



# Management of monomorphic ventricular tachycardia electrical storm in structural heart disease

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Electrical storm (ES) is a life-threatening condition that is defined by three or more episodes of sustained ventricular tachycardia (VT), ventricular fibrillation (VF), or appropriate shocks from an implantable cardioverter defibrillator (ICD) within 24 hours. The most common form of ES is monomorphic VT. It carries poor outcome despite all available intervention therapies. The therapies include rapid recognition of the condition, treatment of the reversible causes, ICD-reprogramming, antiarrhythmic drugs, sedation, and catheter ablation (CA). The first line antiarrhythmic drugs are amiodarone and  $\beta$ -blockers with superiority of propranolol over the others. The long-term use of the antiarrhythmic drugs is limited due to their adverse effects and drug-related proarrhythmic effect. The basic mechanism of monomorphic VT is re-entry pathway which can be targeted by CA. CA should be considered in drug refractory ES and patients should be referred in early course of disease. There are reported studies which showed the superiority of CA over the medical treatment in reducing the arrhythmia burden and ICD appropriate shock. The survival benefit has been reported after successful ablation of ES in case series but to date no randomized control trial shows mortality benefit.

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**Keywords:** Electrical storm, Recurrent ventricular tachycardia

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## 1. Introduction

The most widely used definition of ventricular tachycardia (VT) is three or more consecutive ventricular beats, at a rate >100 per minute [1]. It is classified based on hemodynamic stability, duration (nonsustained if <30 seconds and sustained if >30 seconds or requiring termination due to hemodynamic compromise in <30 seconds), morphology (monomorphic or polymorphic, etc.) or mechanism (scar-related re-entry, automaticity, and triggered activity) [2]. Of all the VTs, 10% occurs in those with structurally normal heart and these are referred to as idiopathic VTs, and the remaining 90% occurs in patients with structural heart disease [2]. Incessant VT is defined as hemodynamically stable VT which persists for longer than 12 hours [3] ES in those without ICD is referred to three or more sustained VT or ventricular fibrillation (VF) occurring within 24 hours [1,4]. In patient with an ICD is best defined as three appropriate VT detections in 24 hours, treated by antitachycardia pacing (ATP), shock, or eventually untreated but sustained in a VT monitoring zone [5].

## 2. Incidence and risk factors

According to the above -mentioned definition of ES, the incidence is ~10–20% in patients who have an ICD implanted for secondary prevention of sudden cardiac death [6,7]. It is lower when ICDs are implanted for primary prevention with an incidence of 4% [8]. Monomorphic VT is the most common form of ES with an incidence of 86–97% [9]. The most common causes of ES include electrolytes (hypo- and hyperkalemia) and metabolic imbalance, myocardial ischemia and infarction, drug-induced proarrhythmia, inherited channelopathies (such as Brugada syndrome), and con-

### Abbreviations

ACLS	advanced cardiac life support
ATP	anti-tachycardia pacing
CA	catheter ablation
ES	electrical storm
ICD	implantable cardioverter defibrillator
ICM	Ischemic Cardiomyopathy
IV	intravenous
NIDCM	non-ischemic dilated cardiomyopathy
VF	ventricular fibrillation
VT	ventricular tachycardia

gestive heart failure [10]. Other risk factors for ES include advanced age, male sex, a low left ventricular ejection fraction, and New York Heart Association functional Class III or IV heart failure, antiarrhythmic agents, hypomagnesemia, hyperthyroidism, and infection or fever [11]. Vicent et al. [12] recently reported six of 108 patients who received sacubitril/valsartan for heart failure with reduced ejection fraction, developed ventricular electrical storm. They concluded that their data are not enough to infer a cause and effect relationship but further investigations are needed to study the proarrhythmic effect of sacubitril/valsartan.

## 3. Prognosis

ES is a life-threatening clinical condition that requires prompt treatment. Eighty percent of the patients are hospitalized especially those who receive a shock from their devices [13]. Moreover, the electrical instability impairs the patient’s quality of life [5]. Most studies suggest that ES is an independent adverse prognostic factor [9]. The mortality is as high as 82% [14]. Also increased mortality has been documented in patients experiencing ES in the (AVID), MADIT (Multicenter Automatic Defibrillator Implantation Trial) II trials [6,8]. In addition, a meta-analysis of 13 stud-

ies revealed that ES was associated with a high mortality rate, increased cardiac transplantation, and hospitalization for acute heart failure [15].

#### 4. Mechanism

The most common mechanism of sustained ventricular arrhythmia in structural heart disease is re-entry [1]. Most monomorphic VTs are due to re-entry mechanism. The structural cardiac changes after an acute myocardial infarction or progresses of nonischemic cardiomyopathy (NICM), lead to scar formation as a consequence to fibrosis [11]. Epicardial patchy fibrosis and abnormal conduction may be detected in 30–50% of patients with NICM [16]. The scar then creates a fixed anatomic barrier to electrical conduction. The survived myofibrils around the border of the scar provide a slow pathway for electrically stable re-entry. A harmless trigger such as premature ventricular ectopic, can initiate monomorphic VT [11].

#### 5. Acute management

The management starts with proper diagnosis of the ventricular arrhythmia. VT should be differentiated from other resembling conditions such as bundle branch block, ventricular preexcitation (Wolff–Parkinson–White syndrome), or supraventricular tachycardia with aberrancy. An undifferentiated wide-complex tachycardia should be treated as VT, particularly in patients who have structural heart disease [11]. Patients with ES should be evaluated immediately for hemodynamic instability and should be treated according to advanced cardiac life support (ACLS) [17]. An algorithm for acute management of ES is suggested in Fig. 1 [18]. Those with VT storm, having pulse and are hemodynamically unstable should receive synchronized cardioversion. Patients who have poor systolic function or rapid VT might require multiple electrical cardioversions or defibrillations [11]. The reversible precipitating factors should be treated aggressively (Table 1) [19].

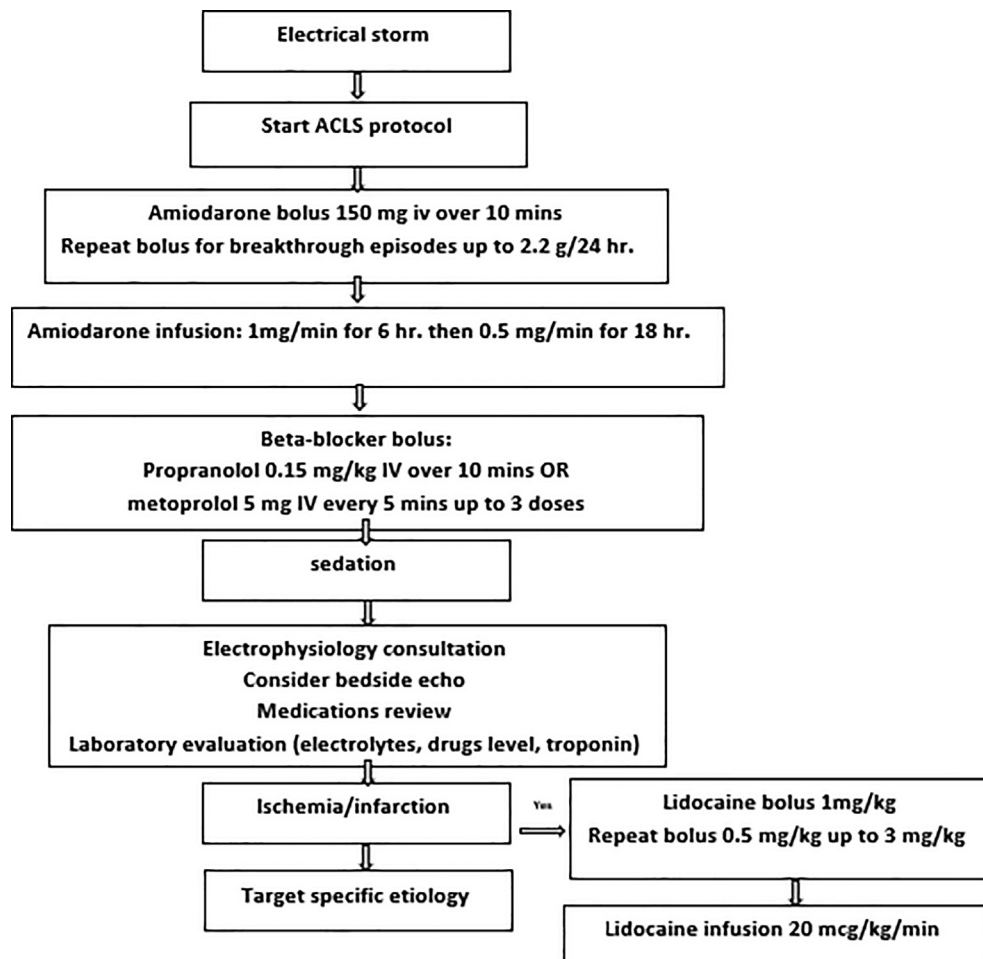


Figure 1. Acute management algorithm for electrical storm [18]. ACLS = advanced cardiac life support; IV = intravenous.

Table 1. Reversible causes of electrical storm.

Acute myocardial ischemia
Electrolyte imbalances
Decompensated heart failure
Hyperthyroidism
Infections, fever
Proarrhythmic drug effects early
Postoperative period

Patients with stable ES and those who received cardioversion due to hemodynamic instability, urgent intravenous (IV) antiarrhythmics drugs, and  $\beta$ -blocker should be started. The recommended approach is IV amiodarone (150 mg IV over 10 minutes, followed by 1 mg/minute IV infusion for 6 hours, followed by 0.5 mg/minute IV infusion for an 18 additional hours). Administration of  $\beta$ -blocker has been shown to improve survival at 1 week and 1 year in ES refractory to amiodarone and/or lidocaine and repeated cardioversions [14]. The preferred  $\beta$ -blocker is propranolol [20,21] (40 mg every 6 hours for the first 48 hours, with additional IV doses as needed for recurrent breakthrough ventricular arrhythmias) [21].

A summary of pharmacologic and nonpharmacologic therapy for acute management of ES is presented in Table 2 [1,19,18].

Early and aggressive use of analgesics and sedative drugs should be considered in patients with

ICD who received multiple shocks. Combination of benzodiazepines such as midazolam and short-acting analgesics such as remifentanyl provide analgesia without negative inotropic effects. Remifentanyl at infusion rate 0.06–0.12  $\mu$ g/kg/min with intermittent boluses of midazolam were used and the patient expressed no discomfort with the external cardioversions [22]. Propofol is a short acting general anesthetic agent that acts through its interactions with the gamma-aminobutyric acid receptor [23,24]. The used bolus doses and infusion rate of propofol that terminated the ventricular arrhythmias were 20–40 mg iv and 40–50  $\mu$ g/kg/min, respectively [23]. However, it should be used carefully because its negative inotropic effects can lead to cardiogenic shock [24]. If an ICD fails to convert a life-threatening rhythm or in state of ICD battery exhaustion, external defibrillation can be used. Interrogating the device helps to distinguish appropriate from inappropriate therapy. If the device reveals appropriate termination of VT or VF, reversible causes such as ischemia, electrolyte imbalances, worsening heart failure, and other causes should be looked for and treated accordingly [11]. Antiarrhythmic medications including amiodarone,  $\beta$ -blockers, and sotalol can reduce the frequency of ICD shocks [25]. Programming ICDs to deliver ATP for fast VT can reduce the need for shocks. Rapid pacing often terminates VT. In PainFREE Rx II Trial, ATP was

Table 2. Antiarrhythmic medications for acute and long-term treatment of ES.

	Acute management	Long-term treatment
<i><math>\beta</math>-blockers</i>		
Propranolol	Bolus: 0.15 mg/kg IV over 10 min then 40 mg every 6 hours for the first 48 h	10–40 mg by mouth 3–4 times a d
Metoprolol	Bolus: 5 mg IV every 5 min up to 3 doses	25 mg by mouth twice a d up to 200 mg a d
Esmolol	Bolus: 0.5 mg/kg IV for 1 min Infusion: 0.05 mg/kg/min	Not recommended
<i>Class III agents</i>		
Amiodarone	Bolus: 150 mg IV over 10 min, up to total 2.2 g in 24 h Infusion: 1 mg/min for 6 h, then 0.5 mg/min for 18 h	Oral load: 800 mg by mouth twice a d until 10 g total Maintenance dose: 200–400 mg by mouth daily
<i>Class I agents</i>		
Lidocaine	Bolus: 1.0–1.5 mg/kg IV, repeat dose of 0.5–0.75 mg/kg IV up to a total dose of 3 mg/kg Infusion: 1–4 mg/min although 1 could start at 0.5 mg/min	
Procainamide	Bolus: 10 mg/kg IV over 20 min	3–6 g by mouth daily fractionated in $\geq 3$ administrations
Mexiletine	Not recommended	200 mg by mouth 3 times a day, up to 400 mg by mouth 3 times a day
<i>Other treatments</i>		
Propofol	Bolus: 50 mg IV propofol infusion: 100 mcg/kg/min	
Overdrive pacing	Start at 90 bpm & titrate upward as needed, usually not faster than 110 bpm	

ES = electrical storm; IV = intravenous.

effective in terminating 229 fast VT of 284 episodes (72%). In comparison to shocks, empirical ATP for fast VT is equally safe, but highly effective, and improves quality of life [26].

## 6. Subsequent therapy

### 6.1. Antiarrhythmics drugs

All antiarrhythmic medications except  $\beta$ -blockers do not improve survival when given for the primary or secondary prevention of sudden cardiac death in patients with ventricular arrhythmia. Such conclusions derived from the randomized controlled trials. However, the use of these medications is essential in some patients to control arrhythmias and improve symptoms [1].

There are four classes of antiarrhythmics drugs according to Vaughan Williams classifications [27]: (1) Class I fast sodium channel blockers; (2) Class II  $\beta$ -blockers; (3) Class III repolarization potassium current blockers; and (4) Class IV nondihydropyridines calcium channel blockers.

### 6.2. Monotherapy

#### 6.2.1. $\beta$ -blocker

Patients who experienced ES especially those who received multiple ICD shocks, will have increased sympathetic tone, which can further induce recurrent arrhythmia.  $\beta$ -blockers can decrease the sympathetic surge. It remains an important treatment, which can reduce the risk of recurrent VT and VF in patients with recent myocardial infarction where there is increased sympathetic activity [14,28]. Propranolol antagonizes both the  $\beta_1$  and  $\beta_2$  receptors compared with metoprolol, which is  $\beta_1$  selective. In addition to its peripheral  $\beta$  receptors blockage, propranolol and due to its lipophilic properties, it can penetrate the central nervous system and block the central and prejunctional receptors [11]. Maybe due to these properties, propranolol appears to be more effective than metoprolol in terminating ES [29]. Chatzidou et al. [21] recently published a single-center, double-blind study of ICD 60 patients with ES. The patients were randomized to two groups and both received IV amiodarone. The propranolol group received 40 mg every 6 hours and the metoprolol group received 50 mg every 6 hours for the first 48 hours. It was shown that the termination of ventricular arrhythmias occurred significantly earlier in the propranolol group (3 hours) compared with the metoprolol group (18 hours). At the end of 24 hours, 27 of 30 patients receiving propranolol free of ventricular arrhythmias compared with 16 of 30 receiving

metoprolol. Also, the period of admission to intensive care unit was significantly lower in the propranolol group (3–4 days vs. 4–6 days) [21]. Moreover, propranolol seems more effective than bisoprolol. Puljević et al. [30] presented cases of five patients in whom the prevention of recidivate VTs was achieved only by using propranolol. In all cases bisoprolol was used. Only after switching from bisoprolol to propranolol, VT suppressed/ICD stopped activating [30]. However,  $\beta$ -blockers should be monitored closely in patients with severe systolic dysfunction especially in the sitting of hypotension that requires inotropic support and decompensated heart failure.

#### 6.2.2. Amiodarone

Amiodarone is considered the most effective antiarrhythmic drug and widely used in the treatment of ES [31,32]. Generally, it is safe to use unless there is contraindication such as hyperthyroidism or QT prolongation. The predominant class action is potassium channel blocker; however, it has a mixed antiarrhythmic class action [19]. It is incompletely absorbed (35–65%) after oral administration. Amiodarone has a true elimination half-life of up to 60 days. Because it has a relatively slow onset of action when administered orally, usually a large loading dose is required (800–1600 mg/day in 3–4 divided doses). In the first months of therapy, arrhythmia can reoccur and should not be considered drug inefficacy. In the setting of ES, IV amiodarone (1 g/day) can be effective in termination of VT and VF during 24-hour period [33]. It can suppress VT in  $\sim$ 40% of patients within 24 hours of IV administration, even when other agents (such as mexiletine, bretylium, lidocaine, or procainamide) are ineffective [34].

Amiodarone showed a higher rate of survival to hospital admission when compared with placebo or lidocaine for out-of-hospital cardiac arrest due to refractory ventricular arrhythmia [35,36]. The usual doses for long-term treatment of supraventricular and ventricular arrhythmia is 200–400 mg/day, however, doses of 100 mg/day have been shown to be effective in some patients [33]. The significant side effects of amiodarone limit its long-term use. They include corneal deposits (90%), photosensitivity (25–75%), pulmonary toxicity (1–17%), hyperthyroidism (0.9–2%), hypothyroidism (6%), blue-gray skin discoloration (4–9%), and liver toxicity [31].

#### 6.2.3. Lidocaine

Lidocaine class IB antiarrhythmic drug, acts as rapid sodium channel blockers binding to the

receptor in a use-dependent fashion. Its effectiveness in terminating ventricular arrhythmias is mostly in ischemia settings [19]. In conditions other than ischemia, the effectiveness of lidocaine in terminating ventricular arrhythmias range from 8% to 30%, which is considered a relatively weak antiarrhythmic effect [11]. It has favorable effects in patients with normal ejection fraction and combination with amiodarone can terminate most refractory arrhythmia [37]. In prospective, blinded, randomized controlled trials, lidocaine was less effective than amiodarone in improving hospital admission rates after out-of-hospital cardiac arrest due to shock-refractory VF or polymorphic VT [36].

#### 6.2.4. Sotalol

Commonly Sotalol is available in a racemic mixture of D-isomer and L-isomer. D-isomer acts as a Class III potassium channel blocker and L-isomer acts as a nonselective  $\beta$ -blocker [19]. In double-blind small trial (33 patients), results showed superiority of sotalol over lignocaine in acute termination of sustained VT [38]. In optimal pharmacological therapy in cardioverter defibrillator patients (OPTIC) trial, it was effective in decreasing ICD shocks but it was not superior to  $\beta$ -blocker alone or combined  $\beta$ -blocker and amiodarone [22]. Based on this trial and other trials, it has failed to demonstrate its superiority to  $\beta$ -blocker therapy in preventing recurrent ICD-shocks [19]. The D-sotalol (pure potassium-channel blocker) was tested in a randomized controlled trial, as a primary prevention of sudden death in patients with systolic dysfunction (ejection fraction  $\leq 40\%$ ) and previous myocardial infarction. There was an increased arrhythmic death in the D-sotalol-treated group and study stopped prematurely [39]. Most likely, the  $\beta$ -blocking effect of the L-isomer has a protective effect [18]. In view of above available data, sotalol should be considered if sustained VT were unresponsive to  $\beta$ -blockers [23,40].

#### 6.2.5. Procainamide

Procainamide is a Class IC agent which acts as fast sodium channel blocker. However, its active metabolite N-acetyl procainamide has potassium channels blockade effect as well. It can acutely terminate ventricular arrhythmias and prevent recurrences [19]. Procainamide was superior to lidocaine and amiodarone in acute VT termination. Hypotension is the most limiting adverse effect that necessitates stopping the medication [41,42]. The systemic side effects such as lupus-

like syndrome, gastrointestinal disturbances, and autoimmune blood impairments limits its long-term use [19].

#### 6.2.6. Combined therapy

Combination therapy is usually required to treat ES [43]. The most effective combination is  $\beta$ -blocker and amiodarone though the data are limited [31]. In OPTIC study, patients were randomized to treatment for 1 year with  $\beta$ -blocker alone (metoprolol, carvedilol, or bisoprolol), amiodarone plus one of the aforementioned  $\beta$ -blockers or sotalol alone. Combination of amiodarone and a  $\beta$ -blocker was the most effective in reducing the number of shocks (10.3%) compared with  $\beta$ -blocker group (38.5%) and sotalol group (24.3%) [25]. Other effective combinations includes Class III agents such as amiodarone or sotalol with a Class IC agent, flecainide [40,43]. Also, it has been reported successful combination of Class III amiodarone and Class IB lidocaine or mexiletine [37,44,45].

In summary, the choice of the antiarrhythmic drugs depends on the efficacy, drug-related proarrhythmic, side effects, and drug interaction.

### 7. Pacing

There are reported cases of successful termination of ES by overdrive pacing which was tried after medical therapy failure [46–48]. This is a temporary method as the ES can reoccur once the pacing is stopped. It can be used as a bridge while waiting for ischemia revascularization or trial of catheter ablation [18].

### 8. Sedation

Patients who experienced ES and received multiple ICD shocks, may have physical and emotional stress and enhanced sympathetic tone, that can further induce recurrent arrhythmia [11]. Case reports showed that short acting anesthetics such as propofol can suppress refractory ES by inhibiting the sympathetic tone [23,24].

### 9. Catheter ablation

The most common form of ES is monomorphic VT and the basic mechanism of it is re-entry pathway, hence catheter ablation (CA) is an important approach to stop the storm. The current guidelines recommend CA in drug refractory arrhythmia or intolerance of drugs therapy due to side effects [1]. With the modern technology and increasing

experience in CA, the success rate is high and the complications of the procedure are low. A meta-analysis of total of 471 ventricular arrhythmias storm patients from 39 publications were collected. Successful ablation was achieved in 72% of all ventricular arrhythmias and 6% patients had a recurrence of ventricular arrhythmias storm. Arrhythmic storm of monomorphic VT had the higher reoccurrence rate. Overall mortality was 17% at 1.2-years follow-up with most of the deaths related to progressive heart failure (62%) [3]. There are reported studies showing the superiority of CA over the medical treatment in reducing the arrhythmia burden [49–52] and ICD appropriate shock [50,53]. Also, prophylactic CA at the time of ICD implantation in patients with history of myocardial infarction, significantly decreased ICD therapies [53,54]. In VANISH trial, 259 patients with ischemic cardiomyopathy (ICM) and an ICD who had VT despite the use of antiarrhythmic drugs, were assigned to either CA with continuation of baseline antiarrhythmic drugs and escalating the antiarrhythmic drugs. There was a significantly lower rate of the composite primary outcome of death, VT storm, or appropriate ICD shock among patients undergoing CA. There was no significant between-group difference in mortality [50]. The survival benefit has been reported after successful ablation of ES in single center and multicenter case series [52,55] but to date no randomized control trial shows mortality benefit.

Most of the studies reported included ventricular arrhythmia in patients with ischemic heart disease. In the setting of non-ischemic dilated cardiomyopathy (NIDCM), there is insufficient information on the role CA for ES [52]. HELP-VT is single center nonrandomized study compared the outcomes of VT ablation in NIDCM and ICM. Two hundred and twenty-seven patients, 63 with NIDCM (34 with ES) and 164 with ICM (67 with ES), presenting with sustained VT were ablated. Complete elimination of any VT was achieved in 66.7% of NIDCM compared with 77.4% of ICM patients. The short-term outcomes were not statistically different between the two groups. At the 1-year follow-up, VT-free survival in NIDCM was 40.5% compared with 57% in ICM. The long-term outcomes were worse in NIDCM group but inability to induce VT at the end of ablation was associated with beneficial long-term outcomes [56]. In a single center case series of 282 patients with NIDCM, underwent endocardial CA with an adjuvant epicardial ablation. At 60 months follow up, nearly 70% of patients had VT-free and 76% had transplant-free survival. Significant VT burden reduction was noted with

repeat of ablation for VT recurrence [51]. Recently, Muser et al. [57] compared long-term outcomes of CA of drug refractory ES in patients with NIDCM with patients with ICM. The study included 267 consecutive patients with NIDCM ( $n = 71$ ; ejection fraction  $32 \pm 14\%$ ) and ICM ( $n = 196$ ; ejection fraction  $28 \pm 12\%$ ). Overall VT-free survival was 48% in NIDCM compared with 54% in ICM at 60 months. Death/transplantation-free survival was 53% in NIDCM and 64% in ICM at 60 months follow up. They concluded that CA of ES was similarly effective in patients with NIDCM compared with patients with ICM, with elimination of ES in 95% of cases and achievement of complete VT control at long-term follow-up in most patients [57].

The ideal time of CA after the onset VT is not clearly defined. In a single center, 98 patients with VT and structural heart disease were studied to look for the outcome of early versus late referral for CA. Late referral was defined as two or more episodes of VT and at least 1 month apart. Thirty-six and 62 patients fit the definition of early and late referral, respectively. Overall acute procedural success was achieved in 89%. One-year VT free survival was superior in the early group. Significant reduction in amiodarone dose is another positive result of this study and best outcome seen in the early group [58].

In conclusion, CA has an important role in ES management. It is superior to medical treatment in reducing arrhythmia burden and ICD appropriate shock. There is growing evidence of survival benefit, however, no randomized control trial to date shows mortality benefit. It should be done early in the course of disease rather than referred late.

## 10. ICD implantation

ICDs are commonly used in patients who are at high risk of sudden cardiac death. These devices do not prevent arrhythmias, and implanting an ICD is contraindicated in the acute phase of ES [59]. The current guidelines recommend implantation an ICD as secondary prevention for sudden cardiac death in patients with structural heart disease who developed sustained VT whether hemodynamically stable or not provided the correctable reversible causes such as acute myocardial infarction, proarrhythmic medication effects, or electrolyte disturbances have been excluded [1].

## 11. Refractory electrical storm

ES in rare situations becomes refractory to medical therapy and CA. There are alternative

approaches which mainly target sympathetic blockade and modulation of the autonomic nervous system. These approaches include stellate ganglion blockade [60], cardiac sympathetic denervation [61], and renal artery denervation [62,63]. Surgical options include postmyocardial infarction left ventricular aneurysmectomy with surgical cryoablation [64] and heart transplantation [52,56].

## 12. Conclusion

ES is a life-threatening condition that requires prompt treatment. It carries poor outcomes despite all available intervention therapies. These therapies include rapid recognition of the condition, treatment of the reversible causes, ICD-reprogramming, antiarrhythmic drugs, sedation, and CA. The first line antiarrhythmic drugs are amiodarone and  $\beta$ -blocker with the superiority of the propranolol over the others. CA should be considered in drug refractory ES and patients should be referred in early course of disease.

## References

- [1] Al-Khatib SM, Stevenson WG, Ackerman MJ, Bryant WJ, Callans DJ, Curtis AB, et al. 2017 AHA/ACC/HRS Guideline for management of patients with ventricular arrhythmias and the prevention of sudden cardiac death: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. *J Am Coll Cardiol* 2018;71:e91-e220.
- [2] Shenthar J. Unusual incessant ventricular tachycardia what is the underlying cause and the possible mechanism? *Circ Arrhythm Electrophysiol* 2015;8:1507-11.
- [3] Nayyar S, Ganesan AN, Brooks AG, Sullivan T, Roberts Thomson KC, Sanders P. Venturing into ventricular arrhythmia storm: a systematic review and meta-analysis. *Eur Heart J* 2013;34:560-71.
- [4] Kowey PR, Levine JH, Herre JM, Pacifico A, Lindsay BD, Plumb VJ, et al. Randomized, double-blind comparison of intravenous amiodarone and bretylium in the treatment of patients with recurrent, hemodynamically destabilizing ventricular tachycardia or fibrillation. The Intravenous Amiodarone Multicenter Investigators Group. *Circulation* 1995;92:3255-63.
- [5] Israel CW, Barold SS. Electrical storm in patients with an implanted defibrillator: a matter of definition. *Ann Noninvasive Electrocardiol* 2007;12:375-82.
- [6] Exner DV, Pinski SL, Wyse DG, Renfroeg EG, Follmann D, Gold M, et al. Electrical storm presages nonsudden death: the antiarrhythmics versus implantable defibrillators (AVID) trial. *Circulation* 2001;103:2066-71.
- [7] Emkanjoo Z, Alihasani N, Alizadeh A, Tayyebi M, Bonakdar H, Barakpour H, et al. Electrical storm in patients with implantable cardioverter-defibrillators: can it be forecast? *Tex Heart Inst J* 2009;36:563-7.
- [8] Sesselberg HW, Moss AJ, McNitt S, Zareba W, Daubert JP, Andrews ML, et al. Ventricular arrhythmia storms in postinfarction patients with implantable defibrillators for primary prevention indications: a MADIT-II substudy. *Heart Rhythm* 2007;4:1395-402.
- [9] Sagone A. Electrical storm: incidence, prognosis and therapy: review article. *J Atr Fibrillation* 2015;8:1150.
- [10] Al-Ahmad A, Shenasa M, Shenasa H, Soleimanieh M. Incessant ventricular tachycardia and fibrillation: electrical storms. *Card Electrophysiol Clin* 2014;6:613-21.
- [11] Eiffling M, Razavi M, Massumi A. The evaluation and management of electrical storm: review. *Tex Heart Inst J* 2011;38:111-21.
- [12] Vicent L, Juárez M, Martín J, García J, González-Saldívar H, Bruña V, et al. Ventricular arrhythmic storm after initiating sacubitril/valsartan. *Cardiology* 2018;139:119-23.
- [13] Proietti R, Sagone A. Electrical storm: incidence, prognosis and therapy. *Indian Pacing and Electrophysiol J* 2011;11:34-42.
- [14] Nademanee K, Taylor R, Bailey WE, Rieders DE, Kosar EM. Treating electrical storm sympathetic blockade versus advanced cardiac life support-guided therapy. *Circulation* 2000;102:742-7.
- [15] Guerra F, Shkoha M, Scappini L, Flori M, Capucci A. Role of electrical storm as a mortality and morbidity risk factor and its clinical predictors: a meta-analysis. *Europace* 2014;16:347-53.
- [16] Kuriachan VP, Sumner GL, Wahab AA, Sapp J, Mitchell LB. Ventricular tachycardia in ischemic and dilated cardiomyopathy: mechanisms and diagnosis. In: *Encyclopedia of Cardiovascular Research and Medicine*. Elsevier: Amsterdam; 2018. p. 690-9.
- [17] Link MS, Berkow LC, Kudenchuk PJ, Halperin HR, Hess EP, Vivek K, et al. Adult advanced cardiovascular life support: 2015 American Heart Association Guidelines update for cardiopulmonary resuscitation and emergency cardiovascular care. *Circulation* 2015;132:S444-64.
- [18] Sorajja D, Munger TM, Shen W-K. Optimal antiarrhythmic drug therapy for electrical storm. *J Biomed Res* 2015;29:20-34. <https://doi.org/10.7555/JBR.29.20140147>.
- [19] Muser D, Santangeli P, Liang JJ. Management of ventricular tachycardia storm in patients with structural heart disease. *World J Cardiol* 2017;9:521-30.
- [20] Gorenek B, Blomström Lundqvist C, Brugada Terradellas J, Camm AJ, Hindricks G, Huber K, et al. Cardiac arrhythmias in acute coronary syndromes: position paper from the joint EHRA, ACCA, and EAPCI task force. *Europace* 2014;16:1655.
- [21] Chatzidou S, Kontogiannis C, Tsilimigras DI, Georgiopoulos G, Kosmopoulos A, Papadopoulou E, et al. Propranolol versus metoprolol for treatment of electrical storm in patients with implantable cardioverter-defibrillator. *J Am Coll Cardiol* 2018;71:1897.
- [22] Mandel JE, Hutchinson MD, Marchlinski FE. Remifentanylmidazolam sedation provides hemodynamic stability and comfort during epicardial ablation of ventricular tachycardia. *J Cardiovasc Electrophysiol* 2011;22:464-6.
- [23] Burjorjee JE, Milne B. Propofol for electrical storm; a case report of cardioversion and suppression of ventricular tachycardia by propofol. *Can J Anaesth* 2002;49:973-7.
- [24] Mulpuru SK, Patel DV, Wilbur SL, Vasavada BC, Furqan T. Electrical storm and termination with propofol therapy: a case report. *Int J Cardiol* 2008;128:e6-8.
- [25] Connolly SJ, Dorian P, Roberts RS, Gent M, Bailin S, Fain ES, et al. Optimal pharmacological therapy in cardioverter defibrillator patients (OPTIC) investigators. Comparison of betablockers, amiodarone plus beta-blockers, or sotalol for prevention of shocks from implantable cardioverter defibrillators: the OPTIC Study: a randomized trial. *JAMA* 2006;295:165-71.
- [26] Wathen MS, DeGroot PJ, Sweeney MO, Stark AJ, Otterness MF, Adkisson WO, et al. Prospective randomized multicenter trial of empirical antitachycardia pacing versus shocks for spontaneous rapid ventricular tachycardia in patients with implantable cardioverter-defibrillators: pacing fast ventricular tachycardia reduces



- shock therapies (PainFREE Rx II) trial results. *Circulation* 2004;110:2591-6.
- [27] Vaughan Williams EM. A classification of antiarrhythmic actions reassessed after a decade of new drugs. *J Clin Pharmacol* 1984;24:129-47.
- [28] Moss AJ, Zareba W, Hall WJ, Klein H, Wilber DJ, Cannom DS, et al.. Multicenter automatic defibrillator implantation Trial II investigators: prophylactic implantation of a defibrillator in patients with myocardial infarction and reduced ejection fraction. *N Engl J Med* 2002;346:877-83.
- [29] Tsagalou EP, Kanakakis J, Rokas S, Anastasiou-Nana MI. Suppression by propranolol and amiodarone of an electrical storm refractory to metoprolol and amiodarone. *Int J Cardiol* 2005;99:341-2.
- [30] Puljević M, Velagić V, Puljević D, Miličić D. Propranolol efficiency in prevention of sustained ventricular tachycardia in patients with implanted cardioverter-defibrillator: a case series. *Croat Med J* 2014;55:75-6.
- [31] Vassallo P, Trohman RG. Prescribing amiodarone: an evidence-based review of clinical indications. *JAMA* 2007;298:1312-22.
- [32] European Heart Rhythm Association, Heart Rhythm Society, Zipes DP, Camm AJ, Borggrefe M, Buxton AE, et al.. ACC/AHA/ESC 2006 guidelines for management of patients with ventricular arrhythmias and the prevention of sudden cardiac death: a report of the American College of Cardiology/American Heart Association Task Force and the European Society of Cardiology Committee for Practice Guidelines (Writing Committee to Develop Guidelines for Management of Patients with Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death). *J Am Coll Cardiol* 2006;48:e247-346.
- [33] Connolly SJ. Evidence-based analysis of amiodarone efficacy and safety. *Circulation* 1999;100:2025-34.
- [34] Levine JH, Massumi A, Scheinman MM, Winkle RA, Platia EV, Chilson DA, et al.. Intravenous amiodarone for recurrent sustained hypotensive ventricular tachyarrhythmias. Intravenous amiodarone multicenter trial group. *J Am Coll Cardiol* 1996;27:67-75.
- [35] Kudenchuk PJ, Cobb LA, Copass MK, Cummins RO, Doherty AM, Fahrenbruch CE, et al.. Amiodarone for resuscitation after out-of-hospital cardiac arrest due to ventricular fibrillation. *N Engl J Med* 1999;341:871-8.
- [36] Dorian P, Cass D, Schwartz B, Cooper R, Gelaznikas R, Barr A. Amiodarone as compared with lidocaine for shock-resistant ventricular fibrillation. *N Engl J Med* 2002;346:884-90.
- [37] Koji Y, Takeshi T, Takahiro T, Ayako O, Takashi M, Hirohiko M, et al.. Renewed impact of lidocaine on refractory ventricular arrhythmias in the amiodarone era. *Int J Cardiol* 2014;176:936-40. <https://doi.org/10.1016/j.ijcard.2014.08.090>.
- [38] Ho DS, Zecchin RP, Richards DA, Uther JB, Ross DL. Double-blind trial of lignocaine versus sotalol for acute termination of spontaneous sustained ventricular tachycardia. *Lancet* 1994;344:18-23.
- [39] Waldo AL, Camm AJ, deRuyter H, Friedman PL, MacNeil DJ, Pauls JF, et al.. Effect of d-sotalol on mortality in patients with left ventricular dysfunction after recent and remote myocardial infarction. The SWORD investigators. Survival with oral d-Sotalol. *Lancet* 1996;348:7-12.
- [40] Priori SG, Blomstrom-Lundqvist C, Mazzanti A, Bloma N, Borggrefe M, Camm J, et al.. 2015 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death. The task force for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death of the European Society of Cardiology (ESC). *Eur Heart J* 2015;36:2793-867.
- [41] Gorgels AP, van den Dool A, Hofs A, Mulleneers R, Smeets JL, Vos MA, et al.. Comparison of procainamide and lidocaine in terminating sustained monomorphic ventricular tachycardia. *Am J Cardiol* 1996;78:43-6.
- [42] Ortiz M, Martín A, Arribas F, Coll-Vinent B, Del Arco C, Peinado R, et al.. PROCAMIO Study Investigators. Randomized comparison of intravenous procainamide vs. intravenous amiodarone for the acute treatment of tolerated wide QRS tachycardia: the PROCAMIO study. *Eur Heart J* 2016;38:1329-35.
- [43] Hsieh J-C, Bui M, Yallapragda S, Shoen K, Huang S. Current management of electrical storm. *Acta Cardiol Sin* 2011;27:716.
- [44] Chia PL, Loh SY, Foo D. Ventricular tachycardia storm: a case series and literature review. *Med J Malaysia* 2012;67:582-4.
- [45] Manolis A, Katsivas A, Vassilopoulos C, Tsatiris C. Electrical storm in an ICD-recipient with 429 delivered appropriate shocks: therapeutic management with antiarrhythmic drug combinations. *J Interv Card Electrophysiol* 2002;6:91-4.
- [46] Kurisu S, Inoue I, Kawagoe T, Ishihara M, Shimatani Y, Mitsuba N, et al.. Temporary overdriving pacing as an adjunct to antiarrhythmic drug therapy for electrical storm in acute myocardial infarction. *Circ J* 2005;69(5):613-6.
- [47] Tanabe Y, Chinushi M, Washizuka T, Minagawa S, Furushima H, Watanabe H, et al.. Suppression of electrical storm by biventricular pacing in a patient with idiopathic dilated cardiomyopathy and ventricular tachycardia. *Pacing Clin Electrophysiol* 2003;26(1 Pt 1):101-2.
- [48] Aguiar Rosa S, Oliveira M, Valente B, Silva Cunha P, Almeida Morais L, Cruz Ferreira R. Ventricular electrical storm after acute myocardial infarction successfully treated with temporary atrial overdrive pacing. *Med Intensiva* 2017;41:252-4.
- [49] Santangeli P, Muser D, Maeda S, Filtz A, Zado ES, Frankel DS, et al.. Comparative effectiveness of antiarrhythmic drugs and catheter ablation for the prevention of recurrent ventricular tachycardia in patients with implantable cardioverter-defibrillators: a systematic review and meta-analysis of randomized controlled trials. *Heart Rhythm* 2016;13:1552-9.
- [50] Sapp JL, Wells GA, Parkash R, Stevenson WG, Blier L, Sarrazin JF, et al.. Ventricular tachycardia ablation versus escalation of antiarrhythmic drugs. *N Engl J Med* 2016;375:111-21.
- [51] Liang JJ, Muser D, Santangeli P. Ventricular tachycardia ablation clinical trials. *Card Electrophysiol Clin* 2017;9:153-65.
- [52] Muser D, Liang JJ, Pathak RK, Magnani S, Castro SA, Hayashi T, et al.. Long-term outcome after catheter ablation of ventricular tachycardia in patients with nonischemic dilated cardiomyopathy. *JACC Clin Electrophysiol* 2017;3:767-78.
- [53] Kuck KH, Schaumann A, Eckardt L, Willems S, Ventura R, Delacretaz E, et al.. Catheter ablation of stable ventricular tachycardia before defibrillator implantation in patients with coronary heart disease (VTACH): a multicentre randomized controlled trial. *Lancet* 2010;375:31-40.
- [54] Reddy Vivek Y, Reynolds Matthew R, Neuzil P, Richardson AW, Taborsky M, Jongnarangsin K, et al.. Prophylactic catheter ablation for the prevention of defibrillator therapy. *N Engl J Med* 2007;357:2657-65.
- [55] Vergara P, Tung R, Vaseghi M, Brombin C, Frankel DS, DiBiase L, et al.. Successful ventricular tachycardia ablation in patients with electrical storm reduces recurrences and improves survival. *Heart Rhythm* 2018;15:48-55. <https://doi.org/10.1016/j.hrthm.2017.08.022>.
- [56] Dinov B, Fiedler L, Schönbauer R, Bollmann A, Rolf S, Piorkowski C, et al.. Outcomes in catheter ablation of ventricular tachycardia in dilated nonischemic cardiomyopathy compared with ischemic cardiomyopathy: results from the Prospective Heart Centre of Leipzig VT (HELP-VT) Study. *Circulation* 2014;129:728-36.
- [57] Muser D, Liang JJ, Pathak RK, Magnani S, Castro SA, Hayashi T, et al.. Longterm outcomes of catheter ablation

- of electrical storm in nonischemic dilated cardiomyopathy compared with ischemic cardiomyopathy. *JACC Clin Electrophysiol* 2017;3:71.
- [58] Frankel DS, Mountantonakis SE, Robinson MR, Zado ES, Callans DJ, Marchlinki FE. Ventricular tachycardia ablation remains treatment of last resort in structural heart disease: argument for earlier intervention. *J Cardiovasc Electrophysiol* 2011;22:1123-8.
- [59] Epstein AE, DiMarco JP, Ellenbogen KA, Estes 3rd NA, Freedman RA, Gettes LS, et al.. American College of Cardiology Foundation; American Heart Association Task Force on Practice Guidelines; Heart Rhythm Society. 2012 ACCF/AHA/HRS focused update incorporated into the ACCF/AHA/HRS 2008 guidelines for device-based therapy of cardiac rhythm abnormalities: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines and the Heart Rhythm Society. *J Am Coll Cardiol* 2013;61: e6-e75.
- [60] Meng L, Tseng CH, Shivkumar K, Ajjola O. Efficacy of stellate ganglion blockade in managing electrical storm: a systematic review. *JACC Clin Electrophysiol* 2017;3:942.
- [61] Vaseghi M, Barwad P, Malavassi Corrales FJ, Tandri H, Mathuria N, Shah R, et al.. Cardiac sympathetic denervation for refractory ventricular arrhythmias. *J Am Coll Cardiol* 2017;69:3070-80.
- [62] Lemery R. Interventional treatment of ventricular tachycardia and electrical storm: from ablation of substrate and triggers to autonomic modulation by renal denervation. *Heart Rhythm* 2014;11:547-8.
- [63] Remo BF, Preminger M, Bradfield J, Mittal S, Boyle N, Gupta A, et al.. Safety and efficacy of renal denervation as a novel treatment of ventricular tachycardia storm in patients with cardiomyopathy. *Heart Rhythm* 2014;11:541.
- [64] Pojar M, Harrer J, Omran N, Vobornik M. Surgical cryoablation of drug resistant ventricular tachycardia and aneurysmectomy of postinfarction left ventricular aneurysm. *Case Rep Med* 2014;2014 207851.