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**Literature search results**

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**Search details**

Brugada Syndrome – anaesthetic conduct and outcome in patients undergoing operative procedures. Pathophysiology and incidence

**Resources searched**

NHS Evidence; TRIP database; Cochrane Library, MEDLINE; EMBASE

“brugada syndrome”; BRUGADA SYNDROME; “sudden unexpected death syndrome” SUDS; anaesthesia; anesthesia; ANESTHESIA; ANESTHESIA, GENERAL; ANESTHESIA, LOCAL; ANESTHESIA, INHALATION; management; Surgery, GENERAL SURGERY; OPERATIVE PROCEDURES, SURGICAL; “operative procedure”; incidence; INCIDENCE; pathophysiology

**Summary**

**Incidence of Brugada Syndrome**

I have looked at incidence of Brugada Syndrome rather than the incidence of co-existing conditions or physiological manifestations. Incidence seems to be higher in men than women 8-10/10,000 and in the Asian population, particularly those from the Far East. Incidence ranges from 1-5/10,000 of the population. In the Japan incidence is 0.1-0.2% of the population. Brugada Syndrome is responsible for 4-10% of all sudden deaths/10,000 population/year. The incidence of sudden cardiac death displays 2 peaks: birth-6 months and 45-75 yrs.
Pathophysiology

Some of the research does review the pathophysiology of Brugada Syndrome, but this is not described in the abstracts. The full article will therefore have to be read. In those abstracts where pathophysiology was described, right ventricular derangements were highlighted in study 29 and autonomic dysfunction in study 36, were highlighted.

Anaesthetic conduct and outcome in patients with Brugada Syndrome undergoing operative procedures

Again many of the papers dealing with this question do not have abstracts or enough detail in the abstract. However parasympathetic dominant condition must be avoided during anesthetic management \(^{11}\), perioperative autonomic imbalance may cause ventricular fibrillation and sudden cardiac arrest \(^{25}\). During surgery, induced ventricular fibrillation was easily controlled by ICD \(^{50}\), and there has been no established consensus on pre-operative risk assessment of patients with Brugada-type ECG \(^{38}\).

Guidelines

European Society of Cardiology

Management of Patients with Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death 2006

Brugada Syndrome mentioned in connection with diagnosis, physiology and management, but not specifically related to operative procedures or anaesthesia.

Evidence based reviews

None

Published research


Author(s): Theodotou N, Cillo JE Jr

Citation: Journal of Oral & Maxillofacial Surgery, September 2009, vol./is. 67/9(2021-5), 1531-5053

Publication Date: September 2009

Source: MEDLINE

Full Text: Available in print at Lincoln County Hospital Professional Library

2. Role of pharmacotherapy in cardiac ion channelopathies.

Author(s): El-Sherif N, Pedalino R, Himel H 4th

Citation: Current Vascular Pharmacology, July 2009, vol./is. 7/3(358-66), 1570-1611

Publication Date: July 2009

Abstract: In the last decade there have been considerable advances in the understanding of the pathophysiology of malignant ventricular tachyarrhythmias (VA) and Sudden Cardiac...
Death (SCD). Over 80% of SCD occurs in patients with organic heart disease. However, approximately 10-15% of SCD occurs in the presence of structurally normal heart and the majority of those patients are young. In this group of patients, changes in genes encoding cardiac ion channels produce modification of the function of the channel resulting in an electrophysiological substrate of VA and SCD. Collectively these disorders are referred to as Cardiac Ion Channelopathies. The 4 major syndromes in this group are: The Long QT Syndrome (LQTS), the Brugada Syndrome (BrS), the Short QT Syndrome (SQTS), and the Catecholaminergic Polymorphic VT (CPVT). Each of these syndromes includes multiple subtypes with different and sometimes complex genetic abnormalities of cardiac ion channels. Many are associated with other somatic and neurological abnormalities besides the risk of VA and SCD. The current management of cardiac ion channelopathy could be summarized as follows: 1) in symptomatic patients, the implantable cardioverter defibrillator (ICD) is the only viable option; 2) in asymptomatic patients, risk stratification is necessary followed by the ICD, pharmacotherapy, or a combination of both. A genotype-specific approach to pharmacotherapy requires a thorough understanding of the molecular-cellular basis of arrhythmogenesis in cardiac ion channelopathies as well as the specific drug profile.

Source: MEDLINE

3. Clinical Case: Anaesthetic management of Brugada syndrome

Author(s): Suarez P.M., Vargas D.V., Infante E.D., Jimenez M.S.C.

Citation: Actualizaciones en Anestesiologia y Reanimacion, 2009, vol./is. 19/1(41-42), 1132-0095

Publication Date: 2009

Abstract: Brugada’s syndrome (BS) was first described at 1992 by P. & J. Brugada’s brothers in patients who died by cardiac arrest without any defect in cardiac structure. They only showed an incompeleted Right Bruch Block (RBB) and ST segment raised at V12 &3 precordials (1). Behind this syndrome it hides a genetic disorder affecting the normal function of sodium channels (2). Its diagnosis is based either on the typical EKG changes or after resuscitated cardiac arrest generally polimorphus ventricular tachycardia who unleash ventricular fibrillation. Due to its lethainess we understand that under the finding of RBB in the preoperative evaluation a more conscientious anamnesis should be done. With the certainty of SB a careful anaesthetic management is obligatory. Copyright copyright 2009 Aran Ediciones, s. L.

Source: EMBASE

4. Cardiac gene defects can cause sudden cardiac death in young people.

Author(s): Kauferstein S, Kiehne N, Neumann T, Pitschner HF, Bratzke H

Citation: Deutsches Arzteblatt International, January 2009, vol./is. 106/4(41-7), 1866-0452

Publication Date: January 2009

Abstract: BACKGROUND: In Europe, sudden cardiac death (SCD) is one of the most common causes of death. Although sudden cardiac death usually happens in older people, 5% to 10% of the affected individuals are young and apparently healthy. Sudden death in infants, children, and young adults is relatively rare, with an incidence of 1 to 5 per 100 000 persons per year. Nonetheless, up to 7000 asymptomatic children die in the USA each year, almost half of them without any warning signs or symptoms. METHOD: Seleective literature review. RESULTS: Although structural cardiovascular abnormalities explain most cases of sudden cardiac death in young people, the cause of death remains unexplained after autopsy in 10% to 30% of cases. Potentially lethal ion channel disorders (channelopathies) such as the long QT syndromes (LQTS), catecholaminergic polymorphic
ventricular tachycardia (CPVT), and the Brugada syndrome (BrS) may account for at least one-third of these unexplained cases. Most of these diseases are hereditary with autosomal-dominant transmission, i.e., there is a 50% chance that the children of affected individuals will be affected themselves. CONCLUSIONS: Post-mortem genetic screening for sequence variations in cardiac ion channel genes has become an important forensic tool for elucidating the cause of sudden cardiac death. Moreover, it allows the identification of other family members bearing the previously undiagnosed gene defect, who can then undergo a cardiological evaluation if indicated by their clinical history.

Source: MEDLINE

Full Text: Available in fulltext at National Library of Medicine

5. Clinical impact of genetic studies in lethal inherited cardiac arrhythmias.

Author(s): Shimizu W

Citation: Circulation Journal, December 2008, vol./is. 72/12(1926-36), 1346-9843

Publication Date: December 2008

Abstract: Over the past decade, molecular genetic studies have established a link between a number of inherited cardiac arrhythmias, including congenital long QT syndrome (LQTS) and Brugada syndrome (BrS), and mutations in genes encoding for ion channels or other membrane components. Twelve forms of LQTS have been identified in 50-70% of clinically affected patients. Genotype-phenotype correlations have been rigorously investigated in LQT1, LQT2 and LQT3 syndromes, which constitute more than 90% of genotyped LQTS patients, enabling stratification of risk and effective treatment of genotyped patients. Genotype-specific triggers for both the cardiac events and the clinical course have been reported, and genotype-specific therapy has been already introduced. More recently, mutation site-specific differences in the clinical phenotype have been reported in LQT1 and LQT2 patients, indicating the possibility of mutation site-specific management or treatment. In contrast, only one-third of BrS patients can be genotyped, and data on genotype-phenotype relationships in clinical studies are limited. A Haplotype B consisting of 6 individual DNA polymorphisms within the proximal promoter region of the SCN5A gene was recently identified only in Asians (frequency 22%). Individuals with Haplotype B show significantly longer duration of both PQ and QRS than those without Haplotype B, indicating that Haplotype B likely contributes to the higher incidence of BrS in Asian populations.

Source: MEDLINE

6. Perioperative management of a 7-year-old child with Brugada syndrome.

Author(s): Baty L, Hollister J, Tobias JD

Citation: Journal of Intensive Care Medicine, May 2008, vol./is. 23/3(210-4), 0885-0666

Publication Date: May 2008

Abstract: Brugada syndrome results from abnormalities in the myocardial transmembrane conduction of sodium, resulting in the characteristic electrocardiographic changes of ST segment elevation in the precordial leads and incomplete right bundle branch block in an otherwise structurally normal heart. Affected patients are frequently asymptomatic until their presentation with potentially lethal arrhythmias including ventricular fibrillation. The youngest reported patient with Brugada syndrome to undergo anesthetic management is presented in this article; the pathophysiology of the syndrome is reviewed, and its perioperative implications are discussed.
7. *Intra day ECG variation after general anesthesia in Brugada syndrome.*

**Author(s):** Brunetti ND, De Gennaro L, Pellegrino PL, Ieva R, Di Nardo F, Cuculo A, Campanale G, Di Biase M

**Citation:** Journal of Interventional Cardiac Electrophysiology, April 2008, vol./is. 21/3(219-22), 1383-875X

**Publication Date:** April 2008

**Abstract:** ECG variations of characteristic coved-type patterns are a common finding in subjects with Brugada syndrome. Few data are available about patients with Brugada syndrome undergoing major surgery and requiring general anesthesia. We reported ECG serial variations in a patient with Brugada syndrome who underwent surgery for tibial fracture. Relevant variations of ST patterns in V1-V2 leads over 24-h ECG monitoring were detectable, albeit these modifications were not associated with incidence of arrhythmias.

**Source:** MEDLINE


**Author(s):** Vaccarella A, Vitale P, Presti CA

**Citation:** Minerva Anestesiologica, April 2008, vol./is. 74/4(149-52), 1827-1596

**Publication Date:** April 2008

**Abstract:** This case report describes an asymptomatic patient with positive familiar anamnesis of Brugada syndrome (BrS) who elected to undergo surgery. The anaesthesiological technique using propofol, fentanyl, atracurium, air/oxygen did not induce any electrocardiographic alteration during the operation; the intraoperation use of a biphasic defibrillator was critical here. The cerebral state index and adhesive plaques connected with a biphasic defibrillator having PM capabilities allowed us to monitor the operation and continually assess the patient's cardiac stability. Afterwards, the patient was transferred to the intensive care unit and was monitored for 24 hours. This anesthesiological technique was performed in place of ARL, which the patient refused.

**Source:** MEDLINE


**Author(s):** Canbay O, Erden IA, Celebi N, AycanIO, Karagoz AH, Aypar U

**Citation:** Paediatric Anaesthesia, December 2007, vol./is. 17/12(1225-7), 1155-5645

**Publication Date:** December 2007

**Source:** MEDLINE

10. *Clinical characteristics and risk stratification in symptomatic and asymptomatic patients with brugada syndrome: multicenter study in Japan.*

**Author(s):** Takagi M, Yokoyama Y, Aonuma K, Aihara N, Hiraoka M, Japan Idiopathic Ventricular Fibrillation Study (J-IVFS) Investigators

**Citation:** Journal of Cardiovascular Electrophysiology, December 2007, vol./is. 18/12(1244-51), 1540-8167
Publication Date: December 2007

Abstract: BACKGROUND: Neither the clinical characteristics nor risk stratification in Brugada syndrome have been clearly determined. We compared the clinical and ECG characteristics of symptomatic and asymptomatic patients with Brugada syndrome to identify new markers for high-risk patients. METHODS: A total of 188 consecutive individuals with Brugada syndrome (mean age 53 +/- 14 years, 178 males) were enrolled in the Japan Idiopathic Ventricular Fibrillation Study (J-IVFS). Clinical and ECG characteristics were evaluated in three groups of patients: Ventricular fibrillation (VF) group: patients with documented VF (N = 33); Syncope (Sy) group: patients with syncope without documented VF (N = 57); and asymptomatic (As) group: subjects without symptoms (N = 98). Their prognostic parameters were evaluated over a 3-year follow-up period. RESULTS: (1) Clinical characteristics: incidence of past history of atrial fibrillation (AF) was significantly higher in the VF and Sy groups than in the As group (P = 0.04). (2) On 12-lead ECG, r-J interval in lead V2 and QRS duration in lead V6 were longest in the VF group (P = 0.001, 0.002, respectively). (3) Clinical follow-up: during a mean follow-up period of 37 +/- 16 months, incidences of cardiac events (sudden death and/or VF) were higher in the symptomatic (VF/Sy) groups than in the As group (P < 0.0001). The r-J interval in lead V2 >/= 90 ms and QRS duration in lead V6 >/= 90 ms were found to be possible predictors of recurrence of cardiac events in symptomatic patients. CONCLUSIONS: Prolonged QRS duration in precordial leads was prominent in symptomatic patients. This ECG marker may be useful for distinguishing high- from low-risk patients with Brugada syndrome.

Source: MEDLINE

Full Text:

Available in fulltext at EBSCO Host

11. [Preoperative evaluation and anesthetic management of a patient with Brugada syndrome-like ECG].

Author(s): Kohda K., Sugano T., Shibama S., Harada M., Satoh Y., Ide Y., Tagami M.

Citation: Masui - Japanese Journal of Anesthesiology, December 2007, vol./is. 56/12(1398-403), 0021-4892

Publication Date: December 2007

Abstract: Brugada syndrome has been known as one of the causes of sudden death due to ventricular fibrillation. We experienced anesthetic management of seven patients with ECG showing Brugada syndrome before surgery, even though they had no symptoms nor family history. All of them showed no problems throughout the operation. Such patients are often untreated, but they have the risks of cardiac accidents such as ventricular fibrillation or sudden death. For preoperative evaluation of patients with Brugada syndrome-like ECG, it is important to ask them their experience of syncope and family history. Ultrasonic cardiography and Holter ECG recording should be done. External defibrillator should be prepared and parasympathetic dominant condition must be avoided during the anesthetic management.

Source: MEDLINE

12. Preoperative evaluation and anesthetic management of a patient with Brugada syndrome-like ECG

Author(s): Kohda K., Sugano T., Shibama S., Harada M., Satoh Y., Ide Y., Tagami M.

Citation: Japanese Journal of Anesthesiology, December 2007, vol./is. 56/12(1398-1403), 0021-4892
Publication Date: December 2007

Abstract: Brugada syndrome has been known as one of the causes of sudden death due to ventricular fibrillation. We experienced anesthetic management of seven patients with ECG showing Brugada syndrome before surgery, even though they had no symptoms nor family history. All of them showed no problems throughout the operation. Such patients are often untreated, but they have the risks of cardiac accidents such as ventricular fibrillation or sudden death. For preoperative evaluation of patients with Brugada syndrome-like ECG, it is important to ask them their experience of syncope and family history. Ultrasonic cardiography and Holter ECG recording should be done. External defibrillator should be prepared and parasympathetic dominant condition must be avoided during the anesthetic management.

Source: EMBASE

13. Prevalence of Brugada sign in a Greek tertiary hospital population.

Author(s): Letsas KP, Gavrielatos G, Efremidis M, Kounas SP, Filippatos GS, Sideris A, Kardaras F

Citation: Europace, November 2007, vol./is. 9/11(1077-80), 1532-2092

Publication Date: November 2007

Abstract: AIMS: The purpose of the present study was to determine for the first time the prevalence of Brugada-type electrocardiographic (ECG) pattern (Brugada sign) in unselected individuals served by an urban Greek tertiary hospital during a 4-year time period. METHODS AND RESULTS: Among 11,488 individuals (6640 males, 4848 females), 25 (23 males, 2 females, aged 36.8 +/- 19.2 years) were found to display the Brugada sign (0.22%). Two cases exhibited the diagnostic type 1 ECG pattern (0.02%) and 23 subjects fulfilled the ECG criteria for type 2 or 3 patterns (0.2%). The incidence of Brugada sign was higher among men (0.34%) than in women (0.04%). Structural heart disease was established in four cases (one of them exhibiting a type 1 ECG pattern). Twenty-one individuals (19 males, 2 females, aged 29.7 +/- 10.7 years) without structural heart disease displaying Brugada-type ECG features (4 cases with spontaneous or procainamide-induced type 1 ECG pattern) were subsequently selected and closely followed up for 24 +/- 12 months. No mortality or life-threatening ventricular arrhythmias were recorded during this period. CONCLUSION: The Brugada-type ECG pattern is infrequently seen in a Greek hospital-based population. All subjects with Brugada sign and structurally normal hearts displayed a benign clinical course without arrhythmic events during a relatively long follow-up period.

Source: MEDLINE

Full Text:
Available in fulltext at Highwire Press

14. Incidence of Brugada electrocardiographic pattern and outcomes of these patients after intentional tricyclic antidepressant ingestion.

Author(s): Bebarta VS, Phillips S, Eberhardt A, Calihan KJ, Waksman JC, Heard K

Citation: American Journal of Cardiology, August 2007, vol./is. 100/4(656-60), 0002-9149

Publication Date: August 2007

Abstract: Brugada syndrome is a genetic dysfunction of the myocardial sodium channel that leads to ventricular dysrythmias. The electrocardiographic (ECG) pattern of Brugada syndrome is occasionally seen after tricyclic antidepressant (TCA) ingestion; however, the
outcome and complication risk for these patients is not clear. The objective of our study was to describe the incidence of Brugada ECG pattern (BEP) and serious complications of these patients in a large case series of intentional TCA ingestions. We also compared the proportion of complications of patients with BEP versus those without BEP. We evaluated 402 TCA ingestions, of which 9 (2.3%) were associated with the development of BEP. We compared the adverse outcomes of all TCA ingestions versus TCA ingestions with BEP. A increase in the adverse outcomes in the BEP group was found: seizures (relative risk [RR] 4; 95% confidence interval [CI] 1.5 to 10.8), widened QRS (RR 4.8; 95% CI 1.8 to 12.9), and hypotension (RR 3.9; 95% CI 2.1 to 7.4). To reduce confounding ingestants, we also compared all patients with an isolated TCA ingestion versus those with BEP. A significant increase in adverse outcomes was again found with the BEP group: seizures (RR 3; 95% CI 1.1 to 8.6), widened QRS (RR 4.8; 95% CI 1.5 to 15.1), and hypotension (RR 3.4; 95% CI 1.9 to 22.3). No deaths or dysrhythmias were found in the BEP group. In conclusion, BEP after TCA ingestion is rare, and death or dysrhythmias did not occur. However, patients with BEP are likely at increased risk for TCA-induced complications.

Source: MEDLINE

15. The Brugada syndrome.

Author(s): Ros senbacker T, Priori SG

Citation: Current Opinion in Cardiology, May 2007, vol./is. 22/3(163-70), 0268-4705

Publication Date: May 2007

Abstract: PURPOSE OF REVIEW: The Brugada syndrome has been an area of intensive investigation since its earliest description in 1992, both on a clinical and on a basic research level. In this review, we will focus on recent achievements in the molecular dissection of the disease pathophysiology and on large multicenter studies dealing with prognostic markers and the natural history of the Brugada syndrome. RECENT FINDINGS: In the past year, two additional genetic pathways have been associated with the disease. Also, an inflammatory or infectious etiology has recently been linked with the Brugada syndrome. The debate on the predictive role of programmed electrical stimulation is still ongoing. Very recently, large follow-up studies questioned the prognostic role of programmed electrical stimulation in this disease. SUMMARY: Knowledge on the genetic determinants of the Brugada syndrome remains limited. Therefore, the management and the risk stratification of patients should be performed on a clinical basis. Sufficient evidence exists to reassure clinicians who feel reluctant to include programmed electrical stimulation in the risk stratification strategy of asymptomatic Brugada syndrome patients.

Source: MEDLINE


Author(s): Makita N, Tsutsui H

Citation: Circulation Journal, 2007, vol./is. 71 Suppl A/(A54-60), 1346-9843

Publication Date: 2007

Abstract: Over the past 10 years, remarkable advances have been made in identifying the genes responsible for primary electrical heart diseases, such as congenital long QT syndrome and Brugada syndrome. Basic and clinical studies on these inherited arrhythmias have provided significant insight into the molecular basis of cardiac electrophysiology and the mechanisms of arrhythmias. However, many studies of genotype - phenotype relationships in these diseases have revealed considerable phenotypic variability in individuals from the same kindred carrying the identical disease-associated DNA variant, as is commonly observed in other polygenic disorders. Furthermore, despite rapid progress in understanding the molecular basis of primary electrical heart diseases, there is little insight
into the genetics of acquired arrhythmias. Recently, it has been recognized that common
genetic polymorphisms in cardiac ion channel and other genes may modify cardiac
excitability, which in turn predisposes affected individuals to arrhythmias in the presence of
triggering factors, such as electrolyte abnormalities or drugs. This paper reviews the current
understanding of the contribution of genetic polymorphisms to the pathophysiology of

Source: MEDLINE

17. Inherited arrhythmic disorders: long QT and Brugada syndromes.

Author(s): Nader A, Massumi A, Cheng J, Razavi M

Citation: Texas Heart Institute Journal, 2007, vol./is. 34/1(67-75), 0730-2347

Publication Date: 2007

Abstract: Inherited arrhythmic disorders comprise a group of syndromes with unique
genetic abnormalities and presentations but with very similar clinical outcomes and
complications, the most terrifying of which are life-threatening arrhythmias and sudden
cardiac death. Advances in molecular biology have enabled us to define and pinpoint many
such disorders, which were previously labeled as idiopathic, to specific genes on various
chromosomes. The current trend in the management of these potentially deadly disorders
is to use pharmacotherapy (antiarrhythmic agents) and defibrillators for the prevention of
sudden death; however, targeted therapy at a molecular level appears to be the path of the
future. Herein, we review long QT and Brugada syndromes and focus on the genetics,
pathophysiology, and clinical manifestations of these inherited arrhythmogenic disorders
that affect patients with structurally normal hearts.

Source: MEDLINE

Full Text: Available in fulltext at National Library of Medicine


Author(s): Hiraoka M

Citation: Circulation Journal, 2007, vol./is. 71 Suppl A/(A61-8), 1346-9843

Publication Date: 2007

Abstract: The incidence of Brugada syndrome (BS) is relatively high in Japan compared
with the rest of the world, ranging between 0.1% and 0.2% in the general population. BS in
Japan, as in other countries, is most prevalent in middle-aged men, and has characteristics
ECG changes, a high recurrence rate in symptomatic patients, and relatively low incidence
of SCN5A mutations. In contrast, both the incidence of a family history of BS and/or sudden
cardiac death and the rate of developing cardiac events in asymptomatic patients are less
in Japan than in other countries. Increased vagal tone and/or decreased sympathetic
activity are suggested as provoking cardiac events. Several factors should be evaluated in
risk stratification for recurrence of life-threatening arrhythmias, because there appears to be
no single determinant for risk stratification: spontaneous ST elevation of coved-type (Type
1), family history of sudden cardiac death, inducible ventricular tachycardia/ventricular
fibrillation and positive late potentials. An implantable cardioverter defibrillator is
recommended for patients with aborted sudden cardiac death. (Circ J 2007; Suppl A: A-61
- A-68).

Source: MEDLINE
19. Genetic and biophysical basis for bupivacaine-induced ST segment elevation and VT/VF. Anesthesia unmasked Brugada syndrome


Citation: Heart Rhythm, September 2006, vol./is. 3/9(1074-8), 1547-5271

Publication Date: September 2006

Abstract: BACKGROUND: Brugada syndrome is an inherited disease associated with sudden cardiac death. The electrocardiographic pattern associated with Brugada syndrome has been linked to the use of sodium channel blockers, including antiarrhythmics, trycyclics and anesthetics. OBJECTIVE: We report a case of bupivacaine-induced Brugada syndrome, in which we investigated the genetic, biophysical and path physiological mechanism involved. METHODS AND RESULTS: The patient developed a Brugada-like electrocardiographic pattern twice under the influence of bupivacaine. The first occurrence was accompanied by ventricular tachycardia (VT) which subsided after withdrawal of the anesthetic. The VT was also observed during co-administration of diltiazem and isosorbide-5-mononitrate, agents thought to facilitate ST segment elevation in the Brugada syndrome. Genetic analysis revealed a missense mutation in the alpha subunit of the cardiac sodium channel, SCN5A. Biophysical analysis by whole-cell patch-clamping revealed a reduction in sodium current as a result of the mutation. The study of bupivacaine in the wedge model revealed use-dependent changes in conduction, heterogeneous loss of the action potential dome in RV epicardium and phase 2 re-entry when the preparations were pretreated with low concentrations of the calcium channel blocker verapamil. CONCLUSION: Our findings indicate that bupivacaine may induce the electrocardiographic and arrhythmic manifestations of the Brugada syndrome in silent carriers of SCN5A mutations. The data have important implications in the management of patients who develop ST segment elevation when under the influence of anesthetics such as bupivacaine.

Source: MEDLINE


Author(s): Herbert E, Chahine M

Citation: Canadian Journal of Physiology & Pharmacology, August 2006, vol./is. 84/8-9(795-802), 0008-4212

Publication Date: August 2006

Abstract: Brugada syndrome (BS) is an inherited cardiac disorder characterized by typical electrocardiographic patterns of ST segment elevation in the precordial leads, right bundle branch block, fast polymorphic ventricular tachycardia in patients without any structural heart disease, and a high risk of sudden cardiac death. The incidence of BS is high in male vs. female (i.e., 8-10/1: male/female). The disorder is caused by mutations in the SCN5A gene encoding Nav1.5, the cardiac sodium channel, which is the only gene in which mutations were found to cause the disease. Mutations in SCN5A associated with the BS phenotype usually result in a loss of channel function by a reduction in Na+ currents. We review the clinical aspects, risk stratification, and therapeutic management of this important syndrome.

Source: MEDLINE

Full Text:
Available in fulltext at EBSCO Host

Author(s): Hayashida H, Miyauchi Y

Citation: British Journal of Anaesthesia, July 2006, vol./is. 97/1(118-9), 0007-0912

Publication Date: July 2006

Source: MEDLINE

Full Text:
Available in fulltext at Highwire Press
Available in print at Lincoln County Hospital Professional Library


Author(s): Hayashida H., Miyauchi Y.

Citation: British Journal of Anaesthesia, July 2006, vol./is. 97/1(118-119), 0007-0912;1471-6771

Publication Date: July 2006

Source: EMBASE

Full Text:
Available in fulltext at Highwire Press
Available in print at Lincoln County Hospital Professional Library

23. Brugada syndrome and anesthetic management.

Author(s): Cordery R, Lambiase P, Lowe M, Ashley E

Citation: Journal of Cardiothoracic & Vascular Anesthesia, June 2006, vol./is. 20/3(407-13), 1053-0770

Publication Date: June 2006

Source: MEDLINE


Author(s): Fujiwara Y, Shibata Y, Kurokawa S, Satou Y, Komatsu T

Citation: Anesthesia & Analgesia, May 2006, vol./is. 102/5(1590-1), 1526-7598

Publication Date: May 2006

Source: MEDLINE

Full Text:
Available in fulltext at Highwire Press
25. [Anesthetic management for patients with Brugada syndrome]

Author(s): Kawaguchi Y, Kushikata T, Hashiba E, Kitayama M, Yoshida H, Ishihara H, Matsuki A, Hirota K

Citation: Masui - Japanese Journal of Anesthesiology, February 2006, vol./is. 55/2(142-9), 0021-4892

Publication Date: February 2006

Abstract: Brugada syndrome should not be neglected in terms of anesthetic management because its perioperative autonomic imbalance may cause ventricular fibrillation and sudden cardiac arrest. Diagnosis of Brugada syndrome is easily made by unique electrocardiographic pattern of right bundle branch block and ST segment elevation in the right precordial leads. Thus the number of patients with Brugada syndrome for anesthetic management tends to increase. We review current concept of anesthetic management for patients with Brugada syndrome including fourteen cases in our institution, two out of which developed VF during operation.

Source: MEDLINE

26. Anesthetic management for patients with Brugada syndrome

Author(s): Kawaguchi Y., Kushikata T., Hashiba E., Kitayama M., Yoshida H., Ishihara H., Matsuki A., Hirota K.

Citation: Japanese Journal of Anesthesiology, February 2006, vol./is. 55/2(142-149), 0021-4892

Publication Date: February 2006

Abstract: Brugada syndrome should not be neglected in terms of anesthetic management because its perioperative autonomic imbalance may cause ventricular fibrillation and sudden cardiac arrest. Diagnosis of Brugada syndrome is easily made by unique electrocardiographic pattern of right bundle branch block and ST segment elevation in the right precordial leads. Thus the number of patients with Brugada syndrome for anesthetic management tends to increase. We review current concept of anesthetic management for patients with Brugada syndrome including fourteen cases in our institution, two out of which developed VF during operation.

Source: EMBASE

27. Perioperative management of patients with Brugada syndrome

Author(s): Kubo K., Nishikawa K., Goto F.

Citation: Japanese Journal of Anesthesiology, November 2005, vol./is. 54/11(1247-1252), 0021-4892

Publication Date: November 2005

Abstract: In 1992, Brugada et al. first reported eight cases of ventricular fibrillation, in which ST-segment abnormalities in leads V1 through V3 along with T-wave inversion, and complete or incomplete right bundle branch block were observed on the standard 12-leads ECG. Since then, this syndrome has been widely recognized as one of important diseases that can produce sudden death in middle aged healthy males. The ECG morphology of
Brugada syndrome is believed to be caused by either an accentuation of the notch in the early phase of the action potential or loss of the action potential dome in the epicardium. Mechanisms of ventricular fibrillation in this syndrome are still unclear, but thought to be phase II re-entry caused by dispersion of the action potentials. It has been shown that mutations of the human cardiac Na+ channel gene (SCN5A) underlie multiple cardiac diseases including Brugada syndrome. In fact, single amino acid substitution within the SCN5A coding region can evoke a cardiac rhythm behavior. In this review, we will focus on recent progress of basic and clinical research of Brugada syndrome and perioperative management of this syndrome.

Source: EMBASE


Author(s): Watrich DG, Woods RA, Steiner IP

Citation: CJEM Canadian Journal of Emergency Medical Care, September 2005, vol./is. 7/5(347-50), 1481-8035

Publication Date: September 2005

Abstract: Brugada syndrome is a potentially lethal and eminently treatable entity that may present with palpitations or syncope. This article presents the case of a young patient with Brugada syndrome and reviews key features in the epidemiology, pathophysiology, diagnosis, treatment and prognosis of this condition.

Source: MEDLINE

29. Pathophysiological mechanisms of Brugada syndrome: depolarization disorder, repolarization disorder, or more?.

Author(s): Meregalli PG, Wilde AA, Tan HL

Citation: Cardiovascular Research, August 2005, vol./is. 67/3(367-78), 0008-6363

Publication Date: August 2005

Abstract: After its recognition as a distinct clinical entity, Brugada syndrome is increasingly recognized worldwide as an important cause of sudden cardiac death. Brugada syndrome exhibits autosomal dominant inheritance with SCN5A, which encodes the cardiac sodium channel, as the only gene with a proven involvement in 20-30% of patients. Its signature feature is ST segment elevation in right precordial ECG leads and predisposition to malignant ventricular tachyarrhythmias. The pathophysiological mechanism of ST elevation and ventricular tachyarrhythmia, two phenomena strongly related, is controversial. Here, we review clinical and experimental studies as they provide evidence to support or disprove the two hypotheses on the mechanism of Brugada syndrome that currently receive the widest support: (1) nonuniform abbreviation of right ventricular epicardial action potentials ("depolarization disorder"), (2) conduction delay in the right ventricular outflow tract ("repolarization disorder"). We also propose a schematic representation of the depolarization disorder hypothesis. Moreover, we review recent evidence to suggest that other derangements may also contribute to the pathophysiology of Brugada syndrome, in particular, right ventricular structural derangements. In reviewing these studies, we conclude that, similar to most diseases, it is likely that Brugada syndrome is not fully explained by one single mechanism. Rather than adhering to the notion that Brugada syndrome is a monofactorial disease, we should aim for clarification of the contribution of various pathophysiological mechanisms in individual Brugada syndrome patients and tailor therapy considering each of these mechanisms.

Source: MEDLINE
30. The surgical patient with Brugada syndrome: a four-case clinical experience.

Author(s): Santambrogio LG, Mencherini S, Fuardo M, Caramella F, Braschi A

Citation: Anesthesia & Analgesia, May 2005, vol./is. 100/5(1263-6, table of contents), 0003-2999

Publication Date: May 2005

Abstract: Brugada syndrome is characterized by a distinctive electrocardiographic pattern (right bundle branch block and ST segment elevation in precordial leads) and a high risk of cardiac arrest for malignant dysrhythmia. The genetic basis is a molecular defect of the cardiac sodium channel and the pattern of inheritance is autosomal dominant. Many factors during general anesthesia (medications, bradycardia, temperature changes) could precipitate malignant dysrhythmia in these patients. Because criteria to identify the surgical patient at high risk for developing malignant dysrhythmia are lacking, we can only speculate about the available studies on nonsurgical patients. We describe four patients during general anesthesia and propose intraoperative and postoperative monitoring (the first 36 h).

Source: MEDLINE

Full Text:

Available in fulltext at Highwire Press

Available in fulltext at Ovid


Author(s): Inamura M, Okamoto H, Kuroiwa M, Hoka S

Citation: Canadian Journal of Anaesthesia, April 2005, vol./is. 52/4(409-12), 0832-610X

Publication Date: April 2005

Abstract: PURPOSE: To review six cases of Brugada syndrome presenting for insertion of a cardioverter-defibrillator under general anesthesia. CLINICAL FEATURES: All patients had a history of syncope, ST segment elevation in the right precordial lead of the electrocardiogram (ECG) which became prominent after a pilscainide challenge test. Routine monitors, right precordial lead of the ECG and an external defibrillator were installed prior to anesthesia. We administered propofol/midazolam for induction, and propofol/sevoflurane combined with fentanyl for maintenance of anesthesia. Atropine and ephedrine were administered to decrease vagal tone. No ECG change or arrhythmia was observed perioperatively. After the successful implantation of the defibrillator, all patients were discharged without any adverse event. CONCLUSION: By avoiding agents or conditions that may exacerbate Brugada syndrome during anesthesia, we were able to manage the patients uneventfully for implantation of a cardioverter-defibrillator.

Source: MEDLINE

Full Text:

Available in print at Grantham Hospital Staff Library
32. The Brugada Syndrome.

Author(s): Brugada P, Brugada R, Antzelevitch C, Brugada J

Citation: Archives des Maladies du Coeur et des Vaisseaux, February 2005, vol./is. 98/2(115-22), 0003-9683

Publication Date: February 2005

Abstract: In 1992 a syndrome was described consisting of syncopal episodes and/or (resuscitated) sudden death in patients with a structurally normal heart and a characteristic electrocardiogram (ECG) displaying a pattern resembling a right bundle branch block with ST segment elevation in leads V1 to V3. The disease is genetically determined with an autosomal dominant pattern of transmission in 50% of the familial cases. Several different mutations have been identified affecting the structure, function and trafficking of the sodium channel. The syndrome is ubiquitous. Its incidence and prevalence are difficult to estimate, but this disease may cause 4 to 10 sudden deaths per 10,000 inhabitants per year representing the most frequent cause of natural death in males younger than 50 in South Asia. The disease has been linked to the sudden infant death syndrome (SIDS) and to the sudden unexpected death syndrome (SUDS) by showing that the electrocardiogram and mutations are the same as in Brugada syndrome. The diagnosis is easily made by means of the ECG when it is typical. There exist, however, patients with concealed and intermittent electrocardiographic forms that make the diagnosis difficult. The ECG can be modulated by changes in autonomic balance, body temperature, glucose level and the administration of antiarrhythmic, neuroleptic and antimalaria drugs. Beta adrenergic stimulation normalizes the ECG. Loss of the action potential dome in right ventricular epicardium but not in endocardium underlies the ST segment elevation. Electrical heterogeneity within right ventricular epicardium leads to the development of closely coupled extrasystoles via phase 2 reentry that precipitate ventricular fibrillation. Antiarrhythmic drugs do not prevent sudden death in symptomatic or asymptomatic individuals. Implantation of an automatic cardioverter-defibrillator is the only currently proven effective therapy. Patients with frequent electrical storms may even need cardiac transplantation as last resort.

Source: MEDLINE


Author(s): Veiser T, Laurent G, Wolf JE

Citation: Casopis Lekaru Ceskych, 2005, vol./is. 144/4(219-23), 0008-7335

Publication Date: 2005

Abstract: Brugada syndrome is believed to be responsible for 4 to 12% of all sudden deaths and for 20% of deaths in patients with structurally normal hearts. As a distinct clinical entity with a high risk of sudden cardiac death it was first described in 1992. The syndrome characterized by ST segment elevation in right precordial leads V1 to V3 unrelated to ischemia and by electrolyte disturbance without obvious structural heart disease. The clinical findings are based on ECG and syncope or sudden death. The arrhythmia leading to sudden death is a rapid polymorphic ventricular tachycardia. The electrocardiographic signature of the syndrome is dynamic and often concealed, but can be unmasked by potent sodium channel blockers such as flecaïnide, ajmaline. The Brugada syndrome is a familial disease displaying an autosomal dominant mode of transmission with incomplete penetration and with incidence ranging between 5 and 66 per 10,000. The syndrome has been linked to mutations in SCNA5, the gene encoding for the a subunit of the sodium channel. Implantation of an automatic cardioverter-defibrillator is the only
currently proven effective therapy.

Source: MEDLINE

34. Anesthetic management of a patient with Brugada syndrome

Author(s): Imai Y., Niwa H., Yamada M., Harada J.

Citation: Journal of Japanese Dental Society of Anesthesiology, 2005, vol./is. 33/2(277-278), 0386-5835

Publication Date: 2005

Source: EMBASE

35. Anesthetic management of a patient with Brugada type ECG

Author(s): Sato Y., Hirai Y., Tachinami Y., Yamaguchi T., Iwatsuki N.

Citation: Journal of Japanese Dental Society of Anesthesiology, 2005, vol./is. 33/2(275-276), 0386-5835

Publication Date: 2005

Source: EMBASE

36. Abnormal myocardial presynaptic norepinephrine recycling in patients with Brugada syndrome.


Citation: Circulation, November 2004, vol./is. 110/19(3017-22), 1524-4539

Publication Date: November 2004

Abstract: BACKGROUND: Life-threatening ventricular tachyarrhythmias can occur in young patients without structural heart disease (idiopathic forms). In many patients, these are typically triggered by an increased sympathetic tone, eg, by physical or mental stress. In contrast, in Brugada syndrome, ventricular tachyarrhythmias more often occur during rest or sleep when the vagal tone is predominant. Furthermore, adrenergic agonists can reduce the level of ST-segment elevation, whereas it is increased by parasympathetic agonists or adrenergic antagonists. The aim of this study was to investigate presynaptic and postsynaptic myocardial sympathetic function in patients with Brugada syndrome.

METHODS AND RESULTS: Nine patients with Brugada syndrome (6 male, 3 female; age, 41+/−13 years) were enrolled in this study. The cardiac autonomic nervous system was assessed noninvasively, quantifying myocardial presynaptic and postsynaptic sympathetic function by means of positron emission tomography with the norepinephrine analogue 11C-Hydroxyephedrine (11C-HED) and the nonselective beta-blocker 11C-CGP 12177 (11C-CGP). Presynaptic sympathetic norepinephrine recycling, assessed by 11C-HED, was globally increased in patients with Brugada syndrome compared with a group of age-matched healthy control subjects (92.9+/−16.2 mL/g versus 69.1+/−14.2 mL/g; P<0.05), whereas postsynaptic beta-adrenoceptor density, assessed by 11C-CGP, was similar in patients and control subjects (10.4+/−6.7 pmol/g versus 10.2+/−2.9 pmol/g; P=NS).

CONCLUSIONS: The present study on autonomic innervation in Brugada syndrome describes an enhanced presynaptic norepinephrine recycling with preserved beta-adrenoceptor density, further supporting the hypothesis of an autonomic dysfunction in Brugada syndrome. This is a further step toward the understanding of the pathophysiology of the disease with potential future impact on therapeutic strategies.
37. **Perioperative approach to a patient with Brugada syndrome.**

**Author(s):** Candiotti KA, Mehta V

**Citation:** Journal of Clinical Anesthesia, November 2004, vol./is. 16/7(529-32), 0952-8180

**Publication Date:** November 2004

**Abstract:** Brugada syndrome is a recently described cardiac anomaly that may be responsible for up to one half of all sudden cardiac deaths in young adults without structural heart disease. It may also be worsened by beta-blockers, and it is almost unreported in the English language anesthesia literature.

**Source:** MEDLINE

38. **Anaesthesia in patients with Brugada syndrome.**

**Author(s):** Kim JS, Park SY, Min SK, Kim JH, Lee SY, Moon BK, Chae YJ

**Citation:** Acta Anaesthesiologica Scandinavica, September 2004, vol./is. 48/8(1058-61), 0001-5172

**Publication Date:** September 2004

**Abstract:** Brugada syndrome is characterized by right bundle branch block, ST segment elevation in the precordial leads and sudden death caused by ventricular fibrillation. We present two successful anaesthetic management cases in patients with Brugada syndrome.

**Source:** MEDLINE

39. **[Anesthetic management of two patients with Brugada-type ECG and of different clinical severity].**

**Author(s):** Hiuge Y, Ohta N, Hirata T, Mori T

**Citation:** Masui - Japanese Journal of Anesthesiology, June 2004, vol./is. 53/6(693-5), 0021-4892

**Publication Date:** June 2004

**Abstract:** Brugada syndrome is an arrhythmia syndrome characterized by typical electrocardiogram (Brugada-type ECG) and development of ventricular fibrillation (Vf) without any distinct structural heart diseases. The essential goal in the management of Brugada syndrome is to avoid the development of Vf. However, there has been no
established consensus on pre-operative risk assessment of patients with Brugada-type ECG. We recently experienced two cases of anesthetic managements for patients with Brugada-type ECG. Based on these experiences and recent cardiological progress on the risk stratification of Brugada syndrome, we thoroughly discuss on the peri-operative managements for patients with Brugada-type ECG.

**Source:** MEDLINE

40. [52-year old patient with recurrent syncope and temporary right precordial EKG-changes with fever]. [German] 52-jähriger Patient mit rezidivierenden Synkopen und temporären rechtsprakordialen EKG-Veränderungen bei Fieber.

**Author(s):** Caliskan R, Zabel M, Witzenbichler B, Sticherling C

**Citation:** Internist, June 2004, vol./is. 45/6(707-12), 0020-9554

**Publication Date:** June 2004

**Abstract:** This case report highlights the importance of considering the differential diagnosis of a primary electrical cardiac disease in a patient with unexplained syncope. In the absence of positive findings in his cardiac and neurological work-up, the presented patient had been diagnosed with "cryptogenic" epilepsy. During a febrile episode, however, his 12-lead ECG showed ST-segment elevations in leads V(1) and V(2), typical for the Brugada-Syndrome. Hence, his antiepileptic medication was discontinued and the patient received an implantable defibrillator. Pathophysiology, diagnosis, risk stratification, as well as the treatment options for this disease of the cardiac sodium channel are reviewed.

**Source:** MEDLINE


**Author(s):** Juang JM, Huang SK

**Citation:** Cardiology, 2004, vol./is. 101/4(157-69), 0008-6312

**Publication Date:** 2004

**Abstract:** In 1992, Brugada and Brugada described 8 patients with a history of aborted sudden death and a distinct ECG pattern of right bundle-branch block with ST segment elevation in leads V1-V3 and normal QT interval in the absence of any structural heart disease. It is called Brugada syndrome now and is believed to be responsible for 4-12% of all sudden deaths and around 20% of deaths in patients with structurally normal hearts. Although this syndrome is observed worldwide and the exact prevalence is unknown, it is more common in the Southeast Asian countries. Repeated syncope, ventricular fibrillation, and sudden cardiac death have been reported in patients with Brugada syndrome. The clinical presentation of Brugada syndrome is distinguished by a male predominance and the appearance of arrhythmic events at an average age of 40 years. The Brugada syndrome is inherited in an autosomal dominant manner with incomplete penetrance and an incidence ranging between 5 and 66 per 10,000. The surface ECG manifestations of the syndrome can transiently disappear, but can be unmasked by potent sodium channel blockers in some cases. Mutations of the cardiac sodium channel SCN5A have been detectable in <20% of patients with Brugada syndrome. Recent genetic studies have confirmed the genetic heterogeneity of the disorder. Antiarrhythmic drugs appear to be of little use in prolonging survival and in preventing recurrences of ventricular arrhythmias. To date, implantable cardioverter defibrillator remains the best therapy to prevent sudden death in these patients. Copyright 2004 S. Karger AG, Basel

**Source:** MEDLINE
42. Pathophysiology of Brugada syndrome

Author(s): Chahine M

Citation: Journal of Cardiovascular Electrophysiology, November 2003, vol./is. 14/11(1257-8; author reply 1258), 1045-3873

Publication Date: November 2003

Source: MEDLINE

Full Text:
Available in fulltext at EBSCO Host


Author(s): Antzelevitch C, Brugada P, Brugada J, Brugada R, Shimizu W, Gussak I, Perez Riera AR

Citation: Circulation Research, December 2002, vol./is. 91/12(1114-8), 1524-4571

Publication Date: December 2002

Abstract: The Brugada syndrome has gained wide recognition throughout the world and today is believed to be responsible for 4% to 12% of all sudden deaths and approximately 20% of deaths in patients with structurally normal hearts. The incidence of the disease is on the order of 5 per 10 000 inhabitants and, apart from accidents, is the leading cause of death of men under the age of 50 in regions of the world where the inherited syndrome is endemic. This minireview briefly summarizes the progress made over the past decade in our understanding of the clinical, genetic, cellular, ionic, and molecular aspects of this disease.

Source: MEDLINE

Full Text:
Available in fulltext at Highwire Press
Available in fulltext at Ovid

44. General anaesthesia in a patient with Brugada syndrome

Author(s): Edge CJ, Blackman DJ, Gupta K, Sainsbury M

Citation: British Journal of Anaesthesia, November 2002, vol./is. 89/5(788-91), 0007-0912

Publication Date: November 2002

Abstract: The successful administration of a combined general and epidural anaesthetic to a patient with Brugada syndrome is reported. A review of the literature is presented.

Source: MEDLINE

Full Text:
Available in fulltext at Highwire Press
Available in print at Grantham Hospital Staff Library
45. The prevalence, incidence and prognostic value of the Brugada-type electrocardiogram: a population-based study of four decades.

Author(s): Matsuo K, Akahoshi M, Nakashima E, Suyama A, Seto S, Hayano M, Yano K

Citation: Journal of the American College of Cardiology, September 2001, vol./is. 38/3(765-70), 0735-1097

Publication Date: September 2001

Abstract: OBJECTIVES: We sought to demonstrate the prevalence, incidence and prognostic value of the Brugada-type electrocardiogram (ECG) in a general population. BACKGROUND: The Brugada syndrome is characterized by evidence of right bundle branch block and ST segment elevation in the right precordial leads, as well as sudden death caused by ventricular fibrillation. However, the natural history of the Brugada-type ECG remains unclear. METHODS: We investigated 4,788 subjects (1,956 men and 2,832 women) who were <50 years old in 1958 and had undergone biennial health examinations, including electrocardiography, through 1999. The Brugada-type ECG was defined as a terminal r' wave in lead V(1) and ST segment elevation > or =0.1 mV in leads V(1) and V(2). Unexpected death was defined as sudden death or unexplained accidental death. RESULTS: There were a total of 32 Brugada-type ECG cases; the prevalence and incidence were 146.2 in 100,000 persons and 14.2 persons per 100,000 person-years, respectively. The incidence was nine times higher among men than women, and the average age at presentation was 45 +/- 10.5 years. The Brugada-type ECG appeared intermittently in most cases and was found in 26% of subjects who died unexpectedly. Cox survival analysis revealed that mortality from unexpected death was significantly higher in subjects with a Brugada-type ECG than in control subjects (p < 0.01). Unexpected deaths were more frequent among subjects with the Brugada-type ECG who had a history of syncope (p < 0.05). CONCLUSIONS: The Brugada-type ECG is not a very rare condition in the adult Japanese population. Subjects with a Brugada-type ECG have an increased risk of unexpected death.

Source: MEDLINE

Full Text: Available in fulltext at Highwire Press

46. Molecular biology and cellular mechanisms of Brugada and long QT syndromes in infants and young children.

Author(s): Antzelevitch C

Citation: Journal of Electrocardiology, 2001, vol./is. 34 Suppl/(177-81), 0022-0736

Publication Date: 2001

Abstract: Sudden cardiac death accounts for 19% of sudden deaths in children between 1 and 13 years of age and 30% of sudden deaths that occur between 14 and 21 years of age. The incidence of sudden cardiac death displays 2 peaks: one between 45 and 75 years of age, as a result of coronary artery disease, and the other between birth and 6 months of age, caused by sudden infant death syndrome. The role of cardiac arrhythmias in sudden infant death syndrome has long been a matter of debate and the role of cardiac arrhythmias in children in general is not well defined. Recent findings point to a contribution of primary
electrical diseases of the heart including the Brugada and long QT syndromes to sudden death in infants and children. Mutations in SCN5A and HERG and KvLQT1 have been shown to be associated with life-threatening arrhythmias and long QT intervals in young infants. These mutations cause changes in sodium and potassium currents that amplify intrinsic electrical heterogeneities within the heart, thus providing a substrate as well as a trigger for the development of reentrant arrhythmias, including Torsade de Pointes (TdP), commonly associated with the long QT syndrome (LQTS). Mutations in SCN5A have also been shown to cause the sodium channel to turn off prematurely and thus to set the stage for the development of a rapid polymorphic ventricular tachycardia/ventricular fibrillation in patients with the Brugada Syndrome. In LQTS, ion channel mutations cause a preferential prolongation of the M cell action potential that contributes to the development of long QT intervals, wide-based or notched T waves, and a large transmural dispersion of repolarization, which provides the substrate for the development of TdP. An early afterdepolarization-induced triggered beat is thought to provide the extrasystole that precipitates TdP. In the Brugada syndrome, mutations in SCN5A reduce sodium current density, causing premature repolarization of the epicardial action potential due to an all or none repolarization at the end of phase 1. The loss of the action potential dome in epicardium, but not endocardium, creates a dispersion of repolarization across the ventricular wall, resulting in a transmural voltage gradient that manifests in the electrocardiogram (ECG) as an ST-segment elevation and in the development of a vulnerable window during which reentry can be induced. Under these conditions, loss of the action potential dome at some epicardial sites but not others gives rise to phase 2 reentry, which provides an extrasystole capable of precipitating ventricular tachycardia/ventricular fibrillation (or rapid TdP). The practical importance of identifying infants and children with Brugada and LQTS syndromes lies in the fact that most deaths due to these congenital defects can be prevented. A simple ECG is often sufficient to permit diagnosis and thus to prevent the development of life-threatening arrhythmic events. Mass ECG screening of neonates and children however has been the subject of debate focused on issues ranging from the emotional impact of dealing with false positives to those concerning socio-economic and medico-legal factors. These issues are discussed in other articles. These concerns notwithstanding, it is important that we continue to question whether the economic inefficiencies of current screening methodologies supersede the value of a young life.

Source: MEDLINE

47. Sudden death in high-risk family members: Brugada syndrome.

Author(s): Brugada P, Brugada R, Brugada J

Citation: American Journal of Cardiology, November 2000, vol./is. 86/9A(40K-43K), 0002-9149

Publication Date: November 2000

Abstract: Brugada syndrome (an electrocardiographic pattern of right bundle branch block, ST segment elevation in leads V1 to V3, and sudden death) is genetically determined and caused by mutations in the cardiac ion channels. The mode of inheritance of the disease is autosomal dominant in half of familial forms. Sudden death may, however, occur from a variety of causes in relatives and patients with this syndrome. Twenty-five Flemish families with this syndrome with a total of 334 members were studied. Affected members were recognized by means of the typical electrocardiogram of the syndrome, either occurring spontaneously or after the intravenous administration of antiarrhythmic drugs. Sudden deaths in these families were classified as related or not to the syndrome by analysis of the data at the time of the event, mode of inheritance of the disease, and data provided by survivors. Of the 25 families with the syndrome, 18 were symptomatic (at least 1 sudden death related to the syndrome) and 7 were asymptomatic (no sudden deaths related to the syndrome). In total, there were 42 sudden cardiac deaths (12% incidence). Twenty-four sudden deaths were related to the syndrome and all happened in symptomatic families. Eighteen sudden deaths (43% of total sudden deaths) were not related to the syndrome (9
cases) or were of unclear cause (9 cases). Three of them occurred in 2 asymptomatic families and the remaining 15 in 5 symptomatic families. A total of 24 of the 50 affected members (47%) and 18 of the 284 unaffected members (6%) had aborted sudden death. This difference in the incidence of sudden death was statistically significant (p <0.0001). Patients with aborted sudden death caused by the syndrome were younger than patients with sudden death of other or unclear causes (38 +/- 4 years vs 59 +/- 3 years respectively; p = 0.0003). In families at high risk of sudden death because of genetically determined diseases, the main cause of sudden death remains the disease itself. However, almost half of sudden deaths are caused by unrelated diseases or from unclear causes. Accurate classification of the causes of sudden death is mandatory for appropriate analysis of the causes of death when designing preventive treatments.

Source: MEDLINE

48. [Anesthetic management of a patient with Brugada syndrome].

Author(s): Sugi Y, Mori M, Ono M, Kurihara Y

Citation: Masui - Japanese Journal of Anesthesiology, August 2000, vol./is. 49/8(884-6), 0021-4892

Publication Date: August 2000

Abstract: Brugada syndrome is characterized by right bundle-branch block, ST elevation in leads V 1 through V 3 and normal QT interval. Ventricular fibrillation frequently occurs in patients with Brugada syndrome. There have been few reports of anesthetic management of Brugada patients. We managed a 47-year-old man with Brugada syndrome, who underwent hemilaminectomy under general anesthesia, without untoward cardiovascular events. Potential problems in anesthetic management of patients with Brugada syndrome are also discussed.

Source: MEDLINE


Author(s): Brugada J, Brugada P, Brugada R

Citation: Revista Espanola de Cardiologia, February 2000, vol./is. 53/2(275-85), 0300-8932

Publication Date: February 2000

Abstract: In 1992 we described a new syndrome characterized by syncopal or sudden death episodes in patients with a structurally normal heart and a characteristic electrocardiogram 9 showing a pattern of right bundle branch block and ST segment elevation in right precordial leads V1 to V3. The disease is genetically determined with and autosomic dominant pattern of transmission. Until now three mutations and one polymorphism in the sodium cardiac channel gene have been identified in two families and one sporadic patient. As in many other genetically determined diseases, the disease is heterogeneous, caused by more than one gene. The syndrome has been identified in almost all countries in the world. Its incidence is difficult to evaluate, but it seems to be responsible for 4 to 10 sudden deaths per year per 10,000 inhabitants in areas like Laos or Thailand, and it represents the most frequent cause of death in young male adults in these countries. Up to 50% of all sudden deaths in patients with structurally normal heart are caused by the disease. The diagnosis can be easily made thanks to the characteristic electrocardiographic pattern. In some patients, the presence of concealed and intermittent forms might make the diagnosis more difficult. The electrocardiogram can be modulated by
Autonomic changes and administration of antiarrhythmic drugs. Beta-adrenergic stimulation normalizes the electrocardiogram, whereas ajmaline, flecainide or procainamide administration increase ST segment elevation. These drugs allow the unmasking of concealed or intermittent forms of the disease. Prognosis of patients with the syndrome is poor without an implantable defibrillator and antiarrhythmic drugs like amiodarone or betablockers do not protect against sudden death. The poor prognosis is similar in patients with a history of aborted sudden death or syncope and in asymptomatic patients in whom the abnormal electrocardiogram characteristic of the syndrome, was identified during a routine examination.

Source: MEDLINE

50. Anesthetic management of a patient with Brugada syndrome for implantable cardioverter defibrillator implantation

Author(s): Jindai R., Tanaki N., Ohmura S., Yamamoto K., Kobayashi T.

Citation: Hokuriku Journal of Anesthesiology, 2000, vol./is. 34/1(21-24), 0367-5947

Publication Date: 2000

Abstract: Brugada syndrome causes idiopathic ventricular fibrillation and sudden death. We report a case with Brugada syndrome who was scheduled for implantable cardioverter defibrillator (ICD) implantation. The patient's electrocardiogram showed right bundle branch block and persistent ST-segment elevation in the right precordial leads. Before anesthetic induction, two adhesive electrodes for external defibrillation were placed for the management of an unexpected ventricular fibrillation. Anesthesia was induced with thiopental, and was maintained with sevoflurane and nitrous oxide in oxygen. During surgery, induced ventricular fibrillation was easily controlled by ICD. The adhesive electrodes for external defibrillation should be placed in implantation of ICD.

Source: EMBASE

51. Anesthetic management of a patient with brugada syndrome

Author(s): Sugi Y., Mori M., Ono M., Kurihara Y.

Citation: Japanese Journal of Anesthesiology, 2000, vol./is. 49/8(884-886), 0021-4892

Publication Date: 2000

Abstract: Brugada syndrome is characterized by right bundle-branch block, ST elevation in leads V1 through V3 and normal QT interval. Ventricular fibrillation frequently occurs in patients with Brugada syndrome. There have been few reports of anesthetic management of Brugada patients. We managed a 47-year-old man with Brugada syndrome, who underwent hemilaminectomy under general anesthesia, without untoward cardiovascular events. Potential problems in anesthetic management of patients with Brugada syndrome are also discussed.

Source: EMBASE

52. The syndrome of right bundle branch block ST segment elevation in V1 to V3 and sudden death--the Brugada syndrome.

Author(s): Brugada J, Brugada P, Brugada R

Citation: Europace, July 1999, vol./is. 1/3(156-66), 1099-5129

Publication Date: July 1999
Abstract: In 1992 a new syndrome was described consisting of syncopal episodes and/or sudden death in patients with a structurally normal heart and an electrocardiogram (ECG) characteristic of right bundle branch block with ST segment elevation in leads V1 to V3. The disease is genetically determined, with an autosomal dominant pattern of transmission. Three different mutations that affect the structure and function of the cardiac sodium channel gene SCN5A have been identified. Two mutations result in total loss of function of the sodium channel. The other mutation results in acceleration of the recovery of the sodium channel from inactivation. The incidence of the disease is difficult to estimate, but it causes 4 to 10 sudden deaths per 10000 inhabitants per year in areas like Thailand and Laos. In these countries, the disease represents the most frequent cause of death in young adults. Up to 50% of the yearly sudden deaths in patients with a structurally normal heart are caused by this syndrome. The diagnosis is easily made by means of the ECG. The presence of concealed and intermittent forms, however, make the diagnosis difficult in some patients. The ECG can be modulated by changes in autonomic balance and the administration of antiarrhythmic drugs. Beta-adrenergic stimulation normalizes the ECG, while intravenous ajmaline, flecainide or procainamide accentuate ST segment elevation and are capable of unmasking concealed and intermittent forms of the disease. Recent data suggest that loss of the action potential dome in the right ventricular epicardium but not the endocardium underlies ST segment elevation seen in the Brugada syndrome. Also, electrical heterogeneity within the right ventricular epicardium leads to the development of closely coupled extrasystoles via a phase 2 reentrant mechanism, which then precipitates ventricular tachycardia-ventricular fibrillation. Right ventricular epicardium is preferentially affected because of the predominance of transient outward current in this tissue. Antiarrhythmic drugs like amiodarone and beta-blockers do not prevent sudden death in symptomatic or asymptomatic individuals. Gene therapy may offer a cure in future years. Implantation of an automatic cardioverter-defibrillator is the only currently proven effective therapy.

Source: MEDLINE

Full Text:

Available in fulltext at Highwire Press