

## Turkish Society of Cardiology consensus paper on management of arrhythmia-induced cardiomyopathy

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### Background

Heart failure (HF) is one of the major causes of mortality and morbidity. The identification of causes of left ventricular (LV) systolic dysfunction is important in terms of initiating causal treatment and improving prognosis. Arrhythmia-induced cardiomyopathy (AIC) is a potentially reversible form of cardiomyopathy (CMP) in which LV dysfunction results from atrial or ventricular arrhythmias (1). It can be resolved by eliminating or effectively treating responsible arrhythmia (2).

### Aim of the document

Early recognition of the relationship between responsible arrhythmia and CMP is of great importance in terms of the improvement of symptoms, LV systolic dysfunction, and functional status with effective treatment. However, in the clinical practice, AIC is often overlooked, and arrhythmias are generally seen as the result of HF. Again, there is a lack of information about the pathophysiology of AIC and the course of the disease after effective treatment of the responsible arrhythmia. This document is written to give clear messages for further recognition and treatment of AIC based on the current literature.

### Definition

AIC is defined as LV systolic dysfunction due to supraventricular or ventricular arrhythmia that can be either sustained

or paroxysmal or is characterized by highly frequent ectopic activity (3). AIC can be divided into two categories. Type 1 AIC (arrhythmia-induced): arrhythmia is accepted as the absolute reason of ventricular dysfunction that returns to normal after successful treatment of arrhythmia. Type 2 AIC (arrhythmia-mediated): arrhythmia exacerbates the LV dysfunction in patients with concomitant heart disease, and treatment of the arrhythmia provides partial improvement (4).

### Epidemiology

The prevalence of HF is increasing worldwide due to better treatment of acute cardiac events, improvements in medical and surgical treatment methods, and aging of the population. Approximately 1%–2% of the general population, and >10% of over 70 years old are affected with HF (5). Cardiac arrhythmias generally occur during the natural course of HF, but sometimes they are the sole etiology of the unexplained systolic HF or dilated CMP. Reliable epidemiological data regarding the AIC are lacking, and the prevalence in general is underestimated, given that arrhythmia is often considered to be a result of rather than a possible cause of CMP.

Although age is the major determinant of incidence and prevalence of overall HF, AIC appears to occur at any age. However, the common types of arrhythmias causing AIC differ among age groups. Focal atrial tachycardia (FAT) (59%) and permanent

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junctional reciprocating tachycardia (PJRT) (23%) are common causes of AIC in children in the largest pediatric series of AIC, whereas ventricular arrhythmias are rare (4). The incidence of AIC was 9%–34% in adult patients with frequent premature ventricular complexes (PVC) and/or nonsustained ventricular tachycardia (VT) referred for electrophysiological evaluation (6).







The most common cause of AIC in adults is atrial fibrillation (AF). Most common arrhythmia coexisting with HF is also AF. The LV systolic dysfunction is found in 20%–30% of all patients with AF, and 10%–50% of patients with HF have AF (7). In the Framingham study, those with AF had a higher risk of developing HF [hazard ratio of 2.22 (CI 1.47–3.34)  $p < 0.0001$ ] (8). Both AF and HF can directly lead to the other, so it is not easy to assess the causal link between AF and systolic dysfunction. The definite diagnosis of AIC in this context can only be made if systolic dysfunction is reversible after restoration of sinus rhythm. Recent ablation studies have revealed that approximately one-third of patients with

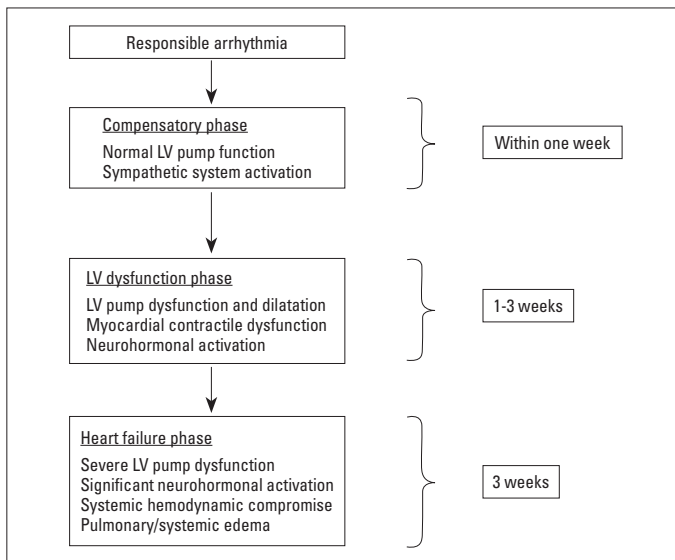
AF and systolic HF had primarily idiopathic dilated CMP, and AIC was detected in 58%–88% of these cases (9, 10).

### Pathophysiology and mechanisms

The main three mechanisms that appear to be responsible for the AIC development are tachycardia, irregular rhythm, and dyssynchrony. There is significant overlap among these mechanisms (11).

In animal models, rapid stimulation has been shown to result in LV dysfunction within weeks after tachycardia begins (4). Three phases have been defined in this situation (Fig. 1). In the compensatory phase (the first 3–7 days of rapid pacing), the LV pump function is normal, and there is an increased neurohormonal activation with early changes in the extracellular matrix. In the LV dysfunction phase (about 1–3 weeks after the onset of rapid pacing), there is cellular remodeling, contractile dysfunction with LV systolic dysfunction, and dilatation. Continued neu-

Consensus statements		References
Main clinical scenarios that increase the suspicion of arrhythmia-induced or arrhythmia-mediated cardiomyopathy (CMP) (AIC) can be listed as follows:		19
<ul style="list-style-type: none"> <li>• Simultaneous presentation of a tachyarrhythmia or frequent ectopy and systolic dysfunction in a patient with no preexisting heart disease.</li> <li>• Asymptomatic CMP in the setting of a persistent arrhythmia or frequent ectopy.</li> <li>• A patient with known structural heart disease now presenting with worsening left ventricular (LV) dysfunction and heart failure (HF) secondary to an arrhythmia.</li> </ul>		
Treatment of AIC should be primarily aimed at eliminating or controlling the arrhythmia using either pharmacological or ablative techniques with the goal of improving symptoms and reversing systolic dysfunction. The exact approach should be selected depending on the underlying arrhythmia. Curative ablation is the preferred method of choice in appropriate patients.		1, 4, 12, 31, 50
Following the normalization of LV function, continuation of HF medication is recommended.		1, 4
Close follow-up is recommended given that recurrent arrhythmia can result in rapid decline in LV function with development of HF.		1, 4
The following laboratory findings can help distinguish AIC from other CMPs: <ul style="list-style-type: none"> <li>• A smaller LV end diastolic diameter and mass index</li> <li>• A more profound reduction in apical longitudinal strain compared to the mid and basal segments</li> <li>• Absence of late gadolinium enhancement and early right ventricular systolic dysfunction on cardiac magnetic resonance imaging</li> <li>• Significant smaller percentage of the LV endocardium with abnormal unipolar voltage</li> <li>• Rapid decline of the N-terminal pro-brain natriuretic peptide levels at one week following control of arrhythmia</li> </ul>		22-28
Implantable cardioverter defibrillator implantation for primary prevention should be delayed for a reasonable period to see the response to the optimal medical treatment in AIC.		4



**Figure 1.** Schematic presentation of the pathophysiology of AIC in animal models

AIC - arrhythmia-induced cardiomyopathy, LV - left ventricle

rohormonal activation and upregulation of the renin angiotensin system are observed. The LV failure phase (>3 weeks) is characterized by severe LV pump failure, severe LV dilatation, significant neurohormonal activation, and defects in  $Ca^{+2}$  handling (4).

Myocardial energy store depletion, increased oxidative stress, blunted response to beta adrenergic stimulation (12, 13), reduced myocardial blood flow (14), and abnormal calcium handling (15-17) have all been implicated in the pathogenesis of AIC.

The proposed mechanisms described above are seen in multiple forms of chronic HF, and they may be related, at least in part, to the effects of elevated filling pressures and decreased cardiac output. Changes seen early after initiation of responsible arrhythmia are more likely to be related to elevated heart rates, whereas later changes are more likely to be due to a combination of the arrhythmia as well as the downstream effects of the HF syndrome (12).

### Causes of arrhythmia-induced cardiomyopathy

There is a wide range of arrhythmias and clinical conditions related with development of AIC. They are listed in Table 1.

### Clinical features

AIC has varied presentations. In fetal life, AIC might be presented with hydrops fetalis (18). In children and adults, clinical picture of AIC can vary from asymptomatic arrhythmia to end-stage HF. Many patients are asymptomatic from arrhythmias or describe subtle or challenging symptoms making early diagnosis more difficult. In this point, AIC could not be detected until manifest HF develops. When symptoms of HF exist, without aggressive treatment of HF and arrhythmia, worsening of HF is inevitable (19). Signs and/or symptoms are related to the tachyarrhythmia (e.g., palpitations, dyspnea, chest discomfort), HF (e.g., dyspnea,

**Table 1. Causes of arrhythmia-induced cardiomyopathy**

#### Supraventricular

Atrial fibrillation  
Atrial flutter  
Ectopic atrial tachycardia  
Atrioventricular nodal reentrant tachycardia  
Atrioventricular reentrant tachycardia  
Permanent junctional reciprocating tachycardia

#### Ventricular

Outflow tract ventricular tachycardia  
Fascicular tachycardia  
Bundle branch reentry ventricular tachycardia

#### Ectopy

Frequent premature ventricular/atrial complexes

#### Pacing

Persistent high rate atrial/ventricular pacing

edema, weight gain, orthopnea), or both. However, shortness of breath was reported as the primary complaint (20). It can be argued that patients with relatively higher heart rates often present earlier with symptoms related to the tachycardia, and they can be detected earlier and treated without development of CMP. Main clinical scenarios that increase the suspicion of arrhythmia-induced or arrhythmia-mediated CMP can be listed as follows (19).

1. Simultaneous presentation of a tachyarrhythmia or frequent ectopy and systolic dysfunction in a patient with no preexisting heart disease.
2. Asymptomatic CMP in the setting of a persistent arrhythmia or frequent ectopy.
3. A patient with known structural heart disease now presenting with worsening LV dysfunction and HF secondary to an arrhythmia.

### Diagnosis

The key diagnostic criterion of AIC is the detection of a pathological tachycardia or persistent arrhythmia in the presence of an otherwise unexplained LV systolic dysfunction (4). The proof of AIC is to document LV systolic dysfunction in serial measurements after pathological tachycardia or resistant arrhythmia in a patient with normal LV functions prior to onset of the arrhythmia (21). However, this proof is rarely seen in clinical practice. A single 12-lead electrocardiogram (ECG) may not diagnose responsible arrhythmia. Continuous ambulatory ECG (Holter) monitoring is able to identify recurrent tachycardia, mean ventricular rate in AF, and the frequency of PVC (1).

Some laboratory tests can help distinguish AIC from other CMPs. Patients with AIC have a smaller LV end diastolic diameter and mass index compared those with preexisting dilated CMP

and secondary tachycardia (22, 23). At baseline, patients with AIC had a more profound reduction in apical longitudinal strain compared to that in the mid and basal segments, while patients with other forms of CMP after arrhythmia correction had a predominant reduction of longitudinal strain in the basal segments (24). Absence of late gadolinium enhancement and early right ventricular systolic dysfunction on cardiac magnetic resonance imaging (MRI) might help to differentiate AIC from other heart diseases (25, 26). Unipolar electroanatomic mapping can be helpful in differentiating AIC from patients with irreversible CMP. Campos et al. (27) have demonstrated that patients with irreversible CMP had a significant larger percentage of the LV endocardium with abnormal unipolar voltage. In addition, a study showed that rapid decline of the N-terminal pro-brain natriuretic peptide levels at one week following control of arrhythmia was associated with AIC (28).

Endomyocardial biopsy specimens from patients with AIC exhibit features distinct from those of other types of CMP, including scant or absent myocardial fibrosis, increased expression of major histocompatibility complex class II molecules, CD68+ macrophage infiltration, and an enrichment of mitochondria in close proximity to intercalated disks (29). However, the potential role of the myocardial biopsy in the diagnosis of AIC requires additional investigations.

When AIC cannot be differentiated from dilated CMP with consequent tachycardia, treatments for both problems are necessary (3). The correct diagnosis can only be established after demonstration of the improvement of LV function within a few weeks or months after successful treatment of arrhythmia (21).

Table 2 shows diagnostic tests, which can help differentiate AIC from other forms of nonischemic dilated CMP.

**Management**

**a) Principles of management**

Treatment of AIC should be primarily aimed at eliminating or controlling the arrhythmia using either pharmacological or ablative techniques with the goal of improving symptoms and reversing systolic dysfunction (Fig. 2). The exact approach should be selected depending on the underlying arrhythmia.

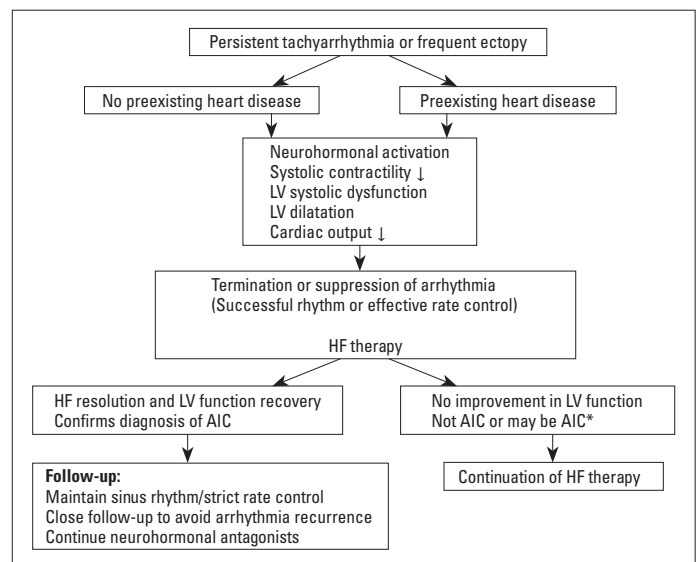
Curative ablation is the preferred method of choice in appropriate patients. Along with arrhythmia control, HF medication is recommended as well (30).

**b) Arrhythmia-specific diagnostic tests and treatment**

**Atrial fibrillation**

AF is the most common cause of AIC in adults (4, 31). AF and CMP often coexist and precipitate one another (31). Figuring out whether LV systolic dysfunction is due to underlying structural heart disease or arrhythmia itself is challenging. Rapid and irregular heart rate and loss of atrial contraction are the proposed pathophysiologic mechanisms for the development of AIC in patients with AF (32, 33).

It was thought that LV systolic function improvement could be achieved with any number of AF treatment strategies, whether that is rate control or pacing and ablation procedure or rhythm control with catheter ablation or antiarrhythmic drugs. In AF-CHF trial, no mortality advantage was observed between patients with



**Figure 2.** Flow diagram of diagnosis and treatment approaches in AIC  
 AIC - arrhythmia-induced cardiomyopathy, HF - heart failure, LV - left ventricle  
 \*: This condition may be related to more extensive myocardial damage produced by longer periods of tachycardia and/or contribution of underlying heart disease

**Table 2. Diagnostic tests that distinguish AIC from other forms of nonischemic dilated CMP**

Tests	AIC	Dilated CMP
LV end diastolic diameter	A smaller LV end diastolic diameter	A larger LV end diastolic diameter
RV systolic dysfunction	Early	Late
Strain distribution	Decreased longitudinal strain in the apical segments	Decreased longitudinal strain in the basal segments
Late gadolinium enhancement on cardiac MRI	No	Yes
NT-proBNP following control of arrhythmia	Fast reduction	Slow or limited reduction

AIC - arrhythmia-induced cardiomyopathy, CMP - cardiomyopathy, LV - left ventricle, MRI - magnetic resonance imaging, NT-proBNP - N-terminal pro-brain natriuretic peptide, RV - right ventricle

AF and HF who were randomized to either rate control or rhythm control (amiodarone with cardioversion) (34). However, AIC can occur in AF and normal ventricular rates given the fact that irregular ventricular contraction is associated with LV dyssynchrony in the long term (35, 36).

In the AATAC-AF trial, 203 patients with persistent AF and HF with LV ejection fraction (LVEF) <40% were randomized to catheter ablation arm or amiodarone arm. Arrhythmia recurrences and hospitalization rates were significantly lower with improved quality of life in the ablation arm. The LVEF improved by  $9.6\% \pm 7.4\%$  in the ablation arm compared to  $4.2\% \pm 6.2\%$  in the amiodarone arm (37). Another systematic review of 19 studies (914 patients) also demonstrated the superiority of catheter ablation to restore sinus rhythm with LVEF improvement by 13.3% (38).

CAMERA-MRI (9) and CASTLE-AF 2 (39) trials also demonstrated the significance of restoring sinus rhythm with catheter ablation with improvements in systolic function and quality of life. In the CAMERA-MRI trial, the primary end point of LVEF improvement was significantly higher in catheter ablation group compared to that in medical rate control group at a median follow-up of 6 months (18.3% vs. 4.4%,  $p < 0.0001$ ). Similarly, in CASTLE-AF 2 trial, LVEF improved by 8% in ablation arm compared with 0% in the medical arm at 60 months.

Briefly, in patients with AIC secondary to AF, restoration of sinus rhythm should be the primary goal. In terms of rhythm control, multiple studies proved the superiority of catheter ablation compared to pharmacological treatment (40-43). Despite primarily successful catheter ablation, recurrence of AF is high, and repeat ablation(s) is/are generally needed. In patients with treatment-refractory AF with a high ventricular rate, atrioventricular node ablation and biventricular pacing seems a rationale option (44, 45).

### Atrial flutter

Atrial flutter is one of the common causes of AIC. In one study, LV systolic dysfunction was observed in 25% of patients presenting with atrial flutter (46). Another study evaluating >1000 patients with atrial flutter found the incidence of AIC nearly 8% (47). Ventricular rate control is difficult in atrial flutter; therefore, rhythm control is often necessary. Although electrical cardioversion is effective in restoring sinus rhythm, recurrences may occur in follow-up period. Given the high success and low complication rate, catheter ablation approach should be primarily considered (48). The LV systolic function improvement was seen in >50% of patients with atrial flutter undergoing ablation (47).

### Supraventricular tachycardias

The AIC has been associated with essentially any frequent and persistent supraventricular tachycardia (SVT). The common reentrant SVTs such as atrioventricular nodal reentrant tachycardia and atrioventricular reciprocating tachycardia are most commonly paroxysmal, but rarely can be incessant. Therefore, they are rarely associated with AIC. The most classic incessant

SVT-mediated AICs in adults are FAT, atrial flutter, dual atrioventricular nodal nonreentrant tachycardia, and PJRT (49, 50). The FAT and atrial flutter are often refractory to pharmacological suppression (50). The radiofrequency ablation of SVTs is effective in over 95% of cases, and it should be recommended as a first-line therapy for SVT-mediated AIC in adults (4).

### Frequent premature ventricular contractions and VT

Idiopathic ventricular arrhythmias in the absence of structural heart disease are considered a benign entity. In a normal healthy population, PVCs have been observed in up to 75% of subjects on 48-hour Holter monitoring (51). However, there is clear association between frequent PVCs and CMP (20, 52, 53).

Underlying pathophysiological mechanism is not entirely clear. Various cellular and clinical mechanisms have been suggested. The excitation-contraction coupling is impaired because of the decreased  $Ca^{+2}$  release from the sarcoplasmic reticulum that results in contractile dysfunction (54). Ventricular dyssynchrony is another suspected mechanism that may also lead to LV impairment.

The predisposition to the development of CMP is another conundrum. PVC burden is the well-accepted risk factor. Burdens above 15%–25% of the total cardiac beats are associated with CMP (20, 52, 53). On the other hand, the vast majority of patients with frequent PVCs will not develop CMP. Although many other risk factors such as lack of palpitations, nonsustained VT, a retrograde P-wave after the PVCs, interpolated PVCs, PVC QRS duration (e.g.,  $\geq 150$  ms), and epicardial origin are suggested, current data are not sufficient for accurate risk prediction (55, 56). Novel imaging modalities such as real-time three-dimensional speckle tracking echocardiography seems promising to detect subtle LV dysfunction.

Regardless of whether PVCs are the cause or the result of CMP, catheter ablation is the preferred treatment option. Successful ablation results in significant improvement of cardiac functions. PVCs burden before and after ablation are the main predictors of LVEF recovery (57). Since frequent PVCs worsen CMP, presence of structural heart disease does not diminish the benefit of ablation.

For patients who require arrhythmia suppression for symptoms or declining ventricular function suspected to be due to frequent PVCs (generally >15% of beats and predominately of one morphology) and for whom antiarrhythmic medications are ineffective, not tolerated, or not the patient's preference, catheter ablation is recommended in the 2017 AHA/ACC/HRS Ventricular Arrhythmias Guideline (Class I recommendation) (56). Pharmacologic treatment has a class IIa recommendation. Beta-blockers and amiodarone are usually preferred. There is a general reservation against other antiarrhythmics because of the increased mortality observed in patients with LV dysfunction and history of myocardial infarction (58, 59). In selected patients suspected of having PVC-induced CMP, Class IC antiarrhythmic drugs effectively suppressed PVCs, leading to LVEF recovery in the major-



ity of 20 patients. No adverse events were reported in this small cohort (60).

Implantable cardioverter defibrillator (ICD) implantation in PVC-induced CMP is not a rare scenario. In these patients, it should be evaluated whether LV function improves after an effective treatment such as ablation.

### **AIC in adults with congenital heart disease**

Cardiac arrhythmias are a major source of morbidity and mortality in adults with congenital heart disease (ACHD). In patients with ACHD, symptomatic tachyarrhythmias may be seen after surgery, or they may be seen in the absence of any intervention, as in Ebstein's anomaly (patients with this anomaly may have accessory connections) (61, 62). Intra-atrial reentrant tachycardia is the most common form of SVT in the population with ACHD (63). Ventricular tachyarrhythmias are well-known late sequelae after surgical repair of a variety of forms of the disease (61). The possible mechanisms for the development of AIC in this group of patients are similar to those in other patients (such as high heart rates, ventricular dyssynchrony) (64-66).

In patients with ACHD with AIC, it is reasonable to apply the principles of rate and rhythm control as in other patient groups (62, 63). Digitalis is less effective in younger, active patients (63). Additional ventricular scarring or systemic ventricular dysfunction/hypertrophy often limits the choice of an antiarrhythmic drug in patients with ACHD. Amiodarone is often the only option for patients with ACHD. However, long-term therapy with amiodarone has severe side effects, particularly in young adults (64). Catheter ablation has additional difficulties due to unusual conduction anatomy and difficulty accessing vessels and intracardiac chambers in such patients (63).

### **AIC in children**

The most common arrhythmias associated with pediatric AIC include FAT and PJRT (67-69). The junctional ectopic tachycardia (JET) is usually seen after congenital heart surgery ("postoperative JET") (58). Atrial flutter, AF or incessant VT may also lead to pediatric AIC although these arrhythmias are typically seen in adults. Beta-blockers are the most common first-line agent in most pediatric AIC (67, 69). There is also a trend toward increased use of flecainide in pediatric CMPs (70). In addition to first-line beta-blockers (67, 69, 71), the combination of sotalol and propafenone has been used in effectively controlling FAT (72). Since the spontaneous resolution is common for young children with FAT (69), the catheter ablation is only recommended in infants and young children if pharmacological treatment is not feasible or unsuccessful (73). However, the FAT is unlikely to resolve spontaneously, and antiarrhythmics are frequently ineffective in children aged  $\geq 3$  years (74). The spontaneous resolution of PJRT is almost unlikely with 12% of reported rate (68), and the antiarrhythmics result in complete control only in few patients. Therefore, the catheter ablation is the primary treatment for PJRT with reported success rates of 90% (68).

### **Follow-up and prognosis**

Following effective and timely treatment of arrhythmia with either elimination or adequate suppression, LV systolic function completely or partially recovers in case of AIC. However, such recovery may be totally absent. This may be related to more extensive myocardial damage produced by longer periods of tachycardia and/or contribution of underlying heart disease (21, 75). Researchers reported that patients with HF with recovered EF have better clinical courses including lower mortality than patients with permanently reduced or permanently preserved EF (76). It may take weeks to months for the recovery of systolic function after treatment of the arrhythmia. This represents the importance of timely interventions before an adverse burden of remodeling goes too far. One study showed that even after years of complete recovery of systolic function, mild LV dilatation and ultrastructural myocardial lesions may be seen (77). This may explain the rare incidences of sudden cardiac death in patients with AIC who had already recovered systolic function (31).

It is also evident that recurrent tachyarrhythmias following initial recovery can cause relapses of AIC (31). Although CMP in response to an arrhythmia may take months to years to develop, recurrent arrhythmia can result in rapid decline in ventricular function with development of HF, suggesting residual ultrastructural abnormalities in the so-called HF with recovered EF. Very close patient follow-up with ambulatory Holter electrocardiogram and imaging studies and aggressive treatment if needed could prevent the devastating consequences of recurrent arrhythmias.

AF-induced CMP is also associated with a more benign prognosis compared to new-onset AF in a patient with established HF because in the latter form, AF is a marker of more advanced HF and associated with a worse outcome (78). Distinguishing AIC in case of AF and HF is very important because aggressive attempts for restoration of sinus rhythm in case of AIC could lead to complete recovery of systolic function and favorable prognosis. Both CAMERA-MRI and CASTLE HF trials have demonstrated that catheter ablation in patients with AF and systolic dysfunction resulted in improvements in HF symptoms, LVEF with reductions in hospitalizations, and total mortality (9, 39). Part of these positive effects of catheter ablation in these studies, although inconclusive, may be attributed to the substantial number of AIC patients presented in these two studies.

Although the decisive treatment of AIC is the control of arrhythmia, treatment with disease-modifying drugs (angiotensin converting enzyme inhibitor, beta-blockers, mineralocorticoid receptor antagonists) still play an important role. The continuation of these medical treatments following recovery of LV systolic function is controversial. It is advisable to continue these medications for HF after recovery of systolic function considering that the persistence of subtle negative remodeling in such patients (31, 77).

### **Summary**

Patients with AIC are the relatively favorable subgroup of the patients with CMP. Correcting the underlying arrhythmia can pro-

vide a dramatic improvement in the LV functions. On the other hand, it may not always be easy to determine whether the arrhythmia is the result or the cause of the CMP. Even though it reminds the chicken and egg conundrum, both groups benefit from appropriate arrhythmia treatment.

Patients with AIC will undoubtedly have the chance to be cured. Regardless of the cause, frequent arrhythmias worsen the preexisting CMP, in which case arrhythmia treatment may lead to partial but important recovery of the LV dysfunction. These patients are those who mostly need sinus rhythm. Catheter ablation is the effective treatment to restore sinus rhythm. The efficacy of drugs that can be used in HF is limited, and the incidence of side effect is high.

Unfortunately, treatment of arrhythmias and restoring sinus rhythm may not always be possible. In this case, efficient rate control is crucial to prevent the development of CMP. Atrioventricular node ablation and pacemaker implantation may be an unpleasant but mandatory solution in patients with atrial arrhythmia progressing to CMP where speed control is insufficient.

It should be kept in mind that many arrhythmias are curable in the ablation era, and LV systolic functions usually improve with an effective arrhythmia treatment. Therefore, the risk assessment of sudden cardiac death and the ICD implantation for primary prevention should be delayed for a reasonable period to see the response to the optimal medical treatment.

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