

## How Many Patients? How Many Doctors?

Julien I.E. Hoffman, MD

One of the fastest growing fields in medicine is the care of adults with congenital heart disease (adult CHD or ACHD). This is mainly a consequence of much longer survival due to advances in pediatric cardiology, interventional catheterization, cardiac surgery, and intensive care. Fifty years ago we could diagnose most forms of moderate and severe CHD, but some anomalies were untreatable and others had relatively low survival after treatment. Today, in-hospital mortality is low for almost all forms of CHD, and most of the survivors can be expected to reach adult life. In addition, a medical system with greater focus on CHD now detects many lesions such as coarctation of the aorta and atrial septal defects that previously went unnoticed. Patients with ACHD need adequate resources, facilities and manpower to care for them, and we in turn need accurate estimates of how many patients have each type and severity of ACHD in order to plan effectively. These estimates are moving targets, because improvements in preoperative, intraoperative, and postoperative care occur continuously. What was good yesterday may not be adequate today.

---

### Articles see pp 2254 and 2264

---

The classic dilemma in studying this problem is how to obtain accurate data on a large number of patients. Studies based on registries showing the incidence of CHD at birth and mortality from CHD may show the big picture, but the details are inaccurate.<sup>1</sup> On the other hand, following a smaller population intensively gives accurate data but runs the risk of being unrepresentative of the population as whole as well as risking excessive stochastic variation, especially for complex anomalies with a low birth incidence.

Several attempts have been made to derive these data. A landmark report of Task Force I of the 32<sup>nd</sup> Bethesda Conference set up by the American College of Cardiology provided some estimates of the numbers of surviving patients with CHD by the year 2000,<sup>2</sup> based on rough estimates of birth rate, birth incidence, and survival rate for patients born between 1940 to 1959, 1960 to 1979, and 1980 to 1989. Because the incidence of specific types of CHD is less important for medical planning, they classified ACHD into three groups by severity<sup>2,3</sup>: Simple, which can be cared for by a well trained physician in the community; Moderate, which should be cared for periodically at a regional ACHD center;

and Severe or complex, which should be cared for regularly at a regional ACHD center. (Table) The ACHD center should have staff well trained in ACHD.

The task force estimated that in the year 2000 there would be 368 800 people with simple, 302 000 with moderate, and 117 000 with severe or complex CHD. With a total population in the USA of about 275 million in the year 2000, and allowing for improvements from earlier to later periods, these figures per million are approximately 1340, 1098, and 425, respectively. Data for specific anomalies were unavailable. Furthermore, as reported in 2 articles published in this issue of *Circulation*,<sup>4,5</sup> substantial improvements in survival have taken place even in recent years.

This conference engendered an attempt to provide better estimates of the prevalence of different types of CHD at various ages.<sup>6</sup> Estimates were made of survival in 5-year age groups since 1940, based on accurately known population birth rates, less accurately known incidences of the main types of CHD, and very rough estimates of the proportion of each age group who were treated and the survival of treated and untreated patients. In addition, about 3 000 000 patients were estimated to have a bicuspid aortic valve, most of which would eventually need treatment. Although this study did consider the prevalence of individual anomalies, the assumptions made were very rough. Nevertheless, the estimates made by the task force fell within the ranges estimated by the later study.

The difficulties of obtaining better data are formidable. Not only are the numbers changing year by year, but there are more serious problems to be faced. There are 2 contrasting approaches to obtaining better prevalence data. One is to follow a select group of patients from birth, as Wren and O'Sullivan did in the Northern Health Region of England,<sup>7</sup> encompassing a population of about 3 million people with 377 310 live births from 1985 to 1994. In this population, about 2000 children with CHD were identified. They collected information about survivors and nonsurvivors, and given the expertise of the involved cardiologists, few errors of misclassification can be expected. Because not all children in this study were followed to adult life, however, some extrapolations had to be made, and variations over the years were not considered. In addition, because even in a population of this size the number of children born with specific types of CHD was often quite small, stochastic variation made accurate predictions difficult.

In this issue of *Circulation*, Moons et al<sup>5</sup> provided more extensive data. They studied 7 497 patients with CHD born between 1970 and 1992 and followed in Leuven, Belgium. These patients constituted about 27% of patients with CHD in Belgium (total population about 10.4 million). Ascertainment and follow up were almost complete. They used the categories of mild, moderate, and severe CHD established by

---

The opinions expressed in this article are not necessarily those of the editors or of the American Heart Association.

From the Department of Pediatrics and Cardiovascular Research Institute, University of California, San Francisco.

Correspondence to Julien I.E. Hoffman, MD, 925 Tiburon Boulevard, Tiburon, CA 94920-1525. E-mail julien.hoffman@ucsf.edu (*Circulation*. 2010;122:2231-2233.)

© 2010 American Heart Association, Inc.

*Circulation* is available at <http://circ.ahajournals.org>  
DOI: 10.1161/CIRCULATIONAHA.110.989350

**Table. Classification of ACHD by Severity (Modified From Warnes et al<sup>2</sup>)**

Simple	Moderate	Severe, Complex
Congenital AS	Ao-LV fistula	Any anomalies with conduits
Congenital mitral disease (except parachute valve)	PAPVC, TAPVC	Any cyanotic heart disease
PFO or small ASD	AVSD, ostium primum	Double-outlet RV or LV
Small VSD	Coarctation of the aorta	Eisenmenger syndrome or pulmonary vascular disease
Mild PS	Ebstein	Fontan procedure
Repaired PDA	PDA	Mitral or tricuspid atresia
Repaired ASD, no residua	Moderate/severe PS, PR	Single ventricle
Repaired VSD, no residua	Sinus of Valsalva aneurysm or fistula	Pulmonary atresia
	Sinus venosus type ASD	Transposition of the great arteries
	Congenital subvalvar or supra-valvar AS (excludes HCM)	Truncus arteriosus
	Tetralogy of Fallot	Ventricular inversion
	VSD with absent pulmonary valve or AR VSD with coarctation of the aorta, mitral disease, RVOT VSD with straddling mitral or tricuspid valve	Heterotaxy, malpositions

Ao indicates aorta; AR, aortic regurgitation; AS, aortic stenosis; ASD, secundum atrial septal defect; AVSD, atrioventricular septal defect (atrioventricular canal); HCM, hypertrophic cardiomyopathy; LV, left ventricle; PAPVC, partial anomalous pulmonary venous connection; PDA, patent ductus arteriosus; PFO, patent foramen ovale; PR, pulmonic regurgitation; PS, pulmonic stenosis; RV, right ventricle; RVOT, right ventricular outflow tract obstruction; TAPVC, total anomalous pulmonary venous connection; VSD, ventricular septal defect.

Connelly et al,<sup>3</sup> and found that for all types of CHD in all ages there had been a decrease in mortality rate. For patients born between 1990 and 1992, survival to 18 years of age averaged 88.6%: 98% for those with mild defects, 90% for those with moderate defects, and 56.4% for those with severe, complex defects. For those born in 1970 to 1974, survival to the age of 18 years for all groups combined was about 81%. As expected, the bulk of the deaths were under 1 year of age, but decreased mortality over the years was seen in all age groups. Not only was survival to 18 years greater in all groups, but better survival was seen disproportionately in those with severe CHD, so that the added care required for patients with ACHD would be increased out of proportion to the raw numbers. Approximating the prevalence per million in Belgium from their data gives 893, 680, and 133 for simple, moderate, and complex CHD, respectively. These figures are somewhat lower than the previous estimates cited above.<sup>2,6</sup> Because they needed fewer assumptions, their data may be more reliable than those described previously.<sup>2,6</sup>

A broader based approach to assessing survival of patients with CHD over the years is to use a comprehensive registry

such as the Multiple Cause of Death public use data files, based on death certificates, compiled annually by the National Center for Health Statistics. Total population age-specific data figures can be obtained from national census data. If the birth incidence of CHD is stable, as it appears to be, a decreasing death rate year by year argues for improved survival. Boneva et al<sup>8</sup> used this registry to examine the standardized mortality rates at all ages from several major forms of CHD between 1979 and 1997. The total number of deaths from CHD was 124 832. They observed a 39% decrease in death rate over this period, and approximately half of the deaths occurred in infancy. They also observed that the median age at death almost doubled over the duration of the study. Importantly, they noted that mortality was higher and declined more slowly in blacks than in whites. Pillutla et al<sup>9</sup> studied patients between 1979 and 2005. They demonstrated that for all CHD, mortality was 48.6% <1 year of age, 12.4% from 1 to 18 years, and 39.5% >18 years of age, and that standardized death rates from all forms of CHD decreased progressively between 1999 and 2006 in age groups <1 year, 1 to 4 years, 5 to 17 years, 18 to 34 years, 35 to 49 years, 50 to 64 years, and ≥65 years. As expected, mortality rates were highest <1 year, quite high from 1 to 4 years, and then essentially <1/100 000 population in each subsequent age group but with a slight increase in those ≥65 years.

The recent study by Gilboa et al<sup>4</sup> used the same Multiple Cause of Death database and census data to derive standardized death rates between 1999 and 2006 for several different major congenital heart anomalies. Even over this short period the average standardized mortality rate decreased by 24%. Just as Boneva et al<sup>8</sup> had done, they also examined differences between ethno-racial groups, but divided them into non-Hispanic blacks, non-Hispanic whites, Hispanics, and other non-Hispanic racial groups. They observed that mortality was higher among non-Hispanic blacks than non-Hispanic whites, with Hispanics similar to non-Hispanic whites.

What are we to make of these trends and differences? It is tempting to ascribe the ethno-racial disparities to socioeconomic differences that affect access to and quality of health care, and this is probably true. If it is, then we need to take these factors into account and find some method of remedying them. However, we should be careful not to ignore the possibility that the same circumstances that may result in poor medical care may also affect the accuracy of filling in birth and death certificates. Gilboa et al<sup>4</sup> listed many of the known deficiencies that affect the accuracy of death certificates. In their study, for example, they commented that 5 deaths over the age of 35 were probably incorrectly attributed to the hypoplastic left heart syndrome. This judgment is almost certainly correct, considering that the oldest reported untreated patient with this disease was 24 years old, surgical treatment for this anomaly began only in 1980, and it was several years before a significant number of patients survived many years beyond surgery. A more serious issue in their study was the finding that patent ductus arteriosus had the lowest median age at death of 11 days. No pediatric cardiologist would accept that these caused death, and in fact at this age these probably represented a normal ductus with delayed closure rather than a congenital anomaly. Without intensive investiga-

tion of the type carried out by Moons et al,<sup>5</sup> we may never know how many errors of classification contribute misleading information.

Nevertheless, the studies described here and others in the literature make it clear that mortality from CHD is decreasing. Because most patients with CHD are improved by treatment but not necessarily cured, for many patients we have merely deferred mortality to later ages. That is not to be discounted, but it emphasizes that more and more patients will be transferred from pediatric to adult cardiologists who will have to bear the burden of an increasingly complex group of patients. Although we certainly need more accurate data, I believe that we know enough now about the likely numbers of patients to make plans for a concerted effort to provide care for this important group of patients.

### Disclosures

None.

### References

- Hoffman JI. Natural history of congenital heart disease. Problems in its assessment with special reference to ventricular septal defects. *Circulation*. 1968; 37:97–125.
- Warnes CA, Liberthson R, Danielson GK, Dore A, Harris L, Hoffman JI, Somerville J, Williams RG, Webb GD. Task force 1: the changing profile of congenital heart disease in adult life. *J Am Coll Cardiol*. 2001;37: 1170–1175.
- Connelly MS, Webb GD, Somerville J, Warnes CA, Perloff JK, Liberthson RR, Puga FJ, Collins-Nakai RL, Williams WG, Mercier LA, Huckell VF, Finley JP, McKay R. Canadian Consensus Conference on Adult Congenital Heart Disease 1996. *Can J Cardiol*. 1998;14: 395–452.
- Gilboa SM, Salemi JL, Nembhard WN, Fixler DE, Correa A. Mortality resulting from congenital heart disease among children and adults in the United States, 1999 to 2006. *Circulation*. 2010;122:2254–2263.
- Moons P, Bovijn L, Budts W, Belmans A, Gewillig M. Temporal trends in survival to adulthood among patients born with congenital heart disease from 1970 to 1992 in Belgium. *Circulation*. 2010;122:2264–2272.
- Hoffman JI, Kaplan S, Liberthson RR. Prevalence of congenital heart disease. *Am Heart J*. 2004;147:425–439.
- Wren C, O'Sullivan JJ. Survival with congenital heart disease and need for follow up in adult life. *Heart*. 2001;85:438–443.
- Boneva RS, Botto LD, Moore CA, Yang Q, Correa A, Erickson JD. Mortality associated with congenital heart defects in the United States: trends and racial disparities, 1979–1997. *Circulation*. 2001;103: 2376–2381.
- Pillutla P, Shetty KD, Foster E. Mortality associated with adult congenital heart disease: Trends in the US population from 1979 to 2005. *Am Heart J*. 2009;158:874–879.

---

KEY WORDS: Editorials ■ adult congenital heart disease ■ mortality