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Original Article

Arrhythmic complications associated with the treatment of patients with congenital cardiac disease: consensus definitions from the Multi-Societal Database Committee for Pediatric and Congenital Heart Disease

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Abstract A detailed hierarchal nomenclature of arrhythmias is offered with definition of its applications to diagnosis and complications. The conceptual and organizational approach to discussion of arrhythmias employs the following sequence: location – mechanism – aetiology – duration. The classification of arrhythmias is heuristically divided into an anatomical hierarchy: atrial, junctional, ventricular, or atrioventricular. Mechanisms are most simplistically classified as either reentrant, such as macro-reentrant atrial tachycardia, previously described as atrial flutter, or focal, such as automatic or micro-reentrant tachycardia, for example, junctional ectopic tachycardia. The aetiology of arrhythmias can be either iatrogenic, such as postsurgical, or non-iatrogenic, such as genetic or congenital, and in many cases is multi-factorial. Assigning an aetiology to an arrhythmia is distinct from understanding the mechanism of the arrhythmia, yet assignment of a possible aetiology of an arrhythmia may have important therapeutic implications in certain clinical settings. For example, postoperative atrial arrhythmias in patients after cardiac transplantation may be harbingers of rejection or consequent to remediable imbalances of electrolytes. The duration, frequency of, and time to occurrence of arrhythmia are temporal measures that further refine arrhythmia definition, and may offer insight into ascription of aetiology. Finally, arrhythmias do not occur in a void, but interact with other organ systems. Arrhythmias not only can result from perturbations of other organ systems, such as renal failure, but can produce dysfunction in other organ systems due to haemodynamic compromise or embolic phenomena.

Keywords: Congenital heart disease; quality improvement; patient safety; outcomes; registry; operative morbidity; paediatric; surgery; congenital abnormalities; cardiac surgical procedures; heart; arrhythmia; dysrhythmia

Historical background

The creation of an accurate, comprehensive, denotative, descriptive, and universal system of nomenclature for arrhythmias has been challenged by the meteoric discovery of mechanisms of arrhythmias, therapeutic options, and the penchant of authors for new names to distinguish one arrhythmia from another.[1] As a result, many synonyms have entered into the literature to describe the same phenomena, but subsequent understanding of the mechanisms of arrhythmias has clarified the diagnostic schema. The community of academic cardiac electrophysiologists has attempted to make sense of this organized chaos and has offered denotative descriptions.[2–8] However, many situations exist that have not been addressed. A distinction exists between the mechanism of the arrhythmia and the aetiology or predisposing cause of the arrhythmia. The purpose of this article is to offer a detailed hierarchal nomenclature

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of arrhythmias and to define its applications to diagnosis and complications.[9]

Consensus definitions

This article will use definition of arrhythmia from The World Health Organization and The International Society of Cardiology Task Force:

> An arrhythmia is defined as "any cardiac rhythm other than the normal sinus rhythm. Such a rhythm may be either of sinus or ectopic origin, and either regular or irregular. An arrhythmia may be due to a disturbance in impulse formation or conduction, or both".[2,10]

The conceptual and organizational approach used in this article for discussion of arrhythmias employs the following sequence:

- location
- mechanism
- aetiology
- duration.

The classification of arrhythmias is heuristically divided into an anatomical hierarchy:

- atrial
- junctional
- ventricular, or
- atrioventricular.

This anatomical hierarchy is followed by the attribution of a mechanism. Mechanisms are most simplistically classified as either

- reentrant, such as macro-reentrant atrial tachycardia, previously described as atrial flutter, or
- focal, such as automatic or micro-reentrant tachycardia, for example junctional ectopic tachycardia.

Reentry accounts for greater than 80% of clinical arrhythmias. By definition, a *reentrant* arrhythmia can be initiated and terminated with pacing. The reentrant mechanism involves unidirectional block and an area of slow conduction; the electrical impulse is blocked in one direction, and due to slowing is able to "reenter" the tissue from the opposite direction. The classic example of a reentrant rhythm is Wolff-Parkinson-White syndrome, with tachycardia utilizing an accessory connection. During sinus rhythm, conduction occurs from atrium to ventricle over both the atrioventricular node and the accessory connection. This fusion of conduction produces a delta wave. At the initiation of tachycardia, conduction is blocked in the antegrade direction over the accessory connection with loss of the delta wave. The electrical impulse progresses from atrium to ventricle via the atrioventricular node, traverses ventricular muscle, and reenters the atrium via the accessory connection. This combination of atria, atrioventricular node, ventricle, and accessory connection results in a circuit with the potential for unidirectional block (accessory connection) and slowing (atrioventricular node) allowing electricity to complete a loop.

Arrhythmias that are not reentrant may be either due to enhanced automaticity or a triggered mechanism. The clinical determination of a triggered mechanism is not usually possible. These arrhythmias are usually *focal* in origin, meaning that there is a discrete origin with locally circumscribed tissue producing a radial spread of electrical activation. The mechanism is described as focal, recognizing that there may be micro-reentry or enhanced automaticity producing the tachycardia.

The aetiology of arrhythmias can be either iatrogenic, such as postsurgical, or non-iatrogenic, such as genetic or congenital. Iatrogenic arrhythmias can be secondary to multiple aetiologies that include:

- postprocedural
- mechanical
- ischemic
- metabolic
- infectious, and
- multi-factorial.

Similarly, non-iatrogenic arrhythmias may also be secondary to multiple aetiologies that include:

- ischemic
- scar-related, such as stretch, fibrosis or associated with jet lesions
- metabolic
- infectious, and
- multi-factorial.

There has been a tendency to describe an arrhythmia that includes an aetiology or a situation, and because the aetiology or situation is suppositional, we recommend that classification of arrhythmias be based on the type or mechanism of arrhythmia alone. Sub-categories indicating predisposing causes can be assigned as desired. The assignment of the possible arrhythmia aetiology should be considered a sub-category, separate from the classification of the arrhythmia.

A variety of temporal descriptors can be used to further subclassify arrhythmias:

- Early arrhythmias are also known as perioperative arrhythmias and occur during the hospitalization or within 30 days of surgery if the patient has been discharged
- Late arrhythmias develop after hospital discharge and more than 30 days after surgery
- Non-sustained arrhythmias have a duration of less than 30 seconds

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- Sustained arrhythmias last for greater than 30 seconds or require immediate termination due to haemodynamic compromise
- The frequency of occurrence of arrhythmias can be described as paroxysmal, recurrent, chronic recurring, or permanent.

Arrhythmias do not necessarily result in harm, and thus are not uniformly adverse events. Clearly, medical error such as inappropriate drug administration can result in an arrhythmia, which may or may not be symptomatic or associated with harm. For instance, excessive potassium administration can result in QRS widening; this is a medical error that may not be associated with a suboptimal outcome.

Controversies

Assigning an aetiology to an arrhythmia is distinct from understanding the mechanism of the arrhythmia. The aetiology can be

- genetic or congenital, as in the case of accessory connections
- acquired, as in the case of atrial reentry tachycardia, or
- multifactorial, as in the case of long standing haemodynamic compromise after a corrective or palliative operation.

Oftentimes it is difficult to assign a primary aetiology because of multiple factors involved.

Availability of observed data can skew the attribution of aetiology. For instance, consider the situation where upon opening the chest, an arrhythmia occurs. The immediate observation is that the arrhythmia is mechanically induced. However, there is simultaneous or co-existent induction of anaesthesia, with the potential for hypoxia, hypotension, and shifts of electrolytes, each of which could result in arrhythmia. In addition, the central venous line, which was just positioned at the junction of the superior caval vein and the right atrium, could mechanically provoke an arrhythmia.

Bradycardia after a Mustard repair was traditionally ascribed to interrupted flow to the artery to the sinus node. However, longitudinal studies show that a gradual progressive development of bradycardia occurs that is more likely multi-factorial. It can be due to multiple aetiologies:

- direct injury to tissue of the sinus node
- injury to the artery to the sinus node
- intrinsic architectural and or haemodynamic abnormalities
- chronic stretch and fibrosis.

Another example can be intraoperative ventricular tachycardia. The heart may be manipulated or retracted in efforts to achieve haemostasis. The physical manipulation of the heart or electrocautery may provoke ventricular arrhythmia. Alternatively, the manipulation may compromise coronary flow or cardiac output, resulting in ischemia or local electrolyte abnormalities, thus provoking ventricular tachycardia. Ventricular tachycardia is the observed arrhythmia, which in this hypothetical case was likely multi-factorial in origin.

Non-sustained ventricular tachycardia can be seen after repair of tetralogy of Fallot with a transannular patch. The immediate assumption is that the ventricular tachycardia is due to the right ventricular incision. However, there also exists the possibility of ischemia from coronary arterial injury, chest tube compression of the ventricle, right ventricular hypertension, hypokalemia, and/or the central line crossing the tricuspid valve. The observed arrhythmia is ventricular tachycardia; the duration is non-sustained, and the aetiology remains to be determined.

The conduction pattern of an arrhythmia is not relevant to the arrhythmia mechanism, although it may certainly be relevant to the patient. Atrial premature beats conducted with aberrancy are no different clinically from normally conducted atrial premature beats. Similarly, atrial premature beats occurring in a bigeminal or trigeminal pattern is a descriptive terminology without implication of mechanism. *Multiform* ventricular premature beats do not necessarily imply *multi-focal* origin of ventricular irritability. The atrioventricular nodal conduction of a rapid atrial tachycardia does not change the classification of the arrhythmia mechanism.

Late occurrence of atrioventricular block in a patient with congenitally corrected transposition of the great arteries can be attributed to surgical closure of the ventricular septal defect, abnormal course or position of the atrioventricular node and conduction system, or perhaps a genetic predisposition such as that seen in patients with the NKX 2.5 gene. In this case, the observed arrhythmia is atrioventricular block, but the etiologic attribution rests between congenital, surgically acquired, and genetic predisposition.

Another diagnostic dilemma is the adult patient with atrial tachycardia after closure of a secundum atrial septal defect. It is well known that adult patients with atrial septal defect may develop atrial tachycardia with or without surgery. In a specific individual, can the attribution of the tachycardia be assigned with assurance to the antecedent disease process or to the surgical atrial incision? No doubt, the speculated answer to this question may be determined by temporal relationships; however, the true answer is moot. There is an arrhythmia – and it is atrial tachycardia.

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The distinction between mechanism and aetiology has important therapeutic implications. Postoperative atrial arrhythmias in transplant patients can be important harbingers of rejection, innocuous consequences of electrolyte abnormalities, a manifestation of pre-existing focal tachycardia from either donor or recipient cardiac tissue, or devastating consequences of coronary artery disease. The arrhythmia mechanism per se is not pivotal, but the proper ascription of aetiology is essential to initiation of appropriate treatment.

Interactions

Arrhythmias can result from perturbations of other organ systems, such as

- renal failure or diarrhea with electrolyte disturbances
- hepatic failure with prolongation of the QT interval
- gastroesophageal reflux or myocardial infarction resulting in vagotonia and bradycardia, or
- pain releasing catecholamines that produce tachycardia.

Conversely, arrhythmias can produce dysfunction in other organ systems, due to haemodynamic compromise or embolic phenomena. Most commonly, hypotension from tachycardia results in decreased cardiac output, poor organ perfusion and resultant organ dysfunction. Pulmonary oedema can result from worsening cardiac function due to arrhythmia. Bradycardia, including that seen with surgical atrioventricular block, can result in decreased cardiac output. In addition, intracavitary thrombus formation from a persistent atrial arrhythmia may result in systemic or pulmonary embolization.

Arrhythmia treatment may result in organ dysfunction, such as dislodgement of a cardiac thrombus with "direct current cardioversion" of atrial fibrillation, producing neurologic injury. Administration of medication may have direct toxic effects to other organ systems, such as amiodarone producing hepatic, thyroid or pulmonary toxicity.

Conclusions

We present a system of classification of arrhythmias meant to produce clarity and help to resolve difficult clinical dilemmas of nomenclature. Postoperative arrhythmic complications may or may not be multi-factorial in origin. The aetiology of an arrhythmia is distinct from the arrhythmia mechanism, although the aetiology may certainly guide therapy.

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