



### **G47 Clinical and Pathological Spectrum of Fatty Cardiomyopathy in Sudden Cardiac Death**

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After attending this presentation, attendees will recognize the pattern of pathology findings as correlated to clinical information from cases within the spectrum of fat cardiomyopathy including arrhythmogenic right ventricular cardiomyopathy.

This presentation will impact the forensic community by reviewing clinical and pathological data for sudden cardiac death cases from the spectrum of fat cardiomyopathy. Forensic and cardiovascular pathologists, as well as other forensic scientists, may find this information useful for comparison with observations from their home institutions and practices.

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a primary heart muscle disease characterized by fibrofatty replacement of the right ventricle (RV) that is commonly associated with sudden death. Infiltration of the RV by fat alone is also believed to be associated with sudden death. However, it is not yet fully known if both conditions are different or similar disease processes in the spectrum of fatty cardiomyopathy and to what extent fatty infiltration of the RV to contribute to sudden cardiac death. In this study, the authors set to characterize the clinical and pathological characteristics of thirteen necropsy hearts collected by the iCAPTURE Cardiovascular (CV) Biobank at St. Paul's Hospital/University of British Columbia and diagnosed with ARVC or right ventricular fatty infiltration from patients that died suddenly and unexpectedly. Each case was referred to a cardiovascular pathologist at the CV Biobank for consultation and patient data were obtained from medical records or the referring pathologist. The CV Biobank, established in 1993, is comprised of cardiovascular tissue specimens from surgery and autopsy (routine hospital and forensic) as well as accompanying annotated data which are securely stored in a database. This unique collection encompasses a wide array of cardiovascular diseases and is a powerful research and educational tool.

These thirteen cases were collected by the CV Biobank during the years 1993 to 2006 and represent approximately 14% of our sudden death cases over this time period. Each of the thirteen cases were assessed in terms of their macroscopic and microscopic features and were found to fit into one of two patterns. Nearly two-thirds demonstrated fibrofatty (six male, two female; age = 17-36 years) replacement of the right ventricular myocardium, while slightly more than 1/3 showed a pattern of predominantly fatty replacement (2 male, 3 female; age = 15-64 years). Within the fatty replacement group, 80% of individuals died following non-strenuous activity and 20% died at rest. Patient histories for this group included one individual with history of fainting and clinical intervention for arrhythmia and one patient with a history of anorexia and bulimia. Within the fibrofatty replacement group, over 57% of individuals died following non-strenuous activity; 28% during strenuous activity and 14% at rest. Patient histories for this group include one individual with documented familial ventricular tachycardia for which he received treatment; one history of dilated cardiomyopathy and mitral valve regurgitation; and one individual with sudden death of a brother due to an unspecified aneurysm. Quantitative computer-assisted morphometric analysis in a subset of seven of the thirteen cases confirmed these two patterns. Of interest, the distribution and extent of involvement differed substantially between fibrofatty and fatty patterns with changes being more extreme and widely distributed in the fibrofatty pattern and localized to the anterolateral apex and lateral base in the fatty pattern.

Thus, fibrofatty replacement of the RV, characteristic of ARVC, and fatty infiltration of the RV alone are both significant findings in cases of sudden cardiac death evaluated at a regional cardiovascular pathology referral centre. The distinctly differing extent and distribution of involvement between the two morphological patterns supports the concept that they represent two different disease processes.

#### **Fat Cardiomyopathy/ARVC, Cardiovascular Pathology, Sudden Cardiac Death**