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PAPER

PATHOLOGY/BIOLOGY

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"Grown-Up" Congenital Heart Disease and Sudden Death in a Medical Examiner's Population*

ABSTRACT: Despite advances in the management of congenital heart disease (CHD), children with CHD who survive into adulthood are at increased risk of sudden death. Sudden death may also be the initial presentation of undiagnosed CHD in some adults. This retrospective descriptive study outlines the spectrum of CHD presenting as sudden death in adults in a medical examiner's population. Despite its rarity (0.2% of all cases investigated between 1991 and 2007), CHD remains an important cause of sudden cardiac death to be recognized at adult autopsy. Bicuspid aortic valve and anomalous coronary anomalies were the most common malformations, comprising 36.9% and 26.2% of cases, respectively. However, a wide spectrum of simple to complex malformations can be seen, with or without prior surgery, and over a wide age spectrum. Once solely a pediatric entity, CHD is now "grown-up" and will likely be diagnosed by forensic pathologists with increased frequency in the future.

KEYWORDS: forensic science, forensic pathology, adult, autopsy, cardiac, sudden death, congenital heart defects, congenital anomalies, cardiac surgery

Congenital heart disease (CHD) is the most common birth defect, with a reported incidence ranging from 6/1000 live births for moderate and severe forms to 75/1000 live births for all forms, the latter including trivial lesions (1). With advances in medical and surgical management, *c.* 90% of children with cardiac defects now survive into adulthood, and by 2005, it was estimated that there were *c.* 1 million adults in the United States living with CHD (2,3). As the number of affected children who survive into adulthood has steadily increased, there are now more adults than children living with CHD, and it is well documented in the clinical literature that adult cardiologists must now be well versed in the management of "grown-up" CHD. Similarly, medical examiners must be knowledgeable in this area as even with appropriate medical management and surgical correction, this population is at increased risk of sudden cardiac death. In addition, previously undiagnosed CHD may initially present with sudden death and thus come to the attention of the medical examiner.

A uniformly accepted definition for "sudden cardiac death" does not exist. It is often described as "unexpected natural death caused by a cardiac cause, within 1 h from the onset of symptoms, in an individual without any prior condition or an individual with a stable medical condition." The World Health Organization extends this definition to include deaths that occur <24 h from the onset of symptoms. In unwitnessed deaths, it is fair to assume that death was sudden if the individual was known to be in good health 24 h before being found dead (4). In individuals over 30 years of age, the most common cause of sudden cardiac death is coronary artery atherosclerosis, which accounts for *c.* 80% of cases (5). CHD is an uncommon finding in autopsy studies of sudden death. In a large population-based study that evaluated mortality in the United States from 1979 to 1997, 0.31% of deaths were caused by CHD; mortality from CHD was highest in infants and children and then rapidly declined with a stable rate between the ages of 15 and 65 years (6).

Congenital heart defects provide numerous mechanical and electrophysiologic substrates for sudden death (7,8). Arrhythmias can arise with any change in the electrophysiologic properties of the cardiac tissue, which may occur at the macroscopic, microscopic, cellular, or ionic level. There may be abnormalities of rhythm or conduction that are inherent (result from hemodynamic or hypoxic stresses of a defect) and that persist after surgical correction. Commonly encountered arrhythmias in patients with CHD include re-entrant atrial and ventricular tachycardias, heart block, and sinus node dysfunction (8). The increased risk of arrhythmia-related sudden death applies mostly to tetralogy of Fallot (TOF), transposition of the great arteries, aortic stenosis, and pulmonary vascular obstruction (5). In adults, nonrhythm-related mechanisms of sudden death in CHD include coronary ischemia, sepsis, thromboembolism, and pulmonary hypertensive crises (9).

Acquired electrophysiologic abnormalities may be the result of surgical repair itself, as actions inherent to some operative procedures may create substrates for sudden death. These include

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complications specific to the anatomic locale of the reconstruction, such as the proximity of intervention to the conduction system, scarring at incision sites, abnormal morphology of reconstructed valves, and the use of prosthetic materials (patches, valves, and conduits) (7,10). Depending on the nature and extent of the surgical repair and the postoperative function of the defect, there may be ongoing or new hemodynamic and hypoxic stresses that can contribute to sudden death. Some surgical procedures are commonly associated with arrhythmias; these include the Mustard or Senning procedure for transposition of the great vessels, Fontan procedure for defects with univentricular physiology, and repair of TOF (8).

A number of long-term population and hospital-based clinical studies have examined the incidence and natural history of CHD and its mortality (6,11–26). These studies have largely focused on pediatric patients and tend to be biased toward complex and syndromic forms of CHD that are most likely to come to the attention of pediatric cardiologists, with the majority of individuals having successful outcomes with medical and/or surgical therapy. One study looked at 197 decedents among a group of 2609 adults who were followed by a specialty CHD clinic (24). The highest mortality was in those with congenitally corrected transposition of great arteries, tricuspid atresia, and univentricular connection, with the most common modes of death being sudden death (26%), progressive heart failure (21%), and perioperative death (18%). For those cases with sudden death, the most common diagnoses were aortic coarctation, Ebstein's anomaly, congenitally corrected transposition of the great arteries, congenital aortic valve (AV) anomaly, and TOF.

Forensic pathologists are exposed to a broad cross-section of the population and as a result see a wide range of anomalies from those that are trivial (with an uncertain contribution toward sudden death) to complex forms that may or may not have been diagnosed prior to death (19). As "sudden death investigators," forensic pathologists will encounter previously undiagnosed (and thus untreated) CHD when a person dies unexpectedly. However, depending on the circumstances of death, they might also be called upon to evaluate a previously treated CHD. Furthermore, despite evidence that CHD has a persistent risk of sudden death, family and/or clinician pressures might result in forensic pathology referral. The likelihood of pressure for coronial or medical examiner autopsy is usually greater if the decedent is many years out from surgery and has had intervening stable cardiac function.

There are various degrees of confidence in assigning the cause of death to a heart defect. For instance, conditions considered to have a high level of certainty of causing death (similar to finding 75% coronary arterial stenosis by atherosclerosis) include the existence of a coronary artery arising from the pulmonary artery; the anomalous origin of the left coronary artery from the right sinus with an associated inter-arterial course; aortic stenosis in association with left ventricular hypertrophy; and operated congenital heart defects. Diagnoses with uncertain significance include minor anomalies of the coronary arteries arising from the aorta and unoperated CHD; however, both findings may be of significance in the absence of other causes of death identified by autopsy, toxicology, and ancillary studies (27).

The forensic literature has a paucity of data on adult CHD beyond case reports; the few larger studies have focused primarily on specific subgroups such as young or middle-aged adults (11,14,25) or sudden cardiac death during recreational or competitive sporting activities (28,29). Although uncommon, forensic pathologists should be aware of the wide spectrum of morphologic defects of spatial arrangement, internal organization, chambers, valves, and vessels of the heart, as well as the findings in the postoperative heart. Forensic pathologists should also learn the special methods of evaluation required to properly evaluate a known or suspected congenitally abnormal heart (see Discussion).

A retrospective review of cases from a large medical examiner's office was conducted to examine the spectrum of structural CHD presenting as sudden death in adults.

Methods

The case files of the Miami-Dade County Medical Examiner Department (DCME) from 1991 to 2007 were reviewed in this retrospective descriptive study. Miami-Dade County has an estimated population of 2.5 million people (30). By Florida Statute 406, all cases of sudden, unexpected, and/or violent death occurring within the county must be reported to the DCME. The files at the DCME contain information on all cases falling under the jurisdiction of the office.

Adults (individuals >16 years of age) whose cause of death was attributed to CHD or its sequelae, such as medical and surgical complications, were included. Congenital lesions of the cardiac conduction system such as Wolff–Parkinson–White syndrome, inherited ionic channel abnormalities such as Brugada and long QT syndromes, and the WHO cardiomyopathies were excluded. Other cardiac lesions such as mitral valve prolapse, which can appear later in life in association with hereditofamilial disorders, were also excluded.

Documents available for review in each case file included investigation report, autopsy report, cardiac pathology consultation report (if applicable), classification of pending case form (if applicable), and autopsy photographs. The following data were collected and stored in a Microsoft Access database: age, sex, race, cause and manner of death, circumstances of death, relevant clinical history, whether autopsy was performed, and autopsy findings related to cardiac and other congenital anomalies. Basic statistics were used to analyze the data.

Results

Of 51,228 adult deaths investigated between 1991 and 2007, 79 cases (0.2%) had a cause of death attributed to structural CHD. The age range was 17–90 years; 65% of decedents were men and 35% were women. Autopsies were conducted in 64 cases (autopsy rate 81%); the remaining cases underwent external examination only. One case was excluded from the study because of documentation that was insufficient to classify the cardiac defect. The frequency and classification of cardiac malformations described at autopsy are detailed in Table 1.

In 34 (53.1%) of the autopsied cases, there was no previous diagnosis of a cardiac defect or history of symptoms attributable to CHD. Of the remainder of the cases, 20 (31.7%) had an antemortem diagnosis of CHD and 11 individuals (17.2%) had a history of symptoms referable to a cardiac defect or were being treated for congestive heart failure that was not otherwise specified or not yet diagnosed. Individuals were involved in various levels of activity at the time of their sudden demise: 22 (34.9%) died while sleeping/resting and 30 (47.6%) were awake at the time of death, of which six (20%) were involved in moderate activity (walking, dancing, yard work, and sexual activity) and three (10%) were involved in vigorous activity (running and competitive sports) at the time of the onset of symptoms leading to sudden death. In 11 cases, there was insufficient information to determine activity level at the time of death.

Eleven cases (17.5%) had a history of previous cardiac surgery. Within this group, the age range was 19–55 years, with the average

TABLE 1—*Classification of cardiac malformation and incidence in autopsied cases.*

Type of Cardiac Malformation	Frequency*
Bicuspid aortic valve	37.5% (*n* = 24)
Obstructive lesions	6.3% (*n* = 4)
Idiopathic hypertrophic subaortic stenosis	1.6% (*n* = 1)
Aortic coarctation	1.6% (*n* = 1)
Ebstein anomaly	3.1% (*n* = 2)
Flow lesions	21.9% (*n* = 14)
Tetralogy of Fallot	4.7% (*n* = 3)
Atrial septal defect	11.1% (*n* = 7)
Ventricular septal defect	4.7% (*n* = 3)
Patent ductus arteriosus	1.6% (*n* = 1)
Other	1.6% (*n* = 1)
Univentricular heart	1.6% (*n* = 1)
Mixed (combination of flow and obstructive lesions)	3 (4.7%)
Subaortic stenosis and ventricular septal defect	1.6% (*n* = 1)
Cor triatriatum sinistrum and ventricular septal defect	1.6% (*n* = 1)
Atrial septal defect, ventricular septal defect, and hypoplastic tricuspid valve	1.6% (*n* = 1)
Coronary artery anomalies	26.6% (*n* = 17)
Single coronary artery	3.1% (*n* = 2)
Hypoplastic coronary artery	3.1% (*n* = 2)
Origin from opposite aortic sinus of Vasalva	3.1% (*n* = 2)
Other forms of anomalous origin from the aorta	17.2% (*n* = 11)
Unclassifiable†	1.6% (*n* = 1)

*% of 64 autopsied cases (*n*, number of cases).

†The unclassifiable case had an anomalous termination of the crista terminalis muscle of the right atrium, associated with abnormally shortened conduction fibers within the musculi pectinate.

age being 34.3 years. In all but four cases, surgery occurred more than 13 years prior to death, with an average span between surgery and death of 18.8 years. In two cases, surgery was performed within 2 weeks of death; one case was that of atrial septal defect (sinus venosus type) repair and the other was a Fontan procedure for a univentricular heart with a hypoplastic pulmonary artery. Of the remaining nine individuals with a remote history of cardiac surgery, one had an AV replacement (Ross Procedure) for bicuspid AV, three had repair of TOF, two had surgery for atrial septal defects (ASDs), and one had surgery for aortic coarctation. The remaining two cases had a history of cardiac surgery (not otherwise specified) in the setting of multiple defects; one had cor triatriatum and ventricular septal defect, and the second individual had atrial and ventricular septal defects associated with a hypoplastic tricuspid valve. One individual with bicuspid AV stenosis died while awaiting surgery.

Five cases (7.9%) had a cause of death related to the secondary complications of CHD. Complications included pneumonia, pulmonary arterial thromboses, endocarditis, exsanguination of aortopulmonary and aortopleural fistulae in the setting of repaired aortic coarctation, and aortic dissection associated with a congenitally bicuspid AV.

Bicuspid AV was the most common congenital heart defect discovered at autopsy (24 individuals; 37.5% of cases). The age range of individuals with bicuspid AV was 32–82 years, with the majority (62.5%) being over the age of 50 years. Only five of 24 cases (20.8%) had an antemortem diagnosis; an additional six cases (25%) had symptoms typically associated with an untreated bicuspid AV including angina pectoris, syncope, and congestive heart failure. Severe calcification of the valvular cusps leading to aortic stenosis was discovered in 22 cases (91.7%), and regurgitant bicuspid valves were discovered in two cases (8.3%). In two cases, the primary cause of death was that of ischemic heart disease with bicuspid AV stenosis listed as a contributing factor. Aortic dissection occurred in one case.

TABLE 2—*Type and frequency of cardiac malformations in cases limited to external examination.*

Type of Cardiac Malformation	Frequency*
Obstructive lesions	26.7% (*n* = 4)
Bicuspid aortic valve	6.7% (*n* = 1)
Idiopathic hypertrophic subaortic stenosis	6.7% (*n* = 1)
Aortic coarctation	13.3% (*n* = 2)
Flow lesions	53.3% (*n* = 8)
Tetralogy of Fallot	13.3% (*n* = 2)
Atrial septal defect	26.7% (*n* = 4)
Ventricular septal defect	6.7% (*n* = 1)
Septal defect NOS	6.7% (*n* = 1)
Unclassifiable	20.0% (*n* = 3)
Congenital heart disease NOS	13.3% (*n* = 2)
Congenital heart disease associated with situs inversus	6.7% (*n* = 1)

*% of 15 cases limited by external examination (*n*, number of cases).
NOS, not otherwise specified.

Anomalous coronary arteries accounted for 17 deaths (26.6%). There were 10 men and seven women who ranged in age from 17 to 90 years of age (mean age = 39 years). Two cases (12%) involved young adult men who were participating in a competitive sporting activity at the time of death. There were two cases of a single left coronary orifice that gave rise to both left and right coronary branches. There were two cases of hypoplastic coronary arteries, both involving the right coronary artery. Of note, one of these cases also had a rudimentary sinoatrial nodal artery and there was fibromuscular dysplasia of the sinoatrial nodal and atrioventricular nodal arteries. In two cases, the origin of the right coronary artery was from the left (opposite) sinus. The remaining 11 cases had other anomalous origins from the aorta involving one or both coronary arteries and with one or more of the following findings: acute angulation of the origin, ostial ridges, narrowing (slit-like) orifices, and/or commissural (near the AV commissure) or high ostial origin (1 cm above the sino-tubular junction). Evidence of previous myocardial ischemia (in the absence of significant coronary atherosclerosis) was documented in three cases (17.6%).

The next most common anomalies were isolated ASDs (seven cases), isolated ventricular septal defects (three cases), and TOF (three cases). The ASDs took various forms: two cases had ostium secundum defects, two cases had sinus venosus defects, and three cases did not specify the type of atrial defect. The remaining 10 cases in the study had an assortment of findings ranging from simple (patent ductus arteriosus [PDA]) to complex (univentricular heart), as well as mixed cases with more than one type of malformation.

Two cases (3.2%) involved women who died in the peripartum period. One woman died during the third trimester of pregnancy, and at autopsy was diagnosed with acute angulation of the right coronary artery. The second woman died 2 days after a Cesarean section for breech presentation, and at autopsy had a PDA. The first case was not suspected clinically. The second woman was thought to have a heart defect as a child, but a diagnosis was not made. During pregnancy, she was documented to have episodes of hemoptysis and intermittent peripheral cyanosis.

Not all cases referred to the medical examiner department require autopsy. When a medical examiner determines that an accurate cause of death and natural manner of death can be determined from an evaluation of the scene, circumstances, and history, as well as an external evaluation of the body, an autopsy might not be performed. Family, religious, and other autopsy objections might also be respected in certain noncriminally suspicious cases. Refer to Table 2 for the causes of death listed in the 15 cases that underwent external examination only.

Discussion

As expected, CHD was an uncommon cause of sudden death in adults in a medical examiner's population. The age range of the overall study population was wide, owing at least partly to the wide age distribution of common congenital heart anomalies such as bicuspid AVs. Furthermore, advances in surgical management have allowed many infants born with heart defects of variable severity to survive into adulthood, albeit with a persistent risk of sudden death. As mentioned earlier, there are various substrates for sudden death in operated CHD related to the residua and sequelae of the primary defect, as well as consequences of the operative procedure. Several population- and hospital-based studies have studied the long-term outcomes of patients who have undergone operative repair of congenital heart defects (24,31–37). In studies that evaluated mortality from the perspective of the terminal circumstances, sudden death accounted for 22–24%, progressive heart failure for 21–40%, and immediately perioperative deaths for 18–26% (24,35). It is accepted that the highest risk of arrhythmias and sudden death is encountered following surgical interventions for uncommon malformations, namely post-Mustard and Senning procedure for transposition of the great vessels, post-Fontan procedure for defects with univentricular physiology, and repair of TOF (8). For the common congenital heart malformations, the risk of sudden death after repair is only slightly higher or comparable to that of the general population (37). In our study population, all the patients dying days, weeks, months, or years following operative procedures experienced sudden (presumably arrhythmic) cardiac death. One individual also had symptoms of congestive heart failure.

Bicuspid AVs were the most common malformation in our study, consistent with the fact that it is the most common congenital heart defect, affecting *c.* 1–2% of the general population (38,39). The fact that many individuals present with sudden death, often at older ages, is entirely consistent with the natural history of the disease as patients can be entirely asymptomatic for years prior to the onset of cardiovascular decompensation (40,41). The most common complication identified in our study was aortic stenosis related to severe calcification of the bicuspid valve and subsequent left ventricular hypertrophy. AV regurgitation and aortic dissection were much less frequent complications. Interestingly, there were no cases of sudden death caused by infective endocarditis from bicuspid AV, another common complication.

Coronary artery anomalies affect *c.* 1% of the population, either in isolation or in combination with other forms of congenital heart defects (42). In our study, roughly one-quarter of all congenital anomalies were isolated anomalous coronary arteries. Although often an incidental finding, variations in the course and origin, intrinsic anatomy, and termination of coronary arteries are increasingly recognized as a potential cause of sudden death, reflected by numerous published case reports and case series (42–54). Specific anomalies most often associated with mortality are anomalous origin from the pulmonary trunk; anomalous origin from the aorta including ostial ridges, acute angulation of the origin, and origin from the opposite sinus of Vasalva; single coronary ostium arising from the aorta; and hypoplastic coronary arteries. Anomalous coronary arteries are thought to lead to sudden death by reduced coronary blood flow and subsequent myocardial ischemia. Origin from the pulmonary trunk leads to shunting between coronaries and coronary steal. Although pulmonary trunk origin of the coronary arteries (the classic coronary malformation associated with sudden death) was not recognized in any of our study cases, it should be a diagnostic consideration for sudden cardiac death in adults. Although it is typically fatal in infancy, it may rarely present in late adulthood with sudden death (46). The various anomalies of origin from the aorta also lead to decreased flow into the affected coronary artery. If the coronary artery takes an unusual course between other cardiac structures, the vessel may be prone to compression or forced to take acute bends in its course, which also puts the myocardium supplied by that artery at risk (4). The origin of a coronary artery from the opposite aortic sinus has most often been associated with sudden death when the left coronary arises from the right coronary sinus. Sudden death associated with the right coronary artery arising from the left sinus has been documented less frequently (4,43,45,51). Evidence for these mechanisms is supported by the documentation of chronic myocardial ischemia in association with coronary artery anomalies.

A retrospective autopsy review of isolated coronary artery anomalies published by the Armed Forces Institute of Pathology (AFIP) illustrated the spectrum and incidence of anomalies diagnosed in their institution: 16.9% of cases had an anomalous origin from the pulmonary trunk, 57.8% of cases had an anomalous origin of a coronary artery from the aorta, 18.2% of cases had a single coronary ostium from the aorta, and 5.4% of cases had hypoplastic coronary arteries. The AFIP study also noted that most lethal anomalies occurred in those under 40 years of age. Furthermore, the AFIP authors felt that coronary arterial anomalies detected during the autopsy of adults older than 40 years were merely incidental and not lethal. In our study, 46.1% of anomalous coronary arteries were diagnosed in adults older than 40 years; we felt that these diagnoses were correct given the totality of available investigative, autopsy and special study data.

The AFIP dataset indicated that sudden death occurred in 32% of cases, with 45% of those deaths occurring during exercise (4). The risk of sudden death occurring during exercise in patients with coronary artery anomalies is well documented by case series of sudden death in young adults during recreational and competitive sporting activities and in military recruits (14,28,29,55,56).

Isolated ASDs are common and can be of various forms including ostium primum, ostium secundum, and sinus venosus. Unless the ASD is significantly large, most individuals are not symptomatic until early adulthood, at which point the left ventricle becomes less compliant resulting in increased left-to-right shunting. Complications of long-standing large ASDs include supraventricular arrhythmias, endocarditis, right ventricular failure, recurrent pulmonary infections, pulmonary hypertension (that predisposes to pulmonary thrombosis), and pulmonary vascular obstruction with Eisenmenger syndrome and reversal of shunting. Sudden death is uncommon in the natural history of ASD, especially if individuals undergo early closure. In our study, all but one decedent had pathologic features of pulmonary hypertension; the one individual who did not have an atrial septal defect associated with extensive right ventricular fibrosis involving both the atrioventricular and sinoatrial nodes. Similar pathophysiology and complications also occur in ventricular septal defects and PDA. Both are common malformations in the general population, and the majority close spontaneously in infancy. Morbidity is high if any of these lesions go undetected as once severe pulmonary hypertension develops, surgical therapy is usually contraindicated; late repair of ASD and VSD is also associated with morbidity (57–61). Uncommonly, PDA may be complicated by aneurysm formation and rupture of the ductus in adults and may be seen following infective endarteritis, surgical closure, or transcatheter coil occlusion (59).

Our study data did not include any cases of untreated aortic coarctation. However, one case of previously treated coarctation was complicated by exsanguination from aortopleural and aortopulmonary fistulae. Complications that could be identified in adults

with late-presentation coarctation include systemic arterial hypertension, left ventricular failure, aortic dissection, premature coronary artery disease, infective endocarditis, and subarachnoid hemorrhages because of berry aneurysms (that occur in association with coarctation). Morbidity is also increased in those with late repair of coarctation (57).

In our study, there were no cases of transposition of the great vessels, but there was one case of an univentricular heart where the individual was in the immediate postoperative period of a Fontan procedure, and three cases of TOF with remote repair. These cases are consistent with the literature on sudden death for these conditions. For repair of hypoplastic left heart syndrome or univentricular heart, the highest postoperative mortality is within the first 48 h after surgery; a significant risk for mortality remains for the first few months and then decreases over time. Unexpected death reportedly occurs in 4.1% of individuals discharged home after stage I Fontan for hypoplastic left heart syndrome (32). For TOF, the long-term survival is poor with a threefold risk of sudden cardiac death within 10 years after surgery, which remains for 30 years, especially if repair occurs at a late age (36). Late sudden death after operative repair has also been observed with aortic stenosis and aortic coarctation (37); one case of each was identified in our study. Three individuals were postoperative for the repair of ASDs; although it is generally thought that those with ASD repair have good long-term survival, given the proximity of the repair site to either the sinoatrial or atrioventricular nodes, it is not entirely unexpected that there is an association with sudden death.

Congenital heart disease is a known cause of maternal mortality during pregnancy. In fact, now that the majority of women born with CHD are surviving to reproductive age in the developed world, combined with the dramatic decline in rheumatic heart disease, CHD is now more common in pregnant women than acquired heart disease (62,63). The majority of women with CHD are able to accommodate the normal physiologic changes of pregnancy without any increased risk of mortality. However, a significant number of women are unable to tolerate the increased cardiovascular demands and physiologic hypercoagulability of pregnancy even if they were clinically stable prior to pregnancy. Furthermore, they may be significantly taxed by the dramatic hemodynamic changes that occur during labor. As with any individual with CHD, the mortality risk is related not only to the primary cardiac lesion, but also with the type of previous surgical intervention and any residua of the primary lesion or sequelae of the primary lesion or surgery.

Although the evidence basis for pregnancy risk in CHD is accumulating and is moving from consensus/expert opinion to valid data from large single centers, more long-term outcome data are needed. Defects currently considered to be low risk (i.e., risk similar to general population) in pregnancy include surgically repaired ASD, VSD, PDA, pulmonic stenosis, and AV stenosis; unoperated bicuspid AV (with no obstruction); small ASD/VSD; and mild pulmonic stenosis. The intermediate category with a mild to moderate risk include unrepaired large ASD/VSD; aortic coarctation; moderate pulmonic stenosis; Ebstein's anomaly and congenitally corrected TGA; and operated aortic coarcation, TOF, and Ebstein's anomaly. Moderate- to high-risk lesions include repaired single ventricle and tricuspid atresia (Fontan procedure), TGA (atrial repair), pulmonary atresia (Rastelli procedure), and ventricular dysfunction. High-risk lesions with very poor tolerance for pregnancy are cyanotic CHD of any type, pulmonary hypertension of any cause, and Eisenmenger syndrome. Complications include endocarditis, embolization, arrhythmias, hypertension, ventricular tachycardias, ventricular dysfunction, venous thromboembolism/pulmonary embolism, and maternal hemorrhage related to delivery. Maternal mortality in Eisenmenger syndrome is estimated to be between 41% and 67%, with fetal loss as high as 50–60% (63).

Our study contained data on one pregnant woman and one woman who died immediately postpartum—neither of whom had an antemortem diagnosis. The woman who died during puerperium had an unrepaired PDA. An unrepaired PDA would fall into the category of low-intermediate risk depending on the size of the shunt; if the shunt is significantly large with the establishment of pulmonary hypertension or Eisenmenger syndrome, then the pregnancy would be contraindicated by the high risk of maternal and fetal morbidity and mortality (64,65). The individual in our study would likely have fallen into the latter group had the diagnosis been made antemortem. The individual who died during pregnancy had acute angulation of her right coronary artery as the only finding at autopsy. Coronary artery anomalies are not any more common in pregnancy; like in any unexplained sudden death of an adult, they should be a diagnostic consideration during pregnancy. A case of sudden death in a woman in her second trimester of pregnancy who had multiple anomalies of her coronary arteries has previously been reported (66).

In addition to the heart defects that occur commonly in the general population, forensic pathologists may also encounter rare and complex malformations. In our study, there was an unusual case of cor triatriatum in association with a ventricular septal defect. Cor triatriatum is a rare malformation where one of the atria, usually the left atrium, is divided by a fibromuscular septum into upper and lower chambers, which communicate through a defect(s) in the septum. The malformation has various forms; in the usual configuration, the pulmonary veins enter the upper chamber and the lower chamber harbors the atrial appendage and the mitral valve. It may occur as an isolated lesion or in combination with other malformations and depending on its form may have a clinical presentation similar to anomalous pulmonary venous return or mitral stenosis. Although usually diagnosed early in life, it has been rarely diagnosed at autopsy in late adulthood (67).

With regard to the practical aspects of the sudden cardiac death autopsy, there are published guidelines that have focused on the approach to CHD (19,27). It is recommended that the congenitally malformed heart not be processed by serial sectioning of the heart—the approach routinely utilized by forensic pathologists who are in search of atherosclerotic coronary artery disease. If one suspects CHD by history or initial inspection of heart *in situ*, the heart should be analyzed *in situ*, noting its relationship to the lungs and thoracic vessels. Clues that there may be a congenital heart defect include malposition of the heart; abnormal heart size or shape; asplenia or polysplenia; malposition of any thoracoabdominal organ, including situs inversus (68); abnormal lobation of the liver or lungs; and malrotation of the bowel. Subsequently, a modified Rokitansky approach with segmental dissection and analysis of the atria, ventricular, and arterial segments and their relationships to each other should be undertaken.

Common mistakes in dissection that limit the ability to properly characterize the malformed heart are separation of the heart from the aorta, pulmonary arteries, or lungs prior to examination; serial sectioning of the coronary arteries without acknowledging the anomalous origins or placement of the vessels; obscuring lesions by opening up the heart along septal defects; poorly placed incisions; and discarding the aortic trunk without careful examination (19). If one is not comfortable with dissection of the congenitally malformed heart and prefers to consult a pathologist with specialized knowledge in cardiac pathology, it is important to consider the above guidelines to maintain the integrity of the cardiopulmonary

block along with the thoracic vessels (including the aortic arch) for complete characterization of the malformation.

Conclusion

Congenital heart disease is a rare but important cause of sudden death in adults, not only for those with previously unrecognized lesions, but also for those with previously diagnosed, stable disease. Although uncommon, CHD should be a consideration for anyone presenting with sudden cardiac death, so that an appropriate autopsy investigation is undertaken to allow for appropriate characterization of defects. Improved surgical techniques and medical management have prolonged the lives of those born with heart defects, including those with complex cyanotic CHD who historically would never have survived beyond infancy. In a large medical examiner's population of adults presenting with sudden death, bicuspid AVs and anomalous coronary arteries are the CHDs most commonly recognized at autopsy. However, a wide spectrum of simple (such as ASDs) to complex (such as univentricular hearts) malformations can be diagnosed. Once a solely pediatric entity, CHD is now grown-up, and in the future will likely be seen with increased frequency in forensic pathology settings.

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