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Interventional Treatment of Patients With Congenital Heart Disease

Nationwide Danish Experience Over 39 Years

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ABSTRACT

BACKGROUND The treatment of congenital heart (CHD) has changed rapidly.

OBJECTIVES The authors reviewed CHD treatment through a 39-year nationwide population-based study on congenital heart surgery and catheter-based interventions, unbiased by referral patterns.

METHODS Using medical registries, the authors identified children (<18 years of age) treated for CHD in Denmark from 1977 to 2015, their need for reinterventions, and their long-term survival. Ten controls per patient, matched by sex and year of birth, allowed comparison with the background population. Survival was described using Kaplan-Meier curves.

RESULTS A total of 9,372 patients underwent 11,968 cardiac surgeries and 1,912 catheter-based interventions. Median age at first procedure decreased from 3.4 years (5th and 95th percentiles: 0.01 to 15.4 years) in 1977 to 1989 (period 1), 0.8 years (5th and 95th percentiles: 0.003 to 13.8 years) in 1990 to 2002 (period 2), and to 0.6 years (5th and 95th percentiles: 0.0 to 14.9 years) in 2003 to 2015 (period 3). More patients were born preterm (<37 weeks) in period 3 compared with those in period 1 (18.5% vs. 6.7%). Catheter-based interventions, not recorded before 1990, were increasingly used as the initial procedure in 5.8% of patients in period 2 and 25.9% of patients in period 3. An increasing part of the population did not undergo surgery at all (4.8% in period 2; 24.0% in period 3). Thirty-day survival increased from 97% (period 1) to 98% (period 2) to 100% (period 3). Ten-year survival increased from 80% (period 1) to 87% (period 2) to 93% (period 3). Compared with the background population, CHD was associated with lower survival in all 3 time periods.

CONCLUSIONS Interventional treatment of CHD has evolved from 1977 to 2015 and is now performed on younger and more preterm patients, often with catheter-based interventions. However, compared with the background population, survival remains significantly reduced. (J Am Coll Cardiol 2017;69:2725–32) © 2017 by the American College of Cardiology Foundation.

The treatment of congenital heart disease (CHD) has changed rapidly. In 1938, Robert Gross was the first to ligate a patent arterial duct (1). In 1944, the Blalock-Taussig shunt (2) introduced palliation of ductal-dependent congenital heart defects, whereas the development of the heart-lung machine in 1953 (3) made it possible to operate directly on the heart. The introduction of prostaglandin in 1974 (4) allowed clinicians to stabilize children with complex CHD before surgery. Echocardiography, with the development of 2-dimensional views and color Doppler (5), provided cardiologists with improved diagnostics. New surgical techniques, including the Fontan (6) and Norwood (7) procedures, enabled staged reconstruction of infants with single ventricles. Catheter-based interventions started in 1966 with balloon atrial septostomy (8), and today these interventions include balloon dilation, stenting, device closures, and transcatheter valve implantation (9). Overall, treatment is now available for most CHDs,

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ABBREVIATIONS AND ACRONYMS

CHD = congenital heart disease

CI = confidence interval

and early survival has increased to nearly 100%; thus, the focus today is on long-term outcomes.

In Denmark, 5 cardiothoracic surgery departments initially conducted treatment for CHD. In recent years, the treatment has been performed in just 2 centers: Aarhus University Hospital (Aarhus, Denmark) and Rigshospitalet (Copenhagen, Denmark). Every Danish citizen has a unique social security number that is used in the health care system from birth until death. This information is collected in nationwide health registries (10,11) and provides a unique opportunity to report long-term outcomes.

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Population-based long-term outcomes following congenital heart surgery have been reported in Norway (12,13) and Finland (14,15). However, this is the first nationwide study that describes the treatment of CHD including both congenital heart surgery and catheter-based interventions over a 39-year period without bias from referral patterns.

TABLE 1 Study Population Demographics

	1977-1989	1990-2002	2003-2015	Total
Patients (‰ of live births)	2,499 (3.4)	3,508 (4.0)	3,365 (4.1)	9,372 (3.9)
Live births in Denmark*	734,517	866,620	801,685	2,402,822
Male	1,274 (51.0)	1,851 (52.8)	1,752 (52.1)	4,877 (52.0)
Age at first procedure, yrs	3.4 (0.01-15.4)	0.8 (0.003-13.8)	0.6 (0.0-14.9)	1.1 (0.003-14.8)
Preterm birth	167 (6.7)	500 (14.3)	621 (18.5)	1,288 (13.7)
Missing data on pre-term birth	1,326 (53.1)	327 (9.3)	213 (6.3)	1,866 (19.9)
Simple diagnosis	1,445 (57.8)	1,777 (50.7)	2,000 (59.4)	5,222 (55.7)
VSD	441 (17.7)	607 (17.3)	579 (17.2)	1,627 (17.4)
PDA	331 (13.3)	327 (9.3)	397 (11.8)	1,055 (11.3)
ASD	274 (11.0)	304 (8.7)	413 (12.3)	991 (10.6)
CoA	342 (13.7)	277 (7.9)	270 (8.0)	889 (9.5)
AS	30 (1.2)	128 (3.7)	133 (4.0)	291 (3.1)
PS	19 (0.8)	108 (3.1)	157 (4.7)	284 (3.0)
MV disease	6 (0.2)	16 (0.5)	27 (0.8)	49 (0.5)
PAPVD	2 (0.08)	10 (0.3)	24 (0.7)	36 (0.4)
Complex diagnosis	865 (34.6)	1,294 (36.9)	1,033 (30.7)	3,192 (34.1)
TGA	264 (10.6)	312 (8.9)	240 (7.1)	816 (8.7)
TOF	256 (10.2)	282 (8.0)	236 (7.0)	774 (8.3)
AVSD	158 (6.3)	293 (8.4)	247 (7.3)	698 (7.5)
UVH	34 (1.4)	196 (5.6)	143 (4.3)	373 (4.0)
PA	119 (4.8)	93 (2.7)	55 (1.6)	267 (2.9)
TAC	29 (1.2)	58 (1.7)	34 (1.0)	121 (1.3)
I/HAA	2 (0.1)	33 (0.9)	39 (1.2)	74 (0.8)
TAPVD	3 (0.1)	27 (0.8)	39 (1.2)	69 (0.7)
Miscellaneous	189 (7.6)	437 (12.5)	332 (9.9)	958 (10.2)

Values are n (%) or median (5th to 95th percentiles). *Data from Statistics Denmark (18).

AS = aortic stenosis; ASD = atrial septal defect; AVSD = atrioventricular septal defect; CoA = coarctation of the aorta; I/HAA = interrupted/hypoplastic aortic arch; MV = mitral valve; PA = pulmonary atresia; PAPVD = partial anomalous pulmonary venous drainage; PDA = patent ductus arteriosus; PS = pulmonary stenosis; TAC = truncus arteriosus; TAPVD = total anomalous pulmonary venous drainage; TGA = transposition of the great arteries; TOF = tetralogy of Fallot; UVH = univentricular heart; VSD = ventricular septal defect.

METHODS

The Danish National Registry of Patients (10) was used to identify children (<18 years of age) treated for CHD in Denmark from January 1, 1977, to December 31, 2015. The International Classification of Diseases-Eighth Revision was used from 1977 to 1993, and the International Classification of Diseases-Tenth Revision was used from 1994 to 2015. Congenital heart surgery was defined as procedures with codes 30 to 33 and codes KF, except for codes specific for catheter-based interventions. Diagnostic catheterizations were not included in the analyses. Specific codes for catheter-based interventions were first introduced to the classifications with aortic balloon valvotomy in 1987, dilation of coarctation and mitral and pulmonary balloon valvotomy in 1990, and atrial septostomy in 1994. Catheter-based interventions before 1990 were not included in the analyses. Finally, reoperations before discharge of initial procedure, extracorporeal membrane oxygenation, use of aortic balloon pump, myocardial biopsies, pacemaker implantations, and ablation procedures were excluded.

The specific CHD of each patient was identified from the primary diagnosis of each hospital contact. When a patient had more than 1 CHD, the most severe defect was chosen according to a hierarchy published previously (16,17). Furthermore, the diagnosis of each patient was subdivided into simple and complex defects (12). Gestational age was identified in the Danish Medical Birth Registry. Prematurity was defined as a gestational age <37 weeks. New operations or catheter-based interventions were defined as new procedures performed during a new admission. All patients were followed from date of surgery or intervention until death, emigration, or end of follow-up (February 24, 2016) according to the Danish Civil Registration System (11).

To compare survival in the CHD population with the Danish background population, the Danish Civil Registration System (11) was used to identify 10 controls per patient, matched by sex and year of birth. The controls were chosen so they were alive at the date of first procedure for each patient. Statistics Denmark was used to identify the number of live births in Denmark from 1977 to 2015 (18).

The study was approved by the Danish Data Protection Agency (record number 2015-41-4034). Because no patients were directly involved in the study, informed consent was not indicated per Danish law.

STATISTICAL ANALYSIS. Data are presented as mean ± SD or median (5th and 95th percentiles), as appropriate. To describe changes over time, the study