Ventricular fibrillation – very rapid beats (near 300 bpm) of the ventricles causing loss of pumping of the blood and it is fatal if not treated quickly with a DC shock to reset the natural pacing rhythm.

Depolarisation- Each beat is started by the spontaneous depolarisation (change from + to – charge) of the pacemaker cells in the sinus node. These trigger neighbouring atrial cells by direct electrical contact and a wave of depolarisation spreads out over the atria to the atrio-ventricular node. Depolarisation causes contraction of the atria, which fills the ventricles with blood. The depolarisation continues from the AV node through the bundle of HIS to the Purkinje fibres which then causes the ventricles to contract.

Repolarisation – The change of polarity of each cell back to a positive charge as the ventricles and atria expand ready for the next beat.

The Electrocardiogram ECG – The P wave starts with depolarisation at the sinus node. The PR segment is the time required for atrial depolarisation. The QRS complex is the spread of electrical activity (depolarisation) over the ventricles. The QT interval is the total time for depolarisation and repolarisation of the ventricles. The ST segment represents the period between ventricular depolarisation and ventricular repolarisation. The T wave is the repolarisation of the ventricles.

Heart block – restriction of the normal electrical pulse through the bundle of HIS, this can cause a slow ventricular contraction rate. The ventricles may only contract once for every two or three contractions of the atria.

References;

Long-Term Prognosis of Individuals With Right Precordial ST-Segment-Elevation Brugada Syndrome; Dr Lars Eckart, Dr Vincent Probst, Dr Jeroen P. P. Smits; American Heart Association Inc, Jan 25th 2005

Natural History of Brugada Syndrome Insights for Risk Stratification and Management; Dr Silva Priori; American Heart Association Inc, March 19th 2002

Proposed Diagnostic Criteria for the Brugada Syndrome; Dr Arthur A. M. Wilde; American Heart Inc, Nov 5th 2002

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Affiliates:

The SADS Foundation USA
www.sads.org
e-mail: sads@sads.org

The Canadian SADS Foundation
www.sads.ca
e-mail: info@sads.ca

SADS UK played a significant role in a campaign to bring about a new chapter in the Department of Health's National Service Framework for Coronary Heart Disease. Chapter Eight, Arrhythmias and Sudden Cardiac Death was published on 4th March 2005.

The chapter includes the following recommendations for good practice for initial treatment.

Patients should receive a hard copy of their ecg which shows their arrhythmia & patients surviving cardiac arrest or having presented with pre-excited atrial fibrillation should be assessed by a heart rhythm specialist.

The following patients should be **urgently assessed** by a heart rhythm specialist:-

Patients with either syncope suggesting an arrhythmia, symptoms of arrhythmia, a personal history of structural heart disease, a family history of early sudden death, recurrent syncope with palpitations, 3rd degree AV block, or with ventricular tachycardia.

The following should be **referred** to a heart rhythm specialist:-

People with suspected ventricular tachycardia, patients with WPW Syndrome, recurrent SVT not controlled by medication, recurrent atrial flutter, symptomatic atrial fibrillation not controlled by medication, first degree relatives of victims of sudden cardiac death who died below the age of 40 years, or patients with inexplicable recurrent falls.

The Recommendations for Ongoing Treatment are:-

Patients with sustained or compromising arrhythmias receive timely referral for appropriate treatment. Those identified at being high risk or with life-threatening ventricular arrhythmias should be considered a candidate for an implantable cardioverter defibrillator (ICD). For patients with sustained SVT catheter ablation should be considered. An outpatient care plan is devised between patient, GP and arrhythmia care team, when further hospital treatment is not recommended.

Please make a donation to SADS UK to enable us to support those affected by cardiac arrhythmia and sudden cardiac death –

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e-mail: sadsau@optushome.com.au

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The Ashley Jolly SAD Trust
Anne Jolly 01277 230642
www.sadsuk.org
e-mail: info@sadsuk.org

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The Brugada syndrome is an inherited cardiac electrical disorder occurring in the absence of obvious structural heart disease and can affect both male and female members of the family. The electrical activity around the ventricles may go in to disarray and cause the heart muscle to beat uncoordinatedly causing ventricular tachycardia. If the tachycardia develops in to ventricular fibrillation, the condition can be life threatening. Brugada syndrome affects 3 in 1000 people in the West and 14 in 1000 in Asia. It is most prevalent in Southeast Asia where it affects more males than females; the ratio is approximately 8:1. It can affect all ages from baby to pensioner but it is more common in years 35 to 45.

Several genes are thought to possibly cause Brugada syndrome but only the *SCN5A* gene on chromosome 3 has been positively identified, where the sodium channels of the cells that make up the heart muscle are affected. Sodium channel deficiency leads to polymorphic ventricular tachycardia and this is probably a direct result of slow conduction through the heart. Mutations in the sodium channel genes that cause Brugada syndrome can also cause LQTS3 and progressive conduction disturbances. Brugada syndrome has autosomal dominant inheritance in most patients.

Brugada syndrome is characterised by episodes of rapid polymorphic ventricular tachycardia, these episodes may self terminate with the patient suffering a syncope episode or possibly the tachycardia continues to degenerate in to ventricular fibrillation resulting in cardiac arrest. A patient with Brugada syndrome will normally have a history of these syncope episodes or cardiac arrests and will have a distinctive ECG pattern showing an appearance not dissimilar to incomplete or complete right bundle branch block and ST segment elevation in the precordial leads V1 to V3 with a coved morphology.

Episodes of dizziness and loss of consciousness sometimes associated with chest pain, sweating and, rigidity and shaking of the limbs associated with Brugada syndrome can sometimes be mistaken for idiopathic epilepsy.

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High temperature during fever can cause a typical Brugada ECG pattern. The high temperature affects the ionic mechanism, it results in premature closing of the sodium channels dramatically reducing the electrical charge carried by the sodium channel.

Unfortunately there are often no symptoms and the victim is found having died in their sleep, this may be referred to as Sudden Unexplained Nocturnal Death Syndrome 'SUNDS'. In the Philippines it is called 'bangungut', in Thailand it is called 'lai-tai' and in Japan 'pok-kuri'. Brugada syndrome episodes normally start when the heart rate is slow which is probably why so many people are affected in their sleep, particularly in the early hours of the morning. A history of sudden death at a young age in the family should be considered seriously and other family members should be referred to an electrophysiologist.

There is substantial evidence that patients with Brugada syndrome have an above average occurrence of supraventricular tachycardia including atrial and atrioventricular re-entrant tachycardia. Monomorphic ventricular tachycardia is not usually present, polymorphic tachycardia is most often detected.

Brugada syndrome can be identified by one of three distinctive ECG patterns. Where structural abnormalities of the heart have been ruled out the Brugada ECG patterns will show abnormalities of repolarisation and depolarisation with ST-segment elevation.

Type 1; Prominent coved ST-segment elevation in the right precordial leads followed by a negative T-wave.

Type 2; High ST-segment take off and elevation followed by a positive T-wave with saddleback configuration.

Type 3; ST-segment elevation in the right precordial leads, it may display saddleback or coved form and has the possibility of displaying both forms.

Care must be taken when connecting the patient to the ECG machine precordial leads, as correct identification of the Brugada ECG pattern is dependent on precise placement of the leads. ECG patterns recorded shortly after resuscitation or immediately after DC shock should not be considered when identifying Brugada syndrome.

Differentiation between Brugada syndrome and the arrhythmogenic right ventricular cardiomyopathy (ARVC) can be very difficult because ARVC can sometimes mimic Brugada syndrome. The structural changes of the heart muscle caused by ARVC may not be identified until autopsy. There are other disorders that can mimic Brugada syndrome that should be considered before making final diagnosis. If the patient has been administered class 1 A or class 1 C antiarrhythmic drugs these may also effect the ECG trace.

For those at high risk, having a history of syncope and a spontaneously abnormal ECG (not provoked with sodium channel blocking drugs) should be considered for an ICD implant. A patient having only spontaneous ST segment elevation without a history of syncope may be considered as low risk and the use of an ICD may be inappropriate as the risk of treatment may outweigh the risk of potential effects of the syndrome.

Definitions

Morphology – form and structure

Monomorphic - having only one form (structure)

Polymorphic – having more than one form (structure)

Precordial – the precordium is the region over the heart and stomach

Autosomal dominant – affects males and females equally with only a single copy of the gene needed to cause the condition.

Tachycardia - a fast heart rhythm above 100 beats per minute

Supraventricular tachycardia – a fast heart rhythm above 100 beats per minute originating from the atria

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