Brugada Syndrome in the Asian Population

1. Is Brugada syndrome more common in Asian patients? If so, why?

Question submitted by: Mona Lee, Vancouver, British Columbia

Brugada syndrome is a rare inherited disorder, characterized by ST elevation in the anterior ECG leads (predominantly in leads V1-V2) and risk of ventricular fibrillation, syncope and sudden death. The presence of the ECG findings without cardiac events is termed Brugada sign.

The diagnosis of Brugada syndrome should be considered in a patient with syncope or resuscitated cardiac arrest with anterior precordial ST elevation in the absence of evidence of coronary artery disease or underlying cardiomyopathy. The majority of patients are male and the average age of symptom onset is 40-years-of-age. The Brugada sign is present in 1:4,000 to 1:10,000 patients and is more common in Asians. However, it has been described in a wide range of ethnic backgrounds including Native Canadians.

The underlying defect leading to Brugada syndrome in approximately 20% of cases is a mutation in the SCN5A gene resulting in a loss of function defect in the cardiac sodium channel. Additional genes that have been implicated involve mutations that encode the α1-(CACNA1C) and β-(CACNB2b) subunits of the L-type cardiac calcium channel.

Genetic testing is available, and detects a mutation in 20% of patients with Brugada syndrome and a positive family history and in < 5% of patients without a family history.

Patients with resuscitated cardiac arrest or syncope suggestive of an arrhythmia are typically managed with an implantable cardioverter defibrillator. Drug therapy with quinidine showed promise in one series, but β–blockers or amiodarone have not been shown to be beneficial. Asymptomatic carriers should avoid drugs with sodium channel blocking effects, including tricyclic antidepressants, certain antibiotics, local anesthetics, propofol, lithium, cocaine and several antiarrhythmic agents.

Answered by:
Dr. Brett Heilbron

Evaluating Exercise Stress Testing in Middle-Aged Women

2. How accurate is a negative exercise stress test in working up a typical chest pain in a middle-aged woman? What features would encourage you to book an imaging scan?

Question submitted by: Dr. W Reimer, Kitchener, Ontario

Middle-aged women being evaluated for possible ischemic chest pain are more likely to present with atypical pain than men. In the largest study evaluating exercise tests in women presenting with chest pain, only 32% had significant coronary artery disease compared with a similar population of men with 72% coronary artery disease. Because of the lower prevalence of coronary artery disease in middle-aged women, a negative exercise test is more likely to be a "true negative."

Exercise tests in middle-aged women have become less popular because of the increased frequency of false positive ECG tests. The ECG, however, is only part of the test. For women, as well as for men, the duration of exercise and the presence or absence of exercise-induced chest pain, as well as, ECG changes should be considered in assessing the level of risk for coronary disease. For middle-aged women who have a "negative exercise test" which implies an exercise duration of more than six minutes, without ECG changes and no symptoms or atypical symptoms, the risk of having significant coronary disease is exceedingly small. This "negative test" is actually better at excluding coronary disease in women than in men.

When assessing a middle-aged woman who presents with atypical chest pain, the presence of resting ECG abnormalities, including bundle branch blocks, would lead me to request a myocardial perfusion scan. Also, poor performance on an exercise test with low duration of exercise, or the development of exercise limiting chest pain in the absence of ECG abnormalities should lead to a myocardial perfusion scan.

Answered by:
Dr. Wayne Warnica

Middle-aged women being evaluated for possible ischemic chest pain are more likely to present with atypical pain than men.

Bundle Branch Blocks

3. When should one worry about bundle branch blocks (BBB)?

Question submitted by: Dr. Claude Leblanc, Calgary, Alberta

As with any laboratory or imaging test, BBB should be interpreted in the light of the clinical situation and previous ECGs. In an observational study, left BBB (LBBB) was seen in 0.4% of men 50-years-of-age and 5.7% of those ≥ 80-years-of-age. For right BBB (RBBB), the figures are 0.8% and 11.3% respectively.

A new LBBB is often indicative of coronary artery disease. Known LBBB interferes with interpretation of the ECG during acute coronary syndromes and, more importantly, the interpretation of the exercise tolerance test. In a patient with heart failure, LBBB is evidence of ventricular dysynchrony.

Such patient may benefit from an implantable cardioverter defibrillator or biventricular pacing. In the Framingham study, only 11% of subjects with LBBB remained free of heart disease > 18-years-of-age.

RBBB can develop in patients with pulmonary embolism and is fairly commonly seen in *cor pulmonale*. It is rarely seen in normal individuals.

Answered by:
Dr. Thomas Wilson

BBB should be interpreted in the light of the clinical situation and previous ECGs.