Assimilating the increasing body of knowledge required for the treatment of patients with cardiac arrhythmias is a substantial task. The trainee is bombarded with chapters and reviews that are important for providing the frame work from which to develop a detailed and nuanced knowledge acquired from clinical experience and ongoing education. Images and Case Reports provide succinct observations of not only a novel or unusual finding, but often of the fundamentals that must be appreciated to place novel findings in context. The Teaching Points series in Circulation Arrhythmia and Electrophysiology provides a more in-depth synthesis of case-based learning building on common and uncommon clinical findings. The purpose of this Case-based Curriculum in Clinical Electrophysiology is to provide the clinician and trainee with an easily accessible body of current reviews, cases and Teaching Points that cover a broad range of the field. Links to the individual articles are provided to facilitate using this article as a spring board for review.

Fundamentals of Electrophysiology, Mapping, Ablation, and Electrocardiography

Emergence of complex behavior: An interactive model of cardiac excitation provides a powerful tool for understanding electric propagation. Spector PS, et al.1

To facilitate an understanding of the relations between electrophysiologic properties and geometry that govern conduction, block and reentry, Spector and colleagues have developed an interactive model that allows the student to study the effects of varying parameters on conduction, block, reentry and even entrainment (http://circep.aha-journals.org/cgi/content/full/CIRCEP.110.961524/DC1).1

Arrhythmogenic implications of fibroblast-myocyte interactions. Rohr S.2

Myocardial fibrosis has major roles in creating the substrate for atrial and ventricular arrhythmias and is reviewed by Rohr.2

Teaching points with 3-dimensional mapping of cardiac arrhythmias: Taking points: Activation mapping. Del Carpio Munoz F, et al.3

Teaching points with 3-dimensional mapping of cardiac arrhythmias: Teaching point 3: When early is not early. Del Carpio Munoz F, et al.4

Teaching points with 3-dimensional mapping of cardiac arrhythmia: Mechanism of arrhythmia and accounting for the cycle length. Del Carpio Munoz F, et al.5

Accurate activation sequence mapping of cardiac arrhythmias is a fundamental skill in cardiac electrophysiology. For complex arrhythmias an electroanatomic mapping system, which allows the mapping data to be displayed on a representation of the anatomy is often employed, as discussed in a series of articles Del Carpio Munoz and colleagues.3–6 Accurate mapping requires selecting an appropriate reference point and mapping window and identification of the electrogram that indicates local activation. In applying a mapping strategy it is important to recognize features of macroreentrant, as compared to focal arrhythmias, and to recognize that seemingly small errors in assigning activation times, reference electrograms, or mapping windows, can render a map uninterpretable or meaningless. These issues are discussed in detail.

Teaching points with 3-dimensional mapping of cardiac arrhythmia: How to overcome potential pitfalls during substrate mapping? Del Carpio Munoz F, et al.6

Endocardial unipolar voltage mapping to detect epicardial ventricular tachycardia substrate in patients with nonischemic left ventricular cardiomyopathy. Hutchinson MD, et al.6

Substrate mapping is a term that refers to localization of the likely arrhythmia substrate, during sinus or paced rhythm rather than during VT, and is often based on concept that areas with low amplitude electrograms are often regions of scar, that contain reentry circuits for scar-related arrhythmias. Del Carpio Munoz and colleagues review substrate mapping and potential pitfalls.6 Hutchinson and colleagues describe the use of unipolar voltage maps, exploiting the “wider field of view” of unipolar as opposed to bipolar recordings, to detect areas of epicardial scar that can overly relatively normal appearing endocardial regions in nonischemic cardiomyopathy.7

Signals and signal processing for the electrophysiologist: Part i: Electrogram acquisition. Venkatachalam KL, et al.8

Signals and signal processing for the electrophysiologist: Part ii: Signal processing and artifact. Venkatachalam KL, et al.9

The pathophyslogic basis of fractionated and complex electrograms and the impact of recording techniques on their detection and interpretation. de Bakker JM et al.10

Recording and interpreting electrograms is fundamental to electrophysiologic studies and mapping. An understanding of how recording techniques influence electrograms, including discussion of artifacts is provided by Venkatachalam and coworkers.8,9 Multicomponent, fractionated electrograms are markers of abnormal conduction and often considered for ablation targets for both ventricular and atrial arrhythmias. De Bakker and Wittkampf discuss the genesis of these signals, how they are influenced by recording techniques, and potential pitfalls related to their use for guiding ablation.10

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All articles referenced have been made free to the reader and accessible from links in the reference list.

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Catheter cryoablation: Biology and clinical uses. Andrade JG, et al.11

Andrade et al review the mechanism of tissue injury, advantages, and limitations of cryoablation.


Signal processing tools are applied not only to electrograms recorded in the electrophysiology laboratory, but also to surface electrocardiograms recorded during ambulatory monitoring and exercise testing. Recognition of the potential for artifacts is important with all recording technologies. Mendenhall et al show how a signal averaging algorithm applied during exercise testing produced artifacts mimicking AV block and Brugada syndrome.13

Atrial Fibrillation and Flutter

Left atrial anatomy revisited. Ho SY, et al.15

Mapping and ablation in the left atrium requires a detailed understanding of left atrial anatomy, which is provided by Ho and colleagues in this review, which also includes discussion of adjacent structures at risk for ablation injury, including the esophagus and phrenic nerves.13

Simultaneous, but dissociated left atrial fibrillation (AF) and pulmonary vein tachycardia: A case of occult pulmonary vein isolation. Miller MA, et al.

Pulmonary vein isolation is a cornerstone of catheter ablation for AF. The correct interpretation of pulmonary vein electrograms and confirming isolation of the vein are important for procedural success. Miller et al show that atrial tachycardia from within a pulmonary vein can make it difficult to recognize isolation when ablation is performed during AF.14

How to approach reentrant atrial tachycardia after atrial fibrillation ablation. Miyazaki S et al.13

Multiple atrial tachycardias after atrial fibrillation ablation: The importance of careful mapping and observation. Miyazaki S et al.16

Restoration of sinus rhythm by incarceration, not elimination, of focal atrial tachycardia in left atrial substrate post atrial fibrillation ablation. Shah AJ et al.17

Interatrial electrical dissociation after catheter-based ablation for atrial fibrillation and flutter. Gautam S, John RM.15

Organized atrial tachycardias are common after catheter or surgical ablation procedures for atrial fibrillation. Their origin can be focal or macroreentrant, and identification and ablation can be difficult. In their teaching rounds paper Miyazaki et al show a case of macroreentrant tachycardia and demonstrate a systematic approach to defining the circuit and site for ablation using a combination of activation and entrainment mapping.15 In a case report Miyazaki et al show that multiple tachycardias and tachycardia mechanisms can exist in a single patient and again demonstrate the use of careful activation and entrainment mapping to sequentially ablate these arrhythmias.16 Shah et al show a case of a focal reentrant tachycardia after prior extensive left atrial ablation, such that ablation of the tachycardia isolated the left atrial appendage.17 Gautam and John show a case in which extensive ablation for AF electrically separated the left and right atria.18

Entrainment with long postspacing intervals from within the flutter circuit: what is the mechanism? Wong KC, et al.16

This case demonstrates how decremental conduction properties in an atrial reentry circuit can increase the post-pacing interval (PPI) following entrainment, producing a misleadingly long “false negative” PPIs at sites in the reentry circuit. Other causes of misleading PPIs are also discussed.

Epicardial-only block during endocardial mitral isthmus ablation facilitated by coronary sinus occlusion. Shah AJ, et al.20

Recurrent perimital tachycardia using epicardial coronary sinus connection to bypass endocardial conduction block at the mitral isthmus. Miyazaki S et al.21

Ablation lines between the mitral annulus and left pulmonary veins are commonly placed to interrupt perimital atrial flutter, but achieving block and confirming block is often difficult. The cases presented in these articles illustrate methods for assessing conduction block and show pitfalls that can occur and how to recognize them.20 Shah and Miyazaki and their coworkers show cases in which the separation of epicardial musculature around the coronary sinus from the left atrial musculature creates the possibility for conduction block in only the epicardial or endocardial component of the “mitral isthmus,” falsely suggesting complete block.20,21

Catheter ablation of atrial fibrillation in transposition of the great arteries treated with musttard atrial baffle. Frankel DS, et al.22

As the number of adults with repaired congenital heart disease increases, AF will be increasing encountered in this population of patients who have complex atrial anatomy. Frankel et al show that catheter ablation for AF is feasible for some of these patients and illustrates the anatomic challenges that can be encountered.22

Congenital sick sinus syndrome with atrial inexcitability and coronary sinus flutter. Varma N, et al.23

Sinus node dysfunction, atrial flutter and fibrillation are rare in children in the absence of structural heart disease. Varma et al present a teaching rounds case of congenital atrial disease with sinus node dysfunction, areas of atrial tachycardia and atrial electrical inexcitability, consistent with extensive fibrosis illustrating the coexistence of multiple brady and tachyarrhythmias in this rare congenital atrial disease.23

Cavotricuspid isthmus ablation guided by real-time magnetic resonance imaging. Piorkowski C, et al.24

Achieving conduction block in the common cavotricuspid isthmus is difficult in some patients due to anatomic constraints, as can also be encountered in other areas of the heart. Piorkowski et al show the first human case of catheter ablation guided by real time magnetic resonance imaging (MRI), which has the promise of facilitating ablation through better understanding of anatomic – catheter relationships during ablation, as well as future potential assessment of tissue injury.24

Remaining ice cap on second-generation cryoballoon after deflation. Bordignon S, et al.25

Cryoballoon ablation is effective for pulmonary vein isolation. Bordignon show a case of ice formation on the balloon, observable with angiography illustrating the relevant anatomy and discuss the potential risk.25

Acute liver failure associated with dronedarone. Joghetaei N, et al.26

Consideration and recognition of toxicities is critically important in selecting therapies for AF. Although rare, dronedarone can cause severe liver toxicity, and monitoring is warranted.26

Pacemakers and Defibrillators

Minimizing inappropriate or “unnecessary” implantable cardioverter-defibrillator shocks: Appropriate programming. Konera JN, et al.27

Posttraumatic stress and the implantable cardioverter-defibrillator patient: What the electrophysiologist needs to know. Sears SF, et al.28
Although patients with ICDs have effective protection from arrhythmic death, recurrent symptomatic arrhythmias can have significant consequences for quality of life and psychological functioning. Optimizing ICD programming, as reviewed by Koneru and colleagues, is important. The recognition of the impact of arrhythmias on patients and the need psychologic support is reviewed by Sears and coworkers.

Biventricular pacing can improve ventricular function and extend survival in selected patients, and the beneficial effects stemming from the change in ventricular activation extends beyond the immediate mechanical effects as reviewed by Cho and colleagues.

Autopsy analysis of the implantation site of a permanent selective direct his bundle pacing lead. Correa de Sa DD, et al.

Arrhythmia management devices have not only improved outcomes for patients with arrhythmias, but also provided substantial insights into pathophysiology. Correa de Sa et al describe an anatomic study of a patient who had effective, chronic His bundle pacing for 2 years and at autopsy the lead position could be directly examined and related to the anatomy of the conduction system.

Shock lead impedance alert: Replace or reconsider? Hilgendorf J, et al.

Charge circuit timeout: A sequence of events leading to failure of an implantable cardioverter-defibrillator to deliver therapy. Catanzaro JN, et al.

Trouble shooting arrhythmia management devices is an important aspect of cardiac electrophysiology. Hilgendorf describe a case of a fall in ICD lead impedance and discuss causes. Catanzaro et al report a case of an unusual ICD software error and discuss the sequence of events in capacitor charging.

Seventh try’s a charm: Ventricular fibrillation terminated by the seventh shock. Li J, et al.

Transvenous implantable defibrillators are highly effective in terminating ventricular tachycardia and fibrillation. Li et al describe a patient who required multiple shocks and review features associated with increased energy requirements for defibrillation.

Right coronary artery fistula as a result of delayed right atrial perforation by a passive fixation lead. Khoeiry G, et al.

Cardiac perforation is an important risk of transvenous lead placement, that usually occurs acutely and often requires only lead repositioning, although tamponade and pericarditis can occur. Khoeiry et al report an unusual case of chronic perforation of an atrial lead ultimately leading to a right atrial coronary artery fistula, and review factors associated with lead perforation.

Paroxysmal Supraventricular Tachycardias


Premature beats and unexpected heart block: An unusual mechanism confirmed by ablation. Morris KE, et al.

AV nodal reentry tachycardia is the most common paroxysmal supraventricular tachycardia encountered in adults and ablation of the slow AV nodal pathway is usually straightforward. The AV node and its connections to the atrium are variable, however, and can be complex. Nguyen show that lower common pathway block can falsely suggest atrial tachycardia, and that some patients require ablation of the leftward extension of the AV node.

Morris et al present a case with complex interaction between an AV nodal slow pathway and fast pathway producing a dual ventricular response in sinus rhythm and interesting episodes of AV block, providing an opportunity to review the physiology of concealed conduction and the distinction of junctional automatic beats from simultaneous dual conduction through the AV node.

Two cases of supraventricular tachycardia after accessory pathway ablation. Wright JM, et al.

Editors’ Perspective: Con founding Eccentricity


Editors’ Perspective: Antioventricular Accessory Pathways and the Atrio-Ventricular Septum

Catheter ablation of an unusual decremental accessory pathway in the left coronary cusp of the aortic valve mimicking outflow tract ventricular tachycardia. Wilsmore BR, et al.

Editors’ Perspective: Teaching Points for Decremental Pathways


Editors’ Perspective: Since We Cannot Directly Measure…We Must Deduce...


Rare forms of preexcitation: A case study and brief overview of familial forms of preexcitation. Koneru JN, et al.

Editors’ Perspective: Difficult Accessory Pathways and Ventricular Diseases

It is important to have a systematic approach to mapping and ablation of accessory pathways causing tachycardias. Most are straightforward, but when difficult and confusing data are observed there are several considerations including: multiple accessory pathways, multiple tachycardia mechanisms, unusually accessory pathway locations, complex septal anatomy and prior ablation altering the atrial activation sequence during orthodromic tachycardia. Mapping the atrial insertion of accessory pathways can be difficult due to the potential for insertion of the pathway into musculature around the coronary sinus, rather than the left atrium, and interpretation of activation may be made more difficult in the presence of prior ablation lesions. These issues are well shown in cases described by Wright and Liu and coworkers.

Antidromic tachycardias use an accessory pathway in the atrial to ventricular direction and can mimic ventricular tachycardias. A systematic approach to exclude ventricular tachycardia and other types of supraventricular tachycardia with aberrancy is needed. Wilsmore and coworkers illustrate the approach to an antidromic tachycardia in a patient with an unusual accessory pathway that mimicked RV outflow tract VT due to its unusual location. In their Teaching Points presentation Thajudeen and coworkers demonstrate these principles in a patient with multiple accessory pathways with antidromic AV reentry and orthodromic tachycardias as well as atrial tachycardia. Leiria and colleagues provide an unusual case illustrating the dangerous rapid ventricular response that can occur during atrial fibrillation in the presence of a rapidly conducting accessory pathway.

Although accessory pathways are most commonly encountered in patients without other forms of heart disease, they are associated with a number of genetic disorders, which are important to consider, as reviewed by Koneru and colleagues.

Atrial tachycardia originating from the junction of the right atrium and a diverticulum of the inferior vena cava. Yamada T, et al.


Focal atrial tachycardias often originate from defined atrial structures. The approach to mapping focal atrial tachycardia is nicely shown in the case report from Yamada et al. Ip and colleagues describe a case of atrial tachycardia precipitated by deglutition that originated from a pulmonary vein.
Sudden Death: Diseases and Syndromes

Long-qt syndrome: From genetics to management. Schwartz PJ, et al.43

Short qt syndrome: From bench to bedside. Patel C, et al.46

Sudden cardiac death in hypertrophic cardiomyopathy. O'Mahony C, et al.47

Arrhythmogenic right ventricular cardiomyopathy. Basso C, et al.48

Brugada syndrome. Mizusawa Y, Wilde AA.49

Catecholaminergic polymorphic ventricular tachycardia. Leenhardt A, et al.50

Dilated cardiomyopathy. Lakdawala NK, et al.51

How to perform and interpret provocative testing for the diagnosis of brugada syndrome, long-qt syndrome, and catecholaminergic polymorphic ventricular tachycardia. Obeyesekere MN, et al.52

Reviews of genetic syndromes causing sudden death and their diagnosis are provided by Schwartz,43 Patel,46 O'Mahony,47 Mizusawa,49 Leenhardt,50 Basso,48 Lakdawala,51 and Obeyesekere.52

Arrhythmogenic right ventricular cardiomyopathy. Basso C, et al.48

Brugada syndrome. Mizusawa Y, Wilde AA.49

Catecholaminergic polymorphic ventricular tachycardia. Leenhardt A, et al.50

Dilated cardiomyopathy. Lakdawala NK, et al.51

Left-dominant arrhythmogenic cardiomyopathy. Smaldone C, et al.53

Smaldone and colleagues present a case of cardiomyopathy with myocardial fibrosis and fat infiltration commonly associated with arrhythmogenic RV cardiomyopathy, but in this case prominently involving the LV, and discuss the difficult diagnosis.53

Does a Brugada pattern ecg precipitated by excessive-dose flecainide provide a diagnosis of a Brugada syndrome patient and/or contraindicate its use? A case study. Reiffel JA.54

Management of an asymptomatic patient with a Brugada type electrocardiogram remains a challenging problem, as the risk of arrhythmic events is low, but an unquantifiable possibility of sudden death often remains. Reiffel present a case that illustrates the conundrum and reviews possible approaches.54

Anomalous right coronary artery and sudden cardiac death. Greet B, et al.55

Congenital coronary anomalies are a potential cause of sudden death, but documentation of the mechanism is rare. Greet and colleagues present a patient who was resuscitated from ventricular fibrillation and found to have anomalous origin of the right coronary artery from the left coronary cusp, and review the relation of coronary anomalies to sudden death.55

Successful catheter ablation of bidirectional ventricular premature contractions triggering ventricular fibrillation in catecholaminergic polymorphic ventricular tachycardia with ryr2 mutation. Kaneshiro T, et al.56

Repetitive episodes of polymorphic VT or ventricular fibrillation (VF) initiated by identifiable PVC foci can be ablated to control the arrhythmia. This situation is most commonly encountered in patients with idiopathic VF, but has also been seen in long QT syndrome, Brugada syndrome, post myocardial infarction, and in cardiomyopathies. Kaneshiro and coworkers report ablation in a patient with catecholaminergic polymorphic VT, showing the features of this rare disease, as well as the approach to focal ablation of PVC triggers.56

Pantoprazole (proton pump inhibitor) contributing to torsades de pointes storm. Bibawy JN, et al.57

A large number of agents can cause QT prolongation and the polymorphic VT torsade de pointes. Bibawy and colleagues report a case linked to a proton pump inhibitor in a patient with other predisposing factors and review issues surrounding evaluation and management of acquired torsade de pointes.57

Ventricular Arrhythmias

For reviews of mapping relevant to ventricular arrhythmias see the Teaching Points articles by Del Carpio3–6 noted above.

Examination of explanted heart after radiofrequency ablation for intractable ventricular arrhythmia. Kelesidis I, et al.58

Left ventricular perforation during cooled-tip radiofrequency ablation for ischemic ventricular tachycardia. Jimenez A, et al.59

Sustained monomorphic VT in patients with structural heart disease is usually due to scar-related reentry. Kelesidis and coworkers compare voltage maps and ablation sites in electroanatomic maps to direct observation of the anatomy after the heart was explanted at transplantation from a patient with nonischemic cardiomyopathy, showing the appears of RF lesions, and anatomic obstacles, including papillary muscles and the thick ventricular wall that can make ablation challenging.54 Irrigated RF ablation often allows creation of deep RF lesions that facilitates ablation, but as shown in the case described by Jimenez, creation of transmural lesions through the myocardium can result in perforation.59

Ablation of ventricular tachycardia in chronic chagasic cardiomyopathy with giant basal aneurysm: Carto sound, ct, and mri merge. Valdigem BP, et al.60

Unusual complications of percutaneous epicardial access and epicardial mapping and ablation of cardiac arrhythmias. Koruth JS, et al.61

Pleuropericardial fistula formation after prior epicardial catheter ablation for ventricular tachycardia. Mathuria N, et al.62

Percutaneous access to the pericardial space for epicardial mapping is required for subepicardial VTs, particularly in patients with nonischemic cardiomyopathies and arrhythmogenic right ventricular cardiomyopathy.61,62 Valdigem and coworkers show a case of endocardial and epicardial mapping for VT due to Chagas disease, with preprocedure imaging that further illustrates the anatomic relations.60 Epicardial mapping and ablation procedure are likely associated with a greater risk of complications than endocardial ablation.63 In a case series Koruth and coworkers describe potential complications related to obtaining epicardial access, mapping and ablation, including pericardial bleeding, subdiaphragmatic bleeding, right ventricular to abdominal fistula, and coronary spasm.61 Mathuria and coworkers report a case of a pleuropericardial fistula related to epicardial mapping and ablation and review the relevant anatomy.62

The left ventricular ostium: An anatomic concept relevant to idiopathic ventricular arrhythmias. Yamada T, et al.64

Supraventricular arrhythmia: Identifying and ablating the substrate. Tabataabeai N, Astrovtham SJ.65

A novel, minimally-invasive surgical approach for ablation of ventricular tachycardia originating near the proximal left anterior descending coronary artery. Mulpuru SK, et al.66

Bipolar ablation of ventricular tachycardia in a patient after atrial switch operation for dextro-transposition of the great arteries. Piers SR, et al.67

VT originating from the outflow tract and supravalvular regions of the heart are encountered in patients with and without structural heart disease. Understanding the anatomy in this region is critical to successful ablation and is reviewed by Tabataabeai and Yamada and colleagues.66,67 Arrhythmias in the protected area anterior to the aorta, beneath the left coronary artery are not approachable with catheter ablation. Mulpuru show a surgical approach to ablation of this region.66 Piers and coworkers present a challenging case of VT originating from tissue between the aorta and RV in a patient with repaired congenital heart disease.66 Integrating CT and mapping data illustrates the anatomic challenges.
Disclosures
W.G.S. is coholder of a patent on needle ablation that is consigned to Brigham and Women’s Hospital. S.J.A. receives no significant honoraria and is a consultant with Abiomed, Atricure, Biotronik, Boston Scientific, Medtronic, Spectranetics, St.Jude, Sanofi-Aventis, Wolters Kluwer, and Elsevier.

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Fundamental Concepts in Electrophysiology in Cases and Reviews
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