Brugada Syndrome was first characterized by Drs. Pedro and Josep Brugada in 1992 as an electrocardiographic right bundle branch pattern with persistent ST-segment elevation in the right precordial leads. It is associated with sudden cardiac death (1), as the electrical abnormality can lead to sudden cardiac dysrhythmias, loss of consciousness, and death. Recent advances in electrocardiogram screening, genetic testing, long-term outcomes, and treatment modalities have established guidelines for evaluating at-risk populations. With careful cardiovascular evaluation, risk assessment, and follow-up, a pathway for Special Issuance of an FAA medical certificate is possible.

Case Presentation

A 35-year-old male commercial airline pilot from the middle east with no significant medical history presented for his annual FAA first-class medical certificate. The pilot reports being in his usual state of excellent health and does not report using prescription medications, over-the-counter supplements, recent surgeries, or medical problems since his last exam. He recently turned 35 years old, and a screening ECG was performed. The ECG showed a right bundle branch (RBBB) pattern with elevated ST segments and positive T wave morphology in leads V2 and V3.

Case Discussion

Brugada pattern and Brugada syndrome are autosomal dominant conditions involving the cardiac sodium channel associated with characteristic ECG pattern of RBBB and ST segment elevations in V1 to V3 leads. Brugada pattern describes the ECG pattern in an asymptomatic patient. The Brugada pattern can be further divided into two main types.

1. Brugada type 1 ECG pattern is characterized by a widened terminal R, ST segment elevation, and terminal T wave inversion that is described as “coved type” pattern
2. Brugada type 2 ECG pattern represents a narrower RBBB pattern with J point elevation and ST segment elevation with a positive T wave deflection, also known as “saddleback” pattern

While three different types of Brugada patterns were initially described, two distinct Brugada ECG pattern types are currently used for assessment, treatment, and risk evaluation (1). Brugada syndrome is characterized by either Brugada type of ECG pattern, along with history of sudden, unexplained death of a family member, history of ventricular fibrillation, or syncope.

Different prevalence rates have been reported. In Japan, the prevalence of Brugada ECG pattern is as high as 1.0%, while prevalence in US urban population ranged from 0.012 to 0.4% (2,3). Early studies investigating sudden, unexpected death looked at structural cardiac abnormalities in post-mortem autopsies. Although some researchers found evidence of cardiac dilation, localized inflammation, and fibrosis, newer studies show associations with SCN5A and SCN10A cardiac sodium channel gene mutations. Of note, the presence of a genetic mutation is associated with only 17 to 30% expression of Brugada syndrome (4).

Brugada syndrome was first described as a clinical entity with specific ECG findings in 1992, but southeast Asian cultures have long recognized sudden unexplained death syndrome known as bangungot (Philippines), pokkuri (Japan), and lai tai (Thailand) as one of the leading causes of death in males younger than 50 years (7). Brugada syndrome is considered an autosomal dominant sodium channelopathy with a population ECG prevalence as high as 1% in Japan and average age of first diagnosis at 41 (2,8). Recent research has linked genetic mutations to Brugada pattern, but less than half of patients that have the mutation manifest as clinical Brugada syndrome.

ECG morphology and clinical history are important factors in determining the category of disease and assessing risk for sudden cardiac events. ECGs with RBBB with prolonged R' and inverted T wave in V1 to V3 are considered to have Brugada type 1 or “coved type” pattern. ECGs with RBBB with J point elevation, ST segment elevation and positive T wave are Brugada type 2 or “saddleback” pattern. History of ventricular arrhythmia, unexplained syncope, and family history of sudden deaths in first-degree relatives with Brugada ECG findings are consistent with Brugada syndrome, rather than Brugada pattern.

FAA risk assessment for Brugada pattern versus syndrome involves ruling out other cardiac disease, obtaining history of unexplained syncope or ventricular arrhythmias, family history of sudden unexpected death, and assessing the Brugada type that could contribute to an incapacitating event. Therefore, an evaluation by a cardiologist or an electrophysiologist with detailed history for ventricular arrhythmias, family history, arrhythmia-related symptoms, and risk assessment for annual recurrence are required for consideration of special issuance of an FAA medical certificate (7). Dysrhythmias should be assessed with a 24-hour Holter monitor study, structural abnormalities evaluated with an echocardiogram or cardiac MRI, ischemia analyzed with a maximal exercise stress test, changing morphology of ECG pattern tested with three serial ECGs done at least 24 hours apart, and classification of Brugada pattern versus Brugada disease by obtaining a detailed personal and family history for symptoms (9).
Aeromedical Issues

The primary aeromedical concern for Brugada pattern and syndrome is related to the risk for sudden incapacitation secondary to ventricular arrhythmias while performing aviation duties. The etiology for ventricular arrhythmias may be due to blunted sodium channel depolarization that results in decreased propagation of the action potential (5). The combination of localized block and short refractory period could serve as the foci for localized reentry and ventricular arrhythmia (5).

Results from long-term Brugada registry showed a 7.7% cardiac event rate per year in patients with Brugada type 1 syndrome with history of ventricular fibrillation, 1.9%-5% with history of syncope, and 0.5% in asymptomatic patients (6).

Risk factors associated with higher rates of cardiac events include male sex, family history of sudden unexplained death, Brugada type 1 ECG morphology, and syncope (7). Currently, opinions differ for the need for provocative testing for stratifying risk in patients with Brugada pattern. While Benito’s group (7) found higher rates of overall cardiac events in electrophysiologically inducible patients, the FINGER long-term registry study did not find increased risk (6).

Overall, patients with Brugada type 1 syndrome may develop ventricular arrhythmia at a rate of 7.7% per year; patients with Brugada type 1 pattern have a rate of 2.3% events per year. Therefore, Brugada type 1 pattern and syndrome are disqualifying for all FAA medical certification (6).

Pilots that have Brugada type 2 syndrome (saddleback ECG findings) and history of ventricular fibrillation or syncope have 1.39% calculated events per year; therefore, the Brugada type 2 syndrome is also disqualifying (8). However, Brugada type 2 ECG pattern without symptoms have less than 0.5% events per year and may be eligible for a time-limited FAA medical certificate (9, 10).

In some patients, the characteristic ECG changes of the Brugada pattern are transient or variable over time. Type 2 pattern initially may subsequently manifest type 1 morphology. Among patients with the type 2 Brugada ECG pattern, the type 1 Brugada ECG pattern can be unmasked by sodium channel blockers (e.g., flecainide, procainamide).

Outcome

The pilot obtained an electrophysiology cardiology consult. A detailed history showed no evidence of syncope, light headedness, palpitations, or nighttime agonal respirations. His family history did not reveal any sudden, unexpected deaths. He denies taking any over-the-counter medications or sports nutritional supplements. Cardiac evaluation found normal echocardiogram, normal maximal exercise stress test without development of arrhythmia or ischemia, and a normal Holter monitor study. Three separate ECGs performed over the course of 7 days showed normal ECG without RBBB pattern. However, the ECGs continued to show ST segment elevation in V2, consistent with Brugada type 2 “saddleback” pattern. The Federal Air Surgeon’s cardiology consultant for EP recommended provocative challenge with flecainide or procainamide, if no conversion to type I then certification.

The airman remains unissued pending the provocative drug challenge.

References


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