What is electrical storm

- Electrical storm refers to a state of cardiac electrical instability characterized by multiple episodes of ventricular tachycardia (VT storm) or ventricular fibrillation (VF storm) within a relatively short period of time, typically 24 hours.*

- The clinical definition of electrical storm is varied, somewhat arbitrary, and is a source of ongoing debate.
Definition

In patients without an implantable cardioverter-defibrillator (ICD), electrical storm has been variously defined as:

- The occurrence of **two or more** hemodynamically stable ventricular tachyarrhythmias within 24 hours
- VT recurring soon after (within **five minutes**) termination of another VT episode
- Sustained and non-sustained VT resulting in a total number of ventricular ectopic beats greater than sinus beats in a 24-hour period.
Definition

• In patients with an ICD, the most widely accepted definition of electrical storm is **three or more** appropriate therapies for ventricular tachyarrhythmias, including antitachycardia pacing or shocks, within 24 hours.

• However, this definition is not comprehensive as it fails to account for:
  • VT that is slower than the programmed detection rate of the ICD
  • VT that fails to terminate with appropriate ICD therapy and remain undetected by the patient
Incidence

- Depends on the population studied
- 10-20% of all ICD recipients
- Incidence lower when ICD implanted for primary prevention versus secondary prevention
- Incidence more in patients experiencing acute MI, past MI and those with inherited channelopathies
Who has an Electrical Storm: Predictors

- **Prior arrhythmia**
  - Previous VT storm
    - Failed ablation therapy
  - VT\(^1,2\) or VF\(^4\) as indication for ICD

- **Poor Cardiac status**
  - EF < 25\(^2,3\)
  - Absence of revascularization after index arrhythmia\(^1\)
  - QRS >120 msec\(^3\)
  - CAD\(^4\)

- **Sub-optimal medication**
  - Absence beta blocker therapy\(^3\)
  - Use of digoxin\(^1\)

- **Co-morbidity**
  - Chronic renal failure\(^2\)
  - Thyrotoxicosis

1. Exner et al. Circ 2001
2. Brigadeau et al. EHJ 2005
3. Arya et al. AJC 2006
4. Verma et al. JCE 2004
Triggers and risk factors

<table>
<thead>
<tr>
<th>Table 1. Causes of electrical storm*</th>
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</thead>
<tbody>
<tr>
<td>1. Enhanced sympathetic tone</td>
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<tr>
<td>2. Ischemia</td>
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<tr>
<td>3. Electrolyte imbalances (potassium, magnesium)</td>
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<td>4. Genetic abnormalities (such as Brugada syndrome, LQTS, CPVT, ERS, etc.)</td>
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<td>5. Iatrogenic (often in the presence of implantable cardioverter-defibrillator)</td>
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<tr>
<td>6. Endocrine disorders (thyroid disorders, pheochromocytoma)</td>
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</tbody>
</table>

*The causes of electrical storm (1-6) are listed in an approximately descending order of occurrence. CPVT, catecholaminergic polymorphic ventricular tachycardia; ERS, early repolarization syndrome; LQTS, long QT syndrome.
Clinical presentation

- Depends on the ventricular rate, the presence of underlying heart disease, the degree of left ventricular systolic dysfunction, and the presence or absence of therapies delivered by an implantable cardioverter-defibrillator (ICD).

- In patients without an ICD
  - Repeated episodes of palpitations, presyncope, or syncope if the patient remains hemodynamically stable
  - Cardiac arrest in those patients with hemodynamically unstable ventricular arrhythmias.

- In patients with a pre-existing ICD
  - Multiple ICD therapies (some combination of anti-tachycardia pacing and ICD shocks).
  - Patients with ventricular arrhythmias that are slower than the detection settings of the ICD may present in similar fashion as patients without an ICD.
Types of arrhythmias

- The frequency of various ventricular arrhythmias is as follows:
  - Monomorphic VT – 86 to 97 percent
  - Primary VF – 1 to 21 percent
  - Mixed VT/VF – 3 to 14 percent
  - Polymorphic VT – 2 to 8 percent

- In patients with documented sustained arrhythmias prior to ICD implantation, there exists a significant correlation between the initial arrhythmia and that recorded during electrical storm.
Clinical syndromes of arrhythmic storms

- Electrical storm develops when a vulnerable anatomic substrate (such as that from structural heart disease or scarring after an MI) is affected by a triggering event, such as premature ventricular contractions (PVCs) or an electrolyte imbalance.

- Determining the cause of electrical storm is essential, because treatment must target the underlying mechanism.
Clinical Causes of Electrical Storm

- Unknown – majority of cases 70-90%
- Decompensated heart failure
- Acute ischemia
- Metabolic disturbances
  - T4, K, Mg, DKA
- Drug proarrhythmia
- Drug overdose
- Fever (DCM and Brugada’s Syndrome)
- Post cardiac surgery
- ICD induced
  - Sub-optimal programming
  - Bi V pacing or pacing induced
  - “Psuedo-Storm” - inappropriate therapies

Approximately 33% cases

Heart failure 19%
CS 14%
Electrolyte 10%
none 57%

Eur Heart J 2006: 700-707
Electrical Storm

Triggers:
- Previous ventricular arrhythmias
- Genetic diseases (e.g., LQT)
- Poor ventricular function
- Electrolyte disturbances
- Ischemia
- Heart failure
- Changes/non-compliance to medications
- Excess alcohol

Structural Vulnerability

Electrophysiologic Vulnerability

Autonomic Vulnerability:
- Autonomic remodeling
- Tissue specific remodeling
Types of rhythm

- Electrical storm can initially be classified on the basis of 3 gross electrocardiographic (ECG) surface morphologies:
  - Monomorphemic VT,
  - Polymorphemic VT, or
  - VF
MONOMORPHIC VT
Monomorphic VT

- In most cases, electrical storm presents as sustained monomorphic VT that is associated with structural heart disease.

- Monomorphic VT occurs when the ventricular activation sequence is the same without any variation in the QRS complexes.

- It is due to electrical wavefront reentry around a fixed anatomic barrier, most commonly scar tissue after MI.

- Monomorphic VT due to wavefront reentry does not require active ischemia as a trigger, and it is uncommon in patients who are having an acute MI.
POLYMORPHIC VT
Polymorphic VT

- Polymorphic VT can be associated with a normal or a prolonged QT interval in sinus rhythm.
- Although polymorphic VT is most often associated with acute ischemic syndromes, it is also seen in the absence of organic heart disease.
- Patients who have acute myocarditis or hypertrophic cardiomyopathy may also present with polymorphic VT.
- Therapy for polymorphic VT and VF varies, depending upon the mode of initiation and the underlying QT interval in sinus rhythm.
- Electrical storm is often the initial manifestation of ischemia.
Targeting Approach

Momomorphic VT
Think reentry, Scar related

Polymorphic Ventricular Tachycardia

Ventricular Fibrillation
Think metabolic, drugs, ischemia, channelopathies brady
Fig. 1 Management of electrical storm.

ACLS = advanced cardiac life support; ECG = electrocardiographic; EP = electrophysiology; IABP = intra-aortic balloon pump; ICD = implantable cardioverter-defibrillator; LSGB = left stellate ganglion blockade; PVC = premature ventricular contraction; Revasc = revascularization; VF = ventricular fibrillation; VT = ventricular tachycardia
Treating Sympathetic over-activity

- Excess sympathetic activity seems to be a common element in factors precipitating and maintaining electrical storm

- Evidence of a role for the sympathetic nervous system:
  - Decreased baroreflex sensitivity in patients in storm\(^1\)
  - Beta blockers are a very effective treatment\(^2\)
  - Peak incidence during the early morning hours\(^3\)

1. Credner JACC 1998
2. Nademanee Circ 2000
3. Fries Int J Cardiol 1997
Reducing Sympathetic Drive: Beta-blockers

- They have the problem of being negatively inotropic
- Give intravenously in the titrated dose to max tolerated
- Use a short acting drug e.g. Esmolol if you are worried about heart failure or hypotension
- Bradycardia not usually a problem as the ICD can operate as a pacemaker
Sympathetic Blockade for Ischemic Electrical Storm
Nadamanee et al. Circ 2000

- 49 patients with electrical storm 10 ± 11 day post MI
- Electrical Storm: ≥20 VT/24 hours
- ACLS protocol:
  - Lidocaine
  - Procainamide
  - Bretylium
  - No beta blocker
- 27 patients had sympathetic blockade after initial ACLS protocol
  - 6 Stellate ganglion blockade
  - 21 beta blockers
Antiarrhythmic Therapy

- Beta-blockade
- Sotalol
- Amiodarone
- Mexiletene
- Quinidine
- Verapamil

OPTIC Trial. Connolly et al. JAMA 2006
Anti-Arrhythmic Drugs

- Amiodarone can be effective, particularly in combination with a beta blocker, avoid in Bradycardia induced VT and Long QT
- Class I drugs don’t work by themselves but are sometimes effective in combination with amiodarone in refractory cases
  - Flecainide, Mexilitine best of a bad bunch
  - Lignocaine for ischaemia
  - Problems with negative inotropic effects
  - Avoid polypharmacy
- Specific drugs
  - Isoprenalin for Brugada and Early Repolarization
  - Quinidine for Brugada, Short QT
  - Flecainide for CPVT and LQT3
  - Verapamil for IVF or fasicular
<table>
<thead>
<tr>
<th>Table 2. Treatment of electrical storm</th>
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<tbody>
<tr>
<td><strong>Pharmacological</strong></td>
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<tr>
<td>Sympathetic blockade</td>
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<tr>
<td>Beta antagonists</td>
</tr>
<tr>
<td>Left stellate ganglion blockade</td>
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<tr>
<td>Propofol</td>
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<tr>
<td>Antiarrhythmics</td>
</tr>
<tr>
<td>Combination therapy</td>
</tr>
<tr>
<td>Beta antagonist + class III antiarrhythmic</td>
</tr>
<tr>
<td>Beta antagonist + class III antiarrhythmic + class IB</td>
</tr>
<tr>
<td><strong>Non-Pharmacological</strong></td>
</tr>
<tr>
<td>Cather ablation</td>
</tr>
<tr>
<td>Overdrive pacing</td>
</tr>
<tr>
<td>Intraaortic balloon pump or extracorporeal life support</td>
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<tr>
<td>Heart transplant</td>
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</table>
How common are ICD storms?

- Seen more as ICD’s means survival to hospital
- Electrical storm affects 10-30% of patients with 2ndry prevention ICDs occurring at 4-9 months post implant.
- The incidence is 4-7% in patients with 1ry prevention ICDs occurring 18-24 months after implant.
- Similar incidence between ischaemic and non-ischaemic patients.
- The average number of tachycardia episodes ranges from 5 to 552²
- Recurrences are common 65% of ICM and 50% DCM

VT/VF Predicts Mortality in Patients with Heart failure – MADIT II

PATIENTS AT RISK

<table>
<thead>
<tr>
<th></th>
<th>Prior to Therapy</th>
<th>Post VT Therapy</th>
<th>Post VF Therapy</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>719</td>
<td>139</td>
<td>30</td>
</tr>
<tr>
<td>Survival</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Years</td>
<td>0.00</td>
<td>0.00</td>
<td>0.00</td>
</tr>
<tr>
<td>1.00</td>
<td>0.82</td>
<td>0.74</td>
<td></td>
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<tr>
<td>0.80</td>
<td>0.61</td>
<td>0.49</td>
<td></td>
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<tr>
<td>0.60</td>
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Aims of Management

• Prevention of future shocks
  – Detection and treatment of precipitants
  – General cardiac management
    • Appropriate HF care
    • Counseling and support
  – Device re-programming
  – Antiarrhythmic drug therapy
  – Catheter ablation
  – Neuraxial modulation
Patient with ICD Shock

Number of ICD Shocks?
- One
- Two or more
  - Assess Remote Monitoring Data (if available)

Worsening Clinical Status?
- (Syncope, Chest Pain, Heart Failure, Recurrent Rapid Palpitations, Confusion)
  - No
    - Schedule ICD Clinic Visit within 24-72h
  - Yes
    - Urgent Assessment Required including ICD interrogation

VT/VF Storm?
- Yes
  - Appropriate ICD Shocks?
    - No
      - Admit to hospital (monitored bed or CCU)
    - Yes
      - Assess need for hospital admission

- No
  - Assess / treat reversible cause
  - Optimize beta-blockade
  - Sedation / Anxiolytics
  - Initiate IV antiarrhythmic agent
  - If recurrent VT, consider general anesthesia, overdrive pacing (PMVT), catheter ablation
  - If refractory VT/VF, consider epidural anesthesia, cardiac sympathetic denervation, LVAD or cardiac transplantation

- Yes
  - Treat reversible cause
  - Optimize ATP programming
  - Ensure optimal beta-blocker dosage
  - Consider antiarrhythmic agent (Sotalol, Amiodarone)
  - Consider catheter ablation
  - Rule out lead malfunction, oversensing
Reverse Likely Precipitants

- Treat underlying ischaemia
- Correct electrolyte disturbance
  - K, Mg, Ca
- Prevent bradycardia
- Turn off LV pacing
- Reverse hyperthermia
- Anti-dotes if drug related
ICD programming during ICD storm

- Disable shocks
  - Avoidance of conscious shocks
  - Avoids treatment of non sustained arrhythmia

- Consider high pacing rates
  - Atrial pacing preferred

- Consider turning off LV pacing in CRT-D

- ATP may be reasonable if MVT
  - Adequate delay in therapy
ICD Reprogramming

• Overdrive pacing;
  – Prevent pause induced arrhythmias
  – Suppress PVCs
• Inactivate proarrhythmic features
  – Bi V pacing
  – Fix sensing issues
• Lengthen detection time – for NSVT
• Alter detection rate – For stable VT
• Turn on ATP, increase first shock
Management of Electrical Storm: Past Approach

- Antiarrhythmic drug therapy: 48 – 91% (Amiodarone)
- No specific action – 29%
- ICD reprogramming – 23%
- Heart failure treatment – 16%
- Ablation – 7%
- Revascularization: 3 – 11%
- Hyperthyroid treatment – 3%

Brigadeau F et al. European Heart J 2006 – 123 patients
Verma et al. JCE 2004 – 208 patients
Advanced Therapies

- Ablation
  - Endo/epicardial
  - Trans coronary ethanol
- Neuraxial Modulation
  - Thoracic epidural anaesthesia
  - Stelate ganglion blockade
  - Cardiac sympathetic denervation
  - Renal sympathetic denervation
- Haemodynamic support
  - IABP
  - ECMO
  - LVAD
  - Transplant

  May be required in acute situation, pre-discharge or electively

  Combination may be required

  Best choice of therapy will depend on aetiology and clinical situation

  Specialised centers
Ablation

- Catheter ablation has been shown to be effective for electrical storm
- Best results are from specialist centers with dedicated multidisciplinary team. On call 24/7.
  - Long procedures (4-6 hours not uncommon)
  - Risk of death, stroke, cardiac tamponade
  - Long term “cure” rates probably only 60-70%
  - Multiple VT morphologies and very rapid VT are the main problems- potential need for haemodynamic support
  - Changing techniques and technology
Fig. 2 Treatment of multiple implantable cardioverter-defibrillator (ICD) shocks.

AF = atrial fibrillation; ATP = antitachycardia pacing; EP = electrophysiology; IABP = intra-aortic balloon pump; SVT = supraventricular tachycardia; VF = ventricular fibrillation; VT = ventricular tachycardia
VT endocardial ablation

Classical activation mapping
Area of earliest activation
Patient needs to be stable haemodynamically
Termination of VT during ablation
Epicardial site

Commonly required in NIDCM, Channelopathies
Specialized approach
Usually under GA
Mapping and Ablation of Triggers Applied to Patients with Repetitive VF

VF not preceded by monomorphich VT
✓ **Purkinje** origin defined by sharp potential preceding wider muscle potential both during VPB and SR

✓ **Muscle origin**: absence of Purkinje activity during ectopy
Ablation Without Ectopy At The Time Of Mapping

Left HIS
RB
Right Purkinje
ICD
VF ASSOCIATED WITH BRUGADA OR LQT SYNDROME

- Follow-up: 17 ± 17 months under ICD monitoring

- Brugada syndrome (7 patients): Importance of RVOT(5/7) Elimination of VF or polymorphic VT in 6/7 patients

- LQT syndrome (5 patients): 3/5 from left Purkinje. No VF or polymorphic VT in 3/5 patients – 1 patient died of a non cardiac cause (AIDS) and one sudden death.
# Haemodynamic Support for Ablation

<table>
<thead>
<tr>
<th>Device</th>
<th>Mechanism</th>
<th>Advantages</th>
<th>Limitations</th>
<th>Contraindications</th>
</tr>
</thead>
<tbody>
<tr>
<td>IABP</td>
<td>Diastolic support and systolic unloading via deflation and inflation of balloon within aorta</td>
<td>Relative ease of insertion, Widespread availability, Lower cost, Smaller size vascular access (7.5–8 Fr)</td>
<td>Modest haemodynamic support (approximately 0.5 L/min), Function dependent on ECG triggers that assume regular stable rhythm—not ideal when in VT</td>
<td>Moderate to severe AI, Severe PVD, Aortic dissection or aneurysm</td>
</tr>
<tr>
<td>Impella 2.5/CP/5</td>
<td>Percutaneous left ventricle-to-ascending aorta axial pump</td>
<td>Single site of peripheral vascular access, No need for trans-septal puncture, Allow augmentation of cardiac output by 2.5 L/min (Impella 2.5), 3.5 (Impella CP) or 5 L/min (Impella 5)</td>
<td>Impella 2.5 and CP provides only partial LV support, Requires large arterial cannula, Electromagnetic interference with magnetic-based mapping systems, May require trans-septal LV mapping</td>
<td>Moderate to severe AI or AS, Severe PVD, Ventricular septal defect, Left ventricular thrombus, Mechanical aortic valve, Right ventricular failure</td>
</tr>
<tr>
<td>TandemHeart</td>
<td>Percutaneous left atrial-to-femoral artery bypass using external centrifugal pump</td>
<td>Can provide up to 5 L/min of cardiac output augmentation</td>
<td>Interference with trans-septal mapping usually mandates retrograde approach for VT mapping and ablation, Requires large venous and arterial accesses</td>
<td>Moderate to severe AI, Severe PVD, Ventricular septal defect</td>
</tr>
<tr>
<td>ECMO</td>
<td>Peripheral CPB using external membrane oxygenator system</td>
<td>Provides complete biventricular support, Useful in severe RV failure, Allows augmentation of cardiac output &gt;4.5 L/min</td>
<td>Increased complexity, requiring additional personnel and resources, large cannulae</td>
<td>Severe PVD, Uncontrolled bleeding diathesis</td>
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Thoracic Epidural Anesthesia Can Be Effective for the Short-Term Management of Ventricular Tachycardia Storm

Duc H. Do, MD; Jason Bradfield, MD; Olujimi A. Ajijola, MD, PhD; Marmar Vaseghi, MD, PhD; John Le, MD; Siamak Rahman, MD; Aman Mahajan, MD, PhD; Akihiko Nogami, MD, PhD; Noel G. Boyle, MD, PhD; Kalyanam Shivkumar, MD, PhD
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A VT Episodes Before and After TEA

B Shocks Before and After TEA

J Am Heart Assoc. 2017;6:e007080. DOI: 10.1161/JAHA.117.007080.
Efficacy of Stellate Ganglion Blockade in Managing Electrical Storm. Lingjin Meng

- The efficacy of percutaneous SGB for managing ES is not well understood.
- RESULTS Of 3,374 publications reviewed, 38 patients from 23 studies met study criteria (52.0 ± 19.1 years of age, 11 women, 17 with ischemic cardiomyopathy). Antiarrhythmics were used in all patients. Mean left ventricular ejection fraction was 31 ±10%. ES was triggered by acute myocardial infarction in 15 patients and QT prolongation in 7 patients. The most common local anesthetic used for SGB was bupivacaine (0.25% to 0.50%). SGB resulted in a significant decrease in VA burden (12.40 ± 8.80 episodes/day vs. 1.04 ± 2.12 episodes/day; p < 0.001) and number of external and ICD shocks (10.00 ± 9.10 shocks/day vs. 0.05 ± 0.22 shocks/day; p < 0.01). Following SGB, 80.6% of patients survived to discharge.
- CONCLUSIONS SGB is an effective acute treatment for ES. However, larger prospective randomized studies are needed to better understand the role of SGB in ES and other VAs.
Efficacy of Stellate Ganglion Blockade in Managing Electrical Storm. Lingjin Meng

A

1st rib
2nd rib
3rd rib
4th rib
Deflated lung
Sympathetic chain

B

Thoracic ganglia

C

6-Month Outcomes in Patients With Implantable Cardioverter-Defibrillators Undergoing Renal Sympathetic Denervation for the Treatment of Refractory Ventricular Arrhythmias

**Figure 2** Burden of Ventricular Arrhythmias Before and After Renal Denervation

![Graph showing burden of ventricular arrhythmias before and after renal denervation.](image-url)
Mortality After Electrical Storm

• Classification Deaths
  – Cardiac non-sudden: 46 - 56%
  – Non-cardiac: 20 - 32%
  – Sudden: 21%

• Increased mortality after Storm likely represents failing heart

Management After Storm

- Post Traumatic Stress
  - Poor QOL after shocks
  - Fear of activity/social situations
  - Anxiety/depression – medical Rx
  - Phantom Shocks/”Afraid to go to sleep” – reassurance
  - Request removal of device – reassurance
Thank you