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Heart to Heart

With Philip S. Chua, M.D.

Brugada Syndrome (Unexplained Cardiac Arrest)

What is Brugada Syndrome?

Brugada Syndrome is a baffling condition where a person, who appears to be healthy, unexpectedly develops cardiac arrest, for no apparent reason. Most victims do not survive the episode. This condition is now believed to be responsible for countless sudden deaths among young people - athletes, students, etc., - who suddenly dropped dead.

Did the victims have any prior symptoms?

No, majority of victims did not have any prior symptoms. They were “in good health” when suddenly they collapsed, went into coma, and later died, or instantly died on the spot. There were some individuals who were lucky enough to have responded to prompt cardiopulmonary resuscitation. Patients like these, with aborted cardiac arrest and death, inspired the three Doctors Brugada (R, P & J) and their colleagues at the Unitat d'Aritmies, Hospital Clinic, in Barcelona, Spain, to pioneer in the study of this baffling condition, and became the first to describe the syndrome in the medical literature as a functional cardiac disorder with unique EKG findings. To honor them for this astute discovery, their peers around the world named the entity Brugada Syndrome.

What were their findings?

The study conducted by the Doctors Brugada in 1992 included 8 patients with a history of aborted sudden death with a distinct and specific EKG tracings that were abnormal (in medical lingo, there were right bundle branch block, ST segment elevation in the right precordial leads, and prolonged QT interval), in the absence of any structural or anatomical heart defect. The physical and clinical examination, biochemical tests, echocardiographic and angiographic tests among these patients yielded normal findings. In 4 of them, a family history of unexplained sudden cardiac arrest was present. The occurrence of ventricular fibrillation (beginning cardiac arrest) in the absence of any structural defect classifies this as a

“primary electrical disease.” These specific and characteristic EKG findings among these patients discovered by the Doctors Brugada, together with the family history of unexplained cardiac arrest, are today the paradigms for the diagnosis of the Brugada Syndrome.

When was this syndrome first discovered?

In science, it is usually hard to accurately date the very first actual discovery of anything, because many scientists/physicians/researchers work simultaneously all around the world. Many of them do not even report their findings right away. But the first officially reported EKG findings, which later was to be known as Brugada Syndrome, was made in the late 1980 by other physicians, where 6 patients with sudden cardiac arrest were resuscitated successfully and found to have these characteristic and unique EKG tracings to be described in 1992 by the Doctors Brugada as a distinct disease entity, which heretofore was undiscovered and unexplained.

How many cases of this syndrome are there?

This entity is increasingly recognized. The Doctors Brugada reported 63 cases by 1998 and a Japanese multicenter reported an additional 63. The past few years, another 163 cases were discovered that satisfy the Brugada criteria, 76 (73%) of whom had actual cardiac arrest and 28 (27%) with syncope (unconsciousness). Ninety five (58%) of them were Asians, with mean age from 22-65. In 36 of them (22%) had a strong family history of syncope, cardiac arrest or sudden death, without prior symptoms or apparent illness. In some cases, the typical abnormal Brugada EKG findings were noted during a routine screening medical exam performed because of a family history of sudden death.

Did Actor Rico Yan die of this syndrome?

This actor in the peak of his career at age 25 when he had sudden explained cardiac death on March 29, 2002 in a resort in Palawan. Police speculated that it was “Bangungot Syndrome”. Since he was young, with no apparent illness, and he suddenly died in his sleep, the cause of his death could well be Brugada Syndrome, alcohol could be a factor. Also, what we have been calling “Bangungot” may actually be Brugada Syndrome. Medical science, especially in the Philippines, has been trying to unravel the mystery of the “Bangungot Syndrome.” Perhaps, this is the same entity. In northwestern Thailand, sudden death, usually occurring during sleep, is the commonest cause of death among young men, and 40% of these victims had a strong family history of sudden cardiac arrest.

How does one prevent Brugada Syndrome?

Since this disease entity has now been “described,” which makes diagnosis of this entity possible, everyone, especially those with a family history of unexplained

sudden cardiac arrest, should be tested, starting with an EKG, and some “provocative” tests (with disopyramide, flecainide, ajmaline, procainamide) done to reproduce the characteristic Brugada EKG findings.

Can genetic testing help?

Since this disease is secondary to a mutation of SCN5A gene of chromosome 3 that has a dominant autosomic transmission pattern, with genetic defect in the alpha subunit of the sodium channel, genetic testing may be in order. However, 30% to 80% of patients will have negative (normal) gene screening in spite of overt or latent clinical Brugada Syndrome.

What’s the treatment for Brugada Syndrome?

If one is diagnosed to have Brugada Syndrome, an automatic implantable cardiac-pacemaker defibrillator (AICD) may be implanted, which will pace the heart and increase the rate if the rate goes down (awake or when asleep, 24/7), and shocks and jolts the heart back to normal rhythm, when the heart goes to ventricular fibrillation. Hundreds of thousands of people worldwide have AICD to prevent sudden death. None of the available drugs today are effective against Brugada Syndrome.