

Heart Disease in Childhood: From Malformed Hearts to the Silent Impact of Unhealthy Lifestyles

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Early recognition of congenital heart disease, coupled with the growth and sophistication of diagnostic, medical, and surgical interventions at early ages, has resulted in significantly improved outcomes. However, the cardiovascular impact of the epidemic of childhood obesity and its related disorders now looms as an even greater threat to the health of children.

Heart disease begins in childhood [1, 2]. Over the past several decades, pediatricians have primarily focused on congenital or structural heart disease, and during that time, advances in surgery, intensive care, cardiac catheterization, cardiac imaging, and medical therapies have significantly lowered mortality rates for infants and children with complex congenital heart disease [3]. An estimated 85% of patients now survive into adulthood; in the United States there are now more adults than children living with congenital heart disease. The number of adults with congenital heart disease is expected to keep growing by about 5% each year [4]. In recent years, however, these remarkable successes have been counterbalanced by an increase in the prevalence of risk factors for early-onset cardiovascular disease, attributable to the epidemic of childhood obesity. A growing number of children who were born with structurally normal hearts are now being diagnosed with hypertension, diabetes, and hyperlipidemia [5].

Congenital Heart Disease

For centuries, heart disease in childhood has been synonymous with "blue babies." However, many infants born with major malformations of the heart and cardiovascular system heart disease, consisting of a significant malformation of the heart and its vascular system, occurs in 8 to 10 out of 1,000 live births. The impact on affected children is profound; in 2 out of 1,000 live births, a child has heart defects severe enough to cause death in the first week of life [7, 8] (Table 1).

In many cases, the potential for a normal life expectancy, the ability to achieve an optimal quality of life, and the availability of sufficient family financial and social resources to support a child with significant congenital heart disease are at best limited. Most families lack the financial resources to

cover all of the expenses of having a child with significant congenital heart disease, which may include but are not limited to the costs of multiple surgeries requiring prolonged hospital stays, multiple other inpatient and outpatient hospital visits, expensive medications, physical and occupational therapy, and additional home support once patients are discharged. In addition, the illness results in time away from work for parents, jeopardizing their economic stability, and the family's social stability may also be disrupted. There is a limited body of research available which addresses the impact of the stresses imposed on families with children who have serious congenital heart defects [11, 12]. Our preliminary experience among the CHD population at Duke suggests that marital disruption occurs at a higher rate than in the general population. Until recently, the possibility of children with congenital heart disease being denied health insurance served as an additional stressor for families. The provision of health care for those children who survive to

TABLE 1.
US Prevalence of Selected Congenital Heart Defects

Type of Defect	Prevalence per 10,000 Live Births
Ventricular septal defect	15.57
Valvar pulmonary stenosis	3.78
Atrioventricular septal defect	3.27
Atrial septal defect	2.35
Tetralogy of Fallot	2.60
Congenital aortic stenosis	0.81
Transposition of the great vessels	2.64
Coarctation of the aorta	1.39
Hypoplastic left heart syndrome	1.78
Patent ductus arteriosus	0.88

Source: Data are from [9, 10].

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adulthood is still problematic, not only because many have been denied health care coverage on the grounds that their congenital heart disease is a preexisting condition, but also because of the cumulative effects of congenital heart defects on a person's educational and vocational preparation to live independently as an adult.

The abnormalities in the development of the heart that constitute congenital heart disease occur between 25 and 60 days after conception. This means that in most cases congenital heart defects are already present by the time pregnancy is confirmed. Despite extensive research, most of the actual causes of congenital heart disease are unknown. The point in heart development at which specific defects occur is known, but the actual triggers causing an arrest in the normal development of the heart have yet to be described. A multifactorial cause for congenital heart disease is hypothesized but has not been confirmed, and the illness does not conform to most generally held genetic theories [8, 13, 14]. Until a cause is identified, prevention is not possible. As a result, the incidence of congenital heart disease has remained stable for centuries.

Sophisticated 2-dimensional cardiac ultrasound imaging capable of resolving to very small structures became available only recently; before that, the diagnosis of congenital heart defects awaited the end of pregnancy. Because fetal circulatory physiology confers protection to fetuses with many of even the most complex congenital heart defects, affected babies survived during pregnancy, only to experience near-fatal or fatal outcomes once fetal circulation was replaced by a more normal circulatory physiology at birth. Most babies with heart defects, because their defects had not been diagnosed before birth, were born in local hospitals that lacked high-tech interventions, and the morbidity and mortality of congenital heart disease was greater than 50% in the first week of life [15, 16]. Heart defects such as transposition of the great vessels, hypoplastic left heart syndrome, congenital aortic stenosis, and coarctation of the aorta were responsible for 95% of deaths in the first week of life.

Certain heart defects cluster among genetic syndromes, some of which are listed in Table 2 [17, 18]. The most common such syndrome is Down syndrome, in which more than 50% of affected babies are born with atrioventricular septal defects (formerly known as endocardial cushion defects).

Congenital heart disease in childhood presents in 1 of 3 ways: central cyanosis (the presence of 3-5 grams of desaturated hemoglobin), congestive heart failure, or a constellation of the following physical exam findings—murmur, lack of pulses, poor peripheral perfusion, hepatomegaly, sustained tachypnea and tachycardia, and diaphoresis with feedings.

The overwhelming majority of patients with congenital heart disease present well before the age of 18 months; most present during the first 6 months of life, and those with critical defects present within the first 3 months of life (Table 3). Early recognition is essential to ensure reasonable outcomes in congenital heart disease. Multiple studies

TABLE 2.
Heart Problems Associated with Selected Genetic Syndromes

Genetic Problem	Associated Heart Problem
Down syndrome	Atrioventricular septal defect, tetralogy of Fallot
Turner syndrome	Coarctation of the aorta, congenital aortic stenosis
Fetal hydantoin (Dilantin) syndrome	Valvar pulmonary stenosis
Trisomy 13 and 18	Ventricular septal defect, double outlet right ventricle
Marfan syndrome	Mitral valve prolapse, mitral regurgitation, aortic insufficiency, aortic root dilation
VACTERL association ^a	Ventricular septal defect, atrial septal defect, tetralogy of Fallot
DiGeorge syndrome	Tetralogy of Fallot, double aortic arch, vascular rings
Noonan syndrome and LEOPARD syndrome ^b	Valvar pulmonary stenosis, hypertrophic obstructive cardiomyopathy
Holt-Oram syndrome	Atrial septal defect (secundum)

^aVACTERL association is defined by the presence of 3 or more of the following congenital malformations, the initial letters of which spell VACTERL: vertebral anomalies, anal atresia, cardiac anomalies, tracheoesophageal fistula, renal or radial anomalies, and limb abnormalities.

^bLEOPARD syndrome, which is associated with Noonan syndrome, is characterized by the following conditions, the initial letters of which spell LEOPARD: lentigines, electrocardiographic conduction abnormalities, ocular hypotelorism, pulmonary stenosis, abnormal genitalia, retarded growth, and deafness (sensorineural).

have documented significantly improved survival in those patients born in, or transferred early to, pediatric cardiovascular centers with high-tech facilities for pediatric cardiovascular interventions, including sophisticated imaging facilities; extracorporeal membrane oxygenation; critical cardiac intensive care units; specialized nursing; physical, occupational, and nutritional therapy; and pediatric cardiovascular surgical and anesthesia programs.

With the discovery of the life-preserving effects of prostaglandin E₁ (alprostadil), the development of sophisticated 2-dimensional echocardiography with Doppler (which is able to identify cardiac defects as early as 16-weeks gestation), the rapid development and miniaturization of specialized cardiac surgical procedures, the sophistication of pediatric cardiac anesthesia support, and the development of extracorporeal membrane oxygenation for pediatrics at major medical centers throughout the country, the outlook for babies with congenital heart disease has significantly improved [18]. Survival for even the most complex and severe congenital heart defects, such as hypoplastic left and right heart syndromes requiring complex surgery, has improved significantly, as has survival for babies born with structurally normal but severely dysfunctional hearts—for example, those with congenital cardiomyopathy requiring neonatal heart transplantation. More than 75% of such children now survive the first month of life. However, early diagnosis and intervention is critical for babies with congenital heart disease. The availability of high-tech resources of this

type is limited primarily to major university medical centers. The specialized technology necessary to diagnose and intervene in critical congenital heart disease is therefore not readily available to many families living in rural towns and remote cities.

Obesity and Hypertension in Childhood: Risk Factors for Early-Onset Cardiovascular Disease

Congenital heart disease is a major challenge to normal cardiovascular function over the long term. However, a new and potentially even more dangerous threat has emerged in the pediatric population: acquired heart disease, like that seen in adults [19-21]. In the past 15 years, with the emergence of childhood obesity as a national health epidemic among children, the incidence of associated hypertension, type 2 diabetes, and hyperlipidemia is soaring, and the clinical pediatric cardiology population is rapidly expanding to include children with structurally normal hearts that have been subjected to the ravages of unhealthy lifestyles. The numbers of children referred to pediatric cardiologists with obesity and hypertension have increased exponentially, fueled by unhealthy lifestyles—physical inactivity combined with excessive dietary intake of sugar, salt, and fat in meals eaten at home, at school, or in fast food restaurants. Misinformation or lack of information about nutrition among parents and caretakers; the proliferation of sedentary alternatives to exercise, such as video and computer games; the lack of easily accessible low-cost after-school physical activities for children; and the absence of regularly required physical education classes in schools have all conspired to make healthy lifestyle choices more difficult to achieve [22]. In turn, we are now facing a generation of children whose life expectancies may be considerably shortened, and the resulting impact on the health care system

from the necessity of managing early cardiac disease will be unprecedented. Many children are the second or third generation in the family to have hypertension, hyperlipidemia, or type 2 diabetes—diseases that will predispose them to early coronary artery disease [20]. The rapid proliferation of conditions predisposing young people to adult heart disease in childhood is a national emergency and threatens an entire generation of our children.

Nationwide, about 1 in 3 children age 2-19 years are estimated to be overweight or obese [23]. Obesity in childhood and adolescence is one of the main predictors of hypertension in adulthood, but it is also associated with other cardiovascular risk factors, such as dyslipidemia, abnormal glucose metabolism, insulin resistance, inflammation, and impaired vascular function (dysmetabolic syndrome). Therapy is largely nonpharmacologic. Major changes in behaviors are needed, including an increase in physical activity and healthier lifestyle choices. Nutrition education also is recommended. The goal is to reduce systolic blood pressures below the 95th percentile for age, gender, and height.

A significant body of work is emerging that describes the demographics of, health care costs, and contributing factors that have led to the growth in childhood obesity [24, 25].

Socioeconomic status is a strong determinant of a young person's potential for obesity. Among children 2-18 years of age, Latino children are most at risk for obesity. Children covered by Medicaid are nearly 6 times more likely to be treated for a diagnosis of obesity than are children covered by private insurance [26].

Children treated for obesity are roughly 3 times more expensive for the health system than is the average insured child. The national cost of childhood obesity is estimated to be approximately \$11 billion for children with private insurance and \$3 billion for those with Medicaid. Annual health care costs are about \$6,700 for children treated for obesity who are covered by Medicaid (compared to \$2,400 for children without obesity) and about \$3,700 for obese children with private insurance (compared to \$1,100 for those without obesity) [26].

Children diagnosed with obesity are 2-3 times more likely to be hospitalized. Children treated for obesity are far more likely to be diagnosed with mental health disorders or bone and joint disorders than are nonobese children [26].

In the 2009 Youth Risk Behavior Surveillance System survey, 13% of high school youth in North Carolina described themselves as obese. Asked whether they had eaten fruits and vegetables 5 or more times per day during the 7 days before the survey, 83% said no. In addition, 32% said they had drunk a can, bottle, or glass of soda or pop (not including diet soda or diet pop) at least 1 time per day during those 7 days. Recommended levels of physical activity were met by only 46% of these adolescents, 35% watched television 3 or more hours per day on an average school day, and 21% played video or computer games or used a computer for something that was not schoolwork for 3 or more hours per

TABLE 3.
Timing of Presentation of Specific Cardiac Lesions in the First Year of Life

Age of Child at Presentation	Type of Cardiac Lesion
1 day	Arteriovenous malformations, tricuspid valve insufficiency, transposition of the great vessels, Ebstein's anomaly of the tricuspid valve
1 week	Obstructions Critical pulmonary/aortic stenosis, coarctation of the aorta, hypoplastic left heart syndrome Ductal dependent lesions Transposition of the great vessels, tricuspid atresia, total anomalous pulmonary venous return with obstruction, tetralogy of Fallot, pulmonary atresia with intact ventricular septum
2-4 weeks	Ventricular septal defect
4-8 weeks	Ventricular septal defect, arteriosclerotic vascular disease, truncus arteriosus
8 weeks to 1 year	Anomalous origin of the left coronary artery from the pulmonary artery, congenital myocarditis, congenital cardiomyopathy

Source: Data are from [17].

day on an average school day [27].

Factors contributing to the emergence of obesity and its related disorders include increased availability and consumption of high-fat, high-calorie foods and soft drinks; a sedentary lifestyle in which a great deal of time is spent watching television and playing computer or video games; a lack of focused physical education programming; a lack of safe places for children to exercise; inability to purchase healthy foods because they are higher in cost or are less readily available; a lack of basic understanding of nutrition; and the proliferation of advertising for fast foods in the media.

In summary, although advances in technology have led to improved survival of children with congenital heart disease, we are now faced with a generation of children burdened early in life by obesity, dysmetabolic syndrome, and hypertension. This is predicted to result in an imminent increase in the incidence of cardiovascular disease and stroke. Reversing this trend will require sweeping policy changes at all levels to make sure that children have access to healthy foods, physical activity, and safe spaces in which to exercise. NCMJ

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