Novel treatment (new drug/intervention; established drug/procedure in new situation)

Isoprenaline and quinidine to calm Brugada VF storm

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Summary

A 3-year-old man with an implanted cardioverter-defibrillator (ICD) for type 2 Brugada syndrome was admitted to coronary care unit with ventricular fibrillation electrical storm and consequent appropriate recurrent ICD shocks. The rhythm did not settle with conventional antiarrhythmics such as β -blockers and amiodarone. An isoprenaline infusion was set up with immediate stabilisation of rhythm. After a period of monitoring, the infusion was stopped and oral quinidine was started. He has remained free of problems for 1 year. This case-report summarises these novel treatments for electrical storm in Brugada syndrome and the mechanism behind them.

BACKGROUND

Electrical storms in patients with implanted cardioverter-defibrillator's (ICD) are very distressing for patients and clinicians alike and will be seen increasingly by non-electrophysiologists, as defibrillators become more common and patients present to non-specialist centres. This patient was given ineffective therapy on admission which may have potentiated the arrhythmia due to a lack of knowledge and experience. The successful treatment of isoprenaline was started on the basis of a case-report. By writing this up further evidence is provided for a very effective treatment that is widely available.

CASE PRESENTATION

A 31-year-old man was admitted outside of normal working hours to the emergency department of a district general hospital with repeated shocks from his ICD.

Eight months earlier, he had suffered an out of hospital ventricular fibrillation (VF) arrest. Resting ECG and echocardiography were normal and angiography revealed no coronary artery disease. A flecainide provocation test was performed and he was given a diagnosis of type 2 Brugada syndrome. An ICD was inserted. He had no significant other medical or family history.

The 8 months following ICD implantation were uneventful until the day of admission, when he had a shock

Figure 1 The resting ECG on admission to coronary care.

Figure 2 Examples of ventricular ectopy.

from his device at rest. On arrival to the emergency department he had a further shock and was moved to the coronary care unit. His admission ECG and routine bloods were normal (figure 1).

The admitting medical team had discussed with the on-call cardiologist and given 25 mg of oral metoprolol. This failed to prevent ongoing ICD shocks and the patient was commenced on intravenous amiodarone and sedated with morphine and diazepam, following further discussion with the on-call general cardiologist. Despite this he had three further shocks and repeated runs of ventricular bigeminy, ventricular ectopy and polymorphic ventricular tachycardia over the next 4 h (figures 2 and 3). Intravenous magnesium was added but the gentleman received further shocks and his rhythm did not settle.

Due to the time of day, we were unable to interrogate his ICD, but subsequent analysis showed that, all shocks were appropriate.

TREATMENT

General anaesthesia was considered as a bridge to the morning, when specialist electrophysiological advice could be sought, but alternative antiarrhythmics were investigated. A literature search revealed that, isoprenaline has been shown to settle electrical storms in Brugada syndrome in the electrophysiology laboratory.¹ It has also been used in acute settings.² An infusion was set up (1.5 mcg/min and titrated up.) There was one further episode of VF as the infusion was commenced but then the rhythm settled. The patient had no further arrhythmias.

After 4 days of observation and monitoring on CCU with no further arrhythmia, we started oral quinidine following specialist advice. The patient was discharged after a further short uneventful period of monitoring. No changes were made to the ICD.

Figure 3 Ventricular ectopy and a salvo of polymorphic ventricular tachycardia.

OUTCOME AND FOLLOW-UP

The patient has remained on quinidine for 1 year at last follow-up and has remained stable with no further electrical storms. Interrogation of his ICD has not revealed any ventricular arrhythmia or device therapy.

DISCUSSION

The Brugada syndrome predisposes patients to ventricular fibrillation, due to cardiac sodium channel abnormalities which tend to be of autosomal dominant inheritance. The sodium channel, voltage-gated, type V, α -subunit gene encoding the α -subunit of the cardiac sodium gene has been identified in 18–30% of Brugada syndrome cases (but not in the case discussed).

The defective myocardial sodium channels reduce sodium inflow currents and consequentially reduce the duration of the normal action potential. Phase 0 of the action potential, influx of sodium, is blunted and this results in reduction of calcium inflow and shortening of phase 2. The cells may therefore fail to conduct the action potential, because dramatically shortened phase 2 recovers excitability quickly. This combination of localised block and short refractory period gives rise to localised re-entry circuits and the potential of arrhythmias in the presence of ventricular premature beats. The effect is more pronounced when there are normal and abnormal sodium channels in the same tissue with heterogeneity of refractory periods. This also occurs between myocardial layers.^{3 4}

Isoprenaline and other sympathomimetic agents (orci-prenaline) to terminate electrical storms in Brugada syn-drome have been described in catheter lab and case studies.^{1 2} The mechanism is thought to be due to increas-ing intracellular calcium, stabilising and restoring the dome in phase 2 of the action potential and reducing the electri-cal heterogeneity responsible for Brugada syndrome. This stability reduces the susceptibility to VF triggered by pre-mature beats.⁵

Quinidine has been used for over 30 years as prophy-laxis in patients with idiopathic ventricular fibrillation⁶ and an 88% success rate has been shown in preventing VF induction in Brugada patients with inducible VF in one study.⁷ Its action is said to be due to inhibiting transient outward (I_{to}) sodium channels, leading to stabilisation of the action potential.³

Learning points

- ▶ VF storm in Brugada syndrome is rare but with increasing use of ICD's and consequent survival, patients are more likely to be seen by the general cardiologist or acute physician.
- ▶ Conventional antiarrhythmic agents have shown no benefit.⁴
- ▶ This case shows the benefit of isoprenaline in Brugada electrical storms acutely and the use of quinidine for prophylaxis.

Competing interests None.

Patient consent Obtained.

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