G120 Isolated Coronary Anomalies and Sudden Death in the Young

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After attending this presentation, attendees will be able to better understand the importance of coronary examination (origin, course, and lumen canalization) during autopsy of young people suddenly dead, to identify coronary artery anomalies.

This presentation will impact the forensic science community by improving knowledge of natural causes of juvenile sudden death (SD).

Background: Coronary artery anomalies (CAAs) had been some of the most confusing, neglected topics in cardiology. In the 1990s, the subject of CAAs underwent to profound evolution related to substantive methodological changes regarding the definition, incidence, morphology, clinical presentation, diagnostic work-up, prognosis, and treatment. CAAs are most frequently found in association with congenital hearth diseases (great arteries transposition, tetralogy of Fallot, common artery trunk) and hypertrophic or dilated cardiomyopathy, but can also occur in the absence of other cardiovascular diseases ("*isolated coronary artery anomalies*"). Isolated CAAs are rare, found in: 0.2–2.2% of autopsies of all ages; 0.5% of pediatric autopsies; 0.6–1.3% of coronary angiograms in adults. CAAs represent common causes of exercise-related sudden death (SD) in young people (< 35 years of age). The mechanism of SD is believed to be episodic myocardial ischemia.

Objectives: The goal of the current study is to detect the frequency, type, and possible pathophysiology due to "isolated" CAAs in an autopsy population of the young, suddenly dead for cardiovascular diseases (CSD).

Methods: In the time interval from January 1990 to December 2009, 236 consecutive cases were collected of cardiovascular sudden death in young people. In all of the cases, the analysis of death circumstance (*most of the deaths analyzed were testified*), the complete autopsy and the toxicological essays let us establish that death was: natural, violent, sudden. The juvenile CSD was defined as unexpected death as a result of natural cardiovascular causes within one hour of initial symptoms in persons < 35 years of age.

Results: Forty nine sudden deaths in young people were reported, identified solely at autopsy and due to: right coronary artery from the left sinus (n = 15); right coronary artery above anterior commissure (n = 11); left coronary artery from right coronary sinus (n = 3); intramyocardial course (n = 10); obstructive valve-like ridge in the Valsalva's sinus and intra-right coronary ostium (n = 9); left anterior descending artery from right coronary sinus (n = 1). The CAAs was either isolated (n = 43, 87.8%) and associated to hypertrophic cardiomyopathy (n = 6, 12.2%). In all patients (43 males and 6 female, age ranging from 13 months to 35 years; median, 22.6), sudden death was the first manifestation of the disease and familial history was negative. The fatal outcome occurred after physical effort (n = 27, 55%), at rest (n = 16, 32.6%), or after emotional stress (n = 6, 12.4%). Unquestionable ischemic damage within the related myocardium, in the absence of obstructive coronary atherosclerosis or other cardiac diseases, was observed in all cases: acute myocardial infarction (n = 29, 59.2%), healing myocardial infarction (n = 4, 8.2%), healed myocardial infarction (n = 16, 32.6%). In this study of Juvenile CSD, death was precipitated by isolated CAAs in 21% of cases.

Conclusion: Data from this collection confirms that isolated CAAs may account for juvenile CSD and that fatal event is frequently the first

manifestation of the disease, it is precipitated by effort and depends on ischemic damage within the related myocardium. Recognition during life of these anomalies, by the use of non-invasive procedures, is mandatory to prevent the risk of SD and to plan the screening in competitive athletes.

Coronary Artery Anomalies, Juvenile Sudden Death, Forensic Pathology

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