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**Article** *in* Inteligencia artificial revista iberoamericana de inteligencia artificial · October 2012

DOI: 10.5031/v3i1.RIA10190



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# Brugada Phenocopy in a patient with surgically repaired Pentalogy of Fallot

**Running Title:** Brugada Phenocopy & Pentalogy of Fallot

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**Disclosures:** None

**Key Words:** Brugada Phenocopy, Brugada Syndrome, Tetralogy of Fallot, Pentalogy of Fallot

**Word Count:** 982

**Abstract**

Brugada Phenocopies are continuing to emerge as clinical entities that are distinct from true congenital Brugada Syndrome (BrS). They are characterized by ECG patterns that are indistinguishable from BrS; however, their etiology and pathophysiology is quite diverse. We report the first documented case of an electrocardiographic Brugada Phenocopy emerging during the immediate postoperative period in a patient who underwent surgical Pentology of Fallot repair. This case suggests that a Brugada Phenocopy ECG presenting within 96 hours of RVOT manipulation negates the requirement of provocative testing with flecainide, procainamide, ajmaline, or other sodium channel blockers.

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**Resumen**

Las Fenocopias del Síndrome de Brugada continúan emergiendo como entidades clínicas que son diferentes de la verdadera forma congénita del Síndrome de Brugada. Las fenocopias se caracterizan por tener un patrón electrocardiográfico que son imposibles de diferenciar de la forma congénita. Sin embargo la etiología y fisiopatología de estas fenocopias es bastante variada. Describimos el primer caso documentado de una fenocopia electrocardiográfica del patrón de Brugada que se manifestó durante el post-operatorio inmediato de un paciente sometido a una corrección quirúrgica de una pentalogia de Fallot. Este caso sugiere que una fenocopia electrocardiográfica del patrón de Brugada se puede observar dentro de las primeras 96 horas después de la manipulación del tracto de salida del ventrículo derecho no requiriéndose pruebas con flecainida, ajmalina o procainamida para descartar disfunción iónica .

## Introduction

Brugada Syndrome (BrS) is a congenital cardiac channelopathy that affects repolarisation in patients with apparent structurally normal hearts predisposing them to malignant ventricular arrhythmias and sudden cardiac death. It is characterized by two distinct ECG patterns. Type-1 has a "coved" ST-segment elevation

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(≥ 0.2 mV) with inverted T-wave in the right precordial leads and Type-2 has a "saddleback" appearance.[1]
Indistinguishable Brugada ECG patterns, referred to as Brugada Phenocopies,[2,3] continue to emerge in the absence of true congenital BrS and their etiology is diverse including metabolic, structural, and ischemic causes.
Pentalogy of Fallot (POF) is a congenital heart defect that includes the 4 classical characteristics of Tetralogy of Fallot (TOF) along with an atrial septal defect.[4] We describe the first case in literature of an electrocardiographic Brugada Phenocopy emerging postoperatively in a patient who underwent surgical POF repair.

**Case Report and ECG Description**

A 7 year-old girl with POF and otherwise unremarkable medical history underwent total surgical repair. Her preoperative 12-lead ECG (Figure 1) showed: sinus rhythm, heart rate 60 bpm, P-wave duration 85 ms, PR interval 167 ms, QRS duration 79 ms, QT interval 380 ms, QTc 424 ms, with extreme right axis deviation (-150°). In lead V1 she had a notched R-wave in the ascending peak.

The surgical technique consisted of right ventriculotomy and resection of the RVOT without valve replacement. A Gore-Tex patch was used to close both ventricular and atrial communications.

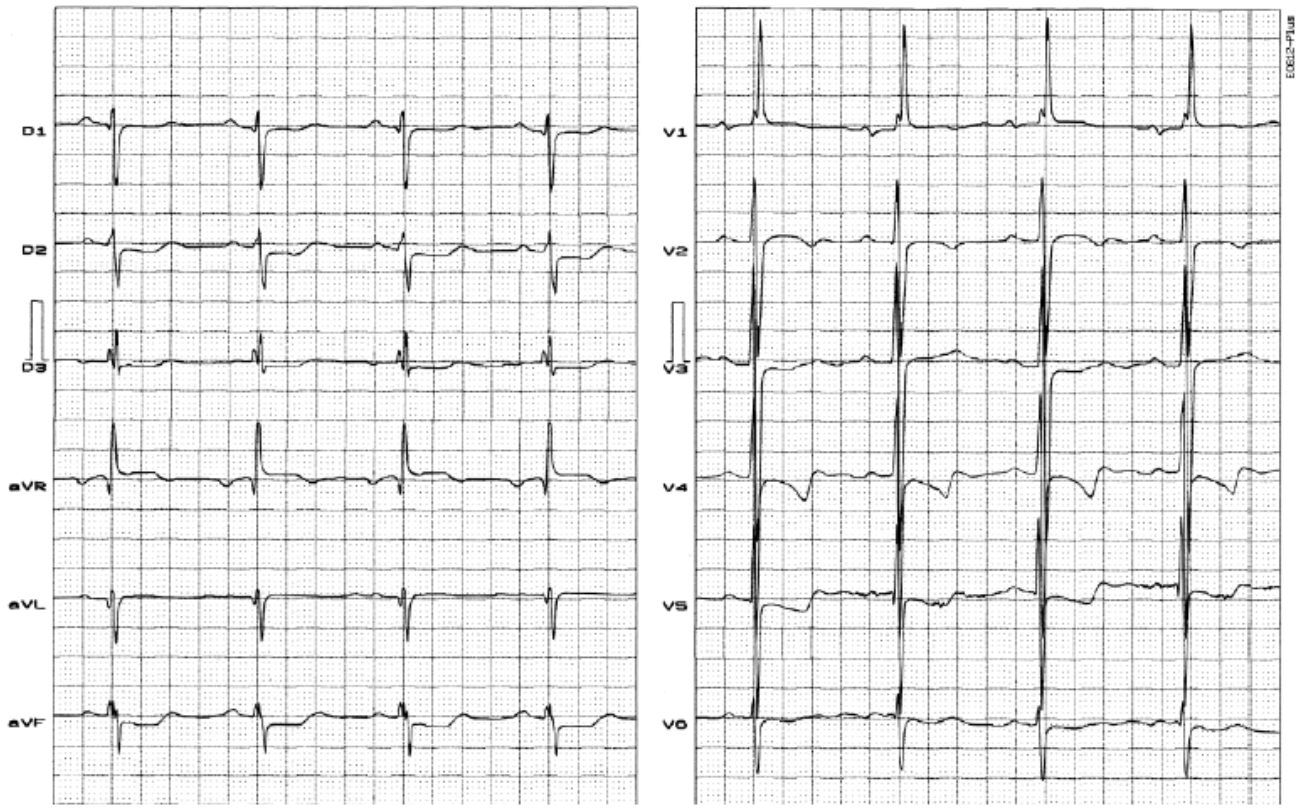


**Figure 1: Preoperative electrocardiogram**
Sinus rhythm, heart rate 60 bpm, P-wave duration 85 ms, PR interval 167 ms, QRS duration 79 ms, QT interval 380 ms, QTc 424 ms, with extreme right axis deviation (-150°). In lead V1 she had a notched R-wave in the ascending peak.

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During the immediate postoperative period her 12-lead ECG (Figure 2) showed: sinus rhythm, heart rate 107 bpm, P-wave duration 85 ms, PR interval 200 ms, QRS duration 120 ms, with extreme right axis deviation (-160°). The ECG pattern was consistent with a Brugada Type-1 ECG pattern (coved). There was also ST-depression in the lateral (V4-V6) and inferior leads (II, III, aVF). Her postoperative hospital stay was unremarkable and she was discharged home after 7 days. During outpatient follow-up an ECG performed at 3 months postoperatively (Figure 3) showed: sinus rhythm, heart rate 65 bpm, P-wave duration 60 ms, PR interval 166 ms, QRS duration 120 ms, with extreme right axis deviation (-160°). The ECG showed resolution of the Brugada Type-1 ECG pattern observed earlier; however, continued to show complete RBBB with an rsR in V1 and V2, and RsR' in V3. There was also prominent R-wave with wide S-wave in I, aVL, V5 and V6. There were negative T-waves in V1-V4 and ST-depression in V4-V6. This pattern was also observed during long-term follow-up.

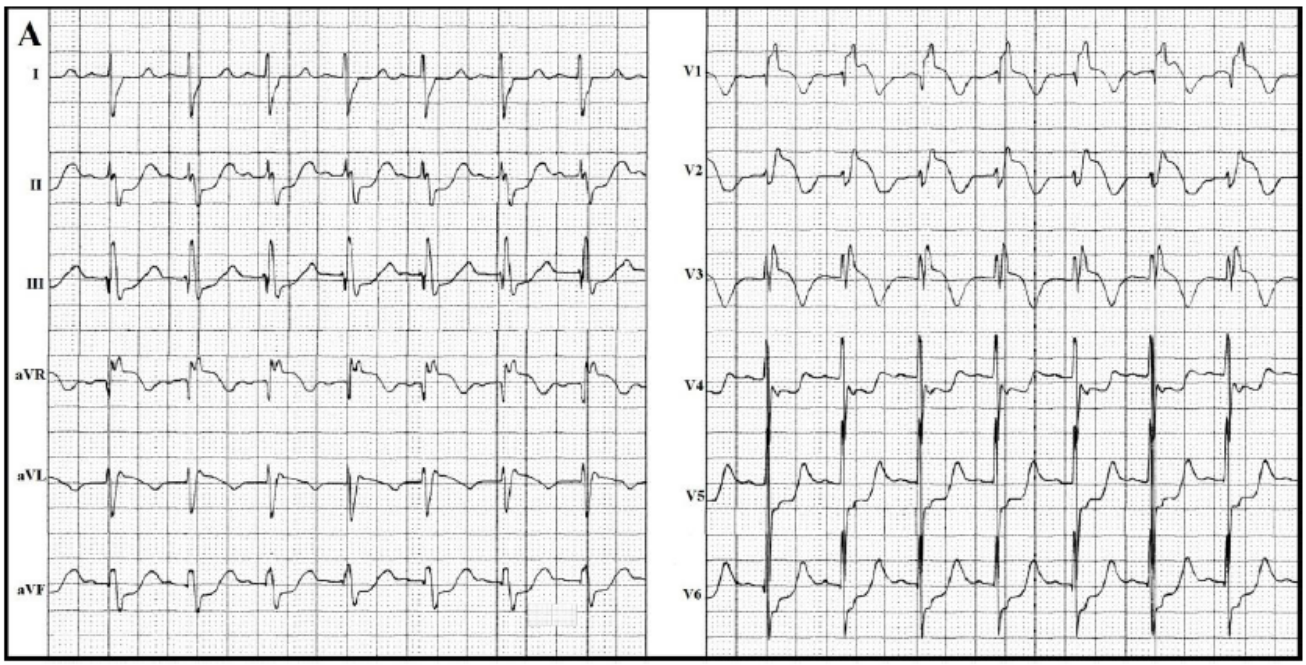


**Figure 2: Immediate postoperative electrocardiogram**
Sinus rhythm, heart rate 107 bpm, P-wave duration 85 ms, PR interval 200 ms, QRS duration 120 ms, with extreme right axis deviation (-160°). The ECG pattern was consistent with complete right bundle branch block (RBBB) with an rsR complex in V1, rsR' in V2 and RSR' in V3. There was a prominent R-wave in I, aVL, V5 and V6. There was ST-elevation and T-wave inversion in the right precordial leads consistent with a Brugada Type-1 ECG pattern. There was also ST-depression in the lateral (V4-V6) and inferior leads (II, III, aVF).

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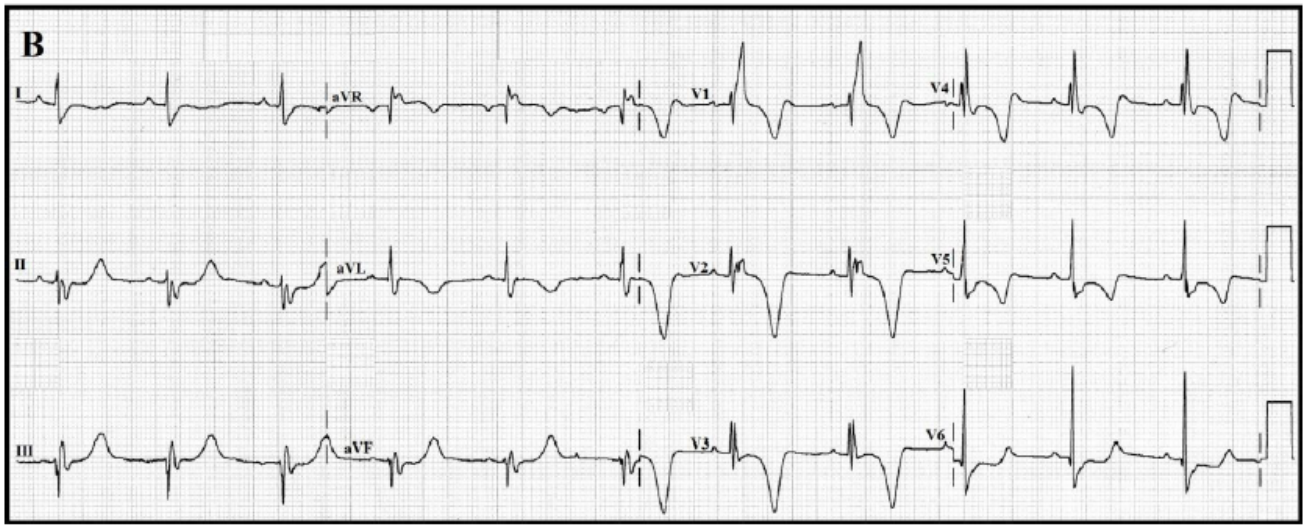


**Figure 3: 3 month postoperative electrocardiogram**
Sinus rhythm, heart rate 65 bpm, P-wave duration 60 ms, PR interval 166 ms, QRS duration 120 ms, with extreme right axis deviation (-160°). The ECG showed resolution of the Brugada Type-1 ECG pattern however continued to show complete RBBB with an rsR in V1 and V2, and RsR' in V3. There was also prominent R-wave with wide S-wave in I, aVL, V5 and V6. There were negative T-waves in V1-V4 and ST-depression in V4-V6.

**Points to Ponder**

TOF is one of the most commonly encountered cyanotic congenital heart lesions with a prevalence of approximately 3.9 per 10,000 live births in the United States.[5] The defect occurs approximately equally in both males and females and accounts for about 7 to 10 percent of all congenital heart lesions.[6] In about 10 percent of cases, an atrial septal defect is associated with TOF[7] resulting in the so-called POF.

Brugada Phenocopies are continuing to emerge as distinct clinical entities. Recently, we updated the criteria for defining a Brugada Phenocopy[3] as follows: (i) the ECG pattern has a Brugada Type-1 or Type-2 morphology; (ii) the patient has an underlying condition that is identifiable; (iii) the ECG pattern resolves after resolution of the underlying condition; (iv) there is a low clinical pre-test probability for true BrS determined by lack of symptoms, past medical history and family history; (v) negative provocative testing with flecainide, procainamide, ajmaline, or other sodium channel blockers; (vi) negative genetic testing (non-mandatory criterion). The patient in this case report satisfied criteria (i) through (iv); however, this case introduces an additional caveat to point (v) in that provocative testing may not be required if there has been recent manipulation of the RVOT.

In this patient, we observed a transient Brugada Type-1 ECG pattern during the immediate postoperative period which resolved at 3 month follow-up leaving the patient with a persistent complete RBBB. Interestingly, Gelband et al[8] demonstrated that surgical right ventriculotomy was strongly associated with RBBB and they attributed the changes in right ventricular activation to the surgical ventriculotomy itself. Their postoperative ECG's, however, were taken 9 days after surgery and not in the immediate postoperative period as in our case. Hence, in their surgical cases the ST-segment elevation in the right precordial leads may have resolved by the time their postoperative ECG's were recorded.

We hypothesize that in our case, manipulation of the RVOT during surgical POF repair may aggravate the anterior RVOT epicardium resulting in a transient Brugada Phenocopy during the immediate postoperative period. As such, we suggest that point (v) of the Brugada Phenocopy definition be revised as follows: negative provocative testing with flecainide, procainamide, ajmaline, or other sodium channel blockers *(non-mandatory if RVOT manipulation has occurred within the last 96 hours)*. We chose the 96 hour cut-off because prior studies have shown that most surgical chest tubes should be removed within the first 24 hours postoperatively[9] and most epicardial pacing wires are removed by the 4th postoperative day because they begin to deteriorate.[10]

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**Conclusion**

Brugada Phenocopies exhibit Type-1 or Type-2 ECG patterns in the absence of true congenital BrS. This is the first case report of a patient with surgically corrected POF demonstrating a Brugada Type-1 ECG pattern during the immediate postoperative period that resolved in follow-up. The mechanism of this transient pattern may be associated with manipulation of the anterior RVOT epicardium.

**References**

**1.** Bayés de Luna A, Brugada J, Baranchuk A,Borggrefe M, Breithardt G, Goldwasser D, Lambiase P, Riera AP, Garcia-Niebla J, Pastore C, Oreto G, McKenna W, Zareba W, Brugada R, Brugada P. Current electrocardiographic criteria for diagnosis of Brugada pattern: a consensus report. J Electrocardiol 2012;45:433-42.

**2.** Baranchuk A, Nguyen T, Ryu MH, Femenía F, Zareba W, Wilde AAM, Shimizu W, Brugada P, Pérez-Riera AR. Brugada Phenocopy: New Terminology and Proposed Classification. Ann Noninvasive Electrocardiol 2012;17:1-16.

**3.** Anselm DD, Baranchuk A. Brugada Phenocopy: redefinition and updated classification. Am J Cardiol. 2012 (In press).

**4.** Brickner ME, Hillis LD, Lange RA. Congenital heart disease in adults. Second of two parts. N Engl J Med 2000;342:334-42.

**5.** Centers for Disease Control and Prevention (CDC). Improved national prevalence estimates for 18 selected major birth defects--United States, 1999-2001. MMWR Morb Mortal Wkly Rep 2006;54:1301-5.

**6.** Report of the New England Regional Infant Cardiac Program. Pediatrics. 1980;65:375-461.

**7.** Rao BN, Anderson RC, Edwards JE. Anatomic variations in the tetralogy of Fallot. Am Heart J 1971;81:361-71.

**8.** Gelband H, Waldo AL, Kaiser GA, Bowman FO Jr, Malm JR, Hoffman BF. Etiology of right bundle-branch block in patients undergoing total correction of tetralogy of Fallot. Circulation 1971;44:1022-33.

**9.** Abramov D, Yeshayahu M, Tsodikov V, Gatot I, Orman S, Gavriel A, Chorni I, Tuvbin D, Tager S, Apelbom A. Timing of chest tube removal after coronary artery bypass surgery. J Card Surg 2005;20:142-6.

**10.** Elmi F, Tullo NG, Khalighi K. Natural history and predictors of temporary epicardial pacemaker wire function in patients after open heart surgery. Cardiology 2002;98:175-80.