Successful resuscitation of out-of-hospital ventricular fibrillation cardiac arrest in an adolescent

一名称青少年院外心室纖維性顫動而心臟停頓的成功復甦

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Cardiac arrest in the paediatric age group is uncommon. Successful resuscitation of out-of-hospital cardiac arrest in children with subsequent hospital discharge and normal neurological outcome is even rarer. A 14-year-old girl who presented with recurrent convulsive syncope was misdiagnosed to be suffering from epilepsy. Subsequently she was witnessed to have cardiac arrest and was successfully resuscitated. Catecholaminergic polymorphic ventricular tachycardia was suspected to be the underlying cause of her symptoms. This case illustrated the important concept of the paediatric chain of survival in the prevention of death and improvement of survival in paediatric cardiac arrest. (Hong Kong j.emerg.med. 2010;17:482-487)

Introduction

Cardiac arrest in the paediatric age group is uncommon. It accounted for 3.6% of critically ill patients aged below 16 years presenting to a local emergency department (ED). The outcome of paediatric cardiac arrest is very poor. Only 4% of paediatric patients with out-of-hospital cardiac arrest could be discharged neurologically intact. In this report we described the successful resuscitation of a 14-year-old girl with out-of-hospital cardiac arrest who was previously misdiagnosed as having epilepsy. It illustrated several critical factors in the successful resuscitation process.

Case report

A 14-year-old girl presented with sudden collapse and twitching of all four limbs for about three minutes in school in 2009. She had up-rolling eyeballs but no cyanosis, tongue bite nor incontinence. She had brief
post-ictal drowsiness. She did not have family history of heart disease or sudden death. She was admitted to hospital for investigation. Baseline investigations, including serum electrolytes, electrocardiogram (ECG) and computed tomography of the brain (CT brain) were normal. She was stable and was discharged.

Twelve days later, she developed another episode of collapse with urinary incontinence. Although electroencephalography (EEG) did not show any epileptiform activity, she was diagnosed to have epilepsy. Sodium valproate was prescribed for “recurrent seizures”.

One month later, she was witnessed by her sister to have difficulty in breathing while walking in school. She became unconscious immediately with cyanotic lips, up-rolling eyeballs, arms twitching and leg extension. The whole event lasted for about 10 seconds. She was then found to be pulseless. Cardiopulmonary resuscitation (CPR) was initiated immediately. The paramedics arrived eight minutes later. Ventricular fibrillation (VF) was detected by the automated external defibrillator (AED) and was converted by one shock (Figure 1). Return of spontaneous circulation occurred soon after the successful defibrillation.

![Pre/Post-Shock ECG](image.png)

Figure 1. Automated external defibrillator record showing ventricular fibrillation converted to sinus rhythm by defibrillation.
She was transferred to the ED. She was comatose on arrival. The Glasgow Coma Scale score was 3/15. Her blood pressure was 99/67 mmHg with pulse rate 78 beats per minute. Her airway was protected by endotracheal intubation. Her 12-lead ECG in the ED showed sinus rhythm with corrected QT interval (QTc) of 459 milliseconds. There were no ECG features of ventricular pre-excitation or Brugada syndrome. The emergency CT brain was unremarkable. She was then transferred to the paediatric intensive care unit (ICU) for further management.

In the paediatric ICU, she had transient ventricular tachycardia (VT). A loading dose of amiodarone was given intravenously and she was sedated with morphine and midazolam infusions. Echocardiography did not reveal any structural cardiac abnormality. However, ventricular bigeminy, bi-directional VT, torsade de pointes (Tdp) and VF recurred when she underwent a painful procedure of putting up a central catheter (Figure 2). She was resuscitated successfully with multiple defibrillations. A total of 27 biphasic DC shocks at 150 to 200J were given. After each shock, polymorphic VT and VF recurred within a few seconds. The post-resuscitation ECG showed normal QTc of 450 milliseconds. Her ECGs were reviewed by a paediatric cardiologist. Catecholaminergic polymorphic ventricular tachycardia (CPVT) was suspected. Atenolol 50 mg daily was prescribed. Her cardiac rhythm was stabilised and mechanical ventilation was weaned off on the next day. Further history did not reveal any recent herbal medicine ingestion or drug overdose. Family ECG screening showed normal QTc. Her heart rate was around 40-50 beats per minute with occasional ventricular bigeminy and non-sustained Tdp after the

Figure 2. Cardiac monitoring record showing bi-directional ventricular tachycardia.
administration of atenolol. With a clinical diagnosis of CPVT, midazolam was continued for sedation.

She was then transferred to a paediatric cardiac centre. Amiodarone was gradually weaned off without any recurrence of ventricular arrhythmias. She did not have any neurological deficit and she was discharged ten days after hospitalisation. She was maintained on atenolol and exercise restriction was advised.

**Discussion**

Cardiac arrest in the paediatric age group is uncommon. The reported incidences of cardiac arrest were 8 to 20 per 100,000 children. Most paediatric cardiac arrests are precipitated by asphyxia or circulatory shock. In about 10% of cases, the presenting rhythm was VT or VF.3

The prognosis of paediatric cardiac arrest is very poor. Only 4% of paediatric patients with out-of-hospital cardiac arrest were discharged neurologically intact.2

As proposed by the American Heart Association, there are four critical interventions to prevent paediatric death in the concept of the Paediatric Chain of Survival.5 The interventions are: (i) prevention of arrest, (ii) early recognition of cardiac arrest and early CPR, (iii) early activation of the emergency medical service, and (iv) early advanced life support.

Since the prognosis of cardiac arrest in children is very poor, implementation of preventive measures may be the most effective way to reduce death in this age group. The patient in our case report presenting with recurrent convulsive syncpe was misdiagnosed as having epilepsy. In retrospect, her symptom was very brief. The post-ictal confusion or drowsiness was also short-lasting. Furthermore, the EEG done was normal. These evidences should alert us that her recurrent symptoms might not be due to neurological problem. With the advantage of hindsight, if the patient was referred for cardiological evaluation, an early diagnosis of ventricular tachyarrhythmia might be arrived at. Preventive treatment, for example antiarrhythmic drugs, may be able to reduce the risk of sudden cardiac arrest.

It was well recognised that it might be difficult to differentiate epilepsy from convulsive syncope. Up to 20% of the patients undergoing long-term follow-up in epilepsy clinics may not have epilepsy.6 Therefore, in patients with epilepsy who have atypical presentation, alternative diagnosis should be searched. For all patients admitted to the ED for seizure, detailed cardiovascular examination to exclude structural heart lesion should be performed. An ECG is mandatory for all cases of first seizure, and it should be considered also for subsequent episodes if the seizure is not typical of epilepsy. These measures aim to reduce misdiagnosis of potentially lethal cardiac arrhythmias, and may reduce sudden death in the young.

The second link in the chain of survival is early recognition of cardiac arrest and early CPR. In out-of-hospital cardiac arrests, only a few victims would receive bystander CPR. In a large study focussed specifically on out-of-hospital cardiac arrest in patients aged 35 years or below, bystander CPR only occurred in 34.4% of all cases.7

It was fortunate that the sudden collapse of the girl in our report was witnessed by her sister and the cardiopulmonary arrest was recognised. Immediate CPR was started by a school worker whose performance was crucial in the chain of survival. If there was no effective CPR, the response to defibrillation would not be the same. CPR improves survival by prolonging the duration of VF before asystole develops and increases the likelihood of successful defibrillation. For every minute without CPR, survival of witnessed VF cardiac arrest decreases 7% to 10%. When bystander CPR is provided, the decrease in survival is reduced by half, averaged 3% to 4% per minute from collapse to defibrillation.8

In Hong Kong, the degree of citizen preparedness in initiating CPR was poor according to a report in 2003.9 In this survey, only 12% of the responders had previous training on basic life support knowledge and CPR skill. To improve the survival of cardiac arrest in young
victims or all population, intensified educational efforts on CPR are warranted.

The third link of the chain of survival is early activation of the emergency medical service and early defibrillation. Our case illustrated the good performance of the emergency ambulance service in Hong Kong. From the activation of the service to defibrillation by AED, the duration was around 10 minutes only. As mentioned above, early CPR alone without defibrillation does not guarantee survival. CPR plus defibrillation within 3 to 5 minutes of collapse can result in survival rates as high as 49% to 75%.

In Hong Kong, training for the use of AED by paramedics was started in 1998. The promotion of AED in public locations for use by non-medical personnel is not without controversy. AEDs are clinically effective in specific locations such as casinos and airports. Their cost-effectiveness, however, remains to be proven.

In schools of Hong Kong, it is not mandatory to install AED. Cardiac arrest in school is rare, therefore, there is no requirement to have an AED for emergency use. In case of sudden collapse that occurs in school, access to AED relies on prompt activation of the emergency ambulance service. In a recent report from the United States, the effectiveness of school-based AED program was examined. It demonstrated that 64% of sudden cardiac arrest victims in school with AED survived to hospital discharge.

The last link of the chain of survival is early advanced life support in hospital. The successful outcome of neurologically intact hospital discharge is dependent on appropriate post-resuscitation care, accurate diagnosis and treatment of the underlying condition. The polymorphic VT and VF in our case were treated empirically by amiodarone after successful DC cardioversion. The differential diagnosis of CPVT was not considered initially. It was not until an unusual storm of VT/VF occurred which required a total of 27 DC shocks, that CPVT was suspected. Careful review of the ECG tracings showed that after each shock, polymorphic VT and VF recurred within a few seconds. This suggested either long QT syndrome or CPVT, both of which would have polymorphic VT or Tdp triggered by sudden adrenergic stimulation. In our patient, the first post-cardiac arrest ECG had borderline increase in QTc, of 459 ms, and all subsequent ECGs showed normal QTc. Family ECG screening also did not identify any first degree relatives having prolonged QT interval. The likelihood of congenital long QT syndrome in our patient is thus very low. On the other hand, the occurrence of frequent and multiform ventricular ectopics, and bidirectional VT was well reported in cases of CPVT.

CPVT was the provisional diagnosis in this case based on the ECG findings and clinical presentation. However, the presentation was not typical of CPVT because the cardiac arrhythmia was apparently not exercise or emotion related. Genetic study is needed for confirmation in our patient. CPVT is a form of malignant channelopathy. Mutations in genes encoding cardiac ryanodine type 2 receptor (RyR2) and less commonly calsequestrin 2 (CASQ2) were identified. The mutations cause inappropriate calcium ion release from the sarcoplasmic reticulum in cardiomyocytes leading to ventricular arrhythmias. Bi-directional VT, polymorphic VT and VF are the common manifestations. In Hong Kong, an adolescent with CPVT has been reported previously. Beta-receptor blocker is the mainstay of therapy in CPVT. The efficacy of beta-blocker to suppress the recurrence of VT is illustrated in our case, while amiodarone therapy failed.

In summary, we reported a 14-year-old girl with suspected CPVT who was misdiagnosed as epileptic. The successful resuscitation of the subsequent cardiac arrest in this case illustrated the importance of early recognition of the emergency, early activation of the emergency medical services, early bystander CPR, early use of AED and early advanced life support in hospital. Since survival to neurologically normal hospital discharge is uncommon in paediatric cardiac arrest,
prevention is the most important link in reducing death. It is recommended that in epileptic patients with atypical presentation, alternative diagnosis such as ventricular arrhythmia should be considered and diagnosed early to prevent sudden death.

References