

Brugada Syndrome

Introduction

The term “Brugada Syndrome” is used when the Brugada ECG is accompanied by symptoms of syncope or cardiac arrest. The majority of patients present with the incidental finding of a Brugada ECG¹. The Brugada pattern is present in 3 out of 1000 persons. The vast majority of patients with the Brugada ECG will not have any symptoms related to it. There are three types of Brugada ECG (Figure). The pattern of clinical concern is the type 1 pattern, which is present in only one out of every 6000 persons. The most common is the type 2 pattern (81% of cases)¹.

Diagnosis

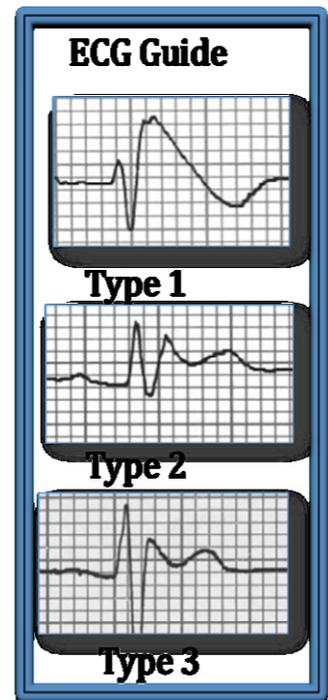
The diagnosis of Brugada ECG is largely made by the resting ECG²⁻⁷. Placing the ECG leads in a higher position on the chest improves the ability to detect the type 1 ECG finding (termed sensitivity)⁸. The ECG often fluctuates over time, and may even be normal at times. Certain drugs bring out the ECG finding, and are used to unmask the type 1 ECG when the diagnosis is suspected or a type 2 or 3 ECG is present. This involves an intravenous infusion of a drug called procainamide^{9,10}. Similarly, the Brugada ECG may be detected when patients are exposed to certain drugs that have sodium channel blocking properties, which should be avoided by Brugada patients. A list of these drugs is available at www.brugadadrugs.org. Fever is another potential precipitant of the ECG¹¹.

Genetic testing is often performed in patients with a type 1 Brugada ECG, primarily for use in screening family members. The yield of genetic testing is only 25%.

A small risk of sudden death exists in patients with Brugada Syndrome, and there may be a very small risk in asymptomatic patients with a type 1 ECG (Brugada ECG, not Syndrome)^{2,12}.

Treatment

Symptomatic patients who have experienced a cardiac arrest or syncope often undergo implantation of an ICD. An asymptomatic patient who shows both spontaneous and/or drug-induced type I ECG should discuss the small risk with their doctor to determine what is best for them. Patients are usually advised to avoid drugs that unmask or worsen the Brugada ECG (www.brugadadrugs.org), and to carry a copy of their ECG with them to avoid misinterpretation when presenting for evaluation of another problem. The Brugada ECG is similar to that seen during a “heart attack” caused by loss of blood supply to the heart, which is often treated with aggressive blood thinners and invasive procedures in an emergency fashion.



Selected References

1. Gallagher MM, Forleo GB, Behr ER, et al. Prevalence and significance of Brugada-type ECG in 12,012 apparently healthy European subjects. *Int J Cardiol* 2008;130:44-8.
2. Probst V, Veltmann C, Eckardt L, et al. Long-term prognosis of patients diagnosed with Brugada syndrome: Results from the FINGER Brugada Syndrome Registry. *Circulation* 2010;121:635-43.
3. Veltmann C, Schimpf R, Echternach C, et al. A prospective study on spontaneous fluctuations between diagnostic and non-diagnostic ECGs in Brugada syndrome: implications for correct phenotyping and risk stratification. *Eur Heart J* 2006;27:2544-52.
4. Antzelevitch C, Brugada P, Borggrefe M, et al. Brugada syndrome: report of the second consensus conference. *Heart Rhythm* 2005;2:429-40.
5. Brugada J, Brugada R, Antzelevitch C, Towbin J, Nademanee K, Brugada P. Long-Term Follow-Up of Individuals With the Electrocardiographic Pattern of Right Bundle-Branch Block and ST-Segment Elevation in Precordial Leads V1 to V3. *Circulation* 2002;105:73-8.
6. Priori SG, Napolitano C, Gasparini M, et al. Natural history of Brugada syndrome: insights for risk stratification and management. *Circulation* 2002;105:1342-7.
7. Brugada J, Brugada R, Brugada P. Right bundle-branch block and ST-segment elevation in leads V1 through V3: a marker for sudden death in patients without demonstrable structural heart disease. *Circulation* 1998 Feb 10;97:457-60.
8. Sangwatanaroj S, Prechawat S, Sunsaneewitayakul B, Sithisook S, Tosukhowong P, Tungsanga K. New electrocardiographic leads and the procainamide test for the detection of the Brugada sign in sudden unexplained death syndrome survivors and their relatives. *Eur Heart J* 2001;22:2290-6.
9. Krahn AD, Gollob M, Yee R, et al. Diagnosis of unexplained cardiac arrest: role of adrenaline and procainamide infusion. *Circulation* 2005;112:2228-34.
10. Brugada R, Brugada J, Antzelevitch C, et al. Sodium channel blockers identify risk for sudden death in patients with ST-segment elevation and right bundle branch block but structurally normal hearts. *Circulation* 2000 Feb 8;101:510-5.
11. Keller DI, Rougier JS, Kucera JP, et al. Brugada syndrome and fever: genetic and molecular characterization of patients carrying SCN5A mutations. *Cardiovasc Res* 2005;67:510-9.
12. Kaplunger JD, Tester DJ, Alders M, et al. An international compendium of mutations in the SCN5A-encoded cardiac sodium channel in patients referred for Brugada syndrome genetic testing. *Heart Rhythm* 2010;7:33-46.