A peer-reviewed, case-based, indexed, on-line, interactive and multimedia, open-access journal that will provide the global electrophysiology community high-quality case reports in a Grand Rounds format.

**CO-EDITORS-IN-CHIEF:**
RANJAN K. THAKUR, MD
ANDREA NATALE, MD

**MANAGING EDITOR:**
BETSY BOGDANSKY

**ASSOCIATE EDITORS:**
NOEL BOYLE
PAUL A. LEVINE
CHANG-SHENG MA
FREDERIC SACHER
PASQUALE SANTANGELEI
ASHOK J. SHAH

**SECTION EDITORS:**
AMIN AL-AHMAD
NOEL BOYLE
ALAN CHENG
TOM KENNY
PAUL A. LEVINE
YASH LOKHANDBALA
ANDRES PEREZ-RIERA
MELVIN SCHEINMAN
MOHAMMAD SHENASA
ALBERT WALDO
ANDREW WIT

Accepting papers at: http://www.arrhythmiaigr.com

Published by

**Science International Corp.**

ISSN 2326-4012
On behalf of the Organizing Committee, it is my great honour to invite you to join the CardioRhythm 2017, to be held in Hong Kong from 10 - 12 February 2017. The Conference is jointly organized by the Hong Kong College of Cardiology and Chinese Society of Pacing and Electrophysiology.

CardioRhythm 2017 will provide an excellent opportunity for you to stay abreast of the latest developments in the field of cardiac rhythm management. Lectures, case presentations and workshops focusing on sudden cardiac death, new drug and ablation treatment for atrial fibrillation, pacing and ICD advances, cardiac resynchronization techniques, remote patient monitoring, and advances in neuromodulation for heart failure and hypertension will be presented by experts from around the world.

Look forward to seeing you in CardioRhythm 2017!

Prof Hung-Fat TSE
Chairman
CardioRhythm 2017
CALL FOR ABSTRACTS:

VA2015 cordially invites you to submit online an abstract to be considered for presentation at our meeting at the Cini Foundation on the Isola di San Giorgio. To inquire about the meeting and the online submission, please contact scientific@venicearrhythmias.org

www.venicearrhythmias.org
Editorial Board

Co-Editors-in-Chief
Ranjan K. Thakur
Andrea Natale

Advisory Board
Masood Akhtar
Serge Barold
A John Camm
Jeremy Ruskin
Mel Scheinman

Section Editors
Amin Al-Ahmad
Noel Boyle
Tom Kenny
Paul A. Levine
Yash Lokhandwala
Alan Cheng

Editorial Board
Albert, Christine
Al-Ahmad, Amin
Antzelevitch, Charles
Asirvatham, Samuel
Auricchio, Angelo
Bhatt, Deepak
Blanc, Jean Jacques
Brachmann, Johannes
Brugada, Pedro
Burke, Martin
Callans, David
Cappato, Riccardo
Chen, Shih Ann
Chinitz, Larry
Chugh, Sumeet
Chung, Mina
Cleland, John
Cummings, Jennifer
Damiano, Ralph

Managing Editor
Betsy Bogdansky

Associates Editors
Noel Boyle
Paul A. Levine
Chang-Sheng Ma
Frederic Sacher
Pasquale Santangeli
Ashok J. Shah

Anders Perez-riera
Melvin Scheinman
Mohammad Shenasa
Albert Waldo
Andrew Wit

Goldberger, Jeff
Gopinathiannair, Rakesh
Grant, Augustus
Greene, Anne
Grubb, Blair
Hayes, David
Hohnloser, Stefan
Israel, Carsten
Jais, Pierre
Jalife, Jose
Kautzner, Josef
Klein, Helmut
Knight, Bradley
Krahm, Andrew
Kusomoto, Fred
Kutalek, Steven
Lampert, Rachel
Lau, Chu Pak
Lebedev, Dmitry
Levy, Samuel
Lewis, William
Link, Mark
Love, Charles
McAnulty, Jack
Malik, Marek
Marchlinski, Frank
Maron, Barry
Michaud, Greg
Moulton, Kreigh
Naccarelli, Jerry
Nademanee, Koonlawee
Narasimhan, Calambur
Narayan, Sanjiv
Nogami, Akihiko
Obel, Pro

Oseroff, Oscar
Oto, Ali
Ovsyshcher, I.Eli
Pogwizd, Steven
Poole Jeanne
Priori, Silvia
Raj, Satish R
Raviele, Antonio
Reddy, Vivek
Revisvilli, Amiran
Ritter, Philippe
Rottman, Jeff
Russo, Andrea
Saksena, Sanjiv
Saliba, Walid
Schuger, Claudio

Shen, Win-Kuang
Shivkumar Kalyanam
Shorofsky, Steve
Singh, Balbir
Vardas, Panos
Varma, M.D., Niraj
Verdino, Ralph
Viskin, Sami
Weachter, Richard
Wellens, Hein
Wharton, Marcus
Widman, Larry
Worley Seth J
Zareba, Wojciech
## Editorial

1 Welcome to Arrhythmia Grand Rounds Journal  
Ranjan K. Thakur, Andrea Natale

## Case Reports

3 Acute Digitalis Delirium Associated with Intravenous Digoxin Administration  
Matthew J. Lengel, Kun Xiang, Vincent F. Mauro, Blair P. Grubb, Christopher J. Cooper

7 Electrical Storm in a Patient with Dilated Cardiomyopathy Suppressed with Cardiac Resynchronization Therapy  
Dimitris Tsiachris, Odysseas Kaitozis, Skevos Sideris, Polychronis Dilaveris, Konstantinos Gatzoulis

11 Syncope Due to Positional Alternating Bundle Branch Block  
Shadi Idris, Howard Weitz, Behzad B. Pavri

16 Malignant Mimicry: An Unusual Cause of VT and AV Conduction Disease  
Yehoshua C. Levine, Rupal Parekh O’Quinn, Paul A. VanderLaan, Peter Zimetbaum

## Case Studies

20 Management of Sinus Node Dysfunction with Accelerated Junctional Rhythms  
Paul A. Levine

26 Method of Assessing the Safety of Functional Single Chamber Atrial Pacing: The AV Nodal Conduction System Stress Test  
Paul A. Levine

## ECG Challenge

33 An Irregular Narrow Complex Tachycardia: What Is Your Diagnosis?  
ECG Challenge with Section Editor Melvin Scheinman, MD  
Mark E. Josephson

## EP Lab Challenge

35 Worth the Delay  
Yash Lokhandwaala
Bench to Bedside

36 Basic Foundations of Clinical Electrophysiology
Andrew L. Wit

38 Bench to Bedside - A Difficult Journey
Negar Salehi, Aravdeep S. Jhand, Vaibhav Satija, Watchara Lohawijarn, Ranjan K. Thakur

Profiles in EP

42 Profiles in Cardiac Electrophysiology
Albert L. Waldo

44 Reflections and Reminiscing on the First Catheter Ablation of the AV Junction in Man
Melvin Scheinman
Welcome to Arrhythmia Grand Rounds

Ranjan K. Thakur, MD¹, Andrea Natale, MD²

¹ Sparrow Thoracic and Cardiovascular Institute, Lansing, Michigan, USA
² St. David’s Medical Center, Texas Cardiac Arrhythmia Institute, Austin, Texas, USA

On behalf of Science International and Dr. Steven Korn, publisher, Betsy Bogdansky, Managing Editor and the editorial board, we welcome readers to the first issue of Arrhythmia Grand Rounds (AGR).

Initially, cardiac electrophysiologic investigations and treatments were only available to patients in advanced economies, but in the last one to two decades, very sophisticated levels of electrophysiologic interventions have become widely available all over the world. This means that sophisticated electrophysiologists are practicing all over the world and they have ongoing educational needs. Many traditional journals are not easily available to readers in parts of the world, largely because of cost. Fortunately, advancements in electronic media make it possible for people all over the world to meet virtually and learn together.

Arrhythmia Grand Rounds was envisioned several years ago as a premier educational venue for clinical electrophysiologists all over the world. It has taken some time to get to the first issue and we are delighted to welcome the readership. We hope to make the journal an ideal teaching and learning environment. We hope to make this process interactive, multimedia and democratic. This evolution will take some time and we encourage your participation and feedback to help us achieve these goals.

We have excellent traditional journals for dissemination of new research findings. However, clinicians enhance their clinical problem solving skills by dissecting and discussing difficult clinical cases with their colleagues and debating therapeutic alternatives – that’s what this journal will aim to do. This journal will provide a case-based approach to clinical cardiac electrophysiology and in due course, we hope to become the preferred destination for unique case reports as well as case reports that offer teachable points. In addition, reviews, editorials, review questions, challenging ECGs, EGMs, and images, etc. from the global electrophysiologic community as well as thought leaders will enhance AGR’s educational mission – to teach electrophysiology, one case at a time. We hope that all those who deal with arrhythmias - electrophysiologists, cardiologist, fellows in training, associated professionals and primary care physicians - will find the journal useful in furthering their knowledge. AGR has one very unique feature to enhance that mission. Case reports and other published material will also be available in slide format so that readers may easily download them and use them for teaching fellows, residents, colleagues and others.

All case reports, editorials, etc. in this journal will be peer reviewed and indexed using the d.o.i. indexing system. For contributors of peer reviewed contents, this will represent a peer reviewed publication. In due course, we will strive for Medline indexing.

The journal has assembled a veritable editorial board. Over the last several months, Dr. Paul A. Levine led the journal and made the first issue possible. Unfortunately, because of his sudden illness, he has asked us to take over as Co-Editors-in-Chief. We offer our best wishes and prayers to Paul and his family as...
they negotiate the difficult days ahead and hope for his full recovery. Dr. Levine will continue to actively participate in helping us shape and guide the journal. The torch has been passed to us and we will endeavor to fulfill the original vision. We shall appreciate your thoughts as to your educational needs and how we should meet those needs.

We thank our colleagues who have guided us in developing and encouraging this journal; they are too numerous to name individually, but we recognize their contributions and appreciate their encouragement. On behalf of the students and teachers of electrophysiology worldwide, we thank our friends in industry who support this educational endeavor. Education makes all of us better!

Again, on behalf of all of us, welcome to Arrhythmia Grand Rounds.

Ranjan K. Thakur, MD

Andrea Natale, MD

Co-Editors-in-Chief
Arrhythmia Grand Rounds
Acute Digitalis Delirium Associated with Intravenous Digoxin Administration

Matthew J. Lengel, PharmD, Kun Xiang, MD, PhD, Vincent F. Mauro, PharmD, Blair P. Grubb, MD, Christopher J. Cooper, MD

1 Department of Pharmacy Practice, College of Pharmacy and Pharmaceutical Sciences, University of Toledo, Toledo, Ohio, USA
2 Division of Cardiovascular Medicine, Department of Medicine, University of Toledo, Toledo, Ohio, USA

Abstract

A 77-year-old woman with a reduced ejection fraction was admitted to the hospital for atrial fibrillation with a rapid ventricular rate. Intravenous digoxin was used to slow her ventricular rate. On multiple occasions in the absence of elevated serum digoxin concentrations, she demonstrated multiple episodes of acute delirium associated with the acute intravenous administration of digoxin.

Case Report

A 77-year-old white woman (84.8 kg) presented with complaints of shortness of breath and an increase in weight of approximately 20 pounds over the past month. Upon presentation, she was noted to be in new onset atrial fibrillation with a ventricular rate of 153 beats per minute. She was subsequently admitted to the hospital. The physical exam and laboratory investigation were both consistent with decompensated congestive heart failure. After initial aggressive diuresis, attempts at heart rate control, and anticoagulation therapy, a transesophageal echocardiogram revealed severely reduced left ventricular systolic function with an estimated left ventricular ejection fraction of 15%.

Two attempts of DC cardioversion failed to maintain the patient in sinus rhythm. The first attempt maintained sinus rhythm for only a few seconds. The second attempt was performed after 72 hours of an amiodarone intravenous infusion. The patient was maintained in sinus rhythm for only 12 hours. Therefore, pharmacotherapeutic management of the atrial fibrillation focused only on rate control.

Due to the relative contraindication of using a beta blocker or calcium channel blocker in a patient with acute decompensated heart failure, digoxin 0.25 mg was given intravenously for ventricular rate control. Immediately following administration, the patient demonstrated new onset transient confusion and
impaired short-term memory (she had no previous history of neuropsychiatric problems). Additional digoxin was not given until hospital day 5 when digoxin was again given for ventricular rate control. A dose of 0.25 mg were given intravenously at 1:28 p.m. and 6:45 p.m. Following the administration of each of these doses, the patient reported a feeling of malaise and both family and staff observed an alteration in the patient’s mental status with delirium and agitation which occurred within minutes after each of these doses. The following day, the patient received an additional 0.125 mg dose of digoxin intravenously at 12:18 a.m. Once again, immediately after administration the patient was noted to be agitated and disoriented. She paced around her room until 5:00 a.m. at which time she sat in a chair and stared into space and was non-communicative. At this time, digoxin-induced toxicity was suspected; however, a serum digoxin concentration obtained at 2:43 a.m. was 1.0 ng/mL. To further test if patient’s mental status changes were related to digoxin, a decision was made to closely monitor the patient when the next digoxin dose was administered later in the day. About 10 minutes after intravenous digoxin 0.25 mg was administered at 1:41 p.m., the patient once again became acutely confused, agitated, and delirious. She exhibited paranoid thoughts as well as a “flight of ideas” (making reference to airplanes on the roof and dogs barking in the room next door). This behavior continued for nearly an hour. A serum determination of all electrolytes obtained during this time period was normal. Seven hours after this digoxin dose, the patient’s serum digoxin level was 1.5 ng/mL. Her mental status improved throughout the night and she was functioning normally the next day. No further digoxin was administered to the patient during her hospital stay. The patient did not demonstrate any further neuropsychiatric symptoms for the remainder of her hospitalization.

No signs of dysrhythmias associated with digoxin toxicity were seen upon a review of telemetry records during the hour that followed each of the digoxin doses. The patient only displayed continuous atrial fibrillation with occasional premature ventricular contractions.

Discussion

Although digoxin toxicity generally results from elevated serum digoxin concentrations, it can occur within concentrations considered “therapeutic,” especially in the presence of electrolyte abnormalities (hypokalemia, hypomagnesemia, hypercalcemia), hypothyroidism, hypoxia, or amyloidosis [2,3].

Digoxin toxicity can have a highly variable clinical presentation but is commonly manifested by gastrointestinal, cardiac, and/or visual symptoms [3]. Altered mental status, sometimes presenting as delirium, is a known but less commonly encountered problem which frequently is not recognized by health professionals as a manifestation of digoxin toxicity.

Cases of delirium associated with digoxin use have been well-documented [4-6], beginning with the initial description by Duroziez in 1874 [7]. The majority of these reported cases were associated with either elevated serum concentrations or supratherapeutic doses of digitalis preparations. One case reported by Grubb [4] described a 68-year-old woman with no history of mental illness who was admitted to the hospital with severe agitation after being started on oral digoxin 0.25 mg daily 6 months prior for her heart failure. The evaluation demonstrated elevated serum digoxin concentration of 3.5 ng/mL. The patient’s mental status improved markedly after the digoxin was discontinued with a full recovery of mental status in 11 days. Shear and Sacks [5] described a 73-year-old woman on digoxin 0.25 mg daily demonstrated disorientation, agitation, and poor memory. A measured digoxin concentration in this patient was also elevated at 3.7 ng/mL.

Our patient demonstrated changes in mental status similar to the cases described above but the symptoms occurred in the absence of an elevated digoxin concentration. Eisendrath and Sweeney [6] described two elderly male patients exhibiting signs of delirium in conjunction with therapeutic serum digoxin concentrations. Both patients exhibited confusion, disorientation, significantly altered affect, and impaired short-term memory days to weeks after digoxin initiation. After the digoxin was discontinued, both patients were improved within 3 days with a complete recovery in 1 to 2 weeks. A possible explanation given for the neurologic events in the face of therapeutic
digoxin concentrations provided by Eisendrath and Sweeney [6] was the modestly low albumin concentration in these two patients. In contrast, the patient we describe above had a normal albumin concentration (3.8 mg/dL) and the confusion occurred within minutes of receiving single intravenous bolus doses of digoxin. Our patient’s mental status returned to her baseline state within 24 hours after the last digoxin dose was administered and sooner after earlier doses.

Our case differs from those previously published in the literature in that the abnormal behavior occurred within minutes of administering intravenous digoxin on more than one occasion and not during any other time. These episodes also occurred in the face of two therapeutic serum digoxin concentrations, suggesting our patient’s behavior was directly related to the intravenous digoxin administration as opposed to persistently elevated serum concentrations. Amiodarone has been implicated in causing altered mental status [8]. However, our patient did not demonstrate any altered mental status while on amiodarone alone, only shortly after being administered digoxin.

Amiodarone also has been implicated in reducing the tissue protein binding of digoxin, thereby reducing the volume of distribution of digoxin and causing an increase in digoxin serum concentrations and the risk for toxicity [9]. However, measured digoxin serum concentrations were not present in our patient and the patient was not on amiodarone the first time she demonstrated altered mental status after receiving digoxin.

A search of the literature did not demonstrate similar cases associated with the acute intravenous administration of digoxin. Due to its relatively rare incidence, the mechanism of acute digoxin induced delirium is not well studied. Martin and Shad [10] did describe a patient who experienced a hypersensitivity reaction to an intravenous bolus dose of digoxin but their patient’s mental status was not altered during the event.

When administered intravenously, digoxin has an approximate 6-hour distribution phase where its serum concentrations are elevated [11]. Toxicity is generally not seen at this time because digoxin is mostly in the systemic circulation rather than within tissues. Our patient appeared to be hypersensitive to the central nervous system effects of the higher digoxin serum concentrations that are present acutely following intravenous administration. A few hours later, once serum concentrations lowered to therapeutic concentrations as digoxin distributed into tissues, the patient consistently became asymptomatic.

In summary, we report a case of acute delirium directly associated with the intravenous use of digoxin on multiple occasions in an elderly woman, which were unrelated to elevated serum digoxin concentrations. Not only do health professionals need to be aware of the potential for digoxin to induce delirium when serum concentrations are elevated but also during acute intravenous administration. Early recognition and avoiding further digoxin administration in such instances is critical for the patient safety and the avoidance of potential adverse clinical outcomes.

Conflict of Interest

The authors have no conflict of interest relevant to this publication.

References


Cite this article as: Lengel MJ, Xiang K, Mauro VF, Grubb BP, Cooper CJ. Acute Digitalis Delirium Associated with Intravenous Digoxin Administration. Arrhythmia Grand Rounds 2015;1(1):3-6. DOI: http://dx.doi.org/10.12945/j.agr.2015.0002-14
Electrical Storm in a Patient with Dilated Cardiomyopathy Suppressed with Cardiac Resynchronization Therapy

Dimitris Tsiachris, MD1, Odysseas Kaitozis, MD2, Skevos Sideris, MD2, Polychronis Dilaveris, MD1, Konstantinos Gatzoulis, MD1

1 University of Athens, First Cardiology Clinic, Athens, Greece
2 Hippokration Hospital, Cardiology Department, Athens, Greece

Abstract
We present the case of a 55-year-old man with dilated cardiomyopathy and a syncopal episode who developed electrical storm with multiple instances of both monomorphic and polymorphic sustained ventricular tachycardia terminated by shocks in the face of worsening heart failure not responding to triple antiarrhythmic drug therapy including sedation and intubation in the coronary care unit. Hemodynamic improvement after cardiac resynchronization therapy with defibrillation backup (CRT-D) implantation brought about a dramatic response in the situation. This case report demonstrates the usefulness and efficiency of CRT-D as an option of treating electrical storm by improving ventricular function in the case of electrical instability being an expression of severe pump failure.

Key Words
Cardiac resynchronization therapy • Electrical storm • Ventricular tachycardia

Introduction
A worrying incidence of proarrhythmic effect in the form of electrical storm (ES) after the institution of cardiac resynchronization therapy (CRT) has been repeatedly reported in the recent past [1]. However there are contradictory reports related to either the proarrhythmic [1] or the antiarrhythmic [2, 3] potential of the CRT as far as the induction or and suppression of ES. In line with these reports, we present a case of ES in a patient with dilated cardiomyopathy not responding to triple antiarrhythmic drug therapy, including sedation and intubation, but successfully suppressed after biventricular pacing.

Case Report
A 55-year-old man with a history of psoriasis and dilated cardiomyopathy was referred to our clinic for the treatment of palpitations and a syncopal episode. His functional status was worsening New York Heart Association (NYHA) class III on optimal medical treatment with ACE-inhibitors, b-blockers, diuretics, and aldosterone antagonists. Amiodarone was also prescribed for a 2-month history of paroxysmal atrial fibrillation and discontinued 1 month prior to his admission for the syncopal episode. The resting electrocardiogram (ECG) showed a normal sinus rhythm with a prolonged PR interval of 210 ms, a wide QRS duration of 184 ms with a left bundle branch block (LBBB) pattern and left axis deviation, as well as a prolonged QT interval of 490 ms (QT corrected=550 ms) (Figure 1). A 24-hour Holter monitoring exhibited more than 3000 monomorphic premature ventricular...
contractions and a mean heart rate of 80 bpm without episodes of bradycardia, heart block or polymorphic ventricular tachycardia (VT).

The two-dimensional echocardiogram documented a marked left ventricular (LV) dysfunction (LV ejection fraction, LVEF of 25%) with a diffuse hypokinesia and increased dimensions of the LV, whereas the right ventricle presented normal dimensions and global function. The results of urine, laboratory tests, including electrolytes, and arterial blood gases were within normal ranges.

An invasive electrophysiological study (EPS) under metoprolol use revealed a normal sinus node function with significant conduction system disease, consisting of atrial-His (AH) interval at 114 ms and a His-ventricular (HV) interval prolonged at 94 ms. During programmed ventricular stimulation of the right ventricular outflow tract, a sustained polymorphic VT causing syncope was induced with triple ventricular extrastimuli and was terminated by shock.

The following days the patient went into ES with multiple and increasingly frequent episodes of both sustained monomorphic VT as well as polymorphic VT degenerating into ventricular fibrillation (VF) interrupted by repeated shocks. Amiodarone, xylocaine, metoprolol, and deep sedation with intubation were required in order to better control the ES. However the VT/VF episodes continued on the next 4 days. Based on telemetry analysis there were no episodes of bradycardia-related VT since heart rate prior to every VT episode exceeded 60 bpm in all cases.

At this stage we hypothesized that the multiple arrhythmic episodes were the result of the deterioration of the unstable electrophysiologic substrate as an expression of severe cardiac pump failure and that improving ventricular function by CRT would probably reduce ventricular arrhythmia burden [4].

A CRT with defibrillation capacity (CRT-D) was thus implanted. The procedure was uneventful without any complications. Sensing and pacing thresholds were optimal. Two days after the implantation an additional episode of VT was interrupted with the first attempt of antitachycardia pacing. Pre-implantation antiarrhythmic drug administration caused a further prolongation of QRS (195 ms) and corrected QT intervals (560 ms).

Eighteen months later the patient gained improvement of activities in daily life and is now in NYHA class II. The LVEF increased from 25% to 30% but most importantly he is free of ventricular arrhythmic events as

---

**Figure 1.** Initial (prior to amiodarone administration) 12 lead ECG showing LBBB with left axis deviation and borderline first-degree atrioventricular block.
no VT/VF episodes were recorded. Post-implantation QRS was 180 ms and corrected QT interval was 520 ms. Treatment with amiodarone remained for the first 12 months and discontinued thereafter (Figure 2).

Discussion

CRT has been well established to have a favorable impact on all-cause mortality and re-hospitalization for heart failure. The current guidelines suggest CRT-D therapy in order to reduce morbidity and mortality in patients with dilated cardiomyopathy with NYHA function class III or IV despite optimal pharmacological treatment, LVEF less than or equal to 35%, sinus rhythm (SR), LBBB, and QRS duration greater than or equal to 120 ms.

Nevertheless the effect of biventricular pacing on arrhythmia events has not been as clear cut. There are a number of reports suggesting that CRT can cause arrhythmia or even induction of electrical storm after the implantation of biventricular pacing [1]. This has been attributed to the altered ventricular activation sequence and the prolongation of ventricular repolarization phase induced by the LV pacing [1]. This is of particular concern in cases when CRT device implantation is not associated with a defibrillator back-up. The incidence reported in limited single series is low, between 3.4% and 4%, with predominance in ischemic cardiomyopathy. On the other hand there is strong evidence that CRT may improve several arrhythmogenic indices in both ischemic and dilated cardiomyopathy patients [5]. In line with these studies, MADIT-CRT trial documented a lower incidence of malignant ventricular tachyarrhythmias in patients responding to biventricular pacing. It has been suggested that reverse LV mechanical remodeling is associated with reversal of electrical remodeling and a lower rate of appropriate defibrillator therapy following CRT.

Similar to our case there are also two other case reports of ES suppression by biventricular pacing in patients with dilated cardiomyopathy and VT [2, 3]. It has also been suggested that the incidence of ES is lower among patients treated with a CRT-D as compared to those treated with an implantable cardiac defibrillator [4]. The full effect of biventricular pacing on the arrhythmogenic substrate of heart failure and especially its acute effect on ES is yet to be fully understood. Focusing on our case, we may recognize

Figure 2. Twelve months postimplant 12 lead ECG showing LBBB with right axis deviation and atrial pacing attributed to sick sinus syndrome secondary to amiodarone.
three different mechanisms that in parallel acted as ES suppressants. Beyond the hemodynamic improvement and the mild QT reduction related with biventricular pacing, late effect of amiodarone (which was administered systematically for the 12 months) might also contribute towards this direction. Our observations suggest that CRT can be an option of treating ES by improving ventricular function in the case of electrical instability being an expression of severe pump failure.

References


Cite this article as: Tsiachris D, Kaitozis O, Sideris S, Dilaveris P, Gatzoulis K. Electrical Storm in a Patient with Dilated Cardiomyopathy Suppressed with Cardiac Resynchronization Therapy. Arrhythmia Grand Rounds 2015;1(Issue 1):7-10. DOI: http://dx.doi.org/10.12945/j.agr.2015.00011-14

Conflict of Interest

The authors have no conflict of interest relevant to this publication.

Case Report

Comment on this Article or Ask a Question
Syncope Due to Positional Alternating Bundle Branch Block

Shadi Idris, MD, Howard Weitz, MD, FACC, Behzad B. Pavri, MD, FACC*

Department of Medicine, Division of Cardiovascular Medicine, Thomas Jefferson University Hospital, Philadelphia, USA

Abstract

We present the case of a 69-year-old female who described recurrent abrupt syncope associated only with recumbency or leaning forward. She had preexisting LBBB on her ECG, and a structurally normal heart. An implantable loop recorder captured episodes that were flagged as “asystole” and “pause detected,” but analysis of the recordings provided the correct diagnosis, and possible insight into the association with recumbency.

Key Words
Alternating bundle branch block • Recumbent syncope

Case Report

Alternating bundle branch block (BBB) is a class I indication for implantation of a permanent pacemaker [1], irrespective of symptoms. It is a harbinger of complete failure of conduction, and can present as unpredictable and sudden syncope. We describe a case where both syncope and alternating BBB were triggered by change in body position.

A 69-year-old female with a history of hypothyroidism and mild bronchospasm was referred for evaluation of recurrent syncope. She described five episodes of syncope over the last 9 months, each triggered by change in body position—either leaning back or lying down on her side. Three of these events occurred during yoga practice. The events were abrupt in onset without any premonitory symptoms; the resulting loss of consciousness was complete but brief, lasting less than 10 seconds in duration; and each episode was self-limited. Three of the five episodes were witnessed by her yoga instructor, and there was no seizure activities, loss of bladder or bowel continence, post event confusion, disorientation, or weakness. She had other episodes of presyncope as well, without loss of consciousness but with greying of vision, also triggered by similar changes in body position.

Her physical examination was completely within normal limits. There was no abnormal orthostatic change in heart rate or blood pressure. Given the positional nature of her symptoms, we performed sequential bilateral carotid message in both supine and seated positions; however, no bradyarrhythmias were provoked. Her resting ECG showed incomplete or complete left bundle branch block, which was known for the past four years, with QRS duration measurements ranging from 110 to 130 ms. An echocardiogram revealed a structurally normal heart.

She was offered the choice of a diagnostic EP study, an extended period of outpatient monitoring, an implantable loop recorder, or an empiric pacemaker. After obtaining a second opinion, she opted for the loop recorder, and a Medtronic Reveal Linq device was inserted subcutaneously at the fourth left intercostal space. The telemetered R waves measured 0.6 to 0.8 mV.

Upon interrogation of the device a week post implant there were multiple instances flagged by the device as “asystole.” Review of the stored ECGs revealed abrupt changes in QRS vector and amplitude resulting in undersensing (Figure 1). Because of her

* Corresponding Author:
Behzad B. Pavri, MD
Department of Medicine, Division of Cardiovascular Medicine
Thomas Jefferson University Hospital
925 Chestnut Street, Suite 200, Philadelphia PA 19107
Tel. 215-955-8882; Fax: 215-503-3976; E-Mail: behzad.pavri@jefferson.edu
history of positional symptoms, we elected to run real-time telemetry from her recorder while the wand was held firmly over the recorder location. Figure 2 shows the ECG changes that were recorded as the patient changed position from sitting to lying on her left side (A), from sitting to lying on her right side (B), and from sitting to leaning forward (C).

A permanent pacemaker was inserted within the next few days.

Discussion

Alternating BBB is a well-known cause of syncope, and reflects bilateral bundle branch disease. ECG manifestations may vary, often resulting in changing patterns of complete and incomplete BBB [2]. Syncope in the setting of alternating BBB is thought to occur when both bundles fail to conduct at the same time, resulting in abrupt asystole. The transition from conduction over one bundle to the other is often interspersed by one narrower QRS complex, which represents equally delayed conduction down both bundle branches, as evidenced in each of the 3 tracings in Figure 2. Conduction over each bundle may be (but does not necessarily have to be) associated with noticeable changes in PR interval [3]; such PR interval changes were not seen on the majority of the ECGs, but one stored example was found that demonstrated a 40 ms increase in PR interval when conduction occurred over the diseased left bundle branch (Figure 3).

Our initial concern was that the changes in QRS complex morphology were due to actual movement of the recorder in the subcutaneous tissues at the base of the left breast, resulting in a change in the perceived QRS vector. However, we excluded that possibility by: (a) ensuring that the Medtronic programmer head (wand) was firmly held against the chest wall, so that there was no displacement of the recorder with body motion; (b) the observation of the narrower QRS complex at the point of transition; (c) the observation that there was no discernible change in the P wave vector, which should have also changed if it was the recorder that was shifting; and (d) the associated PR interval change as shown in Figure 3.

Figure 1. Two representative stored tracings of changing conduction patterns. (A) The ECG changes from a left BBB to right BBB back to left BBB. (B) There are varying degrees of incomplete right BBB, ending with a non-specific conduction delay pattern. Note the intermittent loss of sensing due to changing QRS signal amplitude, resulting in the incorrect detection of “asystole” or “pause” by the device.
Upon reviewing the English language literature, we encountered very few examples of positional changes in the ECG. The earliest such report was prior to the development of precordial or augmented leads, dating back to 1933 [4]; in fact, the authors recommended that no attempt be made to designate the bundle branch involved, and that instead all such cases be called “intraventricular block of the bundle-branch type”. This assertion was then challenged in 1935 based on studies in dogs and monkeys [5]. In the setting of severe acute asthma, new right BBB and other ECG changes have been attributed to changes in right sided filling pressures and hyperinflation of the lungs [6]. We were unable to find any prior report on alternating BBB related to change in body position.

We also considered causes of syncope associated with changes in body position. The commonest etiology would be orthostatic syncope, with gravitational stress associated with arising from recumbency or from the seated position; however, our patient had the opposite situation, with syncope occurring with recumbency or leaning sideways. Carotid hypersensitivity causes syncope with head turning, and this was also excluded. Dizziness with recumbency associated with rotatory symptoms may be a manifestation of vertigo due to inner ear disease, but this too was not applicable to our patient. We came across a report of experimentally induced severe hypoxia causing recumbent syncope in 2% of healthy participants [7]. Seizures may occur during any position; syncopal seizures have been described during recumbency [8].

Finally, we reviewed the data on changes in ventricular filling pressures with changes in body position. In healthy male volunteers, recumbency increased the average right ventricular systolic pressure from 18.6 to 22.8 mmHg and end diastolic pressure from 0.2 to 5.1 mmHg [9]. Although not directly studied with simple recumbency in health, there are data in patients with
though she has not had any recurrence of symptoms in the past 8 months since pacemaker implantation.

In summary, we report an unusual case of changing BBB and syncope related to changes in body position. The exact explanation is unknown, but one may conjecture that it may be related to changes in right and left ventricular filling pressures/stretch, changes in sympathovagal balance, or mechanical effects of cardiac displacement within the chest secondary to changes in body position.

Conflict of Interest

The authors have no conflict of interest relevant to this publication.

References


6. Siegler D. Reversible electrocardiographic changes in severe acute asthma. Thorax. 1977;32(3):328-332. DOI: 10.1136/thx.32.3.328


Cite this article as: Idris S, Weitz H, Pavri BB. Syncope due to Positional Alternating Bundle Branch Block. Arrhythmia Grand Rounds 2015;1(1):11-15. DOI: http://dx.doi.org/10.12945/j.agr.2015.00014-14
Malignant Mimicry
An Unusual Cause of VT and AV Conduction Disease

Yehoshua C. Levine, MD¹, Rupal Parekh O’Quinn, MD¹, Paul A. VanderLaan, MD, PhD², Peter Zimetbaum, MD¹* ¹ Cardiovascular Division, Department of Medicine, Harvard Medical School, Boston, Massachusetts, USA ² Department of Pathology, Beth Israel Deaconess Medical Center, Harvard Medical School, Boston, Massachusetts, USA

The first two authors contributed equally to the manuscript.

Abstract
We present a case of a patient with unexplained asymmetric hypertrophy who presented with ventricular tachycardia and atrioventricular block and was diagnosed with lung disease metastatic to the myocardium.

Key Words
Ventricular tachycardia • Heart block • Imaging

Case Presentation
A 56-year-old female with no known cardiac history and a history of T3N0 NSCLC status post right upper lobectomy and chemoradiation therapy with cisplatin and etoposide in 2010 and a negative screening positron emission tomography (PET) scan 2 months prior presented in 2013 with the acute onset of chest discomfort. Baseline 12-lead ECG was normal (Figure 1 A). Upon presentation to an outside hospital, she had a wide complex tachycardia most consistent with left bundle branch-type VT with a right superior axis and negative concordance in the precordial leads at a cycle length of 250ms (Figure 1 B). Due to hemodynamic instability, she was cardioverted with no recurrence of VT thereafter. Transthoracic echocardiogram (TTE) revealed preserved biventricular systolic function with asymmetric left ventricular septal hypertrophy (Figure 2: PSLA TTE view), and a presumptive diagnosis of hypertrophic cardiomyopathy (HCM) was made. Of note, there was no known family history of SCD, VT, or HCM, and had not had previous ECGs or TTEs. She underwent single-chamber secondary prevention ICD implantation. Two months later, she presented with worsening dyspnea on exertion and was referred for stress testing, during which time she developed AV block with peak exertion (Figure 3: ETT ECG). She was therefore referred to our institution for both endomyocardial biopsy and ICD upgrade to a dual-chamber device, which she underwent without complication. Histopathologic evaluation of the right ventricular endomyocardial biopsy from the intraventricular septum revealed metastatic lung adenocarcinoma (Figure 4 A and B). Subsequent surveillance imaging revealed new lung nodules, and chemotherapy was initiated. To our knowledge, this is the first reported case of metastatic lung adenocarcinoma to the heart presenting with VT and/or AV block that masqueraded as a variant of HCM.

Discussion
Metastatic disease in the setting of breast and lung cancer, lymphoma, leukemia, melanoma, and various sarcomas affect the heart and/or pericardium in approximately 10% of cases [1]. Interestingly,
this incidence is higher now due to improvements in chemotherapy and radiotherapy, likely because of enhanced disease monitoring and longer life expectancy [2]. Adenocarcinoma of the lung is the most common malignancy resulting in metastasis to cardiac structures and is likely due to both the prevalence of the disease and the lungs’ proximity to the heart and pericardium [3].

The clinical profile of myocardial involvement by metastatic disease remains poorly defined and, like our case, may mimic more common pathology upon clinical presentation. Badri et al. recently described a similar patient with a history of lung cancer who presented with MMVT and was noted to have biopsy-proven squamous cell carcinoma of an unknown primary metastatic to the RV [4]. In our case, we felt that a presentation of sustained VT and asymmetric LVH in a middle-aged patient with no personal or family history of cardiac disease and no previous cardiac symptoms whatsoever warranted further evalu-
ation prior to presumptively diagnosing the patient with HCM. Because the patient’s ICD implantation preempted cardiac MRI testing, we felt that the next step would be endomyocardial biopsy, which ultimately revealed the diagnosis.

In patients with metastatic disease with cardiac involvement, palliative chemotherapy should be targeted at the underlying malignancy [3]. After her diagnosis was made, our patient was referred for chemotherapy (pemetrexed and carboplatin) and subsequent radiation. She developed a malignant pericardial effusion that necessitated surgical treatment with a pericardial window. The patient remains in remission 8 months after her initial diagnosis.

This case illustrates that unexplained asymmetric hypertrophy presenting with VT or AV block in a patient with no history of HCM should prompt consideration of infiltrative cardiomyopathies, including the rare possibility of metastatic disease in patients with a history of malignancy.

Conflict of Interest

The authors have no conflict of interest relevant to this publication.
References


Management of Sinus Node Dysfunction with Accelerated Junctional Rhythms

Paul A. Levine MD, FHRS, FACC, CCDS*
Loma Linda University Medical Center and Olive View UCLA Medical Center, Sylmar, California, USA

Abstract
This paper describes the unique use of an atrial overdrive pacing algorithm to manage a patient with symptomatic accelerated junctional rhythms as a component of sinus node dysfunction.

Key Words
Junctional Rhythm • Sick sinus syndrome • Atrial overdrive pacing • Pseudo-pseudo pacemaker syndrome • Pacemakers

Introduction
Pacing therapy is standard management for symptomatic sinus node dysfunction. Not infrequently, sinus node dysfunction is accompanied by either a junctional escape rhythm or less frequently, accelerated junctional rhythms. This case is that of a 56-year-old woman who had severe sinus node dysfunction manifested by persistent sinus rates less than 30 bpm manifesting symptoms of palpitations, weak spells, and exercise intolerance, but no syncope. A St. Jude Medical Identity™ (Sylmar, CA) dual chamber pacemaker was implanted without complication. At the first follow-up visit, the patient reported being improved but not as much as had been expected. Capture and sensing thresholds were stable as were stimulation impedances. There was marked chronotropic incompetence with a good response to rate modulation. After repeated adjustments in the dual chamber pacing mode including long AV delays resulting in functional single chamber atrial pacing and fine-tuning rate modulation, the patient was improved compared to her pre-implant status but continued to be limited by palpitations at rest and exertion. No pathologic atrial or ventricular tachyarrhythmias were recognized by either the pacing system or by a 48-hour Holter monitor despite symptoms occurring during the study.

After approximately 1 year of trying to address her symptoms with multiple pacing system adjustments, she was referred for consultation.
Questions

1. What rhythm could explain the continued symptoms?
2. If the symptoms are due to an abnormal rhythm, how might this pacemaker be programmed to manage this rhythm?

Discussion

In order to develop a therapeutic plan, one needs to have a proven or at least reasonable diagnosis (hypothesis). At a minimum, this patient had severe sinus node dysfunction. It had also been demonstrated that she had a junctional escape focus (Figure 1) and this protected her from profound asystolic episodes and syncope. If the problem was simply sinus node dysfunction, then functional single chamber atrial pacing combined with rate modulation should be sufficient to manage her symptoms. When there are a variety of potential therapeutic options, it is recommended that one approach these in a systematic manner addressing each potential option one at a time. If, on day 1, rate modulation, AF suppression, ventricular autocapture, AMS, and all other diagnostic and therapeutic algorithms were enabled yet the patient remained symptomatic, one would not know what was effective, what was ineffective and which algorithm(s) may have negated the benefit of another feature. This requires more time but over a series of repeated visits, the physician also acquires a better understanding of the patient to allow the physician to “read between the lines” gaining a better understanding of that patient enabling the pulse generator to be better programmed for that patient.

Hence, on day 1 (implant), standard DDD pacing should be utilized. When the patient returned, the event counter diagnostics would be retrieved to provide an overview of how the system behaved since the implant. In addition, the patient history is critical. The patient reported that she was better, she was clearly able to do more but her exercise intolerance was not as good as she had hoped and she continued to experience weak spells. Some of her symptoms may have been due to being physically deconditioned and this would take time however, chronotropic incompetence was confirmed with the heart rate histogram and event record showing predominant pacing at the base rate. The next step was enabling rate modulation. When she was next seen in follow up, her exercise tolerance had improved but she was still limited and she continued to experience weak spells both at rest as well as during walking (the extent of this patient’s usual physical activity).
Knowing that she had junctional escape rhythms, it was presumed that she also had accelerated junctional rhythms [1,2]. This is not uncommon in association with sinus node dysfunction. The junctional (AV nodal) pacemaker focus will respond to increases in sympathetic tone. This may occur with physical exertion as well as any form of stress such as emotional upset, excitement, laughter, etc. As such, an acceleration of the junctional rhythm may also occur at physical rest when rate modulation, even if set optimally, would not be a factor. Even though the QRS complex is normal with a junctional rhythm (presuming there is no bundle branch block), there will be the loss of atrial transport and even retrograde conduction.

The patient's symptoms are suggestive of pacemaker syndrome [3–5]. These same symptoms may occur in the absence of pacing in association with an intrinsic rhythm with the loss of AV synchrony as shown Figure 2 obtained from a different patient. With minimal change in rate, there was a loss of AV synchrony and a marked drop in systolic pressure, pulse pressure, and ejection time resulting in symptoms very similar to VVI pacing with 1:1 retrograde conduction. Since there was no pacemaker present, this is called pseudo-pacemaker syndrome. When a native rhythm such as either an accelerated junctional or idioventricular rhythm is inducing symptoms in a patient with a pacemaker, this is called pseudo-pseudo pacemaker syndrome.

Pseudo-pseudo pacemaker syndrome was my presumptive diagnosis in this patient with the etiology of the symptoms being an accelerated junctional rhythm resulting in a loss of atrial transport. A clue to this as the etiology is the event counts table, which is a component of the heart rate histogram in this model device. There was 97% AP- VS (AR) pacing, and less than 1% ventricular pacing. Most importantly, 2% PVEs were reported (Figure 3).

A PVE is a premature ventricular sensed event. While most of these are true PVCs consistent with the usual experience of the clinician, the definition of a PVC by the pacemaker is slightly different. Any sensed R wave that is not preceded by an atrial event, paced or sensed, is labeled a PVE (PVC). A true PVC that happens to coincidentally follow a native P wave or an atrial output pulse would not be labeled as a PVC. Each QRS associated with atrial undersensing but with intact AV nodal conduction would be labeled as a PVC as would accelerated junctional rhythms. In the case of intermittent atrial undersensing but with intact AV nodal conduction and/or accelerated junctional rhythms, these will usually occur in the normal rate range where as true PVCs tend to occur at a relatively short coupling interval and hence reflect, for that cycle, a relatively high rate. The event counts table (Figure 3) accompanying the event and heart rate histogram provides further insight to the rhythms in this patient.

![Figure 2](image1.png)  
**Figure 2.** A surface ECG is displayed with a simultaneously recorded arterial pressure tracing. This is recorded from a different patient. This individual had sinus rhythm with a left bundle branch block. The fourth complex from the left is a fusion beat, the fifth complex is a ventricular premature beat and there is then sustained ventricular tachycardia with 2:1 exit block out of the VT focus. With the VT, there is a loss of atrial transport without a major change in the effective ventricular rate but a dramatic fall in the systolic pressure, the pulse pressure and the left ventricular ejection time. In that this patient did not have a pacemaker, this would be labeled pseudopacemaker syndrome.

![Figure 3](image2.png)  
**Figure 3.** The event counts table from the event and heart rate histogram diagnostic in the SJM Identity pacemaker. These data represent the pacing state and rates of all four pacing modes (AS-VP or PV; AS- VS or PR; AP-VP or AV, and AP-VS or AR). The fifth column is labeled PVE and represents sensed R waves not preceded by atrial activity, either paced or sensed. The relatively large numbers occurring in the lower rate bins represent either atrial rhythms with intact AV nodal conduction but atrial undersensing or junctional rhythms. The PVEs in the higher rate bins usually represent premature ventricular complexes.
The area of interest is the relatively large number of “PVEs” in the lower rate bins is consistent with either intermittent atrial undersensing combined with intact AV nodal conduction or accelerated junctional rhythms. A PVE is a premature ventricular sensed event or an R wave that is not preceded by atrial activity, what other device manufacturers would label a PVC. Knowing that this patient had severe sinus node dysfunction and was demonstrated during the periodic evaluations to have junctional escape rhythms, the presumptive diagnosis was accelerated junctional rhythms both at rest associated with emotional stress or physical exertion. The junctional focus is influenced by sympathetic and parasympathetic tone.

Junctional rhythms could explain the weak spells at rest due to the loss of atrial transport. If the accelerated junctional rate exceeded the rate-modulated increase in rate during physical exertion, this could also explain the symptoms during exertion. Increasing the aggressiveness of the sensor would be able to manage the symptoms during exercise but not those occurring at rest. Enabling the AF suppression™ algorithm has been reported [6] as a way to manage accelerated junctional rhythms. However, if the concern is management of accelerated junctional rhythms, one must program the pacing mode to AAI(R). If left in the DDD mode, the junctional beat would be sensed on the ventricular channel thus inhibiting and resetting the pacemaker. To eliminate ventricular sensing, it is necessary to program the mode to AAI(R) as well as enabling the AF suppression algorithm.

The AF suppression algorithm [7] will result in an increase in the atrial paced rate when two atrial sensed events occur within a 16-cycle series. These do not have to be consecutive beats. In the usual patient, the trigger for an increased atrial-paced rate in association with this algorithm would be true P waves, even if arising from an ectopic focus. However, any sensed event on the atrial channel would be treated as a P wave by the pacemaker. Many junctional beats will be associated with retrograde conduction but if retrograde conduction does not occur, one then needs to rely on far field R wave sensing of the junctional beat. There was no retrograde conduction associated with the junctional beats and at the nominal atrial sensitivity of 0.5 mV; the far-field R wave was not detected. If the atrial sensitivity was programmed to 0.1 mV, far field R wave sensing was usually present (Figure 4).

Figure 4. This is a sensing threshold test for far-field R wave sensing with the pacemaker programmed to the AAIR mode and a base rate of 75 bpm (upper right on the printout). The top channel is the surface ECG, the second channel is the marker channel, the third is the atrial electrogram and the fourth is the ventricular electrogram. The first part of the rhythm strip prior to the notation “programmed” was recorded with the atrial sensitivity set to 0.1 mV demonstrated consistent sensing of the FFRW wave. With programming, the atrial sensitivity was reduced to 0.2 mV at which point far field R wave sensing was eliminated.
Case Study

Thus, although there is no retrograde conduction, one could take advantage of far-field R wave sensing associated with accelerated junctional rhythms to trigger the AF suppression increase in the atrial paced rate. This may be the only setting in which far-field R wave sensing is desired. For this approach to be successful, it is essential to program the pacemaker to the AAIR mode and this requires an assessment of the integrity of AV nodal conduction. If allowed to remain in the DDDR or DDIR mode, the junctional complex will first be sensed on the ventricular channel, initiate a post-ventricular atrial blanking (PVAB) period and preclude far field R wave sensing while resetting all the timing intervals.

In addition to the AF suppression algorithm being enabled, the base rate was increased to 70 bpm and the rest rate algorithm was enabled at 60 bpm. At the next follow-up visit, the event and heart rate histogram (Figure 5) demonstrated a heart rate distribution following a normal distribution curve that is virtually all atrial paced.

The combination of a higher base rate than the shipped value of 60 ppm, the rest rate algorithm, the AF suppression algorithm and rate modulation resulted in a near “normal” heart rate distribution with effective management of the patient’s symptoms. The key is the patient who reported (a) improved exercise tolerance and (b) resolution of her weak spells both at rest and physical exertion. She still had an occasional very brief palpitation which was believed to be due to the accelerated junctional beats with the transient loss of atrial transport but as the atrial paced rate promptly increased in response to the sensed “P” waves by the AF Suppression algorithm, these episodes were short-lived. Overall, the patient is dramatically improved and will continued to be followed in a routine manner monitoring symptoms, a screening physical exam based on any future symptoms and the event counter diagnostics that the pacemaker provides in a routine manner. It has now been 6 years since this patient was programmed to the AAIR mode with the AF suppression algorithm enabled and she continues to do well.

Conflict of Interest

The author reports a conflict of interest in that he owns publicly traded stock in both St. Jude Medical and Boston Scientific.

Figure 5. The event and heart rate histograms demonstrating that atrial pacing occurred more than 99% of the time and there was a normal distribution curve for heart rate distribution. This was a dramatic improvement from the baseline tracing.
References


Appendix: Special Notes

i. The Event Counts table, the prediction model and the event record, which were very helpful in evaluating and managing this patient, are only available in St. Jude Medical's legacy devices such as Identity and then require use of the SJM 3510 programmer to access and utilize.

ii. The use of the AF suppression algorithm to manage patients with accelerated junctional rhythms is an off-label use of this algorithm and is not specifically endorsed by St. Jude Medical. The author, however, has used it and atrial overdrive algorithms from other manufacturers effectively in similar patients to this one.
Method of Assessing the Safety of Functional Single Chamber Atrial Pacing Mode
The AV Nodal Conduction System Stress Test

Paul A. Levine MD, FHRs, FACC, CCDS*
Loma Linda University Medical Center and Olive View UCLA Medical Center

Abstract
It is important to assess the integrity of AV nodal conduction if one chooses to implant a single chamber AAI pacemaker, program a dual chamber pacemaker to the fixed AAI[R] mode or utilized one of the functional algorithms promoting AAI pacing while allowing for excessively long AP-VS intervals and even low grade AV block. The symptoms associated with low-grade AV block may be subtle. This case demonstrates a method of assessing AV conduction at the bedside at the time of routine follow-up evaluation.

Introduction
In recent years, it has been recognized that unnecessary ventricular pacing with a lead placed in the RV apex may be associated with adverse long-term hemodynamic consequences. Virtually all the manufacturers have introduced algorithms to facilitate functional single-chamber atrial pacing when AV nodal conduction is otherwise intact. There are two groups of algorithms. One is an AV hysteresis algorithm where the selected AV delay is extended by a programmable delta. On a periodic basis, if there is ventricular pacing following either an atrial-sensed or -paced event, the algorithm will extend the AV delay by the programmed delta. If within the extended paced and sensed AV delay, there is intrinsic conduction such that a native R wave occurs, the system retains the extended AV delay until such time as there is AV pacing at the extended interval. When this occurs, the AV delay shortens to the programmed value for a programmable length of time or number of cycles before repeating the search function. There are subtle variations between each of the manufacturers’
The second approach is to provide functional single chamber atrial pacing with back-up ventricular support should overt AV block develop to some degree. The first two such algorithms were Sorin-ELA’s AAIsafeR™ and Medtronic’s Managed Ventricular Pacing™ algorithms. Boston Scientific has recently introduced RhythmIQ™. However, these functional single chamber atrial pacing algorithms may require a relatively high degree of AV block before exiting the AAI pacing state to resume dual chamber pacing and multiple case reports have attested to the weakness of these algorithms in some patients. A small number of physicians also continue to use the true single chamber atrial pacing mode. In these two situations (functional or fixed single-chamber atrial pacing), it is prudent to assess the integrity of the AV conduction system.

**Clinical Case**

The patient, a 63-year-old male with recurrent weak and near-syncopal spells, undergoes implantation of a dual chamber pacing system, Medtronic (Minneapolis, MN) Advisa™ A2DR01 with a presumptive diagnosis of sinus node dysfunction. The implantation was uncomplicated. When seen in follow-up, the patient reports the persistence of the weak spells but the near-syncopal spells had resolved. The system was functioning in the AS-VS pacing state 45% and the AP-VS pacing state 54%. There was 1% AP-VP. The default mode for the Medtronic pacemakers is their proprietary Managed Ventricular Pacing or MVP™ mode [1,2]. In this mode, the system will pace the atrium but withhold the ventricular output even in the presence of low grade second degree AV block. If 2:1 or higher degrees of AV block develop for four consecutive cycles, the system will disengage the functional single chamber atrial pacing mode returning to DDD. After a period of time, the system will again withhold the ventricular output pulse in an effort to determine of the AV block has resolved or persists. The
baseline rhythm at the time of the follow-up evaluation is shown in Figure 1. The pacemaker pocket was well-healed, and the capture and sensing thresholds were excellent as were the lead impedances. There had been no atrial or ventricular high rate episodes according to the event counter diagnostics.

Questions

1. Can the continued symptoms be due to intrinsic rhythms not adequately managed by the pacing system?
2. How might this be further evaluated?

Discussion

The symptoms could be due to intermittent loss of capture, an inappropriate AV delay, intermittent AV block, or be totally independent of the pacing system and his rhythm. The excellent capture and sensing thresholds make intermittent loss of capture unlikely. Medtronic’s Managed Ventricular Pacing mode could allow for a marked first degree AV block [3-5] or intermittent higher grades of AV block [6-8], which could contribute to symptoms but the degree of AV block shown at rest would not be expected to cause a problem if this was the maximum that occurred. To determine whether higher grades of AV block might develop requires further assessment. This could be evaluated with an extended event or Holter monitor or a formal EP study but one would like to accomplish this evaluation while the patient is in the office.

While a Holter or event monitor can commonly be placed that same day, data collection takes a number of days and then the recording needs to be processed with the patient returning for a subsequent evaluation. An EP study to assess AH and HV intervals as well as pace the atrium at increasingly high rates is invasive, needs to be scheduled and is relatively expensive. The following describes a bedside AV nodal conduction system stress test that can be done at the time of the office evaluation and takes only an additional minute or two.

Since the presumption of the MVP mode or the intentional programming of a pacing system to the AAI mode is that AV nodal conduction is intact, it is imperative that the integrity of the AV conduction system be evaluated as even lesser degrees of AV block may be symptomatic [9,10]. This can be done by an “AV Nodal Conduction Stress Test.” Progressively increase the atrial paced rate until either a predetermined maximum rate is reached with intact AV conduction or overt second or higher grades of AV block develop. Even a normal AV conduction system may develop Wenckebach second-degree AV block during incremental atrial pacing at rates that would allow intrinsic conduction during physical exertion. Exercise is associated with increased intrinsic sympathetic activity with enhanced AV nodal conduction.
conduction (a positive dromotropic effect) where as this does not occur when using the pacing system to passively increase the atrial rate with the patient rest.

For this evaluation, the pacemaker is programmed to the AAI mode during the office evaluation starting at atrial paced rates of 100 bpm. The rate is then incrementally increased to progressively higher rates as shown in the following series of printouts (Figures 2, 3, and 4) from a patient with a St. Jude Medical Identity™ dual chamber pacemaker. When performed on this patient, atrial pacing at a rate of 125 bpm resulted in 2:1 AV block (Figure 5). The demonstration of second-degree AV block at a relatively low rate resulted in the disabling of the MVP mode in favor of DDD with Medtronic’s Search AV+™ algorithm. Although it has only been 2 weeks since this patient was seen,
the patient called to report a virtual total elimination of his repeated weak spells and he was ecstatic, suggesting that intermittent low grade AV block was the probable etiology of his continue, albeit minor, symptoms.

Based on the AV Nodal Conduction System Stress Test, the MVP mode was disabled in favor of the standard dual chamber (DDD) pacing mode in conjunction with Medtronic’s AV hysteresis algorithm to promote functional single chamber atrial pacing when AV conduction is intact at a normal or only minimally prolonged AV delay. With higher grades of AV block, dual-chamber (AS-VP or AP-VP) pacing will be restored presumably at a physiologically appropriate AV delay chosen by the clinician.

In patients with a dual-chamber pacemaker who are intentionally programmed to the AAI[R] mode or who are utilizing one of the functional AAI algorithms such as MVP, the AV nodal conduction stress test should be performed at each follow-up visit. Most of the current devices with these algorithms also incorporate an AV hysteresis algorithm such that should AV nodal conduction be demonstrated to be compromised, be it from the progression of disease or as a side-effect of concomitant pharmacologic therapy, the mode is programmed to DDD[R] in conjunction with that manufacturer’s AV hysteresis algorithm.

**Conflict of Interest**

The author has no conflict of interest relevant to this publication.

*Figure 5.* There are two ways to achieve high atrial-paced rates in the Medtronic dual-chamber pacemakers. One is to program the pacing mode to AAI and intentionally choose a high rate, but this requires fixed programming even though the device can then be programmed back to the desired settings. The second way is to utilize the capture threshold test and in the DDD mode, the highest allowed rate is 120 bpm. In the AAI mode which is temporary in that it will be disabled when the test is ended, the atrial paced rate can be programmed as high as 175 bpm. In this case, the selected rate was 125 bpm with the immediate development of 2:1 AV block.

---

**Comment on this Article or Ask a Question**
References


8. Vavasis C, Slotwiner DJ, Goldner BG, Cheung JW. Frequent recurrent polymorphic ventricular tachycardia during sleep due to managed ventricular pacing. PACE. 2010;33:641-642. DOI: 10.1111/j.1540-8159.2010.02655.x


Abstract
An ECG of a patient with 1:2 non-reentrant supraventricular tachycardia is presented and electrocardiographic analysis is discussed. One to two non-reentrant tachycardias may cause a tachycardia-mediated cardiomyopathy because the patient is almost always in a tachycardia. Like other incessant tachycardias, the rhythm and cardiomyopathy are cured with successful ablation.

Interpretation
Analysis of the ECG shows a narrow complex tachycardia in between short periods of sinus rhythm. Analysis of the tracing showed the presence of sinus P waves (arrows) with 2 QRS’s for each P wave except for the last 2 sinus P waves (Figure 2).

The PR interval shows subtle, but progressive prolongation from baseline of 200 ms to 280 ms, prior to block of the first QRS. The PR to the next QRS is mark-

Key Words
Non-reentrant tachycardia • Concealed conduction • Atrio-ventricular nodal reentry • One-to-two tachycardia

An irregular narrow complex tachycardia: what is your diagnosis?

A 35 year-old woman presented with a history of palpitations at rest and exercise for 6 months, becoming incessant over the last 3 months. This was associated with progressive dyspnea on exertion and mild edema. An echocardiogram showed biventricular dilatation with a left ventricular ejection fraction (LVEF) of 37%. A Holter monitor showed a nearly incessant tachycardia, due to recurrent runs of tachycardia, interrupted by a few seconds of sinus rhythm. Her ECG is shown in Figure 1a and Figure 1b.
Josephson, M.E.

A n Irregular Narrow Complex Tachycardia

Electrophysiology study

She was taken to the electrophysiology laboratory where dual atrio-ventricular (AV) nodal pathways with double responses, producing the non-reentrant 1:2 tachycardia shown in figure 1 was demonstrated. Following isuprel and atropine administration, a brief run of slow AV nodal reentrant tachycardia (AVNRT) was inducible, which had not been seen clinically. She underwent radiofrequency ablation of the slow pathway, which prevented recurrent symptoms. Within 6 weeks her ejection fraction normalized.

Figure 1a. 12-lead ECG of a typical run of tachycardia

Figure 1b. Rhythm strip of a typical run of tachycardia

Figure 2. P-waves and PR intervals are denoted

edly prolonged to 610 msec. The best explanation for the 1 to 2 phenomenon is the presence of dual AV nodal pathways with conduction down both the fast (f) and slow (s) pathway. This gives rise to a non-reentrant tachycardia that is double the sinus rate. Conduction down each pathway retrogradely invades the other pathway. This gives rise to subtle prolongation of PR interval to the first QRS until block in the fast pathway occurs and conduction only proceeds over the slow pathway (PR 610 ms). The tachycardia stops when block in the slow pathway occurs and the cycle repeats itself after a few seconds. A schema of the mechanism is shown in Figure 3.
Conclusion

One to two non-reentrant tachycardias are a cause of tachycardia-mediated cardiomyopathy because the patient is almost always in a tachycardia. Like other incessant tachycardias the rhythm and cardiomyopathy are cured with successful ablation.

Conflict of Interest

The authors have no conflict of interest relevant to this publication.

Cite this article as: Josephson ME. An Irregular Narrow Complex Tachycardia: What Is Your Diagnosis? Arrhythmia Grand Rounds 2015;1(1): 32-34 DOI: http://dx.doi.org/10.12945/j.agr.2015.00020-14
Abstract

Presence of manifest pre-excitation suggestive of a right-sided accessory pathway (AP) is not proof that a patient’s symptoms are caused by it. Before ablation is undertaken, tachycardia should be induced and participation of the AP in mediating the tachycardia should be demonstrated.

Right-sided pathways may have Kent-like conduction properties or Mahaim-like conduction properties. A Mahaim-like, decrementally conducting AP during a pre-excited tachycardia can be suspected by measuring the atrioventricular (A-V) interval at the His bundle recording during tachycardia. Proof of AP participation in the tachycardia circuit can be obtained by advancing ventricular activation after a critically timed atrial premature beat (APB) delivered during AV node or His bundle refractoriness. These APBs can be delivered in the right atrial lateral wall and may result in one of 4 types of responses in patients with a decrementally conducting AP. Furthermore, reset of the tachycardia may either result in advancement or delay of ventricular activation. This case illustrates and discusses these principles.

Conflict of Interest

The author has no conflict of interest relevant to this publication.

Comment on this Article or Ask a Question
Basic Foundations of Clinical Electrophysiology

Andrew L. Wit, Ph.D.*
Department of Pharmacology, Columbia University College of Physicians and Surgeons, New York, New York, USA

Key Words
Electrophysiology-Basic • Electrophysiology-Clinical

This section will be a continuing feature of the journal. As the title indicates, it will be composed of a series of articles designed to describe and teach basic electrophysiological concepts underlying clinical electrophysiology. In each issue, the column will elucidate the underlying electrophysiology and how it is translated into the electrophysiological characteristics of one of the case reports in a “user friendly” format. For example, mechanisms of arrhythmias and reasons for response to interventions, mechanisms for conduction abnormalities, mechanisms of effects of drugs, effects of electrical stimulation, and other interventions will be explored as they relate to the clinical case.

Translations from basic to clinical electrophysiology are usually based on properties described in vitro in the basic science lab, which are mimicked in vivo. To take one example, the response of isolated, superfused cardiac cells and tissues to electrical stimulation provides the foundation for understanding the effects of stimulation on clinical arrhythmias. Why are some arrhythmias initiated and terminated and others not? What is stimulation doing to the mechanism causing the arrhythmia, to the electrophysiological properties of the cells causing the arrhythmia? How does cellular electrophysiology enable different mechanisms of arrhythmias to be distinguished one from another by analyzing the effects of electrical stimulation?

Our philosophy is that clinical electrophysiologists need to have a solid foundation of basic electrophysiological knowledge to enable them to understand the causes and response to therapeutic interventions of cardiac arrhythmias. Not only will it help in the treatment of current patients, but it is an absolutely essential requirement for the continued development of this field. It was only because the founding fathers had this knowledge that the discipline has advanced to this stage. Without their intense interests in basic electrophysiology and its application to clinical arrhythmias, the clinical electrophysiology that we know today would not exist. They learned their basic electrophysiology by doing time in an experimental research laboratory, poking microelectrodes into cardiac cells to record...
transmembrane potentials, and designing and studying the electrophysiology of arrhythmia models in hearts of mice, rabbits, cats, dogs, etc. From these experiences, the impetus arose to attempt to record the His bundle electrogram in man after it had been recorded in canine hearts by Brian Hoffman in the 1960s, or to use programmed stimulation to evaluate AV junctional conduction in canine hearts and the mechanism of reentrant AV nodal tachycardia using dual AV nodal pathways in rabbit hearts from the experimental studies of Gordon Moe, around the same time. In parallel with these developments, cellular causes of impulse initiation such as automaticity and triggered activity and their properties were being studied with microelectrodes and translated to the clinical laboratory. The actuality of reentry as a cause of arrhythmias was debated well into the 1960s before laboratory experiments demonstrated its occurrence and proved its importance. The phenomenon of delayed after depolarization induced triggered activity was not translated into triggered clinical arrhythmias until the 1970s and 1980s. More recent laboratory studies have implicated early after depolarizations in the genesis of torsades de pointes associated with genetic and acquired long QT syndromes.

There are too many unsolved problems remaining for us not to take on the responsibility of ensuring that clinical cardiac electrophysiology keeps progressing and progress depends on a thorough understanding of underlying electrophysiological mechanisms. Despite all that has been learned over the past century, we should not become complacent that there is little more to know. I remember having a conversation with a well-known clinical electrophysiologist in the early 1970’s who indicated to me that now His bundle electrograms had been recorded under every conceivable circumstance of conduction disturbances and arrhythmias, there was not much more to do. Subsequent history speaks for itself. Modern therapies such as ablation of AV nodal reentrant tachycardia and pulmonary vein ablation were never even dreamed of at that time and would have seemed like science fiction if someone had proposed that these would become commonly used procedures. Improvements in the approach to catheter ablation of ventricular tachycardia through location of the isthmus region in a reentrant circuit, would have seemed far-fetched to the surgeons trying to prevent arrhythmias by aneurysctomy. Programmed electrical stimulation of the ventricles was strongly condemned by some until the mid to late 1970s. All of these procedures were developed through a thorough understanding of underlying electrophysiological mechanisms. That is the purpose of this column.

A more immediate practicality will be to review basic concepts that appear on the board exams in clinical electrophysiology. This basic electrophysiology that is pertinent to the clinical field is not easily accessible. It cannot be found in one place but must be searched out in numerous review articles and edited book chapters, often written for the basic or clinical research scientist and not for the clinician whose goal is to learn the fundamentals. When time is so valuable, most clinical electrophysiologists do not have the opportunity to search out all this necessary background information that had not been taught in any of the training environments that they have experienced. In this column we will present in a very understandable manner, these important concepts so that a collection of these articles will provide a comprehensive review of the basic electrophysiology necessary for understanding the underlying mechanisms of clinical electrophysiological abnormalities.
Bedside to Bench – A Difficult Journey
An Irregular Wide QRS Tachycardia

Negar Salehi, MD, Aravdeep S. Jhand, MD, Vaibhav Satija, MD, Watchara Lohawijarn, MD, Ranjan K. Thakur, MD, MPH*
Sparrow Thoracic and Cardiovascular Institute, Michigan State University, Lansing, MI, USA

Abstract
A 40 year old man presented with palpitations and was found to have a sustained, monomorphic, irregular wide QRS tachycardia. He had a history of sarcoidosis, based on muscle biopsy. Cardiac MRI, PET scan and electrophysiology study were normal. Potential mechanisms of this tachycardia are discussed.

Key Words
Sarcoidosis • Ventricular tachycardia • Tachycardia mechanism

Introduction
Mechanistic understanding of most clinical arrhythmias are extrapolated based on rigorous bench research and/or established clinical studies. For many arrhythmias, the mechanisms don't change from case to case. For example, atrioventricular reentry, atrioventricular nodal reentry and common atrial flutter are all due to reentry and that fact is invariant. However, for many clinical arrhythmias, the mechanism is not so clear and all the clinical data may not be sufficient to go from “the bedside to the bench” in ascertaining whether the arrhythmia in question is due to reentry, enhanced automaticity or triggered activity. In these cases, the journey from the bedside to the bench may be a difficult, as highlighted by this case.

Case
A 40-year-old Caucasian man with muscle biopsy-proven sarcoidosis presented to the emergency department with new onset palpitations. He denied having had chest pain, dyspnea, lightheadedness or syncope. Past medical history was negative for similar episodes or any cardiovascular history, except hypertriglyceridemia. He had had a skin rash 2 years previously and sarcoidosis was suspected; a muscle biopsy was confirmatory. Physical examination was normal. The initial 12 lead Electrocardiogram (ECG) obtained in the emergency department showed a sustained, monomorphic, irregular wide QRS complex tachycardia (Figure 1).

Subsequent ECG showed normal sinus rhythm (NSR). Ventricular ectopic beats had the same morphology in lead II and V₁-V₆ as the wide QRS tachycardia (Figure 2).

The ECG
The irregular wide QRS tachycardia was thought to be ventricular tachycardia (VT). Atrial fibrillation (AF) was ruled out by 1) the fact that retrograde atrial activity (V-A conduction) was present during tachycardia (Figure 3 - arrows) and 2) wide QRS complexes due to RBBB aberrancy was ruled out because of its atypical nature (positive concordance). Positive concordance in a wide QRS tachycardia may be seen in...
Figure 1. First 12 Leads ECG. Sharp spikes seen in some leads were due to printer artifact; the patient did not have a pacemaker.

Figure 2. Second 12 lead ECG.
the case of a preexcited tachycardia using a posteriorly located accessory pathway (AP) or VT originating in the postero-basal left ventricle. Preexcited AF was ruled out by excluding AF and the absence of slurring of the initial part of the R-waves in V1-V6. An antidromic reentrant tachycardia was ruled out by the irregularity of the tachycardia.

Initial labs including cardiac biomarkers, thyroid function tests and electrolytes were normal. While the patient was being monitored, he had long episodes of palpitations. Rhythm strips showed the same phenomena as the 12-lead ECGs (Figure 3).

A 2-D echocardiogram was normal. Cardiac magnetic resonance imaging (MRI) showed mildly dilated right ventricle (RV) with RVEF of 45% and LV diameter at the upper limit of normal with normal EF, without late Gadolinium enhancement. A detailed electrophysiology study was normal; ventricular tachycardia could not be induced with vigorous programmed stimulation, even with isoproterenol infusion. A fluorodeoxyglucose positron emission tomography (FDG-PET) scan was normal.

Discussion

Sarcoidosis is a systemic granulomatous disease that can involve various organs such as the lung, skin, eyes, liver and the heart. Sarcoidosis is more common in women and African Americans with incidence of 10.9 per 100,000 in whites and 35.5 per 100,000 in African-Americans [1]. The heart can be involved in 25% of patients with sarcoidosis [2]. Cardiac involvement may present with heart block, arrhythmias or heart failure. Ventricular tachycardia is a common arrhythmia in patients with cardiac sarcoidosis and the overall prognosis of sarcoidosis is worse in patients with cardiac involvement [3].

Patients with established extra-cardiac sarcoidosis need comprehensive evaluation to look for any features suggestive of cardiac sarcoidosis (CS). This includes screening tests such as a 12-lead ECG and an echocardiogram [4]. Any abnormalities in the primary tests warrant further investigation with more specific tests like cardiac MRI or FDG-PET scanning. Delayed Gadolinium enhancement in the left or right ventricle might be seen in patients with CS. A FDG-PET scan showing patchy uptake in the myocardium may suggest cardiac involvement [3].

The mechanism of VT in CS is most commonly due to reentry involving a granulomatous scar, although automaticity and triggered activity may play a role as well. Immunosuppression may aid antiarrhythmic therapy and catheter ablation may be an option for refractory cases [5]. As per the guidelines, FDG-PET is necessary to look for myocardial inflammation. Patchy uptake on FDG-PET is suggestive of inflammation and using immunosuppression may be helpful in the treatment of VT [6].

Monomorphic Ventricular tachycardia has been reported as an initial presentation for newly diagnosed CS. Some prospective studies have reported the incidence of CS in patients with unexplained VT to be...
around 25-28%, suggesting that CS should be considered in the differential diagnosis of unexplained, idiopathic VT [7]. In certain cases, unexplained VT was the first sign of an underlying multisystem involvement in sarcoidosis [8].

**Commentary**

This patient presented with a markedly irregular, sustained, monomorphic ventricular tachycardia. What is the mechanism of this tachycardia? This is where we run into difficulty because VT was not inducible during electrophysiology study. So, the arrhythmia mechanism cannot be ascertained based on electrophysiologic maneuvers and can only be inferred based on the surface ECG. While monomorphic, reentrant arrhythmias may be slightly irregular, they don't show the degree of irregularity seen in this case. However, reentrant arrhythmias are more likely to be monomorphic, but this is not exclusive. Recently, Panda, et.al., compared VT morphologies between CS and idiopathic VT and found that pleomorphism during VT was more common in CS than in idiopathic VT [9]. The group from UCLA has proposed a new entity, arrhythmogenic inflammatory cardiomyopathy in patients with occult sarcoidosis [10]. They found evidence of myocardial inflammation (positive FDG-PET scan) in patients with ventricular arrhythmias, suggesting cardiac sarcoidosis as the underlying cause. So, in the final analysis, the journey from the "bedside to the bench," i.e., from the clinical arrhythmia to the specific mechanism that causes it, cannot be made in this case, nor is it possible in every case. To confuse matters more, since the MRI and FDG-PET studies were relatively normal, cardiac involvement with sarcoidosis may be conjectural as well in this case.

**Conflict of Interest**

The authors have no conflict of interest relevant to this publication.
Profiles in Cardiac Electrophysiology

Albert L. Waldo MD, PhD (Hon)
Department of Medicine, Case Western Reserve University; Harrington Heart & Vascular Institute, University Hospitals Case Medical Center, Division of Cardiovascular Medicine; Cleveland, Ohio, USA

Key Words
Advances in cardiac electrophysiology • Biography • Leaders

In our lifetimes, we have witnessed dramatic developments and enormous growth in cardiac EP powered by giants working in institutions that have trained generations of leaders, by scientific discoveries and even by chance events. Fueling this growth have been many things. And interestingly, those behind some of the most groundbreaking events may not have completely grasped the profundity of their ideas at that moment. Nevertheless, these changes almost surely occurred in a community of colleagues who contributed in their own way, and helped provide a platform in which new thinking emerged.

In “Profiles in EP” we plan to invite colleagues to share their stories of how their ideas, discoveries, and developments came to be or to share how their institution came to figure so prominently in developing EP. It is the intent of this column to grasp the essence of discovery from leaders in our profession, and inspire the reader. We also plan to profile the many laboratories and people who have contributed to the advancement of our field.

EP did not happen by accident. How did new ideas and concepts develop? What was the setting? What was the motivation? What was the support? Was it planned, or simply an accident? For those who have advanced the field, upon whose work did they build, and to whom do they owe their success? Do they even consider their work to be a success? What inspired them?

We hope to capture how these Eureka! moments actually happened, and even provide stimulus for continued exploration. Numerous developments have changed our understanding of so many things from basic concepts to nuances of catheter ablation and device development. Nowhere in medicine has innovation and growth been this spectacular, and for so long a period, even preceding our time. The remarkable discoveries date back over a century, and have placed cardiac electrophysiology in a unique place in medicine and science.

For many of us, the scientific and technological developments, and the kind of creative minds behind these discoveries, are part of the reason we have chosen this field. Younger colleagues may wonder how these insights came to be. Is the field now mature and staid, or can a way still be found to contribute to its escalating growth, and if so, how?

The truth of the matter is that we are only at the beginning of a tremendous journey that requires bright minds to explore hitherto unknown possibilities in the field that one day may become reality. We are enriched by having talented friends and colleagues to usher us through this continuing adventure in which a special part is simply getting up in the morning to find new tools to better treat our patients.

This column will focus on the “magic” that leads...
to discovery, and on how we can continue with new stories and new visions. As Marcel Proust said: “The real voyage of discovery consists not in seeing new landscapes but in having new eyes.” We hope this column will provide an important part of the history of our field, and even be an inspiration for its further growth.

Cite this article as: Waldo AL. Profiles in Electrophysiology. Arrhythmia Grand Rounds 2015;1(1): 42-43. DOI: http://dx.doi.org/10.12945/j.agr.2015.00023-14
Reflections and Reminiscing on the First Catheter Ablation of the AV Junction in Man

Melvin Scheinman, MD*
University of California San Francisco, School of Medicine, University of California, San Francisco, California, USA

Key Words
Catheter ablation • AV junction • DC ablation

Introduction

I am delighted to accept the invitation for me to reminisce over my experience relative to the first ablation in a human. It has been a source of great pride and fulfillment to watch the amazing growth and development of this methodology to where ablative procedures are now used to control or cure virtually all cardiac arrhythmias. I have previously described my experience in a scientific communication [1]. I am happy to elaborate in a less structured format.

In the 1970s, patients with supraventricular tachycardia (predominately atrial fibrillation) with drug refractory rapid ventricular response underwent cardiotomy with direct cryo or ligature ablation of the AV junction. I conceived of the notion of attempting to achieve the same endpoint but with use of a catheter technique.

In researching the pertinent literature I found that a Mr. Beazell had described preliminary reports of a catheter technique that used high voltage direct current (DC) shocks to the region of the AV junction localized by fluoroscopy was successful in producing AV block in canines. In addition, another fortuitous event was the acceptance of the Dr. Rolando Gonzalez to our cardiac fellowship program. In 1979, Dr. Gonzalez and I visited Mr. Beazell (who worked for Telectronics) at Harbor General Hospital in Los Angeles and observed his techniques.

Our experimental work: 1979

We tried the same technique, starting after our visit to Mr. Beazell’s lab, but had difficulty replicating his results, no doubt due to our own faulty knowledge of the fluoroscopic landmarks in dogs. In any event, we decided to use an electrode catheter in order to record the His bundle potential so as to clearly localize the AV junction. We learned that application of shocks were more successful where a large Hus bundle potential was inscribed along with a large atrial signal.

Dr. Gonzalez performed all the animal studies including application of the DC shock and production of AV block. These studies included right heart catheterization as well as exercise studies [2]. I am very much indebted to his dedication and hard work. The animal hearts were then delivered to Drs. Lev and Bharati [3] who performed careful histologic studies and reassured us that the technique did not result in perforation of the cardiac chambers or heart valves and that the coronary circulation was not affected. We also learned early on from experiments delivering high energy shocks into a saline beaker that not all catheters could withstand the high energy (100-200 Joules) delivered. Of note during these experiments, Mr. Rubenstein, an electrical engineer working at San...
Francisco General Hospital, suggested use of radiofrequency (RF) energy and we did use the RF unit in the operating theatre but unfortunately abandoned this approach after we caused ventricular fibrillation in several dogs. This occurred, no doubt since we were totally ignorant in the proper usage of RF energy. It was not until several years later when Dr. Jonathan Langberg joined our program and taught about the proper electrode size and RF setting that produced safe and effective lesions in canines [4].

After several years of research to allow for proper use of the appropriate catheter and assurance of both efficacy and safety we proceeded with application to our IRB as well as to the FDA for use of this method in humans.

The first patient: 1981

We are sincerely indebted to the determination and courage of Mr. Paul Anderson, a retired oil refinery worker who volunteered to be the first patient in the world to receive this treatment. Mr. Anderson suffered from heart failure and with atrial fibrillation would develop pulmonary edema. He also had severe rheumatoid arthritis and when presented to our thoracic surgical colleague (Dr. Arthur Thomas), he was deemed too high a risk for thoracotomy and direct surgical ablation. The experimental nature of the new procedure was discussed with the patient and his wife who were very enthusiastic about participating.

I felt that due to my attachment to the new technique, it was not appropriate for me to obtain informed consent. The task was left to one of my colleagues on the cardiac EP service, Dr. David Hess who was very knowledgeable about both the patient’s clinical problem as well as our experimental work. I remember Dr. Hess calling me the evening before the scheduled procedure asking that I consider doing additional animal work. I refused because we had spent a number of years with canines and I felt there was really little more to learn. In any event, when I approached the patient the morning of the procedure, he was eager to get on with the ablation.

I remember a host of individuals in the laboratory that day including Drs. Morady, Hess, Ruey Sung and several cardiologists and EP fellows. We should give due credit to Mr. Booker Pullin who brilliantly fabricated all the connectors from catheter to the defibrillator.

Finally, after placement of the catheter in the proper position adjacent to the His bundle signal, the D/C shock was delivered accompanied by a flash and loud sound. The patient immediately went into complete AV block without any untoward effects. A temporary pacemaker had been placed and when AV block ensued, a permanent pacemaker was inserted.

Of interest, AV conduction resumed and a repeat procedure was required along with drug therapy. He subsequently died of congestive heart failure in 1986. He was a wonderful and courageous individual and I will never forget his admonition to “get on with this” on the morning of the procedure. The report was published in 1982 [5]. This was followed by successful ablation of a posteroseptal accessory pathway in 1984 and published the following year [6].

It has been nothing short of incredible for me to watch the remarkable evolution of the catheter ablation technique to all manners of complex arrhythmias. The most remarkable part of this as a clinician is to tell a patient that they are actually cured of their arrhythmia problem and can lead a normal life.

Conflict of Interest

The authors have no conflict of interest relevant to this publication.
References


Cite this article as: Scheinman M. Reflections and reminiscing on the first catheter ablation of the AV junction in man. Arrhythmia Grand Rounds 2015;1(1): 44-46. DOI: http://dx.doi.org/10.12945/j.agr.2015.00022-14