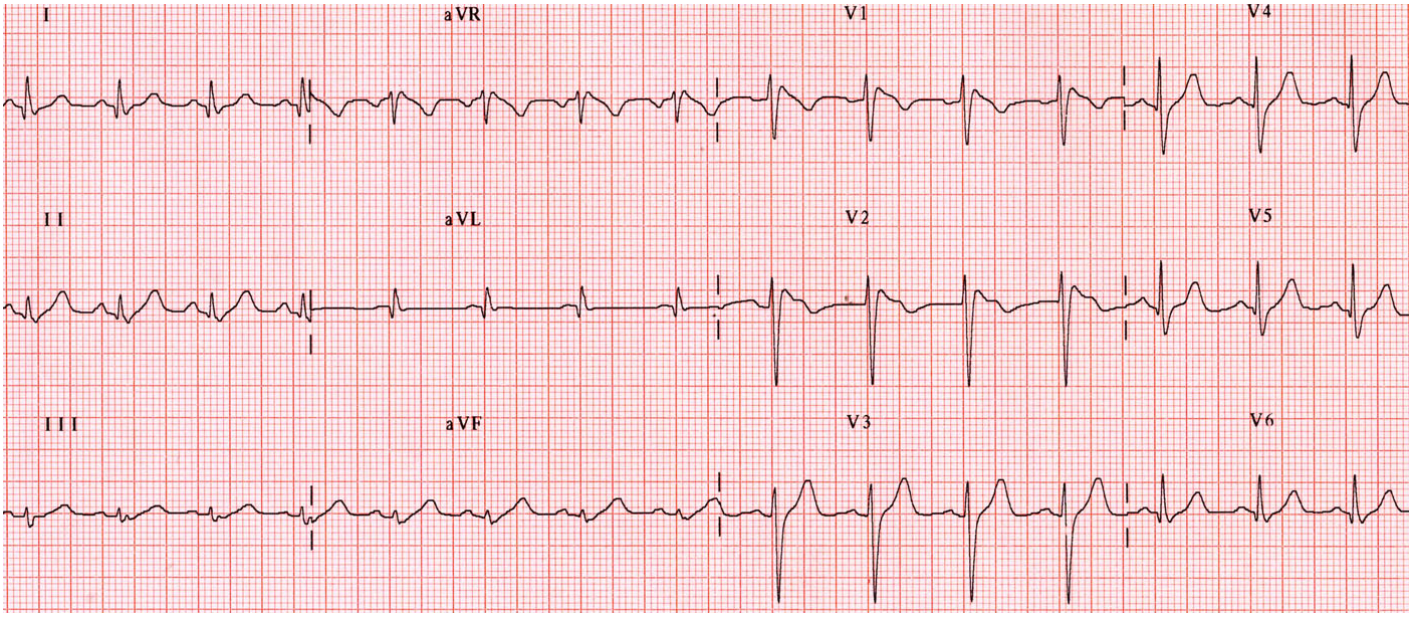
The first case in the world of Brugada Phenocopy due to coronary anomaly

Brugada Syndrome (BrS) is an arrhythmogenic disease responsible for at least 20% of sudden deaths in patients without structural heart disease and is the leading cause of death in subjects under the age of 40 years. Currently, the cornerstone of therapy for the prevention of sudden cardiac deaths from BrS is an implantable cardioverter defibrillator.

Brugada Phenocopies (BrP) are clinical entities that present with identical ECG patterns to those of true BrS but are elicited by various other clinical circumstances. They form a group of heterogeneous conditions that are perhaps the most difficult to differentiate from true congenital BrS due to identical ECG patterns. There is growing interest in the mechanisms responsible for this acquired ECG pattern and all these conditions must be carefully considered and excluded.



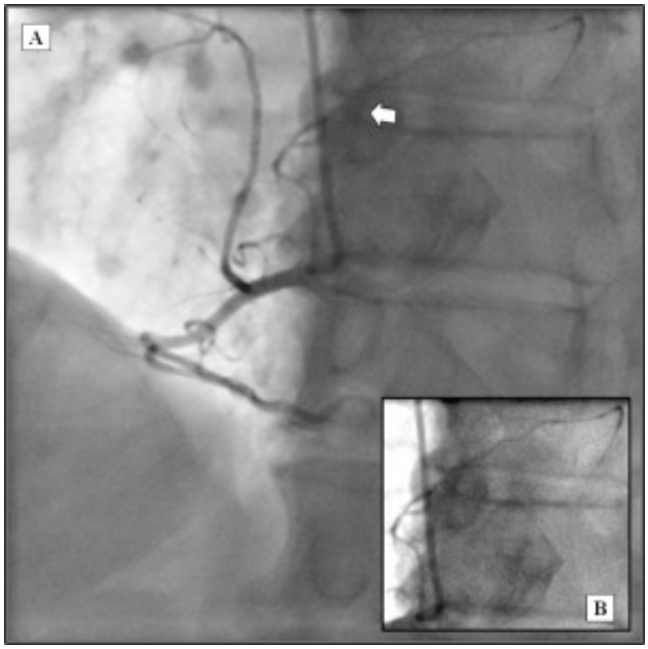
ECG at the admission with Brugada type I pattern.

A thirty-eight years old man was admitted to our department for an acute coronary syndrome. A Brugada type I ECG pattern was found. The percutaneous coronary angiography, performed with right femoral artery access, showed no coronary atherosclerotic lesions and two fistulous communications, the first arising from the conus artery with probable drainage located at the mediastinal vessels and the second arising from the atrial artery with probable drainage located at the right bronchial circulation.

Given the headquarters of this fistula, a temporary ischemia at the level of the right ventricular outflow tract (RVOT) resulted in a temporary transmural dispersion of repolarization/slow conduction, which could be at the base of the ECG manifestations. At the restoration of a regular coronary artery flow at the level of the RVOT the ECG manifestations did not appear more.

On the basis of good general clinical condition, the patient refused any further investigation and intervention for closure of the fistulas, hence he was treated conservatively with medical therapy.

Many diseases and conditions can lead to a BrP, including electrolyte disturbances, hypothermia, acute myocardial infarction/ischemia, acute pericarditis, right ventricular cardiomyopathy, acute stroke, central and autonomic nervous system abnormalities, muscular dystrophy, etc.



Panel **A**: coronary artery angiography showing two fistulous communications. The first arising from the conus artery with probable drainage located at the mediastinal vessels (arrow), the second arising from the atrial artery with probable drainage located at the right bronchial circulation. Panel **B**: enlarged detail of the fistula arising from the conus artery.

Currently in literature there are a limited number of BrP cases associated with ischemia and recently Dr. G. Dendramis have published an association between the presence of a coronary artery fistula at the level of the conus artery and Brugada type I ECG pattern in a patient without personal or family history suggestive of BrS and negative provocative challenge. As said by Byron H Gottschalk et al., this is the first description present in the literature to date of confirmed BrP due to an anomaly in the coronary circulation and it provides evidence that atypical causes of

myocardial ischemia may induce BrP and the diagnosis should be considered in such cases.

This case teaches us as, at the finding of a Brugada ECG pattern and before anything else, we must exclude any other structural heart diseases or others factors that may also lead to a BrP.

At the manifestation of a BrP, patients should be carefully studied because different therapeutic approaches are required for patients with BrP than in patients with true BrS. We hope that in the future there will be more attention when clinicians will be faced with an ECG with the typical Brugada pattern so we can exclude first of all causes of BrP and only subsequently direct our diagnostic doubt on the real BrS.

An international registry and online educational portal provides an updated registry of BrP cases along with the diagnostic criteria (Brugada phenocopy international registry and online educational portal, available online at www.brugadaphenocopy.com).

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Publication

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