H73 Sudden Death (SD) in Adults With Congenital Heart Disease (CHD): A Retrospective Review of Cases in the Cook County Medical Examiner's Office

Lorenzo Gitto, MD, State University of New York Upstate, Department of Pathology, Syracuse, NY 13210; Steven M. White, MD, PhD, Cook County Medical Examiner's Office, Chicago, IL 60612; Ponni Arunkumar, MD, Cook County Medical Examiner's Office, Chicago, IL 60612; Serenella Serinelli, MD*, State University of New York Upstate, Department of Pathology, Syracuse, NY 13210*

Learning Overview: After attending this presentation, attendees will better understand the causes of SD in adults with diagnosed and undiagnosed CHD in Cook County, IL.

Impact on the Forensic Science Community: This presentation will impact the forensic science community by providing information about the anatomic defects (simple or complex, single or multiple) and the medical history in adult CHD cases in a medical examiner's population.

CHD in adults is associated with a high rate of SD, despite improved life expectancy because of better surgical and interventional therapies. Most congenital heart defects are repaired in childhood; however, there may be less severe defects that go undiagnosed, resulting in SD in adulthood. The annual incidence of SD in the entire adult CHD population is relatively low, but because the CHD population is growing and aging, the risk profile for SD may have changed.

The electronic database of the Cook County Medical Examiner's Office in Chicago, IL, was examined using the keyword "congenital," searching for cases of CHD as a primary or contributory cause of death, in the period between July 2008 and July 2018. Cases were reviewed for age, sex, race, cause and manner of death, medical history, type of congenital heart disease, and gross and microscopic cardiac findings. Only subjects between 18 and 99 years of age were included in this study.

A total of 84,820 deaths were reported in the studied period in the Cook County Medical Examiner's Office. Of these, there were 15 cases (0.02%) of adult CHD identified, including 14 natural deaths and 1 accidental death. The age ranged between 21 and 62 years. Seven subjects were Caucasian, six African-American, one Hispanic, and one Asian. The male/female ratio was 2.7/1. A complete autopsy was performed in 14 out of 15 cases with consultation with a cardiac pathologist requested in 7 cases.

Of the 14 hearts examine, 12 hearts were enlarged, 8 were dilated, and 9 showed hypertrophy. Simple cardiac defects were observed in 7 cases (50%): 2 cases with isolated Atrial Septal Defects (ASD); 2 cases with isolated Ventricular Septal Defects (VSD); 1 case with VSD combined with pulmonary stenosis, 1 case of bicuspid aortic valve, and 1 case of congenital anomaly of the left coronary ostium. Complex defects were observed in 7 cases (50%): 2 tetralogies of Fallot, 1 Ebstein's anomaly, 1 double outlet right ventricle, 1 transposition of the great vessels alone, 1 transposition of great vessels combined with tricuspid atresia, and 1 Total Anomalous Pulmonary Venous Return (TAPVR).

Diagnosis of CHD was known in 12 cases out of 15 (80%) and, among these, 11 subjects had undergone prior (single or multiple) surgical repair. In 13 out of 15 cases, CHD was the primary cause of death, while it was a contributory cause in 2 cases. Toxicological analyses were negative in 14 out of 15 cases, while in 1 case the subject was found to be positive for a non-toxic level of ethanol.

Despite the limited number of cases, CHDs were very heterogeneous and SDs were more common in adult males (73%). Almost all the CHDs were previously known. The three unknown CHD cases consisted of valvular diseases or coronary defect, which could explain the absence of clinical signs and, therefore, the lack of diagnoses. It is questionable whether these minor abnormalities can cause SD, but in the absence of other potentially lethal pathology at autopsy, they may be used as a cause of death.

In 11 out of the 12 cases (92%) with a known diagnosis of CHD, surgical correction was performed in childhood and 3 of them required additional surgical interventions in adulthood. Since subjects suffering from these complex CHD may require multiple surgical corrections during their lifetimes, there could be misinterpretation of the original cardiac defect at autopsy because of the distortion of the usual anatomy due to the surgeries. It is important for the forensic pathologist to be trained in recognizing CHDs and the surgical procedures used to treat them. When the diagnosis is not clear, the forensic pathologist should request a consultation with a cardiac pathologist experienced in CHD.

Congenital Heart Disease, Adult Cardiac Pathology, Sudden Death