Correspondence

Lessons learnt from a tragic loss

Dear Sirs,

Sudden cardiac arrest (SCA) without overt heart disease is thankfully rare but nevertheless an incredibly emotive condition principally because of its inherent predilection for younger patients.[1] The recent case report by Westaby *et al.* with editorial by Sedgwick *et al.* highlights an important case whereby a young woman died following prolonged cardiopulmonary resuscitation attempts for refractory ventricular arrhythmias. The article concentrates on lessons learnt and potential improvements during the acute resuscitation phase. Principally the team examined the role and timing of the external LUCAS cardiac compression, ECMO, levosimendan and intra-aortic balloon counterpulsation. These technologies have a limited evidence base in this situation,[2–4] perhaps a reflection of the complexity of planning high quality studies in such circumstances.

A diagnosis of idiopathic ventricular fibrillation was reached yet the authors do not elaborate further into the differential diagnosis of refractory, catecholamine-sensitive ventricular arrhythmias pertinent to this case. In particular, one must question a diagnosis of catecholaminergic polymorphic ventricular tachycardia (CPVT) which can present not only with its hallmark of adrenaline sensitive bidirectional VT but also with cardiac arrest. Some authors have suggested that CPVT is the underlying pathology in up to 56% of cases of unexplained cardiac arrest.[5] Genetic mutations, principally of ryanodine or calsequestrin receptors, are also found in only 55–60% of CPVT cases[6] so even molecular autopsy can be unhelpful in nearly half of CPVT presentations. It is also noted that this girl had a history of syncope with a normal echocardiogram and 12-lead ECG, all hallmark features of CPVT.

In times of austerity one might therefore raise the question of cost efficacy of these expensive resuscitation technologies, particularly their widespread inclusion in the cardiac arrest situation. Possibly a more cost efficacious method would be that of screening for and increased education regarding inherited cardiac conditions causing SCA. In particular the repeated use of beta receptor agonists during the arrest situation only serves to worsen arrhythmias in CPVT as was seen in this case. Obviously this can be difficult to avoid and can potentially lead to a downward spiral of arrhythmia, poor cardiac output and further beta agonist use. Provocation testing at index syncope presentation in at risk individuals and their families is potentially more rewarding and should certainly be included in the lessons learned from this report.[7]

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The authors reply

Catecholaminergic polymorphic ventricular tachycardia (CPVT) was indeed considered in the differential diagnosis and a recommendation was made to the family to consider screening for the recognised ryanodine and calsequestrin receptor mutatations. However, none of the antecedent syncopal episodes had been adrenergically triggered (which would have constituted a potential 'red flag').

Furthermore, the fact that this patient's refractory VF was eventually controlled by a combination of beta blockers (plus lidocaine) and the withholding of further adrenaline administration does not point specifically to CPVT, as adrenergically-mediated recruitment of the Purkinje network may be an important perpetuating mechanism in electrical storms of diverse aetiologies. Interestingly, in this patient it was subsequently necessary to withhold the beta blocker and re-introduce inotropic support with adrenaline 48 hours later to facilitate weaning off ECMO support, but that unintended "catecholamine challenge" did not provoke any ventricular ectopy or non-sustained/sustained VT, which would make CPVT less likely as the underlying diagnosis.

As regards cost efficacy, we agree that patients with suspected ion channel disorders should receive appropriate investigation but that is quite separate from the goal of offering optimal circulatory support in rare cases like our patient who suffered haemodynamic collapse following refractory electrical storm. This patient presented with recurrent syncope but no 'red flags' in her baseline assessment and tests, and she was being (appropriately) investigated along conventional lines by prolonged ambulatory ECG monitoring with the option of an implantable ECG loop recorder if the symptoms continued and no diagnosis had been made.

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Provocative testing is only indicated for patients with syncope/TLOC if there is some suspicion of an inherited cardiovascular disorder based on ECG and/or clinical clues (e.g. adrenergic-triggering or a family history of premature sudden cardiac death). We are not aware of any evidence supporting its blanket use in patients with recurrent syncope/TLOC who are deemed at 'low risk' on the basis of current clinical algorithms. Unfortunately, this case is also a timely reminder of a key limitation of all such risk-stratification schemes in clinical practice, namely that 'low risk' is not the same as 'zero-risk'.

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