Treatment of the arrhythmic storm

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Arrhythmic storm (AS) is defined as three or more episodes of sustained ventricular tachycardia (VT) or ventricular fibrillation (VF) occurring during a 24 h span.1 This condition occurs more frequently in patients with dilated cardiomyopathy, both ischaemic or idiopathic, as an evolution of the arrhythmic substrate of this pathology. Before the introduction of implantable cardioverter–defibrillator (ICD), AS was considered an event statistically irrelevant, but the more widespread application of the device has changed the epidemiological appreciation of the problem, attesting now an incidence of the condition in 10–15% of the patients with ICD.2 The main predictive factors leading to AS are severe left ventricular dysfunction, advanced age, and previous episodes of VT/VF.

Arrhythmic storms carry elevated mortality both in the acute and in the mid-long-term settings, higher than the one recorded for ventricular arrhythmias not part of an AS. The hospitalization rate is 50–80% for patients surviving AS, and they are also at higher risk of hospital admission for heart failure, heart transplantation, and death.3 Physiopathology of AS is complex and still not completely understood.

Among the main precipitating factors are: adrenergic hypertonia, acute ischaemia, heart failure, abnormalities of the intracellular calcium currents, and electrolytes unbalance. Nonetheless, as indicated in the SHIELD (SHock Inhibition Evaluation with Azimilide) trial, in only a small portion of patients (13%), a reversible cause can be clearly identified (Table 1).4 Management of AS begins with the evaluation of the haemodynamic and metabolic state of the patient, and with the continuous monitoring of the electrocardiogram (ECG), and vital parameters, in an intensive care setting. Of the utmost importance is the documentation of the arrhythmia responsible for the AS (e.g. trigger, VT morphology, baseline ECG), also in consideration of possible ablation treatment. Patients’ stabilization, prevention of the arrhythmic recurrences, sedation, and containment of the adrenergic tone, are the leading goals of the acute management of AS.

Amiodarone is the preferred drug, unless contraindicated (e.g. hypothyroidism, prolonged QTc) for prevention of arrhythmia recurrences.

Lidocaine and azimilide are second choice drugs used when there are contraindications for Amiodarone and beta-blockers. Verapamil should be used when extrasystole originates from the His-Purkinje system. As far as the non-pharmacological adrenergic block, left cardiac sympathetic denervation has been effective in reducing the incidence of ventricular arrhythmias and sudden cardiac death in patients with malignant ventricular arrhythmias.

As a result of significant progress in electrophysiological mapping and ablation techniques, transcatheter ablation procedures have become the cornerstone of AS treatment and are now regarded as a potential lifesaving therapy (Figure 1).

The majority of the ablation procedures are carried out through an endocardial approach (left ventricular access...
with a trans-septal puncture or retrograde transaortic. For structural cardiomyopathies (e.g. coronary artery disease, dilated cardiomyopathy, right ventricular arrhythmogenic dysplasia), once determined the scar area with three-dimensional electroanatomic mapping, radiofrequencies are delivered in the areas with late potentials or ‘local abnormal ventricular activities’. This ablation strategy, aiming at modification of the arrhythmic substrate without the need for induction of the arrhythmia, has been effective and often is a forced choice when arrhythmias are haemodynamically not well tolerated.\(^5\)

A systematic review of ablation treatment in patients with AS has been conducted by Nayyar et al.\(^6\) Of the 471 patients identified, 88% underwent endocardial radiofrequency ablation; the remaining 12% had surgical ablation, ethanol ablation, electrofulguration, cervical sympathetic gangliectomy, or renal denervation. For patients undergoing endocardial radiofrequency ablation the most frequent arrhythmia (83%) was VT with a scar-dependent re-entry mechanism. In the 38 studies, reporting data on arrhythmias recurrences, 91% of the patients had successful ablation of the clinical arrhythmia. Multiple procedures were required in 19% of the patients (1.3 ± 0.4/patient).

During the last few years, the role of epicardial substrate in AS has been appraised, particularly for patients with ischaemic cardiomyopathy, and in larger groups of patients with non-ischaemic conditions, 50% of whom requires an epicardial approach. It should be emphasized that these procedures are challenging, are burden with a 4% complication rate (mainly epicardial bleeding and coronary stenoses), and that in patients with previous cardiac surgical procedures are often unsuccessful.\(^5\) Despite the treatment of patients with Brugada syndrome is typically pharmacological (antipyretics and intravenous isoprotanol), new ablation strategies have recently been suggested.

There have been instances of AS described in the contest of myocarditis treated with transcatheter ablation. Dello Russo et al.\(^8\) reported on a series of 20 patients with viral myocarditis, confirmed by biopsy, and refractory VT. In these patients, ablation was safe (no major complications during or after the procedure), and reasonably effective clinically.\(^8\)

When standard ablation procedure fails, either epicardial or endocardial, alternative procedures have been suggested such as alcoholic (ethanol) transcoryonary ablation, epicardial ablation with surgical pericardial window, or surgical cryoablation (under direct visualization and with a specialized cryoablation probe).

Mechanical circulatory support devices, such as intra-aortic balloon counterpulsation, left ventricular assist device, and extracorporeal membrane oxygenation, have been successfully used in the management of AS, essentially as ‘bridge’ to ablation procedures or transplantation.

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