G90 A "Café Coronary" in a 2-Year-Old: Case Report

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After attending this presentation, attendees will understand the history of the term "café coronary" and the mechanisms, genetics, presenting signs and symptoms, and pathologic findings of abnormal cholesterol metabolism involved in familial hypercholesterolemia.

This presentation will impact the forensic community by reviewing the role genetic diseases play in fatal, premature pediatric coronary artery disease. During café coronary events, myocardial ischemia should be considered as a cause of death, even in the pediatric population, and especially if there is a family history of premature coronary artery disease or familial dyslipidemia.

"Café coronary" is a term used to describe a sudden attack resulting in death that occurs during or shortly after eating, often in the elderly, and is sec- ondary to choking; however, the death is erroneously attributed to coronary artery disease. In children and adolescents, the opposite scenario, death in a suspected choking victim having a final diagnosis of myocardial ischemia secondary to coronary artery disease, is extremely rare.

Childhood is a critical period in which dietary and lifestyle patterns have long-term implications for coronary heart disease risk in adult life. Smoking, high intake of dietary total fat and saturated fat, low exercise level, and excessive alcohol consumption are correlated with elevated serum cholesterol, obesity, and hypertension in children, as well as a predisposition to premature death from coronary heart disease.

Children and adolescents can be at an even higher risk of cardiovascular disease if there is a family history of premature coronary artery disease or familial dyslipidemia. Of the primary hyperlipidemias, familial hypercholesterolemia (FH) is the most common and the most documented to have important cardiovascular consequences beginning in childhood. FH is an inherited dominant condition due to a defect in the LDL receptor gene and is usually discovered when there are increases in plasma total and LDL cholesterol in the child and in at least one of the parents. More than 600 different LDL-receptor mutations have been described. Mutations of the LDL-receptor cause significantly elevated LDL levels. This inability for cholesterol uptake leads to premature atherosclerosis and a very high risk of early cardiovascular disease and myocardial infarction. Patients with homozygous FH manifest cardiovascular disease within the first two decades of life, and may present within the first decade of life with physical findings related to cholesterol deposition, such as tendon xanthomata, cutaneous xanthelasma, or corneal arcus. FH heterozygotes usually present with problems in early to mid-adulthood.

A 2-year-old Hispanic male appeared to be suffering from a "café coronary" while eating, but was actually suffering from acute myocardial ischemia secondary to >90% stenosis of multiple coronary arteries. Initial responders and emergency department personnel proceeded with resuscitative procedures/protocols in response to a presumed choking/ asphyxia event. Autopsy revealed extensive cholesterol deposition in the coronary arteries with additional deposits found throughout the aorta and within the skin (xanthomas). The decedent's family history was significant for a father and 12-year-old sister with hypercholesterolemia. A recent visit to the pediatrician revealed fasting plasma total cholesterol >400 mg/dl. It is recommended that medicolegal death investigators become familiar with the possibility of an acute cardiac death in young children with a family history of abnormal cholesterol metabolism.

Café Coronary, Children, Familial Hypercholesterolemia

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