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THE PRESENT AND FUTURE

JACC STATE-OF-THE-ART REVIEW

Multidisciplinary Critical Care Management of Electrical Storm

JACC State-of-the-Art Review

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ABSTRACT

Electrical storm (ES) reflects life-threatening cardiac electrical instability with 3 or more ventricular arrhythmia episodes within 24 hours. Identification of underlying arrhythmogenic cardiac substrate and reversible triggers is essential, as is interrogation and programming of an implantable cardioverter-defibrillator, if present. Medical management includes antiarrhythmic drugs, beta-adrenergic blockade, sedation, and hemodynamic support. The initial intensity of these interventions should be matched to the severity of ES using a stepped-care algorithm involving escalating treatments for higher-risk presentations or recurrent ventricular arrhythmias. Many patients with ES are considered for catheter ablation, which may require the use of temporary mechanical circulatory support. Outcomes after ES are poor, including frequent ES recurrences and deaths caused by progressive heart failure and other cardiac causes. A multidisciplinary collaborative approach to the management of ES is crucial, and evaluation for heart transplantation or palliative care is often appropriate, even for patients who survive the initial episode. (J Am Coll Cardiol 2023;81:2189–2206)

Electrical storm (ES) is a life-threatening state of cardiac electrical instability characterized by repetitive clusters of sustained ventricular arrhythmias (VAs) over a short period.[1,2] More than 80% of ES episodes are caused by monomorphic ventricular tachycardia (MMVT), but polymorphic ventricular tachycardia (PMVT) and ventricular fibrillation (VF) may cause ES.[3,4] Although VA may self-terminate, medical intervention or external defibrillation is usually required in the absence of a functioning implantable cardioverter-defibrillator (ICD). A standard clinical definition of ES is 3 or more sustained VA episodes (including appropriate ICD shocks) separated by at least 5 minutes over 24 hours, recognizing that other definitions have been reported in the literature.[1,2]

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ABBREVIATIONS AND ACRONYMS

AAD = anti-arrhythmic drug
ATP = antitachycardia pacing
CAD = coronary artery disease
CS = cardiogenic shock
ECMO = extracorporeal membrane oxygenator
ES = electrical storm
GDMT = guideline-directed medical therapy
HF = heart failure
ICD = implantable cardioverter-defibrillator
ICM = ischemic cardiomyopathy
IV = intravenous
LVAD = left ventricular assist device
LVEF = left ventricular ejection fraction
MCS = mechanical circulatory support
MI = myocardial infarction
MMVT = monomorphic ventricular tachycardia
NICM = nonischemic cardiomyopathy
PMVT = polymorphic ventricular tachycardia
PVC = premature ventricular contraction
TdP = torsades de pointes
UNOS = United Network for Organ Sharing
VA = ventricular arrhythmia
VF = ventricular fibrillation

ES may occur frequently during long-term follow-up in patients receiving an ICD for secondary prevention, up to 28% over 33 months.[3-6] The contemporary multicenter OBSERVO-ICD (Observational Registry on Long-term Outcome of ICD Patients) demonstrated an overall incidence of ES of 4.7% over a median of 39 months, with a higher risk in secondary prevention patients than primary prevention patients (10.5% vs 3.9%).[7] Most patients with ES (77%-94%) have underlying structural heart disease such as ischemic or nonischemic cardiomyopathy, although ES can occur in patients with macroscopically normal hearts.[7-9] Most patients with ES have a pre-existing ICD because of an underlying cardiomyopathy, and an ICD can abort a VA, resulting in survival to hospital admission.[9] ES risk factors include lower left ventricular ejection fraction (LVEF), older age, prolonged QRS duration, lack of appropriate guideline-directed medical therapy (GDMT), chronic kidney disease, and previous VA episodes (especially MMVT as the presenting rhythm).[5,6,9-11] ES is a risk factor for both sudden and nonsudden cardiac death and generally portends a poor outcome analogous to worsening heart failure (HF).[3,4,7-10,12,13] ES carries a worse prognosis than isolated episodes of VA, although any occurrence of VA in patients with ICDs carries high short-term and long-term mortality from both arrhythmic and nonarrhythmic causes (particularly pump failure).[3,4,7,10,12-15]

Overall, ES represents a medical emergency, foreshadows an increased risk of death, and requires a multimodality therapeutic approach typically necessitating cardiac intensive care unit admission. This *JACC* State-of-the-Art Review describes the pathophysiology, diagnostic assessment, medical management, and interventional management of ES.

HIGHLIGHTS

- ES is a life-threatening condition characterized by recurrent VAs.
- Management of patients with ES integrates antiarrhythmic drugs, beta-blockade, sedation, and catheter ablation.
- Comprehensive evidence-based treatment algorithms are needed to define best practices for managing patients with ES.

PATHOPHYSIOLOGY OF ES

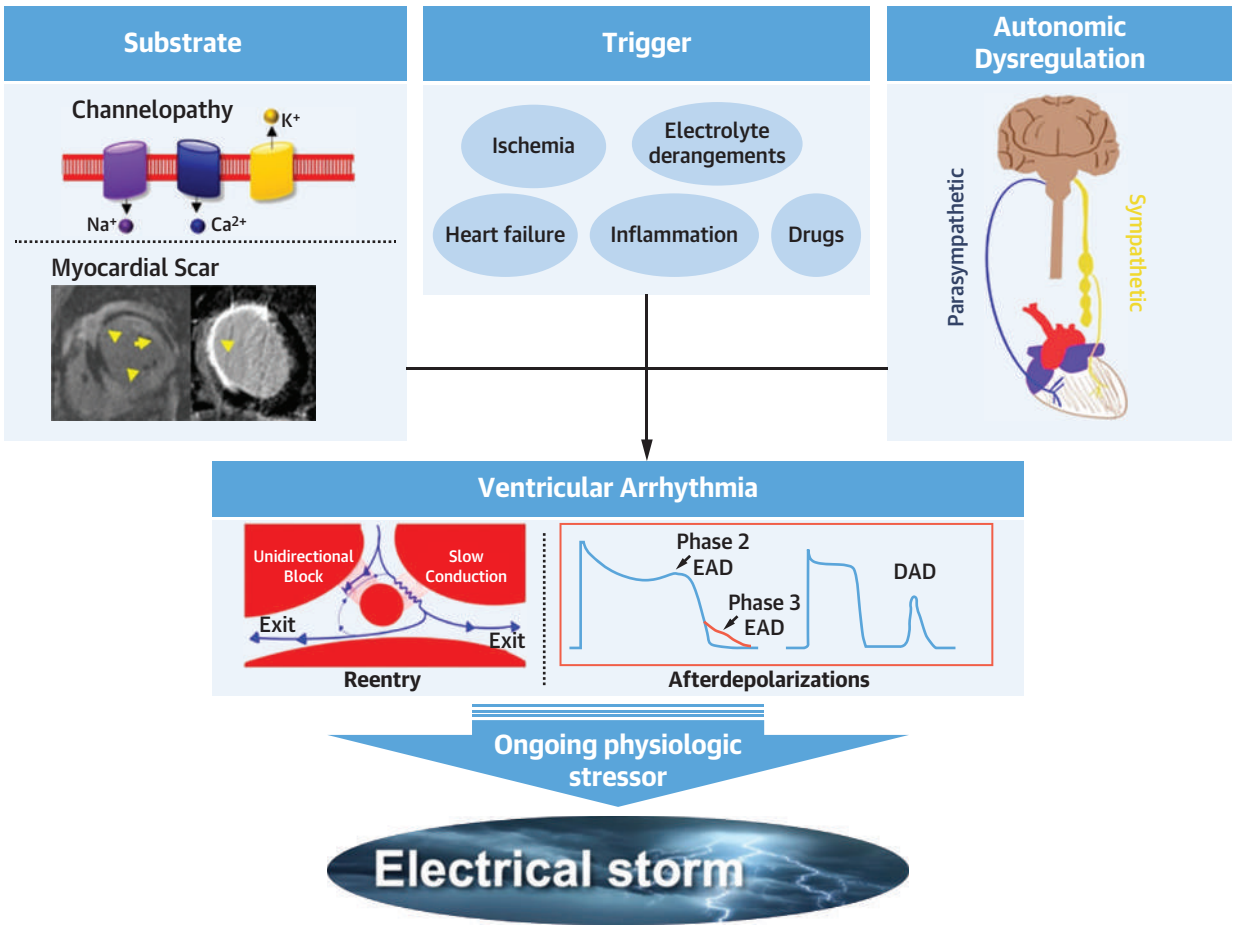
MECHANISMS OF VAs AND ES. Development of ES usually requires both an arrhythmic substrate and a proarrhythmic trigger (**Figure 1**). Most ES develops on a backdrop of either structural heart disease or pathogenic ion channel defects (channelopathies) (**Table 1**). Structural heart disease can cause arrhythmogenic remodeling with the development of myocardial scar that forms the basis for re-entry and may impair the expression and function of ion channels resulting in proarrhythmic alterations in ion handling.

The most common mechanism for VA during ES is macro-re-entry caused by slow conduction through surviving tissue channels in scar resulting in MMVT.[1,16] Additionally, micro-re-entry can result from focal inflammation or interstitial fibrosis and functional re-entry can occur via heterogeneously impaired excitability and decreased repolarization reserve, which may be augmented by myocardial stretch and elevated sympathetic tone during decompensated HF.[1,16-18] Re-entry requires an area of anatomic or functional conduction block, an electrical pathway with unidirectional block, and a pathway with slow or heterogeneous conduction. This substrate is present in patients with myocardial scar from a prior myocardial infarction (MI), interstitial fibrosis from dilated nonischemic cardiomyopathy (NICM), focal inflammation, or infiltrative cardiomyopathy.[16]

Less commonly, ES is secondary to triggered activity from early afterdepolarization or delayed afterdepolarization. Primarily driven by a reduction in repolarization, early afterdepolarizations are the primary mechanisms for PMVT and torsades de pointes (TdP) in congenital or acquired long QT syndrome.[1] Delayed afterdepolarizations are secondary to increased intracellular calcium concentrations from sarcoplasmic reticulum calcium release and can occur in myocardial ischemia, digoxin toxicity, excessive beta-adrenergic stimulation, or catecholaminergic PMVT.[1,17,19]

Re-entry and triggered activity are not mutually exclusive for ventricular arrhythmogenesis, and patients typically have an underlying myocardial substrate allowing re-entry with VA initiated acutely by triggered activity from ischemia, electrolyte abnormalities, or drug toxicity.[1,2] Sympathetic activation decreases the VA threshold by increasing

FIGURE 1 **Mechanisms of Arrhythmogenesis in Electrical Storm**



Triggers such as myocardial ischemia, inflammation, or hemodynamic decompensation, as well as drug and electrolyte effects, often with accompanying autonomic nervous system imbalance, can lead to sustained ventricular arrhythmia caused by re-entry and/or afterdepolarizations in those with vulnerable anatomic or electrical substrates, including myocardial scar promoting re-entry, early afterdepolarizations (EADs) from long QT syndrome, or delayed afterdepolarizations (DADs) from myocardial ischemia. Perpetuation of the inciting trigger and the resulting sympathetic nervous system response leads to recurrent ventricular arrhythmia and electrical storm (ES).

afterdepolarizations and causing dispersion of action potential duration (heterogeneity of repolarization) in myocardial tissue allowing for higher susceptibility to VA.[17] In diseased myocardium, a triggered short-coupled premature ventricular contraction (PVC) often initiates re-entrant VT or VF from conduction block in one limb of the re-entry circuit and slow conduction in the other limb.

SUBSTRATES FOR ES

STRUCTURAL HEART DISEASE. Patients with chronic infarcts and ischemic cardiomyopathy (ICM) most frequently present with MMVT caused by re-entry through subendocardial scar (Figure 2).[1] Less commonly, PVCs arising from Purkinje fibers in the scar border zone cause PMVT and VF.[1] Acute myocardial ischemia (and reperfusion itself) can be arrhythmogenic through several mechanisms, including functional re-entry from myocardial tissue repolarization heterogeneity and depolarization of the ischemic tissue as well as triggered activity from delayed afterdepolarizations.[1]

Patients with NICM, particularly inherited cardiomyopathy syndromes, may have distinct scar regions that facilitate re-entrant VT or trigger VF.[16] Certain NICM substrates have a higher propensity for VA and ES, including arrhythmogenic (right ventricular)

TABLE 1 Structural and Electrical Substrates Predisposing to Electrical Storm, With Associated Disease-Targeted Therapies[1,2]

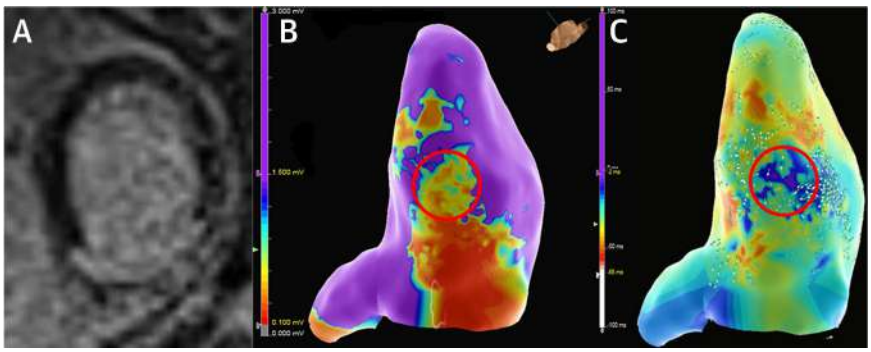
Substrate	Triggers for ES	Disease-Targeted Therapy
Structural heart disease		
ICM	Acute ischemia Sympathetic tone Decompensated HF	Revascularization if indicated Catheter ablation
NICM	Sympathetic tone Decompensated HF	Hemodynamic support Consider catheter ablation
Arrhythmogenic cardiomyopathy	Sympathetic tone	Catheter ablation
Cardiac sarcoidosis	Granulomatous inflammation Initiation of immune suppression (occasionally)	Immune suppression if active inflammation Catheter ablation
Chagas disease	Inflammation	Antitrypanosomal therapy for active infection Autonomic modulation Catheter ablation
Viral or lymphocytic myocarditis	Inflammation	Immune suppressive therapy for selected patients Hemodynamic support
Giant cell myocarditis	Inflammation	Immune suppressive therapy Hemodynamic support
Conduction defects (channelopathies)		
Congenital long QT syndrome	QT-prolonging agents Sympathetic tone	Avoid QT-prolonging agents Beta-blockers Atrial pacing Autonomic modulation
Acquired long QT syndrome	QT-prolonging agents Bradycardia	Avoid QT-prolonging agents IV magnesium Atrial pacing Isoproterenol
CPVT	Sympathetic tone ICD shocks	Beta-blockers Flecainide Autonomic modulation
Brugada syndrome	Parasympathetic tone Fever Excessive alcohol intake	Avoid sodium channel blockers Avoid provoking drugs/conditions Isoproterenol or quinidine Consider catheter ablation
Early repolarization syndrome or idiopathic VF	Parasympathetic tone	Isoproterenol or quinidine Consider catheter ablation for PVC triggers
Short QT syndrome	Parasympathetic tone	Isoproterenol or quinidine
Idiopathic or short-coupled VF	Parasympathetic tone	IV verapamil Isoproterenol or quinidine Consider catheter ablation for PVC triggers
Idiopathic (outflow tract) VT	Sympathetic tone	Beta-blockers or verapamil

CPVT = catecholaminergic polymorphic ventricular tachycardia; ES = electrical storm; HF = heart failure; ICD = implantable cardioverter-defibrillator; ICM = ischemic cardiomyopathy; IV = intravenous; NICM = nonischemic cardiomyopathy; PVC = premature ventricular contraction; VF = ventricular fibrillation.

cardiomyopathy, cardiac sarcoidosis, and Chagas cardiomyopathy.[20-22] In these diseases, acute myocardial injury/inflammation and chronic myocardial scarring can cause re-entrant MMVT or PVC-mediated VF. Acute (fulminant) myocarditis, especially giant-cell myocarditis, can provoke ES. Nearly one-half of patients requiring venoarterial extracorporeal membrane oxygenator (ECMO) support for fulminant giant-cell myocarditis may present with ES.[23,24] In cardiac sarcoidosis and acute fulminant myocarditis (particularly giant cell), immunosuppression is essential, recognizing that initiation of immunosuppression can trigger VA.[1,20,23,24]

STRUCTURALLY NORMAL HEARTS. Idiopathic MMVT arising in patients with structurally normal hearts, including outflow tract VT and fascicular VT, is typically considered benign but may rarely result in ES.[1] Inherited or acquired alterations in ion channels and transporters causing impaired depolarization and repolarization can provoke VA despite a structurally normal heart.[25,26] Ion channel mutations and medications that impair repolarizing currents or augment depolarizing currents can prolong the QT interval and predispose to bradycardia-dependent TdP caused by triggered early afterdepolarizations.[26] Idiopathic VF can be initiated by a short-coupled triggering PVC (often with the PVC arising from structures dense with Purkinje fibers such as the moderator band and papillary muscle), often resulting in ES.[27,28] Malignant early repolarization syndromes can be associated with VF, but ES is rare.[25] Specific antiarrhythmic drugs (AADs) and other therapies to use or avoid in

FIGURE 2 Substrate for Ventricular Tachycardia During Electrical Storm



A patient with an ischemic cardiomyopathy presenting with electrical storm (ES) secondary to inferior myocardial infarction, resulting in inferior left ventricular scar as shown by **(A)** late gadolinium enhancement on cardiac magnetic resonance. **(B)** The imaging correlated with areas of low voltage **(red circle)** on electroanatomical mapping during ventricular tachycardia ablation in the inferior left ventricle. **(C)** Activation mapping in sinus rhythm demonstrated areas of late activation and slow conduction **(red circle)** that corresponded to the area of scar. VA = ventricular arrhythmia.

the setting of VA have been identified for many of these conditions (Table 1).[26]

DIAGNOSTIC ASSESSMENT AND RISK STRATIFICATION. The initial diagnostic evaluation for ES patients focuses on evaluating the clinical context, arrhythmia characteristics, triggers, cardiac substrate, hemodynamic state, and risk profile (Figure 3).

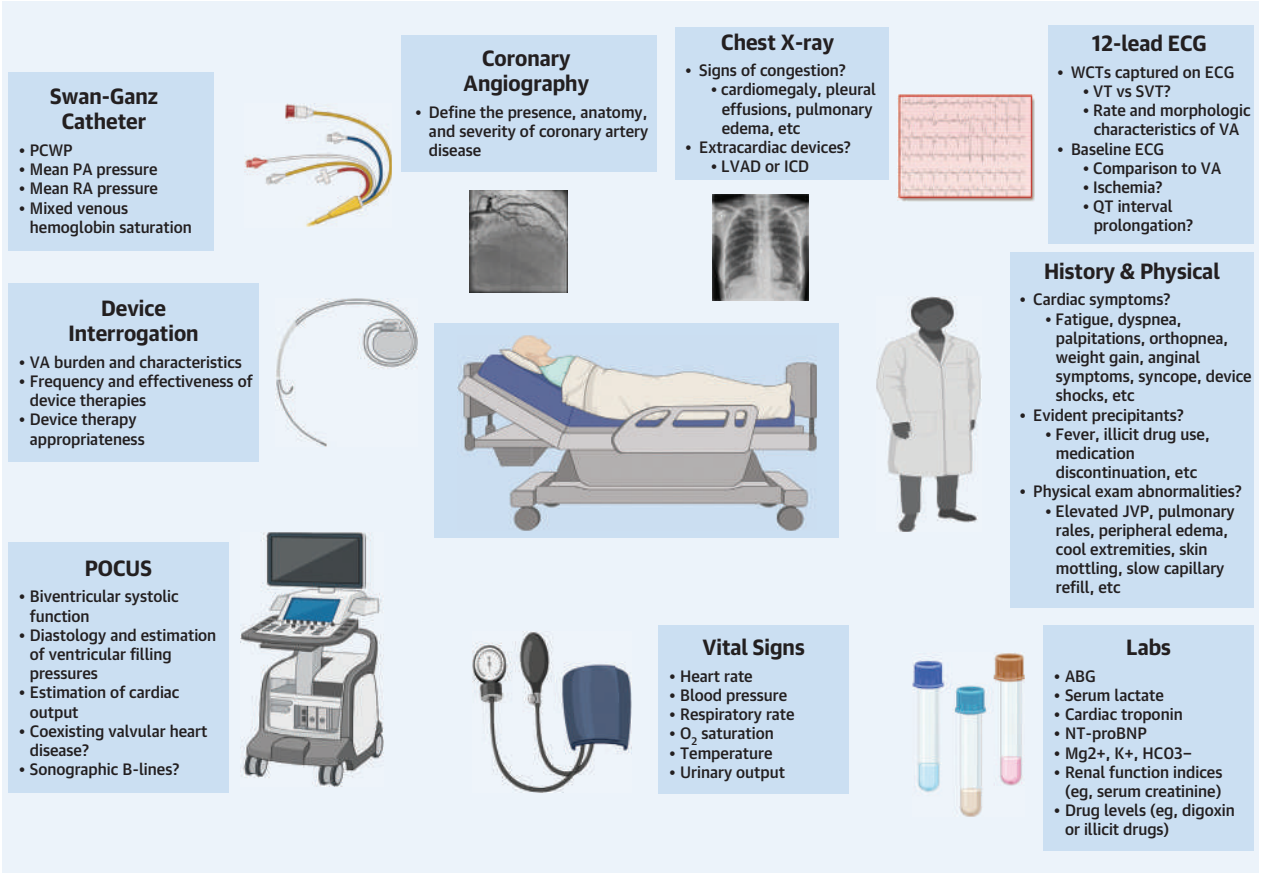
CLINICAL CONTEXT. A crucial distinction between ES patients with and without an ICD is that those with ICDs often present in a comparatively stable state, whereas patients without an ICD are often critically ill after cardiac arrest, necessitating standard advanced cardiac life support measures.[2] The patient's history of VA and prior therapies including AADs or catheter ablation can predict the likelihood of treatment success. In our experience, patients who are naïve to AADs may have a more favorable response to medical therapy, whereas those presenting with ES despite long-term AAD therapy are more likely to require catheter ablation and patients with drug-refractory VA despite prior ablation attempts are less likely to respond to repeat catheter ablation.

ARRHYTHMIA CHARACTERISTICS. A 12-lead electrocardiogram should be obtained during both the native rhythm and VT if possible, and initiation of continuous (ideally 12-lead) electrocardiographic monitoring is invaluable.[1] Information regarding the frequency and duration of VA episodes from ICD interrogation and cardiac telemetry (when available) is important for risk stratification. Higher-risk VA features that may justify a more aggressive initial strategy include VF/PMVT, faster ventricular rate, more frequent or incessant VA episodes, a tendency to degenerate to VF, failure of ICD therapies, and VA triggered by short-coupled PVCs.

IDENTIFICATION OF TRIGGERS. Reversible triggers are identified in a minority of ES patients, including myocardial ischemia, worsening HF or volume overload causing myocardial stretch, infection, medication changes causing drug toxicity or QT prolongation, imbalances in autonomic activity, noncardiac organ failure, thyrotoxicosis, and electrolyte derangements (particularly hypokalemia and hypomagnesemia).[2,3,11] Discontinuation of offending proarrhythmic drugs is important.[2] Elevated sympathetic tone and adrenergic excess often drive ES.[17,19] The initial laboratory evaluation to identify triggers includes serum electrolytes, lactate, kidney/liver/thyroid function, and cardiac biomarkers.[2]

CARDIAC SUBSTRATE EVALUATION. It is essential to exclude myocardial ischemia as a trigger for ES, particularly for patients with established coronary artery disease (CAD) or ICM. A coronary angiogram is often indicated to identify obstructive CAD even for patients without clear evidence of acute MI.[1] Computed tomography coronary angiography can be considered in selected stable patients when the clinical suspicion for CAD is low or to exclude a coronary anomaly.[1] Echocardiography is a first-line test to evaluate the underlying cardiac substrate by identifying structural heart disease and performing noninvasive hemodynamic assessment.[1] Cardiac magnetic resonance or positron emission tomography can identify occult structural heart disease or active

FIGURE 3 Diagnostic and Clinical Assessment for Patients With Electrical Storm



Evaluation of the patient with ES incorporates history and physical examination, laboratory analysis, cardiac diagnostic testing, and hemodynamic assessment. A comprehensive assessment is essential to identify triggers, understand the cardiac substrate, and recognize hemodynamic decompensation. ABG = arterial blood gas; ECG = electrocardiogram; ICD = implantable cardioverter-defibrillator; JVP = jugular venous pressure; LVAD = left ventricular assist device; NT-proBNP = N-terminal pro-B-type natriuretic peptide; PA = pulmonary artery; PCWP = pulmonary capillary wedge pressure; POCUS = point-of-care ultrasound; RA = right atrial; SVT = supraventricular tachycardia; VA = ventricular arrhythmia; VT = ventricular tachycardia; WCT = wide complex tachycardia.

myocardial inflammation.[20] Acute myocarditis with ES is a potential indication for endomyocardial biopsy to identify giant-cell myocarditis.[24]

HEMODYNAMIC ASSESSMENT. ES can result from or herald hemodynamic destabilization in patients with cardiomyopathy.[3,13] Identifying coexisting decompensated HF or cardiogenic shock (CS) is crucial during the evaluation of ES and may impair the ability to tolerate recurrent VA or standard ES therapies. Initial clinical evaluation should include assessment of perfusion and volume overload, potentially supplemented by invasive hemodynamic measurements. New, worsening, or severe ventricular dysfunction suggests that progressive cardiomyopathy may be the primary driver of ES, and evidence of advanced HF should be sought.[29]

ICD MANAGEMENT DURING ES

IMPORTANCE OF ICD IN PATIENTS WITH ES. The presence of an ICD substantially mitigates the risk of arrhythmic death during ES.[1,2] Most patients with ES have an ICD, making appropriate management of the ICD during ES crucial to patient care.[3,5-8,11,14] In ICD patients, ES can trigger multiple ICD therapies including antitachycardia pacing (ATP) or ICD shocks depending on the device programming and the VA rate.[7] ICD shocks can further exacerbate ES by provoking pain and emotional distress (including anxiety, depression, phantom shocks, and post-traumatic stress disorder), which stimulate sympathetic drive and increase the risk of subsequent VA episodes, potentially leading to a vicious cycle of recurrent VA and ICD therapies.[2,17,30] An ICD that can successfully

terminate VT using ATP is an important protective factor during ES, and patients without an ICD or whose ICD is ineffective for terminating VA (or requires multiple shocks to succeed) are at higher risk and require a more aggressive initial approach to therapy.[7]

IMMEDIATE MANAGEMENT. The immediate goal of treatment in ICD patients with ES is to avoid or minimize repetitive, ineffective, or inappropriate shocks (including those for hemodynamically tolerated or nonsustained VA). This can be achieved acutely by ICD reprogramming or applying a magnet over the ICD, which suspends VA detection and therapies while maintaining the pacing function.[2] If the clinical VT is below the ICD detection rate, ICD therapies can be manually administered through the device to terminate the VT as appropriate. It is advisable to place external defibrillator pads on all patients (including those with a functioning ICD) for external cardioversion if ICD therapies do not terminate the VT.

ICD interrogation. ICD interrogation can quantify the frequency of VA and ICD therapies, determine whether the ICD therapies were appropriate, assess the VT morphology, identify the mechanism of initiation, recognize failure to abort VA episodes, and allow reprogramming to optimize detection and treatment of VA. If the VT rate is below the ICD therapy zones, the rate cutoffs can be adjusted. Contemporary ICDs allow for programming different therapies into 2 or more zones based on rate, allowing ICD therapies to be tailored to the observed VA with the goal of avoiding ICD shocks for slower VA.

APPROPRIATE VS INAPPROPRIATE THERAPIES. It is crucial to determine whether the ICD therapies are appropriate (ie, for sustained VA). Inappropriate ICD therapies include those administered for arrhythmias such as nonsustained VT or supraventricular tachycardia (eg, atrial fibrillation with rapid ventricular rate), or those caused by artifactual signals such as T-wave oversensing or lead noise (eg, lead fracture).[31] Programming strategies using supraventricular tachycardia discriminators can decrease the incidence of inappropriate ICD shocks.[31]

ICD REPROGRAMMING TO PREVENT ICD SHOCKS. If the VT is well tolerated without significant symptoms or hemodynamic compromise, the ICD can be reprogrammed by increasing the detection time or programming an ATP-only zone to prevent shocks. ATP is effective for shock-free termination of VT in approximately three-quarters of MMVT episodes.[32] ATP can be potentially optimized to improve MMVT termination by:

1. Increasing the number of cycles of ATP therapies;
2. Increasing the number of bursts per ATP cycle;
3. Decreasing the ATP cycle length (ie, a lower percentage of VT cycle length);
4. Decreasing the ATP coupling interval with every subsequent burst ("scan" programming); or
5. Progressively decreasing the R-R interval during an individual ATP burst ("ramp" programming).

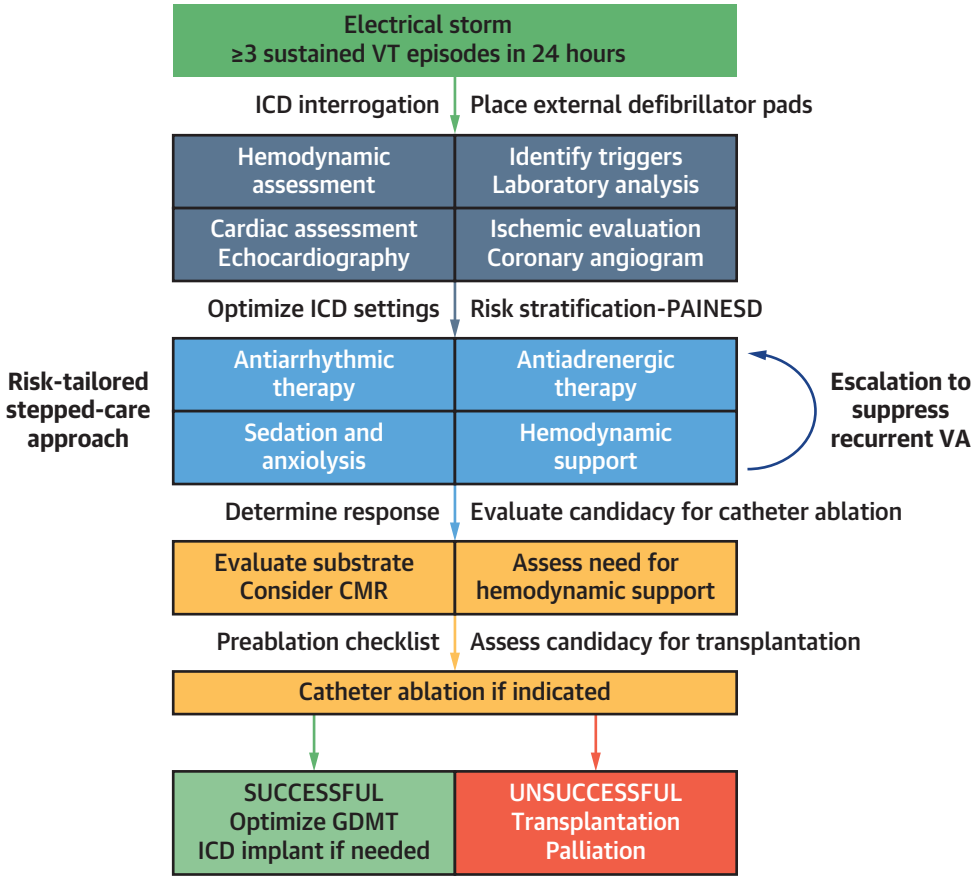
Device proarrhythmia. Whereas an ICD can effectively treat VA, there is an inverse relationship between the aggressiveness and safety of ATP, and ICD therapies can be proarrhythmic.[33] More aggressive ATP therapies (shorter coupling intervals, ramp ATP, more ATP attempts, more bursts per attempt) can be more effective at VT termination but may risk accelerating a well-tolerated VT or degenerate VT into VF. Acceleration of VT is more likely with shorter or variable cycle lengths of VT and less likely in the presence of AADs.[33] Low-energy ICD shocks can potentially lead to VT acceleration or degeneration to VF, making it important to program backup high-energy ICD shocks following ATP or low-energy ICD shocks.[33] Cardiac resynchronization therapy may trigger ES immediately after implantation by provoking re-entry via LV pacing, and this can be ameliorated by turning off the LV lead or changing to backup VVI pacing mode may be appropriate, followed by lead revision.[34]

DEVICE IMPLANTATION OR UPGRADE. ES survivors without an ICD should generally receive a secondary-prevention ICD during hospitalization, whereas an upgrade to cardiac resynchronization therapy (if indicated) may reduce the subsequent risk of VA in responders.[1,2,34] Placing an ICD during ES can provoke multiple ICD shocks, so this is typically considered only at the time of hospital discharge. ICD implantation is contraindicated for incessant VA or when the patient has advanced HF, unless they are being bridged to transplant or left ventricular assist device (LVAD).[1,2] Complete deactivation of ICD therapies is justified for patients pursuing a palliative care approach.[2]

ACUTE MEDICAL MANAGEMENT

GENERAL PRINCIPLES. ES spans a spectrum of acuity and associated risk, necessitating a flexible management strategy tailored to the severity of the presentation. The core elements of medical management for ES include AADs, beta-adrenergic blockade, sedation, and hemodynamic support, with individual treatments ranging in potential efficacy, invasiveness, and risk of complications (**Central Illustration**).

CENTRAL ILLUSTRATION **Global Approach to the Evaluation and Management of Electrical Storm**



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Comprehensive management of electrical storm includes integrated diagnostic assessment, risk stratification, stepped-care medical therapy, and catheter ablation. Escalation of treatment intensity is needed for higher-risk presentations or recurrent ventricular arrhythmias (VAs). Patients who fail or are not candidates for catheter ablation may be considered for heart transplant or palliation. CMR = cardiac magnetic resonance; GDMT = guideline-directed medical therapy; ICD = implantable cardioverter-defibrillator; PAINESD = Pulmonary Disease, Age >60 Years, Ischemic cardiomyopathy, NYHA Functional Class III-IV, Ejection Fraction <25%, VT Storm, Diabetes; VA = ventricular arrhythmia; VT = ventricular tachycardia.

The European Society of Cardiology guideline recommendations for the management of ES are summarized in Table 2, recognizing that development of a comprehensive evidence-based treatment algorithm for ES remains a crucial unmet need.[2]

The most effective intervention for acute termination of VA is synchronized electrical cardioversion (for MMVT) or unsynchronized defibrillation (for PMVT or VF), either externally or using an existing ICD.[1,2] Immediate electrical cardioversion or defibrillation is always preferred for patients with hemodynamically unstable VA and is appropriate for hemodynamically stable VT when the risk of sedation is low.[1,2]

AADs. AADs targeting cardiomyocyte ion channels can achieve chemical cardioversion, facilitate the success of electrical cardioversion or ATP, and reduce the risk of VA recurrence.[1,2] AADs commonly used for typical ES caused by MMVT in patients with structural heart disease include Class I AADs (eg, lidocaine and procainamide) that inhibit VT by reducing electrical excitability and slowing conduction and Class III AADs (eg, sotalol and amiodarone) that inhibit VT by prolonging the refractory period and hindering

re-entry.[1,2] It is essential to recognize that these standard AADs may be ineffective or even harmful in less common causes of ES, particularly channelopathies, and alternative first-line AADs for these conditions are shown in Table 1.

Intravenous (IV) amiodarone, lidocaine, procainamide, and sotalol can effect acute termination of MMVT in patients with structural heart disease, each having important strengths and limitations.[1,2] The acute efficacy for VT termination appears to be greatest for procainamide, intermediate for amiodarone and sotalol, and lowest for lidocaine.[1,2,35-38] Due to its higher acute efficacy, procainamide carries a Class IIa recommendation for acute termination of MMVT (particularly hemodynamically stable MMVT), compared with a Class IIb recommendation for amiodarone or sotalol.[1,2] However, procainamide is contraindicated in severe structural heart disease, decompensated HF, acute MI, and advanced kidney disease, all of which are common in ES populations. Therefore, IV amiodarone is generally preferred in most patients with ES caused by structural heart disease, particularly for facilitating ATP or electrical cardioversion and for preventing recurrent VT.[2] AADs can cause dose- and infusion rate-dependent hypotension via vasodilation from alpha-1 blockade, negative inotropy via beta-1 blockade, and other mechanisms; the incidence of hypotension appears lowest with lidocaine.[35-38]

After acute termination of VT, it is logical to continue AAD therapy to prevent recurrence, recognizing that an AAD that was not effective for termination of VT may still prevent VT recurrence. Amiodarone IV is recommended as the first-line AAD in typical patients with ES caused by structural heart disease based on its greater efficacy for preventing recurrent VT and suppressing VT refractory to other AADs.[2,35-37] The antiarrhythmic efficacy and receptor-binding profile of amiodarone differs with IV and oral administration, and accumulation of an active metabolite during oral amiodarone loading increases its efficacy.[39] When chronic amiodarone is appropriate for preventing recurrent VT, IV amiodarone is typically continued during initial oral amiodarone loading (eg, 800-1,600 mg/d up to a total of 10-20 g) until the patient has been free from VA for ≥48 hours.[1,2,39] Sotalol can occasionally be substituted when amiodarone is not desired because of concerns about long-term toxicity (eg, younger patients who are naïve to AADs).[1,2] To avoid proarrhythmia, sotalol and procainamide should only be considered when the baseline QT interval is not prolonged; serum potassium and magnesium are normal; kidney function is not severely impaired; and the patient is not concomitantly receiving QT-prolonging drugs (eg, amiodarone).

When amiodarone is ineffective as monotherapy or for higher-risk presentations, lidocaine is often added as a second-line AAD to suppress VA during further amiodarone loading. Lidocaine is more effective in ischemic myocardium and can be safely combined with QT-prolonging drugs but can accumulate during decompensated HF or CS. Procainamide can synergistically block sodium channels and suppress VT but is usually a third-line AAD in ES because of its potential toxicity, risk of accumulation of the active QT-prolonging metabolite N-acetylprocainamide with kidney dysfunction, and lack of an oral equivalent (although quinidine may be substituted).[38] Due to additive QT prolongation, amiodarone is typically discontinued when procainamide is added; however, adding procainamide at a low dose to ongoing amiodarone therapy can be considered in selected patients with close monitoring. Serum drug concentration monitoring is necessary for patients receiving lidocaine or procainamide, along with QTc monitoring for patients receiving QT-prolonging AADs. Quinidine blocks the transient outward potassium current (I_{TO}) and may be effective for suppressing VA in Brugada syndrome and other inherited arrhythmia syndromes, as well as for selected patients with VA that are refractory to other AADs (particularly for suppressing

TABLE 2 ESC Guideline Recommendations for Electrical Storm[2]

Class I Recommendations
Mild to moderate sedation is recommended in patients with ES to alleviate psychological distress and reduce sympathetic tone (LOE: C)
Antiarrhythmic therapy with beta-blockers (nonselective preferred) in combination with intravenous amiodarone is recommended in patients with structural heart disease and ES unless contraindicated (LOE: B)
IV magnesium with supplementation of potassium is recommended in patients with TdP (LOE: C)
Isoproterenol or transvenous pacing to increase heart rate is recommended in patients with acquired long QT syndrome and recurrent TdP despite correction of precipitating conditions and magnesium (LOE: C)
Catheter ablation is recommended in patients presenting with incessant VT or ES caused by MMVT refractory to AADs (LOE: B)
Class IIa Recommendations
Deep sedation/intubation should be considered in patients with an intractable ES refractory to drug treatment (LOE: C)
Catheter ablation should be considered in patients with recurrent episodes of PMVT/VF triggered by a similar PVC, nonresponsive to medical treatment or coronary revascularization (LOE: C)
Class IIb Recommendations
Quinidine may be considered in patients with CAD and ES caused by recurrent PMVT when other AAD therapy fails (LOE: C)
Autonomic modulation may be considered in patients with ES refractory to drug treatment and in whom catheter ablation is ineffective or not possible (LOE: C)
Institution of mechanical circulatory support may be considered in the management of drug-refractory ES and cardiogenic shock (LOE: C)

AAD = antiarrhythmic drug; CAD = coronary artery disease; ESC = European Society of Cardiology; LOE = Level of Evidence; MMVT = monomorphic ventricular tachycardia; PMVT = polymorphic ventricular tachycardia; TdP = torsades des pointes; VT = ventricular tachycardia; other abbreviations as in Table 1.

TABLE 3 MCS Devices and Their Role in Catheter Ablation for Electrical Storm[46]

Modality	Function	Advantages	Disadvantages
IABP	Inflation in diastole increases coronary perfusion; deflation during systole reduces afterload	Easy to place and widely available Lower risk of complications Unloads LV by reducing afterload	Primarily effective in sinus rhythm Ineffective at higher heart rates or during VA Contraindicated in patients with AI (as with all other devices) Modest hemodynamic support
Impella	A continuous-flow pump placed across the AV provides LV unloading and augments cardiac output	Relatively easy to place and widely available Significant increase in cardiac output Some support during VA Directly unloads LV	Contraindicated in mechanical AV (must cross aortic valve) Crowded LVOT limits retro-aortic approach to ablation Can cause ventricular ectopy More complications than IABP
TandemHeart	Arterial bypass system with transseptal LA access and external pump	Full cardiac output support Addition of oxygenation circuit is possible Some support during VA	Not widely available Requires transseptal access (leaving a residual ASD) Large arterial and venous sheaths with risk of vascular complications
Venoarterial ECMO	Portable complete cardiopulmonary bypass	Full biventricular cardiac and pulmonary support Support during VA or cardiac arrest Can be placed at bedside without fluoroscopy	Large arterial and venous sheaths with risk of vascular complications Thromboembolic and bleeding risks Standard configurations increase LV afterload

AI = aortic insufficiency; ASD = atrial septal defect; AV = aortic valve; ECMO = extracorporeal membrane oxygenation; ES = electrical storm; IABP = intra-aortic balloon pump; LA = left atrial; LV = left ventricle; LVOT = left ventricular outflow tract; VA = ventricular arrhythmia.

PVCs that trigger PMVT or VF).[1,2,26] IV magnesium sulfate is recommended for TdP, even when the serum magnesium level is normal, and increasing the heart rate via transvenous pacing or isoproterenol is indicated for bradycardia-dependent TdP.[1,2]

ADRENERGIC BLOCKADE. Recurrent VT in ES is often promoted by stimulation of cardiac beta-adrenergic receptors, and beta-adrenergic blockade is a crucial component of ES management (especially in the setting of acute myocardial ischemia).[1,2,17,19] Sotalol and amiodarone (particularly IV amiodarone) have beta-blocking properties, but adding another beta-blocker can enhance their efficacy.[35,36] Adding or up-titrating a GDMT beta-blocker (eg, metoprolol succinate, bisoprolol, carvedilol) can be considered, although the alpha-1 blockade produced by carvedilol often causes dose-limiting hypotension.[1,40] Whereas the beta-1 blocker metoprolol tartrate can be initiated and rapidly titrated for patients who are beta-blocker naïve (particularly for amelioration of myocardial ischemia), nonselective beta-1/2 blockers (eg, propranolol) are preferred during ES.[1,2] Propranolol should be considered for patients with ES, either as initial therapy or when a beta-1 blocker is ineffective.[2,41] In the first randomized controlled trial comparing pharmacologic regimens in ES, propranolol (160 mg/d) displayed better efficacy than metoprolol tartrate (200 mg/d) in preventing recurrent VT in 60 ICD patients with ES receiving IV amiodarone, including higher freedom from VA at 24 hours (47% vs 10%).[41] Theoretical advantages of propranolol over metoprolol include blockade of both beta-1 and beta-2 adrenergic receptors, strong central nervous system penetration that may reduce sympathetic outflow, more comprehensive beta-receptor inhibition via inverse agonism, and mild sodium channel blockade at very high doses.[41] An ultrashort-acting IV beta-1 blocker (eg, esmolol or landiolol) can be added to oral beta-blockers as a second-tier therapy in ES, having the advantage of rapid onset and easy up-titration with quick offset in case of hypotension.[2,19]

When beta-blockers are ineffective or not tolerated for suppressing recurrent VA, inhibition of cardiac sympathetic innervation via percutaneous cervical sympathetic (stellate) ganglion blockade is potentially beneficial.[2,19,42] Stellate ganglion blockade can be performed quickly and easily at bedside by providers with expertise in ultrasound-guided jugular venous access.[42] Left-sided stellate ganglion blockade is performed first, with bilateral stellate ganglion blockade reserved for intubated patients because of the potential risk of phrenic nerve paresis that could compromise respiration.[42] Surgical cardiac sympathetic denervation may be considered for refractory ES in patients who respond favorably to stellate ganglion blockade.[1]

SEDATION. The risk of further VAs triggered by anxiety and post-traumatic stress can be mitigated by appropriate sedation/anxiolysis to reduce central sympathetic outflow.[2,30] Benzodiazepines are first-line drugs for anxiolysis and can induce amnesia surrounding cardioversion or ICD shocks. Opioids are indicated to treat pain from electrical therapies or chest compressions. Dexmedetomidine is a second-line IV sedative medication that exerts specific anti-adrenergic effects by decreasing central sympathetic

outflow via alpha-2 receptor activation, potentially resulting in a reduced risk of tachyarrhythmias; low doses of dexmedetomidine generally do not cause respiratory depression.[43] For severe or refractory ES, endotracheal intubation with general anesthesia (eg, using propofol) may prevent recurrent VT and mitigate the traumatic experience of repeated defibrillations but can worsen hemodynamic instability.[2,44] Initiating dexmedetomidine prior to weaning general anesthesia may reduce the risk of rebound sympathetic activation.

HEMODYNAMIC SUPPORT. Medical treatments for ES have vasodilatory, negative inotropic, and negative chronotropic effects that can cause hypotension, low-output HF state, or overt CS. GDMT often must be held or reduced in the acute phase of ES caused by dose-limiting hypotension from beta-blockade and AAD therapy. Fluid resuscitation should be performed cautiously, because ES patients are often volume-overloaded, and this may promote recurrent VAs. Most vasopressors and inotropes have proarrhythmic effects mediated by direct activation of beta-adrenergic receptors (eg, dopamine, epinephrine, dobutamine) or augmentation of their downstream second messenger systems (eg, milrinone), and these drugs should be avoided or used at the minimum dose that restores organ perfusion.[45] Pure vasoconstrictors without inotropic effects such as phenylephrine or vasopressin can be antiarrhythmic by promoting central sympathetic withdrawal via the baroreflex but will reduce cardiac output and should be avoided in low-output states or CS.[45] Norepinephrine has better hemodynamic efficacy and can be substituted despite a slight risk of proarrhythmia.[45]

When vasoactive therapy is either ineffective for restoring hemodynamic stability or results in proarrhythmia, temporary mechanical circulatory support (MCS) should be considered.[46] In the context of ES, MCS can serve as a bridge to catheter ablation, durable LVAD, or heart transplantation and may be used to support patients during catheter ablation. Each MCS modality has advantages and disadvantages related to use during ES or catheter ablation (**Table 3**).[46] Unlike other temporary MCS devices, venoarterial ECMO can entirely replace the native cardiac output even during cardiac arrest.[47] Among patients receiving venoarterial ECMO, those with ES generally have better outcomes because of their reversible etiology and limited end-organ failure. Any decision regarding the use of temporary MCS in ES, particularly venoarterial ECMO, should occur in the context of the reversibility of the patient's hemodynamic compromise, treatability, and candidacy for advanced HF therapies, ideally using a multidisciplinary "Shock Team" approach.

STEPPED-CARE ALGORITHM. A stepped-care approach to ES management can tailor the initial intensity of the 4 central management components (and potential risk of complications) to the acuity and risk of the presentation and then escalate proportionately in case of recurrent VAs (**Figure 4**). A low-risk ES patient (eg, hemodynamically stable MMVT, functioning ICD) starts at step 1 for each component, with standard initial therapies including IV amiodarone monotherapy plus an oral beta-blocker and an oral benzodiazepine.[2] A higher-risk ES patient (eg, no ICD, hemodynamically unstable VA) starts at step 2 with standard add-on therapies, including a second AAD (typically lidocaine), esmolol, and dexmedetomidine, in addition to step 1 initial therapies. Patients with recurrent VAs escalate to the next higher step, and rescue therapies that can be added include stellate ganglion block, third-line AADs (eg, procainamide), general anesthesia, and escalating degrees of hemodynamic support. The threshold for escalation should be lower for low-risk interventions (eg, dexmedetomidine or stellate ganglion block) vs high-risk interventions (eg, general anesthesia or venoarterial ECMO).

THE ROLE OF CATHETER ABLATION IN ES

Catheter ablation is essential to consider for patients with ES to either terminate incessant VAs or prevent recurrent VAs after medical stabilization.[1,2,48] Catheter ablation carries a Class I indication in patients with ICM and ES with failure or intolerance of AADs and a Class IIa indication for patients with refractory VA and NICM.[1,2,48] In ES patients with structural heart disease and MMVT without reversible causes, catheter ablation during the index hospitalization should be strongly considered, with urgent catheter ablation appropriate for incessant VA despite the potentially greater risk of complications in unstable patients.[49]

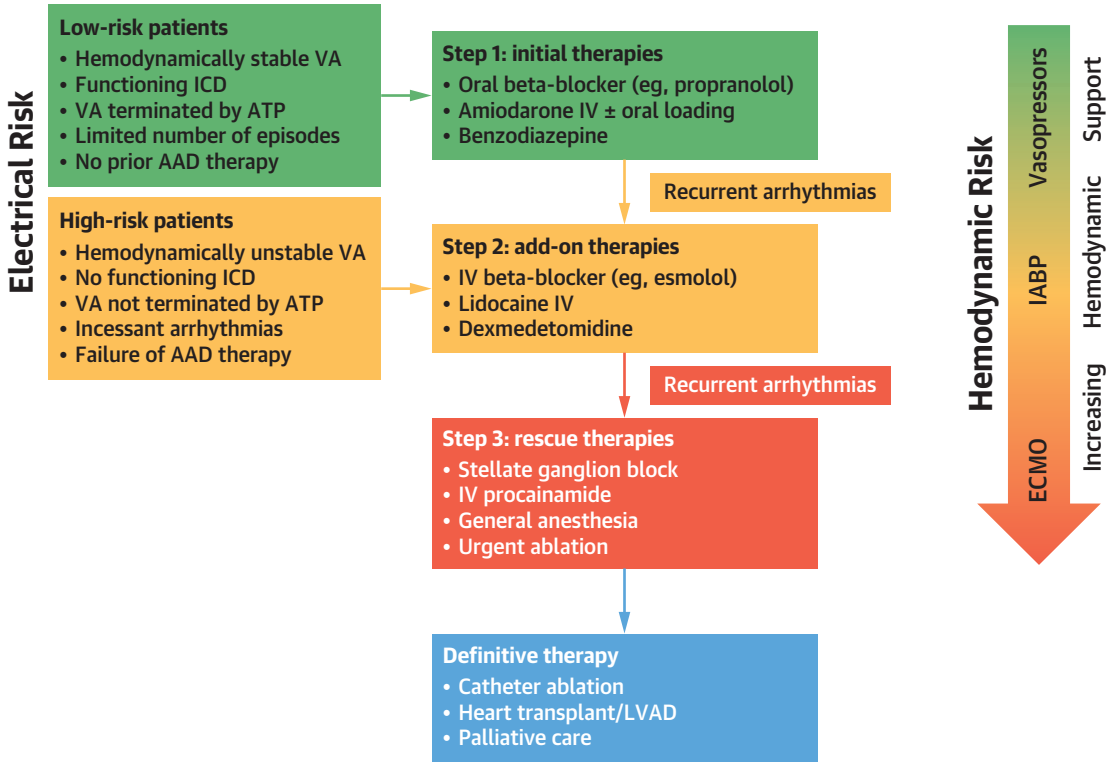
In the VANISH (Ventricular Tachycardia Ablation vs Escalated Antiarrhythmic Drug Therapy in Ischemic Heart Disease) trial, approximately one-fourth of patients presented with ES, and this subgroup appeared to derive similar benefit from catheter ablation vs intensification of AAD therapy.[50] Observational studies report better outcomes after catheter ablation compared with medical therapy in ES cohorts; however, these are limited by small size and potential selection bias.[51,52] Recurrence of VT and 1-year mortality after catheter ablation were higher in

FIGURE 4 Stepped-Care Algorithm for Rational Escalation of Medical Therapy in Electrical Storm

Intensity	Antiarrhythmic Drugs	Adrenergic Blockade	Sedation/Anxiolysis	Hemodynamic Support
Step 1	**Amiodarone IV*** • Bolus 300 mg (max 5 mg/kg) over 20 min • Repeat 150 mg bolus over 10 min for recurrent VA • Infusion 1 mg/min until free from VA ≥6 hours (may continue for longer) • Continue 0.5 mg/min until ES resolves	**Oral beta-blocker** • Propranolol 20-40 mg Q6h (preferred) • Metoprolol tartrate 25-50 mg Q6h (may be less effective) • May instead increase GDMT beta-blocker (eg, bisoprolol, carvedilol, metoprolol succinate) for selected low-risk patients	**Benzodiazepine** • Lorazepam 1 mg Q4-6h PRN • Diazepam 5 mg Q4-6h PRN • Midazolam 2 mg Q1-2h PRN	**Vasopressors** • Phenylephrine 0.1-2.0 μg/kg/min • Vasopressin 0.01-0.04 U/min • Norepinephrine 0.02-0.2 μg/kg/min

Step 2	**Lidocaine IV†**
	• Bolus 1-1.5 mg/kg (max 100-120 mg)
	• May repeat 0.5-0.75 mg/kg Q5-10 min x1-2 doses (max 300 mg or 3 mg/kg)
	• Infusion 1-2 mg/min (max 4 mg/min)
	• Goal serum procainamide concentration:
	  • 1.5-5 μg/mL
		IV beta-blocker
		• Esmolol
		  • Bolus 0.5 mg/kg (may repeat Q5 min x2)
		  • Infusion 50-300 μg/kg/min
		• Propranolol 1-3 mg Q5 min (max 5 mg)
		• Metoprolol 2.5-5 mg Q5 min (max 15 mg)
			Dexmedetomidine
			• Bolus (optional) 0.5-1 μg/kg over 10 min (typically not recommended due to risk of hypotension)
			• Infusion 0.2-0.7 μg/kg/h (maximum 1.0-1.5 μg/kg/h)
				Intra-aortic balloon pump
				• Contraindicated with aortic aneurysm/dissection, severe aortic insufficiency, or peripheral vascular disease
				• Less effective with tachycardia or atrial fibrillation

Step 3	**Procainamide IV‡**
	• Bolus 10-15 mg/kg (max 17-20 mg/kg, usually 1 g total) over 30-60 min
	• Infusion 1-2 mg/min (max 4 mg/min)
	• Goal serum procainamide concentration:
	  • 4-8 μg/mL (up to 10 μg/mL)
		Stellate ganglion blockade
		• Left stellate ganglion blockade
		  • 20 mL of 0.25% bupivacaine without epinephrine
		• Bilateral blocks if intubated
			General anesthesia
			• Endotracheal intubation
			• Propofol infusion often used, titrated to RAAS goal of −3
			• Opioid typically added (eg, fentanyl infusion)
				Advanced MCS
				• ECMO preferred
				• Percutaneous LVAD can be considered for selected patients
				• Contraindicated with severe aortic insufficiency or peripheral vascular disease



Medical management of ES requires tailoring the intensity of each component to both the initial risk assessment and response to treatment. Antiarrhythmic drugs (AADs) and adrenergic blockade reduce the risk of recurrent VA, augmented by sedation; hemodynamic support is often needed because of hemodynamic compromise as a cause of VA or consequent of therapy. Low-risk patients can start at step 1, whereas higher-risk presentations may justify starting at step 2 and increasing to the next step is warranted in case of recurrent VA. *Higher doses of IV amiodarone have been reported (ie, 2 mg/min for 6 hours, then 1 mg/min for 18 hours), but IV amiodarone doses >2.1 g/d increase the risk of adverse events such as hypotension. †Serum lidocaine concentrations may be increased by beta-blocker/amiodarone therapy, impaired liver function, or reduced liver blood flow, as occurs during decompensated HF/shock (especially infusion >2 mg/min). ‡Serum concentrations of procainamide and NAPA can be increased by kidney dysfunction or amiodarone, and the total procainamide + NAPA level should be 10 to 20 μg/mL (max 30 μg/mL); additive QT prolongation can occur with accumulation of NAPA (especially with concomitant amiodarone), so maintaining a serum procainamide + NAPA concentration in the lower end of the therapeutic range may be prudent. ATP = antitachycardia pacing; ECMO = extracorporeal membrane oxygenator; GDMT = guideline-directed medical therapy; HF = heart failure; IV = intravenous; NAPA = N-acetylprocainamide; PRN = as needed; Q = every (indicates dose frequency); RAAS = renin-angiotensin-aldosterone system; other abbreviations as in Figures 2 and 3.

FIGURE 5 Preprocedural Planning Prior to Catheter Ablation

Painesd Score		Points
P	Pulmonary disease	5
A	Age >60 years	3
I	Ischemic cardiomyopathy	6
N	NYHA functional class III-IV	6
E	Ejection fraction <25%	3
S	VT Storm	5
D	Diabetes	3
Risk of hemodynamic decompensation during VT ablation		
≤8 points		Low risk
9-15 points		Intermediate risk
≥15-17 points		High risk

Catheter Ablation for Electrical Storm: Periprocedural Checklist

Categorize

Primary arrhythmia	MMVT: EPS to identify scar, circuits, or areas of automaticity. PMVT/VF: EPS to identify triggers.
Structural substrate	Consider echo, CT and/or CMR to identify cardiomyopathy type and characterize areas of abnormal myocardium.

Optimize

Clinical stability	Optimize volume status, renal function, and electrolytes.
Triggers	Identify and stabilize acute triggers: adrenergic stimuli, electrolyte abnormalities, ischemia, etc.

Procedural Planning

Safety considerations	Hemodynamic stability achieved? Choice and timing of procedural anticoagulation. Left ventricular thrombus ruled out?
Anesthesia plan	Partner with anesthesia team to optimize plan based on anticipated procedure length, inducibility of arrhythmia, etc.
Hemodynamic support	Use PAINESD score for risk stratification. Consider benefits of each MCS modality.
LV access plan	Mechanical aortic or mitral valve? Anticipated need for epicardial access? Prior cardiac surgery or pericarditis?

Prior to catheter ablation, risk stratification using the PAINESD (Pulmonary Disease, Age >60 Years, Ischemic cardiomyopathy, NYHA Functional Class III-IV, Ejection Fraction <25%, VT Storm, Diabetes) score **(left)** is recommended. Patients with a high PAINESD score are at higher risk of hemodynamic decompensation, and preprocedural initiation of temporary mechanical circulatory support should be considered.[49,65] A preprocedural checklist **(right)** for planning prior to catheter ablation can ensure consistent care. CMR = cardiac magnetic resonance; CT = computed tomography; EPS = electrophysiological study; LV = left ventricular; MMVT = monomorphic ventricular tachycardia; PMVT = polymorphic ventricular tachycardia; VF = ventricular fibrillation; VT = ventricular tachycardia.

those with ES, highlighting the substantial risks even among survivors.[53] Elimination of clinical VT and postprocedural noninducibility after catheter ablation predict lower VT recurrence, lower ES recurrence, and higher survival rates.[51-54] Risk factors for VT recurrence after ablation include lower LVEF, previous ablation, and presence of an ICD.[12]

PREPROCEDURAL EVALUATION. Important preprocedural considerations are described in **Figure 5**.

Hemodynamic stabilization. Catheter ablation involves moderate sedation or general anesthesia as well as repeated induction of VT for mapping purposes, which can lead to hemodynamic decompensation even with previously tolerated VT. Patients should receive preprocedural hemodynamic stabilization, as well as risk stratification for intraprocedural decompensation.[55]

Addressing triggers. Initial efforts should focus on correcting identified triggers.[2] Catheter ablation may still be beneficial for identification and elimination of the underlying VT substrate; however, mapping and assessment of procedural success will be more straightforward without residual triggers. Delaying ablation may be necessary to allow myocardial healing after acute MI or during immunosuppression for cardiac sarcoidosis or myocarditis.

Substrate. VT circuits are often easier to localize in ICM than in NICM.[56] Small studies in ES report similar short- and intermediate-term outcomes after ablation in ICM and NICM populations.[54,57] Successful catheter ablation after ES has been reported in arrhythmogenic cardiomyopathy, cardiac sarcoidosis, and Chagas disease; epicardial ablation is often required in these patients.[20-22,56,58] Successful catheter ablation has been reported targeting right ventricular outflow tract substrate in Brugada syndrome and VF-triggering PVCs in early repolarization syndrome and idiopathic VF.[18,25,27,56]

CATHETER ABLATION STRATEGIES IN ES

MAPPING AND ABLATION FOR VT. VT ablation in structural heart disease is performed using a combination of 2 primary strategies. Activation mapping involves definition of the full activation circuit during VT, often with the aid of entrainment mapping, followed by ablation targeting the critical isthmus.[16,18,56] Substrate modification includes delineation of scar borders and all potential critical channels through scar while in sinus rhythm, followed by ablation aimed at eliminating identified channels and abnormal signals within the scar.[16] The major limitation of activation mapping is the need to induce and maintain VT for the duration of mapping, leading to potential hemodynamic compromise. Limitations of substrate modification include a longer procedure, less certainty regarding ablation targets, and potential for proarrhythmic effect if incomplete ablation is performed within heterogenous scar tissue. Substrate modification in addition to activation-based ablation was shown to be superior to activation mapping alone for preventing VT recurrence.[59]

MAPPING AND ABLATION FOR VF. VF is not characterized by a consistent re-entrant circuit, but VF driver activity at scar border zones may be mapped and targeted for ablation.[16,18] Triggering PVCs from the outflow tract, papillary muscles, or Purkinje system may be reproducibly identified and targeted for ablation with success rates exceeding 80%.[16,18,52,56] Catheter ablation of PVC triggers in patients with PMVT or VF refractory to medical therapy carries a Class IIa indication.[2]

MEASURES OF PROCEDURAL SUCCESS. Benchmarks for procedural success in VT ablation include termination of VT during ablation, noninducibility of VA, elimination of all abnormal signals within scar, or elimination of the triggering PVCs.[48,60-62] Multiple clinical or inducible VT morphologies are often observed in advanced cardiomyopathy, and elimination of the clinical VT during ES is more important than elimination of all possible VTs. In a large multicenter series of catheter ablation in ES, 87% had elimination of clinical VT and 64% had complete noninducibility of VT at the end of the procedure.[53] Up to 35% of the ES population may be too unstable to perform postprocedural programmed stimulation, and recurrence is high in those with residual inducibility of VT.[53]

MECHANICAL CIRCULATORY SUPPORT FOR CATHETER ABLATION IN ES. Peri-ablation acute hemodynamic decompensation is associated with higher likelihood of procedural failure, more VT recurrence, and increased in-hospital and long-term mortality.[49,55,63] Interventions to minimize harm related to the catheter ablation procedure include preprocedural optimization of hemodynamics, avoidance of general anesthesia if possible, and choosing substrate modification over VT induction in higher-risk patients. MCS can provide intraprocedural hemodynamic support while mapping unstable VTs, particularly in patients with prior failed substrate-based ablation or extensive scar with anticipated long duration of ablation.[47,64]

MCS during catheter ablation is used more frequently for ES patients, although in-hospital and short-term mortality remains high among ES patients requiring MCS.[47,53,63,64] Use of MCS improves hemodynamic stability during catheter ablation but has not been shown to reduce VT recurrence or improve long-term outcomes.[51,63,64] Small propensity-matched analyses showed lower risk of hemodynamic decompensation, higher likelihood of postprocedure noninducibility, and lower mortality with up-front MCS compared to rescue or no MCS, although outcomes are mixed.[55,63,64]

The PAINESD (Pulmonary Disease, Age >60 Years, Ischemic cardiomyopathy, NYHA Functional Class III-IV, Ejection Fraction <25%, VT Storm, Diabetes) score (**Figure 5**) is a risk-stratification tool developed to predict acute hemodynamic decompensation during VT ablation and postprocedural mortality; ES is among the risk factors.[49,64,65] A PAINESD score of ≥17 (≥15 when general anesthesia is excluded as a risk factor) is associated with higher hemodynamic risk and greater need for MCS and has been proposed as a criterion to select patients for pre-emptive MCS during ablation.[49,64,65] When using MCS to support ablation, initiation prior to the procedure should be considered to avoid acute hemodynamic decompensation and the need for bailout MCS.[47,55,64,65]

ADVANCED HF EVALUATION

RECOGNITION OF ADVANCED HF. The development of VA and ES may be a symptom of progressive cardiomyopathy heralding the transition to advanced HF.[3,4,13-15,29,52] Indeed, ES survivors most often succumb to worsening pump failure, with long-term outcomes as poor as patients hospitalized with decompensated HF.[4,13] Patients with advanced HF and ES should be considered for advanced HF therapies, such as heart transplantation or durable LVAD.[29,40] This can be performed most efficiently via a multidisciplinary team-based approach analogous to the "Shock Team" strategy used for patients with cardiogenic shock, including members with expertise

in electrophysiology, advanced HF, and critical care cardiology. The decision to list a patient for heart transplantation or proceed with LVAD depends on the patient's probability of recovery, suitability for transplant, degree of hemodynamic compromise, and anticipated waiting time for transplantation. Decision making is particularly challenging in the setting of ES, when a patient may not have the severe ventricular dysfunction typical of advanced HF and might have myocardial recovery if arrhythmias are suppressed. In most cases, heart transplantation is preferred for advanced HF patients with refractory VA, and LVAD is reserved for those patients who are not favorable candidates for transplantation or who are too unstable to survive until transplantation.

HEART TRANSPLANTATION. ES is rarely the sole reason for heart transplantation, with refractory VA historically accounting for fewer than 1% of adult heart transplants.[66] In October 2018, a new heart allocation system was instituted in the United States that prioritizes patients with refractory VA.[67] Under the revised heart transplant allocation system, patients with MCS and life-threatening VA are prioritized as a status 1 (highest priority) and those with life-threatening VA without MCS can be listed as status 2 (high priority) to facilitate urgent heart transplantation for patients with ES and refractory VA.

LVAD. LVAD implantation during ES is fraught with challenges, because recurrent VA episodes in LVAD patients can lead to right ventricular dysfunction and inadequate LV preload, which can impair device flow and produce complications.[68-70] LVAD patients possess an arrhythmogenic substrate from ventricular scar and remodeling caused by advanced cardiomyopathy, plus unique proarrhythmic mechanisms such as apical scarring from the LVAD cannula and hemodynamic perturbations such as hypovolemia triggering suction events.[69] More than one-third of LVAD recipients develop VA during follow-up (particularly those with VA before LVAD) and this portends a worse prognosis.[29,68-70] ES occurs in up to 10% of patients after LVAD and is associated with high 1-year mortality; risk factors for ES after LVAD included prior VA, prior VT ablation, AAD use, and perioperative MCS.[70] Nonetheless, patients with a history of VA before LVAD implantation may have comparable 1-year survival when stratified by INTERMACS (Interagency Registry for Mechanically Assisted Circulatory Support) profile.[69,71] Ablation at the time of LVAD implantation can be considered for selected patients.[72]

PALLIATION. Given the high short-term risk of death and poor long-term outcomes among patients who survive ES, timely palliative care consultation during hospitalization is important to establish overall goals of care. Among ES patients with advanced HF, comparatively few will receive heart transplant or LVAD, and hospice care may be an appropriate option. Ongoing palliative care follow-up after discharge can be beneficial in patients with advanced HF and should be considered for ES survivors with HF.[40]

MANAGEMENT OF PATIENTS AFTER RECOVERY FROM ES

ANTIARRHYTHMIC DRUGS. Oral amiodarone is recommended for long-term management in patients with ES caused by MMVT or repeated ICD discharges with a low VA burden, following ICD optimization.[2] Higher doses of amiodarone may be needed chronically to suppress VT (eg, 300-400 mg/d), recognizing the greater toxicity and potential excess risk of noncardiac death in patients with more severe HF.[15,39] If amiodarone is not desirable or tolerated, then guidelines recommend alternate AAD therapy according to underlying disease and cardiac function.[2] Oral mexiletine is often added to amiodarone, although ablation was superior in the VANISH trial.[1,73,74] Ranolazine has antiarrhythmic effects and showed some efficacy as add-on therapy for refractory VA in case series; however, ranolazine was not effective for prevention of ICD shocks in a randomized trial.[1,75,76] AADs are usually continued in patients with a successful ablation because of the substantial risk of recurrent VAs, with composite event rates after ablation as high as 59.1% (vs 68.5% with medical therapy) in the VANISH trial.[50,62] Patients with an unsuccessful ablation or those who are not candidates for ablation often receive combination AAD therapy including amiodarone, but rates of recurrent VAs are high (exceeding 40%).[53,55,62,63,74,76] Cardiac stereotactic body radiotherapy may be an alternative for highly selected patients with refractory VA who fail or are not candidates for repeat catheter ablation.[77]

GDMT. Optimization of GDMT is an important step after recovery for ES patients with underlying cardiomyopathy.[40] Reinstitution of GDMT prior to hospital discharge is essential given the elevated risk of death caused by HF in patients with VA and the recognition of inadequate GDMT as a risk factor for ES.[4-6,14,15,40,52] Whereas angiotensin-converting enzyme inhibitors, angiotensin receptor blockers,

and angiotensin-neprilysin inhibitors may not have a specific effect on VA, aldosterone antagonists (and perhaps SGLT2 inhibitors) appear to reduce the risk of sudden cardiac death and it is essential to initiate all indicated GDMT classes in patients with cardiomyopathy.[2,40] Digoxin may be proarrhythmic and is often discontinued. A crucial unanswered question is whether ES survivors with reduced LVEF who receive propranolol should be switched to a GDMT beta-blocker, recognizing that propranolol has not been studied for HF with reduced LVEF.[2,40]

FOLLOW-UP. ES survivors remain at substantial risk of VA, HF, and other adverse events, justifying close multidisciplinary follow-up after hospital discharge.[8,9,52,54] Recurrent ES is common, occurring in up to one-third to one-half of ES survivors or more, particularly those with MMVT, lower LVEF, older age, and not receiving angiotensin-converting enzyme inhibitors.[3,6,52-54] Frequent contact with a cardiologist is warranted, including an electrophysiologist and often an advanced HF expert. Remote ICD monitoring can alert clinicians to recurrent VA, including asymptomatic nonsustained or ICD-treated events. Diligent monitoring for noncardiac toxicities is necessary for patients receiving chronic amiodarone.[39]

CONCLUSIONS

ES is a heart rhythm emergency with a high risk of morbidity and mortality. ES management requires an integrated multidisciplinary team, including providers with expertise in critical care cardiology, electrophysiology, and advanced HF. ES typically develops in patients with cardiomyopathy and may be a manifestation of cardiac deterioration reflecting a transition to advanced HF that can require heart transplantation or LVAD. Most ES patients have a pre-existing ICD for primary or secondary prevention, and diligent ICD programming is helpful. Suppression of arrhythmias in ES patients integrates AADs (typically amiodarone), adrenergic blockade, and sedation/anxiolysis tailored to the severity of the clinical presentation in a stepped-care paradigm. Propranolol and stellate ganglion blockade may be beneficial. Many patients require catheter ablation to resolve ES and reduce the risk of further VA. Hemodynamic compromise is common, potentially requiring hemodynamic support before, during, and after catheter ablation. Optimization of GDMT during hospitalization is essential along with close multidisciplinary follow-up, because many ES survivors will develop complications from progressive HF. Collaborative multicenter clinical trials are needed to define best practices for ES patients, recognizing the wide spectrum of severity that can manifest.

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