CASE REPORTS

Myocardial hamartoma as a cause of VF cardiac arrest in an infant

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Cardiac arrests in children are fortunately rare and the presenting cardiac rhythm is often asystole. However, ventricular fibrillation (VF) can occur and may respond favourably to defibrillation.

CASE REPORT

A 7 month old girl was sitting in her high chair when she was witnessed by her parents to collapse suddenly at 1707 hours. They attempted cardiopulmonary resuscitation (CPR) and the ambulance crew arrived 7 minutes later, the cardiac monitor displaying VF. No defibrillation or medications were administered and she was rapidly transferred to her nearest emergency department (ED), arriving at 1723. She had been previously fit and well, and had not had respiratory difficulties prior to her arrest.

She was in cardiorespiratory arrest on arrival to the ED. She was promptly endotracheally intubated and intraosseous access obtained. Cardiac rhythm on arrival was asystole. She responded with a return of spontaneous circulation following 30 minutes of resuscitation with atropine, adrenaline, and sodium bicarbonate. She was transferred to the paediatric intensive care unit (PICU) on an adrenaline infusion.

On arrival in the PICU, further investigation, including a transthoracic echocardiogram, revealed a right atrial mass suspicious of a right atrial tumour, which was confirmed by transoesophageal echo. Following a stable night, the patient had a further VF arrest at 0700 the following morning, which responded favourably to adrenaline and a single 2 J/kg shock.

The patient was then transferred urgently to cardiac theatre and underwent resection of the tumour from the anterior leaflet of the mitral valve. The valve was replaced and a pericardial patch sited on the ventricular septum. Frozen sections taken peri-operatively confirmed a diagnosis of a myocardial hamartoma.

The infant returned to PICU paced and on inotropic support, and was discharged from PICU to the cardiac ward 2 weeks later. She was subsequently re-admitted to hospital 2 months later following a further VF arrest at home from which she was successfully resuscitated, and she was eventually discharged with an implantable defibrillator in situ. At the time of writing, she has a good neurological outcome.

DISCUSSION

VF as a presenting rhythm in adult cardiac arrest is common. In adult patients, arrhythmias are generally secondary to acquired cardiac lesions resulting from chronic problems such as hypertension and coronary artery disease. In children, most arrhythmias result from primary cardiac lesions.¹ A prejudice exists that ventricular fibrillation in children is rare; however, it may be the presenting rhythm in up to 10% of paediatric cardiac arrests.²

Current recommendations in paediatric patients do not emphasise early defibrillation because VF/ventricular tachycardia (VT) is thought to be much less common in these

patients. However a review by Young *et al*² found that the survival to hospital discharge in infants and children presenting with VF/VT was of the order of 30%, compared with only 5% of patients whose initial presenting rhythm was asystole. VF has also been demonstrated to be relatively more common in infants than any other paediatric age group ($p < 0.006$).¹ One study of over 500 000 children presenting to an ED over a period of 5 years found VF to be the third most common presenting cardiac arrhythmia of any origin in infants below the age of 1 year.¹ It has been suggested that a subgroup of patients (those < 1 year old) that would benefit from early defibrillation can be identified.²

In a study documenting the terminal rhythms of 100 paediatric patients, 22 had VF/VT, of whom 16 deteriorated to bradycardic/asystolic arrest as the final electrical event.³ Therefore, although children may appear to have VF/VT as a terminal rhythm less frequently than bradycardic/asystole, VF/VT may precede bradycardic/asystole in some, and may represent a window of opportunity for bystander CPR and defibrillation.² This was the case for our patient; her first arrest deteriorated to asystole, but prompt defibrillation during the second arrest in PICU resulted in a rapid return of cardiac output.

Current ambulance service and paramedic guidelines in our region prevent paramedic crews from defibrillating infants even if they record VF as the primary rhythm, because of the perceived rarity and possibility for misdiagnosis. However, following our patient's initial presentation and because of her high risk of recurrent VF arrest, associated with the myocardial hamartoma, it was felt that an exception should be made and this infant may now be defibrillated by paramedic crews in the event of a further VF arrest.

The current guidelines for the defibrillation of children using a monophasic defibrillator are 2, 2, and then 4 J/kg. As with adults, the higher energy is continued in the following cycles, unless a cardiac output is obtained. The use of automated defibrillators and biphasic defibrillators in children, however, remains controversial. The development of automated external defibrillators has not yet addressed the energy levels required to treat VF or VT in children or the reliability of these devices in the detection of VF and VT.⁴ Similarly, there are no current guidelines on the safety and energy required to defibrillate small children using a biphasic defibrillator. The use of lignocaine or other anti-arrhythmics in children with resistant VF is also controversial and current guidelines are not consistent worldwide.

Myocardial hamartomas are extremely rare. In a series of 40 infants and children presenting with primary cardiac tumours described by Takach *et al*,⁶ 16 were found to have myocardial hamartomas. Their ages ranged from 3 to 20 months, with a mean age of 1.1 years. Of the 16 patients, 15 presented with symptoms of cardiovascular collapse on admission (cardiac arrest or hypotension). All had ventricular tachycardias that were unresponsive to medical therapy. The infant described in our report presented with the

typical clinical and pathological features of a myocardial hamartoma.

The origin of these lesions is controversial and they have variously been described as histiocytoid cardiomyopathy,⁶⁻⁸ Purkinje cell tumours,^{9,10} arachnoidosis of heart muscle,⁸ myocardial hamartoma,¹¹ a developmental disorder of the Purkinje cells,¹² and viral induced lesions.¹³ It has been suggested that these tumours may serve as sites of abnormal or enhanced automaticity, or that they may distort the spread of excitation with possible facilitation of re-entry.¹⁰ The predominant subendocardial location, frequent involvement of the conduction system, positive histochemical staining for cholinesterase, and clinical presentation with uncontrollable arrhythmias have all been cited as supportive features of a developmental disorder of the Purkinje cells.¹⁰

The lesions have been described most commonly arising in the left ventricle, although atrial lesions have previously been described, and lesions on all four heart valves,⁷ most commonly the mitral valve.¹¹

In some patients (such as the infant described in this case) with extensive hamartomatous infiltration of the endomyocardium, complete resection cannot be accomplished. These patients may be at risk from recurrent ventricular tachycardias. Cryoablation is a technique that has been used to extend the area that can be effectively treated in this situation. In these patients ventricular tachycardia has not recurred in late post-operative follow up, and it seems that the mitotic potential is minimal and the recurrence of further infiltration unlikely.¹⁴

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A case report of an unusual sternal fracture

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Most sternal fractures are transverse, and a lateral chest radiograph is diagnostic. We report a case of vertical sternal fracture that was not seen on plain radiographs but was revealed using computed tomography (CT). Thoracic CT with coronal reformatted images can also demonstrate sternal fracture lines, supernumerary synchondrosis, and costosternal joint abnormalities.

Up to 8% of patients admitted with blunt chest trauma have sternal fractures,¹ caused primarily by anterior blunt chest trauma, usually from motor vehicle accidents when the chest strikes the steering wheel.² Mandatory seatbelt laws, now common in many parts of the world, are associated with an increased incidence of sternal fractures.³

Most sternal fractures are transverse, and a lateral radiographic view is diagnostic.² These fractures are often missed radiographically because lateral plain chest radiograph is not

usually obtained during the initial trauma evaluation. Using a computed tomography (CT) scan, we diagnosed a vertical midline sternal fracture that was not visible on standard posteroanterior and lateral chest radiographs.

CASE REPORT

A 36 year old man who had been involved in a motor vehicle accident 2 days previously was transferred to our emergency department (ED) from a rural state hospital because of persistent chest pain. He had reported that his car had hit road barriers while he was driving at a speed of approximately 90 km/h and using a restraining seat belt. He was alert and oriented. Vital signs on presentation were in normal limits.

The patient complained of substernal chest pain that increased with inspiration, and of mild abdominal pain. On physical examination, there was a 4-cm, sutured incision on the right temporal scalp area. Pain and tenderness upon palpation of the anterior chest wall over the sternum and slight bruising from the seat belt across the sternal area were