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Electrical storm management in structural heart disease

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Electrical storm (ES) is a life-threatening condition characterized by at least three separate episodes of ventricular arrhythmias (VAs) over 24 h, each requiring therapeutic intervention, including implantable cardioverter defibrillator (ICD) therapies. Patients with ICDs in secondary prevention are at higher risk of ES and the most common presentation is that of scar-related monomorphic VAs. Electrical storm represents a major unfavourable prognostic marker in the history of patients with structural heart disease, with an associated two- to five-fold increase in mortality, heart transplant, and heart failure hospitalization. Early recognition and prompt treatment are crucial to improve the outcome. Yet, ES management is complex and requires a multidisciplinary approach and well-defined protocols and networks to guarantee a proper patient care. Acute phase stabilization should include a comprehensive clinical assessment, resuscitation and sedation management skills, ICD reprogramming, and acute sympathetic modulation, while the sub-acute/chronic phase requires a comprehensive heart team evaluation to define the better treatment option according to the haemodynamic and overall patient's condition and the type of VAs. Advanced anti-arrhythmic strategies, not mutually exclusive, include invasive ablation, cardiac sympathetic denervation, and, for very selected cases, stereotactic ablation. Each of these aspects, as well as the new European Society of Cardiology guidelines recommendations, will be discussed in the present review.

Definition and epidemiology

Electrical storm (ES) refers to a life-threatening condition characterized by multiple episodes of ventricular tachycardia (VT storm) or ventricular fibrillation (VF storm) within a relatively short period of time, typically 24 h. The exact definition of ES is still debated, but the most widely accepted is ≥ 3 VT/VF episodes over 24 h, each requiring therapeutic intervention [including implantable cardioverter defibrillator (ICD) therapies], or an incessant VT lasting ≥12 h. Notably, a recent analysis of various combinations of clustered VAs revealed that additional combinations, such as even two VA events within 3 months carry meaningful prognostic implications.[1]

In ICD recipients, ES incidence varies greatly according to the indication: from 4% within 2–3 years in primary prevention to 10–28% in secondary prevention, with an associated two- to five-fold mortality, heart transplant, and heart failure (HF) hospitalization increase.[2] These data mostly stem from early ICD studies, when routine programming was very aggressive, with lower VF detection rates, shorter detection times, and no anti-tachycardia pacing (ATP) therapies during capacitor charge. The Italian OBSERVO-ICD registry[3] clearly showed that this kind of set-up increases both the risk of inappropriate

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and appropriate (but potentially unnecessary) ICD interventions and of ES.

Electrical storm is mostly due to monomorphic VT (MVT): VF alone accounts for 1–21% of episodes, VF combined with VT for 3–14%, and polymorphic VT (PVT) alone 2–8%.[4] These data mostly reflect the higher prevalence of coronary artery disease (CAD, at higher risk of scar-related MVT) compared with non-ischaemic cardiomyopathies (NICMs) where PVT and VF episodes are more represented. Male gender, advanced age, reduced left ventricular ejection fraction (LVEF), MVT, class I antiarrhythmic drugs (AADs), and comorbidities increase the susceptibility to ES.[2]

Pathophysiology

The probability of ES in structural heart disease (SHD) depends on the complex interaction of different elements:

- A susceptible electrophysiological substrate: (usually myocardial scar) predisposing to macroscopic or microscopic re-entrant circuits. The circuit may be functionally favoured by myocardial disarray or increased dispersion of ventricular repolarization due to ion channel remodelling.
- Precipitating factors or triggers (rarely identified): acute myocardial ischaemia, acute decompensated HF, ongoing infections, endocrine emergencies (e.g. thyrotoxicosis), electrolyte imbalances, and poor compliance with AADs.
- Modulating factors, most notably autonomic imbalance, represented by a various combination of sympathetic hyperactivity and vagal withdrawal, typically characterizing all types of SHD and HF patients. Particularly unfavourable is the over-imposition of acute sympathetic stressors on a chronic autonomic imbalance. For instance, in the TEMPEST study,[5] the prevalence of ES was significantly higher on working days and during daytime hours.

Clinical presentation and first assessment

Electrical storm may have heterogeneous clinical presentations, depending on underlying myocardial function, VA cycle length and duration, and the presence of an ICD. In the case of ventricular dysfunction and/or fast VAs, arrhythmias may be poorly tolerated leading to syncope, haemodynamic deterioration, or cardiac arrest. Otherwise, patients may report palpitations or light-headedness or develop HF and low-output symptoms (confusion, dyspnoea, nausea and vomiting, abdominal pain). In ICD recipients, device programming is crucial in determining clinical presentation since patients may be asymptomatic if ATPs are quickly effective in terminating VAs, while repeated ICD shocks might cause worsening systolic function and psychological disorders. In the case of slow VTs below the ICD treatment threshold, clinical manifestations may replicate those of patients without ICD.

Initial assessment must include haemodynamic evaluation and advanced cardiac life support (cardioversion, resuscitation, and defibrillation) if appropriate. Simultaneously, a systematic evaluation and correction of overt reversible causes (acute myocardial ischaemia, electrolyte imbalances) is essential; K^+ should be >4 mEq/L, $Mg^{2+} > 2$ mg/dL, and total Ca^{2+} corrected for albumin should be within the normal range. Other possible triggers such as decompensated HF, hypoxia, acidosis, hyperthyroidism, toxins, or drug intake need to be appraised and specifically managed. However, reversible causes in ES are identified in only 10% of patients.

Initial risk stratification should consider the haemodynamic tolerability of the arrhythmia and the presence of pre-existing significant comorbidities. Patients at risk for cardiogenic shock or showing signs of hypoperfusion (e.g. reduced central venous oxygen saturation, increased lactates) and end-organ damage must be triaged to critical care units.[6]

Cardiac rhythm monitoring including a 12-lead ECG should be obtained as soon as possible. In case of uncertainty, wide QRS tachycardias must be treated as VT until proven otherwise. Early detection of haemodynamic changes is key to perform timely treatment adjustments; therefore, a urinary catheter, an arterial line, and a central venous catheter should be positioned promptly. Swan-Ganz catheter insertion may help in monitoring patients and guiding haemodynamic stabilization.

Echocardiography is essential for initial evaluation, overall management, and non-invasive monitoring. The assessment of aortic regurgitation and of right ventricular function is crucial to guide decisions on potential haemodynamic support devices. Close echocardiographic monitoring provides useful information on patient's haemodynamic status, for instance, by evaluating variations in the LV outflow tract velocity-time integral.

The choice on the possible acute use of inotropes, vasopressors, inodilators, or vasodilators in unstable ES patients depends on clinical presentation and multiparametric assessment and physicians should always consider the possible pro-arrhythmic effect of such drugs.

Figure 1 summarizes the overall proposed management of patients with ES and SHD.

Implantable cardioverter defibrillator interrogation and reprogramming

Implantable cardioverter defibrillator interrogation and reprogramming by expert personnel are of pivotal importance. Once confirmed that ICD interventions were appropriate, the risk of shock should be minimized, and reversible triggers should be identified. Higher VF detection rates, longer detection times (in both VT and VF zones), and ATP therapies significantly reduce the risk of appropriate but potentially avoidable ICD shocks. Accordingly, 2022 European Society of Cardiology (ESC) guidelines recommend (Class I, LOE, A) detection times $\geq$6–12 s or 30 intervals.[7]

In haemodynamically tolerated MVT, ATP success probability should be maximized by proper programming; on-demand manual ATP could be considered in centres with 24/7 available expert personnel. The likelihood of ATP success depends on the presence and duration of an excitable gap, conduction time from the pacing site to the re-entrant circuit; the presence of anatomic/functional barriers and autonomic balance. Accordingly, in the case of MVT from the basal LV in a biventricular device carrier, pacing from the LV catheter, when possible, may increase the probability to reach the circuit. Likewise, longer trains

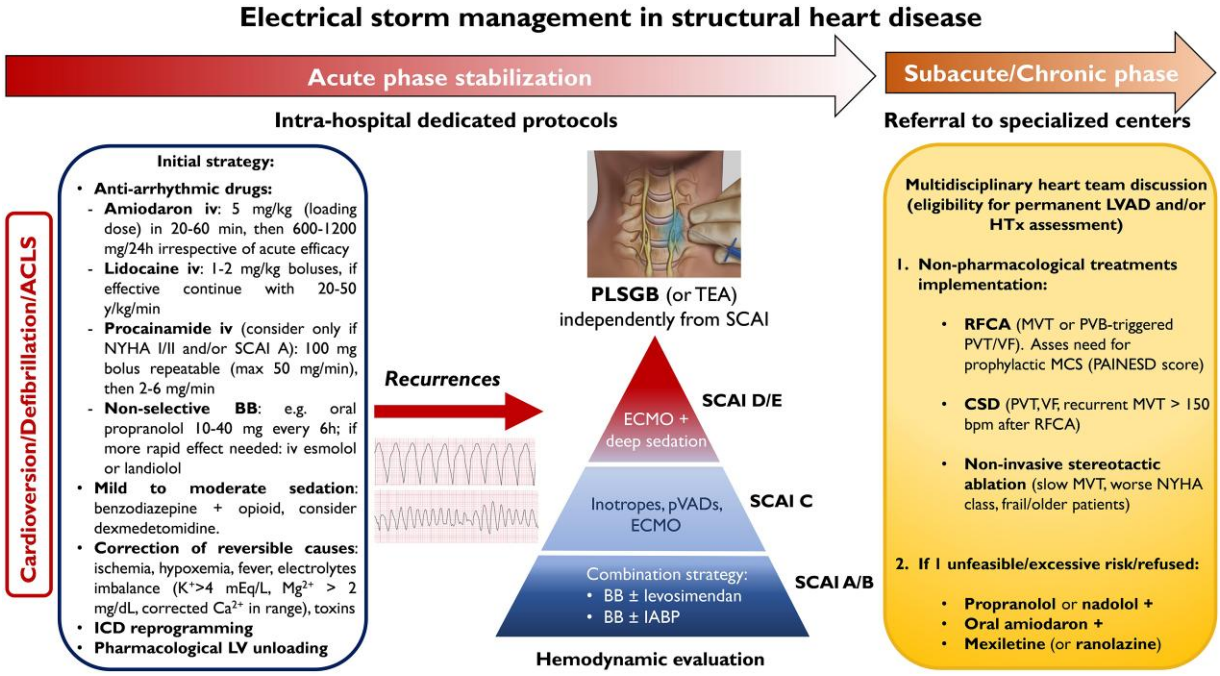

Figure 1 Proposed management of patients with electrical storm and structural heart disease. ACLS, advanced cardiovascular life support; BB, beta-blocker; CSD, cardiac sympathetic denervation; ECMO, extracorporeal membrane oxygenation; HTx, heart transplant; ICD, implantable cardioverter defibrillator; LVAD, left ventricular assist device; PLSGB, percutaneous left stellate ganglion block; pVADs, percutaneous ventricular assist devices; RFCA, radiofrequency catheter ablation; SCAI, Society for Cardiovascular Angiography and Interventions; TEA, thoracic epidural anaesthesia.

and shorter ATP cycle lengths (<84%) are more likely to enter the circuit, albeit at the expense of a higher risk of VT acceleration. Ramps also carry a higher risk of VT acceleration and should only be considered in burst non-responders (ESC 2022, Class I, LOE B).[7]

Some device algorithms or set-ups may favour VAs development. For instance, algorithms minimizing RV pacing, particularly in case of frequent premature ventricular beats (PVBs), may favour pauses and long-short-long sequences that in turn may trigger VAs (mostly PVT/VF but also MVT); even threshold tests may rarely be pro-arrhythmic, as well as LV epicardial pacing.

Anti-arrhythmic drugs

Anti-arrhythmic drugs are the cornerstone for ES management, although an impact on mortality has never been proven. Anti-arrhythmic drug choice should consider the aetiology and mechanism of VAs, the severity of the underlying cardiac dysfunction, and the potential risk of adverse, including proarrhythmic effects. Discussion of all AADs is beyond the scope of the present work; essential elements only will be highlighted.

Due to the pivotal role of adrenergic activation in the genesis and maintenance of VAs leading to ES, beta-blockers (BBs) play a central role. Unless contraindicated, they should always constitute the first-line pharmacological approach. Non-selective BBs such as propranolol or nadolol should be preferred over selective ones (Class I, LOE B, ESC 2022[7]); the former being preferable in case of significant LV dysfunction for its short half-life. This preference is justified by the fact that in SHDs, there is a chronic down-regulation of B1 receptors in favour of B2. Accordingly, in a recent Greek randomized study[8] including 60 ICD carriers, oral propranolol (160 mg/day) was significantly better than oral metoprolol (200 mg/day), both associated with i.v. amiodarone, in reducing VAs among patients presenting with ES and severe LV dysfunction. Unfortunately, i.v. propranolol is no longer available in several European countries, limiting the available i.v. BBs to metoprolol, esmolol, and landiolol (all B1 selective; landiolol being super selective: 100 times more than metoprolol and 8 times more than esmolol). The last two appear particularly useful in the case of initial haemodynamic compromise thanks to their short half-life (9 and 3-4 min, respectively).

In addition to BBs, i.v. amiodarone and/or lidocaine are the two drugs that are more commonly used in the acute phase. Amiodarone use is often limited by the hypotensive effect largely related to the solvent employed (unfortunately) in commonly used pharmaceutical formulations, but still strongly recommended in MVT storm with a high arrhythmic burden (Class I, ESC 2022[7]). Although lidocaine is particularly useful in acute myocardial ischaemia, its effectiveness can be tested in any VAs condition thanks to its very quick onset of action and neglectable negative inotropic or hypotensive effect. A randomized Spanish study, the PROCAMIO study,[9] recently compared i.v. bolus of procainamide (10 mg/kg/20 min) and amiodarone (5 mg/kg/20 min) for the treatment of haemodynamically tolerated wide QRS tachycardias in 62 patients with an average LVEF of 40%, demonstrating a lower incidence of major adverse cardiac events (9 vs. 41%, almost all represented by severe

hypotension) and a greater cardioversion rate (67% vs. 38%) at 40 min with procainamide. The lower incidence of severe hypotension was confirmed in the subgroup of patients with SHD, in which, however, the difference in efficacy was not significant, albeit with a trend in favour of procainamide. Current ESC 2022 guidelines recommend considering i.v. procainamide, in patients presenting with haemodynamically tolerated sustained MVT and known or suspected SHD (Class IIa, LOE B). Yet, procainamide should be used with caution due to its potency as both sodium and potassium channel blocker and negative inotropic effect (contraindicated in severe HF, acute myocardial infarction, and end-stage renal disease) and is not easily available in several European countries.

Following acute stabilization of the patient, oral mexiletine (particularly in case of a good acute response to lidocaine) or ranolazine can be started in association with BBs and amiodarone (unless contraindicated) while the patient is waiting for advanced VAs management, or if further strategies (VT ablation and/or denervation) are contraindicated or refused. Ranolazine for VAs is still off-label despite several small studies and a large randomized clinical trial (RCT) including 1012 ICD recipients, the RIAD trial,[10] showed its safety and moderate efficacy in preventing recurrent ICD interventions in patients with SHD. High dosages are required for an effective anti-arrhythmic activity (1000 mg b.i.d. in the RIAD trial).

Sedation

In all patients, benzodiazepines (e.g. midazolam) and short-acting opioid analgesics (e.g. remifentanil) should be considered as first-line therapy to reduce adrenergic overdrive and to control the pain and discomfort related to defibrillations without negative inotropic effect (Class I, LOE C, ESC 2022). Propofol should be used with caution due to a significant risk of negative inotropic effect; nevertheless, data support its efficacy in refractory ES.

Dexmedetomidine, a selective alpha-2 adrenoceptor agonist, has sedative, analgesic, and anti-inflammatory properties and reduces catecholamine release, prolongs ventricular refractory period, and increases vagal tone. It requires close monitoring as it may reduce blood pressure and heart rate in a dose-dependent manner.[6]

Finally, the use of deep sedation associated with mechanical ventilation should be reserved for haemodynamically unstable patients for direct suppression of VAs and as a bridge to definitive treatment (e.g. catheter ablation, coronary revascularization, therapeutic optimization) or resolution of possible reversible causes.[11]

Acute autonomic modulation

The direct interaction with cardiac sympathetic output is a powerful weapon for VAs refractory to AADs, mild sedation, and/or catheter ablation. Based on the strong pathophysiological rationale and the very promising preliminary clinical data, autonomic modulation is now recommended by 2022 European Guidelines (Class IIb, LOE B).[7] Several techniques can be used, but only two are bedside available: percutaneous left stellate ganglion block (PLSGB) at the cervical level and thoracic epidural anaesthesia (TEA).

Most of the cardiac sympathetic efferent fibres arise from preganglionic neurons located in the spinal cord from T1 to T4 and then synapse with postganglionic neurons in the paravertebral thoracic sympathetic ganglia from T1 to T4. C8 and T1 ganglia generally fuse in the stellate ganglion, which is percutaneously accessible from the base of the neck. The blockade of sympathetic fibres from the upper half of the stellate ganglion directed to the eye determines the so-called Horner's syndrome (miosis, ptosis, and enophthalmos).

Percutaneous left stellate ganglion block (PLSGB) with a local anaesthetic can be achieved using the anatomical, or anterior, approach (Moore technique), or the lateral, ultrasound-guided approach. Both can be performed by trained physicians while the patient is lying supine, with no major safety concerns despite ongoing antithrombotic therapy. The combination of a fast-acting drug (e.g. lidocaine) with a long-acting one (e.g. bupivacaine or ropivacaine) has the advantage of combining a rapid anti-arrhythmic effect onset with a longer-lasting protection, although several studies showed that the protection of PLSGB may go far beyond the half-life of the anaesthetic used, underlying the importance of the interruption of the acute vicious cycle of sympathetic activation. PLSGB was proved to be extremely effective in reducing the arrhythmic burden independently from the type of SHD and the type of VAs. Preliminary data suggest that, in case of recurrences after PLSGB, the right PSGB does not provide additional benefit,[12] while the repetition of PLSGB a second and eventually even a third time may improve the arrhythmic outcome.[13] Horner syndrome, which reflects ocular fibres block, is not related to PLSGB anti-arrhythmic efficacy.[13] PLSGB using a catheter for continuous infusion of lidocaine or another anesthetic may be particularly helpful in case there is a need to stabilize the patient for a longer timeframe.

Thoracic epidural anaesthesia provides a more extensive sympathetic block than PLSGB (bilateral and from T1 to T4) but requires anaesthesiologic skills and the lateral decubitus to be performed. Furthermore, it should not be performed in patients on dual antiplatelet and/or anticoagulation therapy and carries a non-trivial infective risk.

Mechanical circulatory support

Ventricular arrhythmias and decompensated HF form a vicious circle, each condition favouring the other; therefore, it is crucial to understand when traditional therapies are failing, and an upgraded treatment is needed. When AADs are ineffective or harmful (e.g. negative inotropic effect), the use of mechanical circulatory support (MCS) guided by haemodynamic assessment and in association with acute autonomic modulation may become necessary in pursuing haemodynamic stabilization and ensuring organ perfusion. Occasionally, haemodynamic stabilization might even restore sinus rhythm.

Multiple MCSs are available and the choice on which system to prefer should be based on patient characteristics, haemodynamic condition (SCAI classification), and centre availability and expertise.[14]

Intra-aortic balloon pump (IABP) represents a relatively simple and diffuse system with rare vascular complications but a modest increase in cardiac output (0.5–1 L/min), inversely related to heart rate. The use of IABP should be

considered in the setting of acute-on-chronic heart failure, particularly in the early stages.

Escalation to percutaneous ventricular assist devices (pVADs), such as the Impella and the TandemHeart systems, can offer greater support in more advanced clinical scenarios. The Impella can maintain a cardiac output of 2.5–5 L/min at the cost of more invasive vascular accesses while the TandemHeart unloads the LV bypassing blood from the left atrium, through a transseptal cannula, to the iliofemoral arterial system and can guarantee a flow of 3.5–5 L/min. Both devices reduce myocardial wall stress and oxygen demand.

For deteriorating or extreme cardiogenic shocks (SCAI D/E), the only device providing complete biventricular circulatory support is venous-arterial extracorporeal membrane oxygenation (VA-ECMO). In contrast with previous systems, ECMO increases afterload and myocardial wall stress. Thus, percutaneous (IABP/Impella) or invasive (LV cannulation) LV venting appears indicated.

Unfractionated heparin is required for pVADs and ECMO to maintain an activated clotting time of 250–300 s. Several complications may burden the use of pVADs such as bleeding, vascular injuries, or limb ischaemia (potentially reduced with antegrade perfusion).

Sub-acute/chronic phase

Multidisciplinary heart team discussion

Although specific recommendations in current guidelines are still lacking, once acutely stabilized, patients with moderate to severe LV dysfunction especially if aged <65, with a history of previous ablation and/or with NICM, should be referred to specialized centres with the possibility of a global patient care, including not only VAs but also HF advanced programmes. In these patients, eligibility for permanent LV assist devices and/or heart transplants should be carefully evaluated by the heart team before proceeding further in VAs management.[15]

Chronic autonomic modulation

Permanent cardiac sympathetic blockade is obtained through surgical cardiac sympathetic denervation (CSD). The procedure typically consists in the removal of the lower half of the stellate ganglion (T1), together with the sympathetic ganglia from T2 to T4. Left CSD is an established therapy for the prevention of malignant VAs in channelopathies.[16] In the last decade, the usage of CSD has been increasingly reported also for patients with SHD and refractory VAs (both PVT and MVT). In this setting, bilateral CSD demonstrated a greater efficacy than left CSD in reducing the arrhythmic burden, with a 1-year ICD shock-free survival around 50%,[17] and the modest price to pay of longer procedural times and a potential reduction in heart rate and chronotropic competence since the sinus node is primarily innervated by right-sided nerves.[18] In patients with SHD, longer arrhythmia cycle length, and NYHA ≥ 3 are independent predictors of VAs recurrences after CSD, although the patient may still reduce the arrhythmic burden. Notably, post-CSD recurrences show a significant lengthening of the arrhythmia cycle[19] that may improve the outcome of a subsequent further attempt of VT ablation, if required. Cardiac sympathetic denervation is performed through video-assisted mini-invasive thoracoscopic surgery during general anaesthesia and single-lung ventilation. According to the published data and our experience (the largest in Europe),[20] the procedure can be considered safe even in advanced patients, provided that the patient is managed by a multidisciplinary dedicated team of surgeons, cardiologists, and anaesthesiologists. The true efficacy is probably still largely underestimated at present since most of the cases reported have been performed as a bailout strategy on almost terminal patients.[21]

Radiofrequency catheter ablation

In patients with recurrent scar-related MVT, radiofrequency catheter ablation (RFCA) was superior to AAD titration in preventing recurrences.[7] Although RCTs are lacking, retrospective studies suggest a survival benefit of RFCA, particularly in patients with a history of ES.[22] An early referral lowers both recurrences and acute complication rates.[23] The effectiveness of RFCA is higher in patients with CAD, while results in patients with NICM are generally worse and highly variable across centres, possibly also because of smaller numbers and heterogeneity of both the arrhythmic substrate and the approaches used. In general, deep intramyocardial (typically septal) and/or difficult to access epicardial substrates (e.g. near to the course of coronary arteries, or in case of adhesions after cardiac surgery) are associated with a greater risk of recurrences.[24] Accordingly, in the ESC 2022 guidelines,[7] there is a Class I, LOE B recommendation for RFCA in specialized centres in patients with CAD and recurrent MVT on amiodarone, and a Class IIa, LOE C recommendation for patients with CAD and recurrent MVT on BBs or sotalol and for patients with NICM and recurrent MVT in whom AADs are ineffective, contraindicated, or not tolerated. Finally, in case of ES or incessant VT due to MVT refractory to AAD, there is a Class I, LOE B recommendation for RFCA independently from the underlying aetiology. These strong recommendations underline the importance to organize dedicated pathways for the referral of patients from spoke to hub centres for RFCA, and to implement hospital protocols (including neuromodulation) in both settings, as a stabilization bridge to possible RFCA.

In patients with SHD and a history of ES, survival free from ES recurrences after RFCA is generally good (>90% at 1 year,[25] in a single-centre large study also at 5 years[26]), although acute non-inducibility rate of any form of VT at the end of the procedure has been <80% despite extensive ablation. Persistent inducibility of any form of VT with cycle ≥ 250 ms was an independent predictor of VT recurrence;[26] therefore, in these patients, CSD could be considered before discharge.

Despite its effectiveness, RFCA in patients presenting with ES, even when performed in high-volume centres, is challenging, and is associated with an increased risk of peri-procedural morbidity and mortality compared with patients without ES. Furthermore, failure of RFCA in this group of patients portends a high risk of mortality. Mechanical circulatory support in this contest has been shown to be feasible and to allow safer and prolonged mapping and ablation of unstable VTs. Chronic obstructive pulmonary disease, age >60, CAD, NYHA class ≥ III, LVEF <25%, ES at presentation, and diabetes mellitus predict acute decompensation during RFCA in the PAINESD score. Patients with a score >15 were shown to benefit from

prophylactic haemodynamic support (compared with rescue MCS).[6] In this context, pVADs are superior to IABP, but in case of biventricular dysfunction, haemodynamic support with ECMO should be considered.[6]

Finally, RFCA should also be considered in selected cases of PVT/VF storms non-responsive to AADs, where a monomorphic PVB (often related to the Purkinje network) can be identified as a trigger and targeted for ablation (Class IIa, LOE C, ESC 2022).[7]

Stereotactic radiotherapy

Stereotactic radiotherapy (or STAR, STereotactic Arrythmias Radioablation) is a new anti-arrhythmic treatment option based on the non-invasive administration of a single fraction, high-dose radiotherapy (RT) to a relatively small cardiac volume,[27] derived from the established oncological approach. STAR may potentially overcome one of the main limitations of conventional VT ablation associated with recurrence, i.e. the accessibility to regions difficult to reach even with the latest generation tools (e.g. bipolar/needle ablation), such as deep intramural or subepicardial sites.[28] To date, clinical experiences with STAR for refractory VAs are mostly limited to case reports and case series and there is a lack of safety data, especially at the cardiac level, in the long term, as well as a full understanding of the radiobiology of cardiac effects and therefore of the optimal dose. However, preliminary results are promising. Except for one patient treated with protons,[29] all the others reported received photons-based STAR. Particle therapy, compared with photons, has the potential to further improve the safety profile of STAR by concentrating the therapeutic dose to the target while reducing off-target side effects; with last-generation active scanning particle therapy the potential to interfere with ICDs is very limited.[30] A European consortium named STOPSTORM (https://stopstorm.eu/) has been recently constituted to collect the limited experiences of each centre with the aim to expand the sample size and defined shared operational protocols for patient selection and therapy implementation, which requires a very close collaboration between radiotherapists, radiologists, and electrophysiologists. Currently, STAR should be considered as a bailout therapy for patients with MVT and a dominant clinical morphology either refractory and/or non-candidate to invasive ablation or CSD due to excessive operative risk or other contraindications.

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