‘Born’ with a ‘dead’-end-tract resulting in arrhythmias in the aorto-mitral continuity: coincidence, causation, and ‘commensuration’

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Ventricular arrhythmias originating from the aortomitral continuity: an uncommon variant of left ventricular outflow tract tachycardia by J. Chen et al., on page 388

The word coincidence is derived from the Latin cum- (‘with’, ‘together’) and incidere (a verb composed from ‘in’ and ‘cadere’: ‘to fall on’, ‘to happen’). As such, a coincidence occurs when something uncanny, accidental, and unexpected happens under conditions named, but not under a defined relationship. On the other hand, causation (also referred to as causality) is the relationship between an event (the cause) and a second event (the effect), where the second event is understood as a consequence of the first one.

One of the most challenging types of research in biology is to determine the causation between observed phenomena. In order to prove causation, sophisticated animal or in vitro experiments are often required. However, invariably the first step is an observation which draws attention to an unexpected phenomenon occurring simultaneously in patients.

In this issue of the Journal, Chen et al. present their experience with ventricular and supraventricular arrhythmias originating from the region called aorto-mitral continuity (AMC). The primary aim of the manuscript was to describe the electrocardiographic (ECG) characteristics of premature ventricular complexes (PVCs) originating from this region. The ECG characteristics of AMC-related ventricular tachycardia (VT) are very similar to those of other left ventricular outflow tract tachycardias and they may even overlap because they all originate from closely adjacent cardiac structures, such as the left coronary cusp, the anterior side around the mitral annulus, the superior basal septum and epicardium. In order to safely perform radiofrequency catheter ablation, it is important to differentiate the ECG characteristics of left ventricular outflow tract VTs, as has already been pursued by other authors who published similar results. Dixit et al. showed PVCs from AMC to have a qR pattern in V1 without rS. The mean QRS duration was also consistent with those observed by Kumagai et al., suggesting that the origin may be located deeply in the subepicardium, which was also supported by recording late activation of the ventricular area during sinus rhythm. Moreover, this is the first study to divide AMC into three parts and describe the VT/PVC origins as anterior, mid-, and posterior AMC.

The ECG differences have only been described between anterior and mid-AMC-VTs as none of the patients have experienced VT or PVCs that originated from the posterior area, which is consistent with the development theory.

Much more interesting is that in some patients atrioventricular nodal reentrant tachycardia (AVNRT) was also observed. In order to determine whether this is a coincidence or causation we should look more closely into the unique structure and the embryogenetic development of AMC.

Aorto-mitral continuity, which is also called the left fibrous trigone, consists of the aortic annulus and the anterior side around the mitral annulus. Although this is a fibrous structure, it is known to be a potentially arrhythmogenic area for atrial as well as ventricular tachyarrhythmias. Systematic studies investigating the arrhythmogenic potential of AMC are very scarce. In order to better understand possible aetiologies, we must take a closer look at the development of the atrioventricular conduction. It is hypothesized that during the maturing of the atrioventricular conduction system the left and the right bundle branches, as well as a third septal branch, develops from a ‘specialised’ interventricular ring. This ring initially encircles the junction of the developing ventricles. This third, ‘non-branching’ bundle moves into the smooth septal surface of the left ventricle and encircles the aortic root as the so called ‘dead-end-tract’, first described by Kurosawa et al. in 1985. Under normal circumstances, the third branch...
disappears at maturity; however, sometimes it persists as a remnant of the atrioventricular conduction system closely located to or even reaching the AMC. This unique finding was supported by McGuire et al. who found cells in AMC that histologically and electrophysiologically resembled the atrioventricular junctional cells, as well as sleeves with nodal-type tissue around the mitral annulus. Such AMC cells might be responsible for genesis of non-reentrant tachycardias and automatic triggering.

Two cell types play an important role in the development of the heart and its conduction system. The first group consists of cardiac neural crest cells (CNCs) originating from the neural crest and invading the heart via the aortic arches. These cells have mostly an instructive and signalling function and only a limited constructive function as the majority of them are destined for apoptosis. Via the atrial pole they contribute to the bundle branches and via the venous pole to the sinoatrial and atrioventricular node areas. They also play a role in formation of the vestibular spine, the membranous part of the ventricular septum, septation of the common truncus arteriosus, smooth muscle tunics of great arteries, and the epicardial cushions at the future atrioventricular junction and outflow tract. The second group is constituted of the epicardium-driven cells (EPDCs) which originate from the proepicardium and the subepicardium. They have both constructive and instructive roles in heart morphogenesis and are often seen around CNCs. These CNCs invade the myocardium and subendocardium via small fenestrations in the myocardial layer and migrate into different structures such as the atrioventricular cushions and subendocardium of the ventricular trabeculae and atria. These trabeculae form papillary muscles, ventricular septum, and Purkinje fibers. Purkinje fibers maintain automaticity, which later may trigger ventricular arrhythmia. Migration and differentiation of these cells in the AMC region needs further investigation and may contribute to new understandings concerning possible ectopic foci in this area.

Right- and leftwards atrioventricular nodal extensions have been described. The leftward extension, however, is not consistently described in humans and its role is less clear. It expresses less connexin 43, would therefore conduct slowly and may form a slow conducting pathway in cases of AVNRT where more than one slow pathway is observed, in intranodal reentry, or may even provide a slowly conducting pathway between the left atrial side of the intraatrial septum and the nodal tissue. Whether this leftward extension may be long enough to reach AMC and, therefore contribute to reentrant arrhythmias involving the AMC region, has not been reported and also requires further research.

What makes the occurrence of AVNRT in this patient population especially intriguing is the lack of any evidence suggesting ‘commensuration’ (association) between the findings mentioned above. Commensuration is a relation between events. Basically, events are commensurate with one another when one contains all that is required for the occurrence of the other, and as little as possible that is not required. Certainly, the other end of this spectrum is over-determination. Over-determination in the present context implies a causal relation between the above-mentioned events. However, these may simply be coincidental.

The authors of this manuscript as well as others describe patients with AMC-VTs and occurrence of AVNRT without clear evidence of a causative or exclusive relationship. Until now, literature does not provide clear histological, developmental, or electrophysiological explanation for these findings.

To follow our initial philosophy at this point in time we are unable to determine whether there is any causality between these arrhythmias. We have no evidence showing commensuration between these effects. In order to reveal possible commensuration or to translate this into biological terms, further investigations are required. Specifically, pathological and probably imaging studies such as molecular imaging might give answers. Regardless of the possibility of coincidence, the study by Chen et al. further helps determine the ECG characteristics of AMC-VTs and PVCs and may improve the safety of ablation interventions.

Conflict of interest: none declared.

References