

Idiopathic Ventricular Fibrillation

Definition

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Definition: Idiopathic ventricular fibrillation (IVF) is defined as unexplained sudden cardiac death due to ventricular fibrillation (VF) without any identifiable structural or electrical cause after extensive investigations (no phenotype). SCD is commonly defined as death from an unexpected circulatory arrest, occurring within an hour of the onset of symptoms.

When VF has been documented by electrocardiography during resuscitation maneuvers, unexplained SCD is termed as IVF. (**Visser M, van der Heijden JF, Doevendans PA, Loh P, Wilde AA, Hassink RJ. Idiopathic Ventricular Fibrillation: The Struggle for Definition, Diagnosis, and Follow-Up. *Circ Arrhythm Electrophysiol.* 2016 May;9(5):e003817. doi: 10.1161/CIRCEP.115.003817.**), (**Krahn AD, Healey JS, Chauhan V. Systematic assessment of patients with unexplained cardiac arrest: cardiac arrest survivors with preserved ejection fraction registry (CASPER). *Circulation* 2009;120: 278–85. 14.**).

It is very important a careful 12-lead ECG documentation of premature ventricular contractions (PVCs) morphologies and couplings characteristics because CA among patients with IVF has a very distinctive mode of onset: with very short coupling intervals. The PVC initiating VF had a coupling interval of 302 +/- 52 msec (297 +/- 41 ms) and a prematurity index of 0.4 +/- 0.07. These PVCs occurred within 40 ms of the peak of the preceding T wave. VT that is not pause dependent is of diagnostic value. (**Viskin S, Lesh MD, Eldar M, et al. Mode of onset of malignant ventricular arrhythmias in idiopathic ventricular**

fibrillation. J Cardiovasc Electrophysiol. 1997 Oct;8(10): 1115-20. doi: 10.1111/j.1540-8167. 1997.tb00997.x.) PVCs are originated from the distal Purkinje conducting system in » 50% of patients. (These sources can be eliminated by focal energy delivery (**Michel Haïssaguerre 1, Morio Shoda, Pierre Jaïs, Akihiko Nogami, Dipen C Shah, Josef Kautzner, Thomas Arentz, Dietrich Kalushe, Dominique Lamaison, Mike Griffith, Fernando Cruz, Angelo de Paola, Fiorenzo Gaita, Mèlèze Hocini, Stéphane Garrigue, Laurent Macle, Rukshen Weerasooriya, Jacques Clémenty Mapping and ablation of idiopathic ventricular fibrillation Circulation. 2002 Aug 20;106(8):962-7. doi: 10.1161/01.cir.0000027564.55739.b1.**), from the left ventricular septum (in » 23% of cases), from the RVOT in >1%, and from both in <1% of cases.

No clinical evidence for drug abuse/intoxication at the time of initial presentation;

No clinical evidence for electrolyte disturbance at the time of initial presentation;

No identifiable Structural Heart Disease (SHD) demonstrated by normal echocardiographic and absence of delayed gadolinium-enhanced MRI. (before ICD);

No detectable coronary artery disease on coronary angiography or exercise testing;

No known repolarization abnormalities associated with long or short QT interval or J-wave syndromes;

Negative pharmacologic testing with infusion of a Class I sodium channel blocker (ajmaline, flecainide, or procainamide) for to exclude Brugada syndrome;

Negative catecholamine infusion with isoprenaline and adrenaline to exclude catecholaminergic polymorphic ventricular tachycardia (CPVT) or ARVC and to confirm the absence of LQTS. In both IVF and CPVT, no baseline ECG abnormalities can be detected, (**Christian Wolpert 1, Mara Vogel 2, Christian Nagel 2, Claudia Herrera-Siklody 2, Norman Rüb 2**Ventricular

arrhythmias in ion channel diseases. *Herzschrittmacherther Elektrophysiol.* 2017 Jun;28(2):169-176. doi: 10.1007/s00399-017-0510-6.)

The spectrum of IVF leading to SCD in apparently normal hearts, is divide in 2 categories, based on the primary pathogenesis.

- 1) Conduction abnormality: heterogeneity of depolarization with localized structural abnormality and
- 2) Excitation abnormality with primary VF substrate of rapid or multifocal ectopic hyperactivity without SHD and from Purkinje or myocardial foci.