional echocardiography</td>
<td>244</td>
<td>9.7</td>
</tr>
<tr>
<td>Cardiac catheterization</td>
<td>303</td>
<td>12.1</td>
</tr>
<tr>
<td>Surgery</td>
<td>565</td>
<td>22.5</td>
</tr>
<tr>
<td>Autopsy</td>
<td>327</td>
<td>13.0</td>
</tr>
</tbody>
</table>

Southwestern Medical Center is the sole tertiary center for pediatric catheterization and surgical services. The closest competing university-affiliated center is more than 240 miles away. All local private-practice pediatric cardiologists participated in this study. The data on all infants and children seen by any of the pediatric cardiologists in the area were reviewed. Although there was no set age limit at diagnosis, all infants were born between 1971 and 1984. The hospital records, office records, autopsy reports, and county medical examiner’s records were available for review. The state health department provided a listing of all deaths in Texas attributed to CHD from 1971 through 1986. This listing was cross-referenced with the Texas State Birth Index to identify infants with CHD born in Dallas County who died out of county or who died without referral to a pediatric cardiologist. Records of these cases were obtained from the place of death to verify the presence of CHD.

The diagnosis of CHD was made on the basis of the clinical diagnosis by a pediatric cardiologist, two-dimensional echocardiographic studies, cardiac catheterization, observations at surgery, or observations at autopsy. Cases in which clinical examination suggested “possible” CHD but were not confirmed by subsequent examination or laboratory evaluation were excluded. Cases were categorized according to accuracy of method of diagnosis, ranging in order of accuracy from increasing clinical evaluation to echocardiographic evaluation, cardiac catheterization, surgery, and autopsy (the most accurate) as shown in Table 1. Cases were divided into mild and severe levels; severe cases were those that required cardiac catheterization and/or surgery or had autopsy.

Diagnosis Classification

CHD was defined as a structural abnormality of the heart or great vessels that is potentially of functional importance. Children with only a bicuspid aortic valve or cardiac arrhythmias or infants with estimated gestational age of less than 38 weeks with only a patent ductus arteriosus were excluded. An anatomic coding system was used based on that developed by the International Society of Cardiology.2 Each case was classified into a single diagnostic category. Those cases with multiple cardiac defects were classified according to a hierarchical assignment used by the Baltimore-Washington Infant Study.3 For example, by this scheme, an infant born with d-transposition, ventricular septal defect, and pulmonary stenosis would be classified under d-transposition.

Statistical Considerations

The prevalence rate of CHD was based on the number of cases born alive from January 1, 1971, through December 31, 1984, (irrespective of the age at diagnosis) divided by the number of live births in that same period. The time trend analysis was based on a 12-month moving average of the cohort prevalence rate calculated by dividing the number of cases from each 12-month birth cohort by the number of live births in that 12-month period. The moving average gives a rate for all 12-month periods, thereby removing the restriction of calendar years. The analysis of temporal trends in the CHD data was based on estimated month of conception because timing of the malformation is related to first trimester events rather than the date of birth. This was computed for the CHD cases from the birth certificates. For the cases with unknown gestation, the average gestation of 39.7 weeks (278 days) was used. The denominator birth data were also adjusted back 278 days so that rates of CHD by conception month could be computed. Temporal trends of rates were analyzed for total number of cases of CHD, for severe cases, and for cases of ventricular septal defect, the most common lesion. Additionally, the trend in the percentage of mild cases was examined.

The cohort prevalence rates of CHD by ethnic groups were based on the frequency of cases assigned to the specific ethnic group divided by the number of live births assigned to that ethnic group. The birth certificate data on parental ethnicity were used to assign the infant’s ethnicity using the Texas health department’s algorithm.

Results

During the period from January 1, 1971, through December 31, 1984, 2,509 cases of CHD were diagnosed among 379,561 live births, giving an overall cohort prevalence rate of 6.61/1,000 live births. These cases included those diagnosed by clinical evaluation, two-D echocardiography, cardiac catheterization, surgery, or autopsy. The prevalence of those with more severe defects requiring cardiac catheterization and/or surgery or who had autopsy was 3.15/1,000 live births.

The time trends in Figure 1 indicate the 12-month moving average of the cohort prevalence rates of CHD. Note that in the period 1970–1978, the overall rate appears to be rising; this may reflect a true increase in the rate or merely lower ascertainment of cases during the early years. Note the time trend for the more severe forms of CHD, that is, those having cardiac catheterization, surgery, or autopsy, is relatively flat throughout the period of observation. Examination of rates by month of conception failed to reveal seasonal trends.

Because previous reports have suggested a striking increase in incidence of ventricular septal defects in
the 1970s, we looked at this trend in Dallas County. Figure 2 shows the time trends for cases of ventricular septal defect. This graph suggests a progressive increase in overall rate during the first 8 years and then a leveling-off. This apparent increase is due to an increase in the number of mild cases not having cardiac catheterization, surgery, or autopsy.

An increasing rate of mild cases also was found when all types of CHD were combined. Figure 3 displays an increasing percentage of mild cases in the later years.

The ethnic distribution of the normal newborn population was 60.7% white, 23.6% black, and 14.7% Mexican-American. The overall prevalence rates of CHD was 7.18/1,000 live births for whites, 5.63/1,000 live births for blacks, and 5.88/1,000 live births for Mexican-Americans. The rate of CHD was significantly higher in the white population than in the black (p<0.001) or Mexican-American (p<0.001) populations. The rates for severe cases having cardiac catheterization, surgery, or autopsy, was 3.27/1,000 for whites, 2.91/1,000 for blacks, and 3.01/1,000 for Mexican-Americans; the differences among ethnic groups were not statistically significant. These findings suggest that the higher overall prevalence rate of CHD in the white population is due to a higher rate for the milder forms.

The frequency distribution of specific cardiac diagnoses is listed in Table 2. Note that isolated ventricular septal defect was the most common primary diagnosis. Acyanotic congenital heart lesions comprise the seven most prevalent lesions. The most common cyanotic lesions were transposition, tetralogy of Fallot, and hypoplastic left heart syndrome. Ten lesions accounted for 88% of the cases in the present series. Comparable data are presented from three other studies, which selected cases on the basis of clinical evaluation as well as results from catheterization, surgery, or postmortem examination.4–6 In the present series, 52% of all the diagnoses were made on the basis of noninvasive clinical evaluation and is similar to the 44% in the NIH Collaborative Study and the 52% in the San Francisco Kaiser Plan Study.

Discussion

In epidemiologic studies of birth defects, the ascertainment of cases may be incomplete for several reasons: 1) cases in abortuses and stillborns may not be included; 2) infants with severe cases may have died before referral; 3) some cases may have been referred to outlying centers; and 4) mild asymptomatic cases may not be referred to tertiary centers. In this study, abortuses and stillborns were not included, and prevalence rates were calculated using only live-born frequencies. This seems justified because live birth frequencies also represent the population at risk of having CHD that would be detected clinically by pediatric cardiologists. With regard to severe cases that might have been missed, review of records from the County Medical Examiners Office reveal that less than 0.7% of all deaths in the pediatric age group died with CHD and were not previously ascertained by our case detection methods. With regard to the problem of referral of cases to outlying cardiac centers, the geography of the region minimizes this problem because the nearest other university-
TABLE 2. Distribution of Diagnoses of Congenital Heart Disease

<table>
<thead>
<tr>
<th>Cardiac lesion</th>
<th>Dallas County (%)</th>
<th>Omistaed County (%)</th>
<th>San Francisco Kaiser Plan (%)</th>
<th>NIH Collaborative Study (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ventricular septal defect</td>
<td>42.8</td>
<td>33.4</td>
<td>31.3</td>
<td>29.5</td>
</tr>
<tr>
<td>Pulmonic stenosis</td>
<td>8.9</td>
<td>4.8</td>
<td>13.5</td>
<td>8.6</td>
</tr>
<tr>
<td>Atrial septal defect</td>
<td>7.0</td>
<td>7.0</td>
<td>6.1</td>
<td>7.4</td>
</tr>
<tr>
<td>Patent ductus</td>
<td>5.3</td>
<td>10.2</td>
<td>5.5</td>
<td>8.3</td>
</tr>
<tr>
<td>Endocardial cushion defect</td>
<td>5.2</td>
<td>4.3</td>
<td>3.7</td>
<td>3.6</td>
</tr>
<tr>
<td>Aortic stenosis</td>
<td>4.4</td>
<td>5.9</td>
<td>3.7</td>
<td>3.8</td>
</tr>
<tr>
<td>Coarctation</td>
<td>3.8</td>
<td>5.4</td>
<td>5.5</td>
<td>7.1</td>
</tr>
<tr>
<td>Transposition</td>
<td>3.5</td>
<td>7.5</td>
<td>3.7</td>
<td>2.6</td>
</tr>
<tr>
<td>Tetralogy</td>
<td>3.4</td>
<td>4.8</td>
<td>3.7</td>
<td>3.8</td>
</tr>
<tr>
<td>Hypoplastic left heart</td>
<td>3.4</td>
<td>4.3</td>
<td>0.6</td>
<td>3.1</td>
</tr>
<tr>
<td>Hypoplastic right heart</td>
<td>2.5</td>
<td>3.2</td>
<td>0.6</td>
<td>2.8</td>
</tr>
<tr>
<td>Total anomalous pulmonary veins</td>
<td>1.4</td>
<td>2.7</td>
<td>0.6</td>
<td>...</td>
</tr>
<tr>
<td>Double-outlet right ventricle</td>
<td>1.2</td>
<td>...</td>
<td>0.6</td>
<td>1.0</td>
</tr>
<tr>
<td>Single ventricle</td>
<td>0.9</td>
<td>...</td>
<td>0.6</td>
<td>0.7</td>
</tr>
<tr>
<td>Truncus arteriosus</td>
<td>0.6</td>
<td>...</td>
<td>2.5</td>
<td>1.7</td>
</tr>
</tbody>
</table>

affiliated center is more than 240 miles away and all local pediatric cardiologists participated in this survey. With regard to mild asymptomatic cases not being referred to pediatric cardiologists, the data do reveal that in later years more mild cases were seen by pediatric cardiologists, suggesting the rates for earlier years may have been underestimated.

A potential limitation of this study is the use of a prevalence rate for each birth cohort. Prevalence is the number of cases in a specific population, in this instance, the birth cohort. One would expect that the earlier birth cohorts would have a higher prevalence than the later birth cohorts because of their longer period of follow-up. Previous studies have also reported variable lengths of follow-up: the Collaborative Study,6 1 month to 9 years; the Olmstead County Study,4 1–19 years; and the Kaiser Study,5 5–13 years. In the present study, the minimum length of follow-up was 4 years (for those born in 1984). We found that 95% of the cases were diagnosed by 4 years of age; hence, the 1984 rate may be underestimated by 5% and the 1982 and 1983 rates by less than this.

The reported rates of CHD vary substantially from study to study (Table 3). Differences in these rates are due to differences in case definition, methods of diagnosis, case detection rates, and length of follow-up. In the studies listed, CHD was defined in a similar fashion, but inclusion criteria differed. For example, in the first three reports, premature infants with patent ductus arteriosus were included, whereas in the last three reports, they were excluded. Furthermore, the methods of diagnosis have changed over the past 3 decades. The studies that survey cases born since 1980 have required confirmation by two-dimensional echocardiography, whereas this technique was not available in previous years. The prev-

TABLE 3. Major US Surveys of Congenital Heart Disease

<table>
<thead>
<tr>
<th>Years</th>
<th>Study population</th>
<th>Study design (case selection)*</th>
<th>All cases</th>
<th>Severe cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>1956–1965</td>
<td>12 centers6</td>
<td>Follow-up 1 mo–9 yr (clin, cath, surg, pm)</td>
<td>420</td>
<td>219</td>
</tr>
<tr>
<td>1959–1966</td>
<td>San Francisco Kaiser Plan5</td>
<td>Follow-up 5–13 yr (clin, cath, surg, pm)</td>
<td>163</td>
<td>79</td>
</tr>
<tr>
<td>1959–1969</td>
<td>Olmstead County, Minn4</td>
<td>Follow-up 1–19 yr (clin, cath, surg, pm)</td>
<td>186</td>
<td>126</td>
</tr>
<tr>
<td>1969–1977</td>
<td>New England7</td>
<td>Follow-up 1 yr (cath, surg, pm)</td>
<td>...</td>
<td>3,357</td>
</tr>
<tr>
<td>1981–1982</td>
<td>Baltimore-Washington, DC3</td>
<td>Follow-up 1 yr (echo, cath, surg, pm)</td>
<td>664</td>
<td>428</td>
</tr>
<tr>
<td>1981–1984</td>
<td>Alberta, Canada8</td>
<td>Follow-up 1 yr (echo, cath, surg, pm)</td>
<td>573</td>
<td>347</td>
</tr>
<tr>
<td>1971–1984</td>
<td>Dallas County</td>
<td>Follow-up 4–17 yr (clin, echo, cath, surg, pm)</td>
<td>2,509</td>
<td>1,195</td>
</tr>
</tbody>
</table>

*Clin, clinical; echo, echocardiography; cath, cardiac catheterization; surg, surgery; pm, postmortem examination.
†Per 1,000 live births.
alence rates in several studies\textsuperscript{4–6} included mild cases made by clinical examination alone. Prevalence rates also depend on the length of follow-up. Several studies have shown that the prevalence increases as the follow-up period lengthens due to delay in diagnosis.\textsuperscript{4,5} Hoffman and coworkers\textsuperscript{5} report the observed prevalence rate was 5.2/1,000 after 1 month's observation, 7.8/1,000 after 11 months' observation, and 8.3/1,000 after 23 months' observation. Studies that include only infants less than 1 year old cannot be compared with studies having longer follow-up periods. Therefore, comparisons between various studies are limited by differences in study design.

The prevalence rate of severe cases with cardiac catheterization, surgery, or autopsy is also shown in Table 3. Because more precise methods of diagnosis were used, it is unlikely that these figures are influenced by false-positive diagnoses. Also, ascertainment of severe cases should be more complete because they exhibit congestive heart failure, cyanosis, or a prominent heart murmur. Undetected cases with severe heart disease most frequently are accounted for by those infants who die before referral to the cardiac center. The Baltimore-Washington Infant Study reported that out of 598 infants with severe CHD, only 23 (3.8\%) failed to be referred to a pediatric cardiac center.\textsuperscript{9} These clinically undetected cases would have increased the rate of severe CHD by only 0.06/1,000 live births. Two thirds of these infants died within the first 24 hours of life, and most had other major associated problems. In the Dallas study inspection of the yearly frequency of CHD cases ascertained by autopsy only revealed a progressive decline in later years. For example, for the cohort born between 1971 and 1975, autopsy-diagnosed cases accounted for 14.3\% of the cases, whereas in the 1980–1984 cohort, they accounted for only 5.6\% of the cases. The data suggest that in the earlier years, infants with severe birth defects were not promptly referred for cardiac evaluation.

A higher prevalence rate of mild cases of CHD was found in the white population compared with the rates in the black or Mexican-American populations (4.0/1,000 vs. 2.7/1,000 and 2.9/1,000). This may reflect true ethnic differences or differences in case detection. Because the prevalence rates for severe CHD were similar for the various ethnic groups, we speculate that case detection in the minority groups was lower due to social factors that interfere with routine health care in asymptomatic children.

Several reports have suggested that the prevalence rate of CHD has increased over the past 2 decades.\textsuperscript{1,8–12} Two birth defect surveillance systems directed by the Centers for Disease Control have reported a twofold increase in the incidence of ventricular septal defect between 1970 and 1977.\textsuperscript{1} The Metropolitan Atlanta Congenital Defects Program reported the rate of ventricular septal defect remained relatively stable from 1970 to 1973 at approximately 1.0/1,000 live births and then increased to 2.5/1,000 in 1976 and 1977. The National Birth Defects Monitoring Program of the Centers for Disease Control also reported a twofold increase in the rate of ventricular septal defect during this same period. Although this program has the advantage of being nationwide, it has the disadvantage of being based on the neonatal discharge diagnosis.

Spooner and coworkers\textsuperscript{10} have evaluated temporal increases in the prevalence rate of ventricular septal defect in counties surrounding Albany, New York. They found the estimated rate of ventricular septal defect increased 2.6-fold from 1970 to 1981. For infants diagnosed when less than 1 year old, the rate increased by more than fourfold. The authors ascribe this dramatic increase to changes in patterns of referral (i.e., in the past, many murmurs in infants may have been ignored unless cyanosis or heart failure was present). The increase in referral of small ventricular septal defects is reflected in the increase in spontaneous closure of these defects from 7\% in 1970 to 35\% in 1982. We also found an increase in referral of small ventricular septal defects (Figure 2). These data suggest that either more children are being born with mild CHD or more children are diagnosed with mild CHD.

Grabitz and coworkers\textsuperscript{8} reported the time trend for rates of CHD diagnosed in the first year of life between 1981 and 1984. The Alberta Heritage Pediatric Cardiology Program included cases confirmed by echocardiography, catheterization, surgery, or autopsy. They found an overall prevalence rate of 5.8/1,000 in 1981, 6.1/1,000 in 1982, 6.9/1,000 in 1983, and 7.4/1,000 in 1984. However, this apparent increase in the rate could be explained by the increased use of echocardiography for diagnostic confirmation. Recently, a 60\% increase in prevalence of ventricular septal defects was noted in the Baltimore-Washington Infant Study between 1981 and 1984, which was ascribed to an increase in echocardiographic diagnosis.\textsuperscript{12}

In the present study, the apparent increase in prevalence of CHD in the 1970s predates the advent of two-dimensional echocardiography. This increase in prevalence is explained by an increase in referral of mild cases to a pediatric cardiologist. The rates for more severe cases remained rather stable (Figure 1). One factor that would affect the rate of severe cases and the percentage of mild cases (Figure 3) is more infants being diagnosed by two-dimensional echocardiography who previously would have undergone cardiac catheterization. Because at this center the rate of cardiac catheterization had not fallen between 1983 and 1987 despite the use of two-dimensional echocardiography and because most cases underwent catheterization as a prelude to surgery, it is unlikely that a significant number of severe cases were classified as mild cases.

In summary, data from this study and other recent surveys indicate an apparent increase in the rate of CHD has occurred because of better detection of mild cases and increased use of echocardiography to confirm cases.\textsuperscript{8,10,12} The increase in detection of mild
cases has probably resulted from a heightened awareness and better diagnostic skills of referring physicians. The fact that the prevalence rate between 1971 and 1984 of more severe cases of CHD has remained relatively stable suggests that the true prevalence probably has not increased significantly.

Acknowledgments

We acknowledge the invaluable assistance of the Texas State Department of Health, Bureau of Vital Statistics, the Dallas County Clerks Office, and the Dallas City Bureau of Vital Statistics in obtaining birth and death certificate information. The authors also acknowledge the cooperation of Drs. Edgar A. Newfeld, W. Pennock Laird, and Damaris Wright as well as the Dallas County Medical Examiners Office for making available their clinical files.

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KEY WORDS • congenital heart disease • epidemiologic methods
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Circulation. 1990;81:137-142
doi: 10.1161/01.CIR.81.1.137

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

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