Don't Forget The Bubbles

CARDIAC RHYTHMS

Facilitators Guide

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Topic: **Cardiology & the ECG**
Author: **Anna McCorquodale**
Duration: **1-2 hours**
Facilitator level **ST4+ level used to seeing children acutely**
Learner level **Anyone involved in initial assessment of children with cardiac symptoms; paediatric trainees, emergency trainees, foundation doctors**

OUTLINE

- **Basics: 15 min with sharing of any departmental guidelines that exist**
- **Main session: 2x 15min case discussions**
- **Covering key take home messages**
- **Advanced session: 2x 30min grey case discussions, thoughts around follow up/review timelines, decisions around transfer, how to manage children with a complicated cardiac history**
- **Summary: 5 min**

PRE-READING FOR LEARNERS

The list of links should be sent out in advance to allow time for people to access the resources at a convenient time

https://www.gosh.nhs.uk/conditions-and-treatments/conditions-we-treat/supraven-tricular-tachycardia
patient information and includes a downloadable leaflet

https://dontforgetthebubbles.com/donovan-dwyer-svt-in-infants-at-dftb17/ (25 min)
lecture around treating SVT in children and what happens when it doesn't work

https://www.apls.org.au/algorithm-svt
standard ALSG guideline for UK management of SVT

https://www.escardio.org/static-file/Escardio/Guidelines/publications/NEONAT-guidelines-neonatal-slides.pdf
European guidelines on interpretation of neonatal ECGs

https://www.ajemjournal.com/article/S0735-6757(08)00240-4/pdf
interpreting the paediatric ECG including age related changes

https://www.youtube.com/watch?v=1o2vSR206Ts
(53 minutes) - US lecture on children's arrhythmias. Prioritise listening to the first 30 minutes which given a good overview of aetiology and treatment.

Palpitations are a common reason for children to present to the emergency department, the majority of these will be benign from a cardiac perspective and instead related to stress or anxiety. Appropriate management at the front door will reduce unnecessary investigations in those who can be reasonably reassured but ensure adequate investigation and/or onward referral in those where an underlying cardiac issue is more likely.
Palpitations (or descriptions thereof) are common but true arrhythmias are not. Add into this that the majority of children will be in normal sinus rhythm (NSR) by the time of assessment so to truly identify those who have something wrong we have to be confident in identifying arrhythmias where they are present and critical when analysing an ECG in NSR.

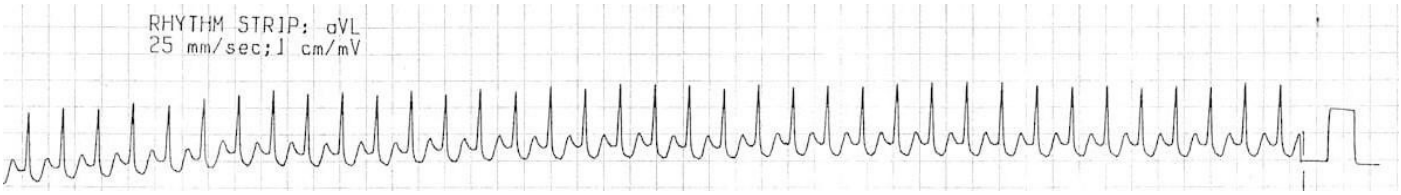
An approach to possible cardiac rhythm presentations:
1. Initial triage/ABC assessment
2. Immediate arrhythmia management where this is required
3. Thorough history: how long, how sudden, how often, how is it triggered?
4. Background medical history. This becomes of pivotal importance where there is a personal cardiac history and in some cases it can be quite historic.
Consider a teenager who had heart surgery as an infant, not all rhythm complications occur immediately. In some specific congenital heart diseases, the prevalence of arrhythmias is now being seen as a late surgical complication as children now survive into adulthood.
5. Family history. A cardiac family history should of course be sought, but think a little outside the box as well. The majority of the inherited rhythm disorders are autosomal dominant but their diagnosis relies on an electrically active heart. Some families have unexplained deaths during sporting events, an unusual number of car accidents, seizures in someone without a formal diagnosis of epilepsy. Where there are a number of unexplained deaths, start to be more critical about what this could mean.
A full examination is necessary but can be completely normal following the event.

If we look at some London based data: Palpitations – A cause for concern? **Stredder et al, Archives of Disease in Childhood 2016** we see that 58% were not asked about triggers, 30% were not asked about duration and, importantly for risk stratification, 59% were not asked for a family history of cardiac disease. All were examined and 98% had an ECG. Are we focussed on the immediate presentation and therefore not using all the possible information to hone in on the possibilities?

MAIN SESSION

CASE 1 (15 MINUTES)

A 5 year old girl presents feeling 'butterflies'. She has recently started school, her mother initially thought it was related, however, she has now mentioned this around once a fortnight and not necessarily on school days.
Today she complained of the same feeling and mum thought she looked pale so brought her to the ED for an opinion.
In triage her heart rate (HR) was 220 so she was taken to resus. Blood pressure (BP) is normal although she does look pale. She is talking to you about butterflies and points to her sternal notch.
This is her 3 lead ECG:



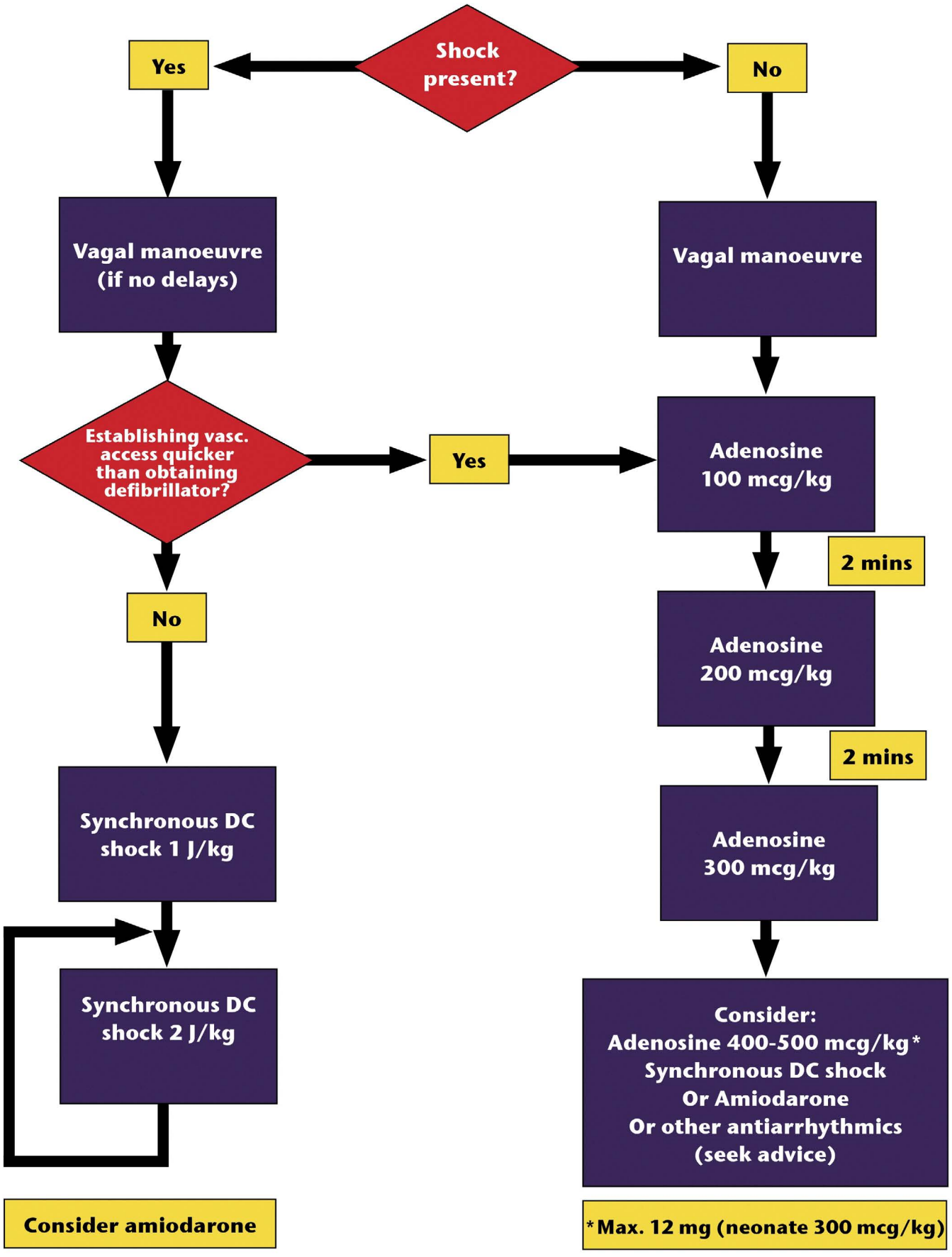
Discussion points:

What are your immediate management steps?

Due to her initial observations, this child has been triaged to resus and needs to be seen quickly. For anyone approaching a structured ABC would be a priority to establish whether shock was present or not.
A further part of the ABC assessment is the rhythm identification as this will guide ongoing management. SVT is actually an umbrella term for all arrhythmias arising from above the AV node, but what is typically meant when the phrase is used and indeed is also the most common subtypes of SVT is AVNRT/AVRT and this is depicted above. In young children it can be difficult to differentiate from sinus tachycardia. Typical features include:

1. HR >220bpm
2. Narrow complex regular tachycardia
3. P waves difficult to identify

In this case there is no shock present so we can proceed to the right branch of the ALSG guideline.



How often does the Valsalva manoeuvre work?

REVERT trial (adults); Appelboam et al, Lancet 2015 showed that by modifying the traditional valsalva manoeuvre to be done semi-recumbent with a passive leg raise immediately afterwards increased the success from 17% cardioversion to 43%. This has been trialled (out of hospital) in known children with SVT. **Upside-down position for the out of hospital management of children with SVT, Bronzetti et al, International Journal of Cardiology 2017** this showed that a modified position (upside down) improved cardioversion rates from 33% to 67%.

Assuming a progression to pharmaceutical therapy is required, what do you do and what do you do if the first attempt does not work?

We now have a patient who is in sustained SVT but with no features of shock. Continuing down the right branch of the ALSG guideline the first line pharmaceutical therapy is adenosine at 100mcg/kg.

What if this does not work? Here we are referring to the DFTB lecture link. Consider the 5 Ds:
Delivery – the half life is short, it needs to be given quickly and with a fast flush
Distance – the closer to the heart the better as it needs to be active at the point it reaches the heart.
Dose – the starting dose is 100mcg/kg but this may need increasing
Drugs – theophylline competes at the adenosine receptor
Diagnosis – is this really SVT?
The approach to adenosine dosing is clearly set out by ALSG, however, we do have an idea of how effective different doses are.

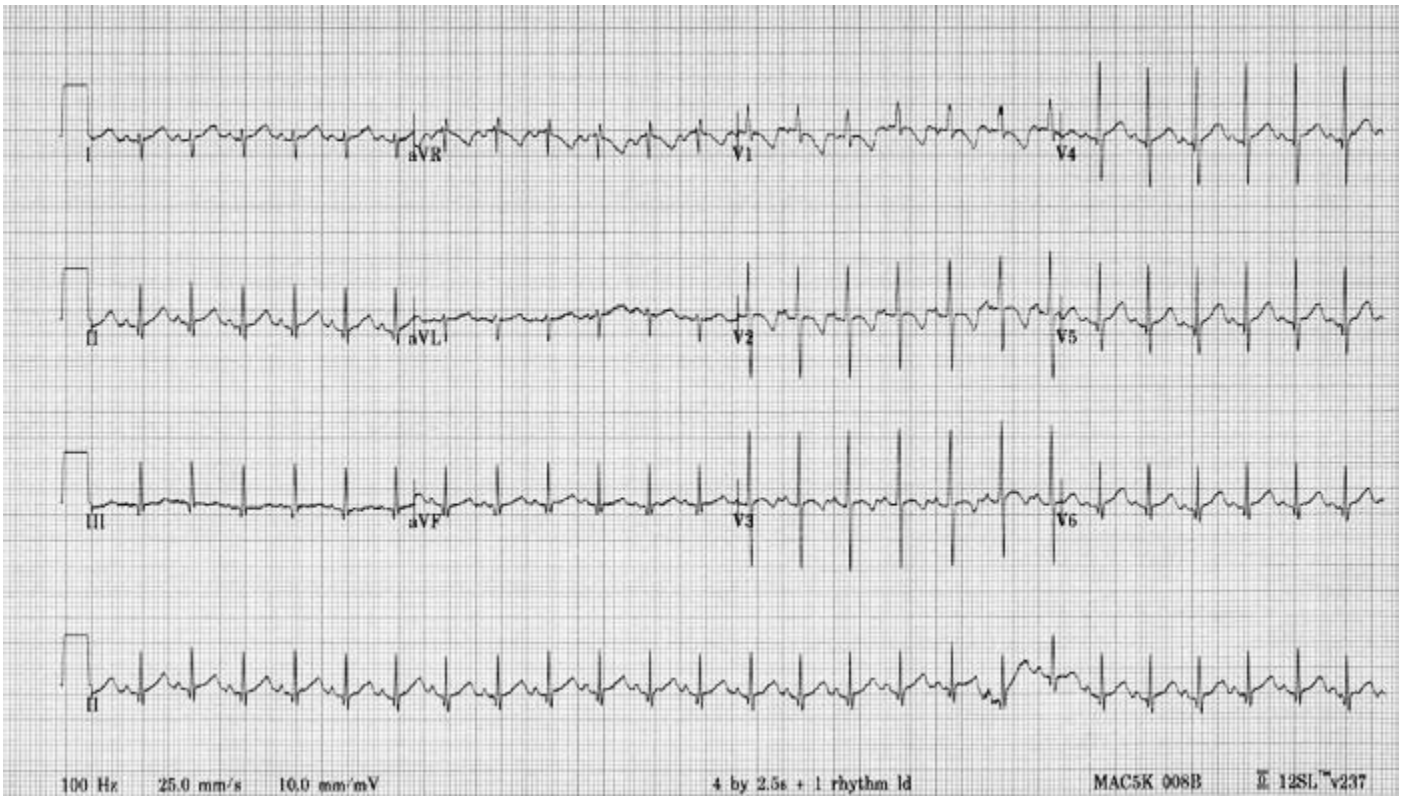
She cardioverted (phew!), what now? Discharge, if not now, then how do you plan towards this?

Many units will have different approaches to the management of these patients after cardioversion and a discussion around what your local policy is might be useful. Certain features are likely to guide the discussion:

1. Age of the child. There are two likely age groups for SVT, infants and early school years
2. 1st vs 2nd (or more) episode
3. Presence of a visible accessory pathway on the resting ECG i.e. a delta wave. Where this occurs, the chance of recurrence and worsening is higher. Cardiology services would be more likely to consider early EP study with ablation.
4. Anxiety/education of the family
5. Where do you consider the best place for follow up
a. Local general paeds vs PEC
b. Tertiary cardiology services, how do you link into this service?

CASE 2 (15 MINUTES)

A 2 month old (ex 34/40) boy presents with an episode at home of perioral cyanosis, with a possible 5 second lapse in breathing and vacant staring. The episode was reported to last around a minute. On arrival at hospital the child was well with normal observations.
Here is his ECG:



How do you classify a BRUE?

In 2016 the American Academy of Paediatrics issued a clinical guideline on brief unresolved unexplained events (BRUEs) which had previously been known as ALTEs. This guideline was condensed and summarised; **Tate et al, Archives of Disease in Childhood Education and Practice 2017.**
Definition:
BRIEF – duration < 1 min & age < 1 year
RESOLVED – patient returned to normal state in both observations and appearance
UNEXPLAINED – no identifiable medical condition
EVENT – one or more of cyanosis, pallor, breathing change (absent decreased or irregular), change in tone, altered responsiveness
The change in terminology and guideline change was to improve research opportunities and tighten up the objective criteria on which a diagnostic description is based. Choking or gagging in the history now precludes this

description being applied. The guidelines focus on stratifying children into high vs low risk and investigating accordingly. It should be emphasised that the evidence for what investigations to undertake is weak and based on the cumulative evidence from pre-existing ALTE literature.

What are the important investigations?

The important part in this decision is to be clear about the risk group into which a patient fits. Review of the literature surrounding ALTEs has revealed a subset of patients who met the BRUE criteria but were unlikely to have any serious underlying condition.

Low risk BRUE:

Age >60 days

Gestational age >32/40

Post conceptual age >45 weeks

First episode

No CPR given (by a trained medical provider)

No concerning history or examination findings (eg FHx sudden cardiac death)

Where the above features are present then investigation and hospital stay can be limited. Recommendations based on weak evidence include: a short period of monitoring (but not overnight hospital stay) and 12 lead ECG.

Do all infants with ALTEs need to be admitted? Claudius et al, Pediatrics 2007 here a US based study showed that of all admitted patients only 14% had a subsequent clinical course requiring admission. All of the cases had features which put them in the high risk subgroup of the new guidelines.

What pertinent parts of the ECG would you focus on and what are you trying to rule out?

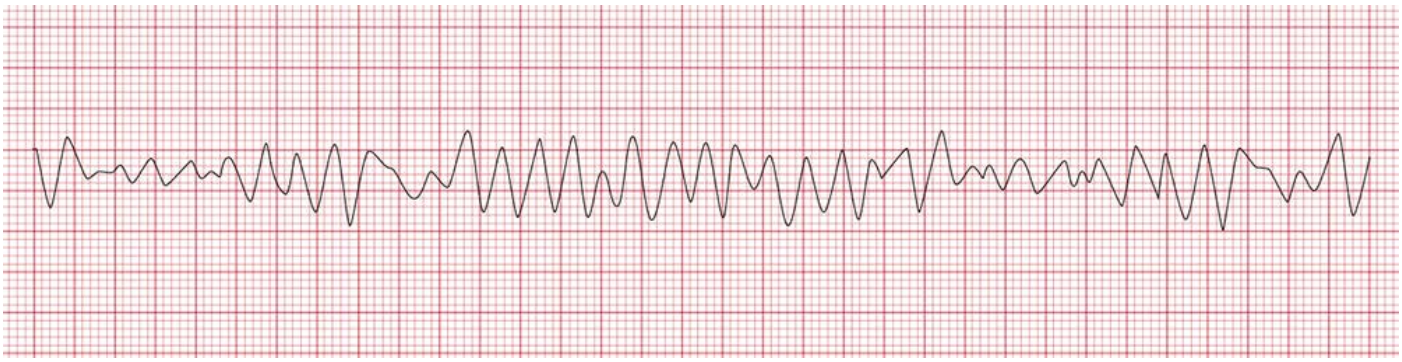
If the consensus guideline suggests (on weak evidence) that a 12-lead ECG is required, what is the chance of a positive yield and what are we looking for?

Cardiac testing and outcomes in infants after an ALTE, Hoki et al, Archives of Disease in Childhood 2012 showed significant cardiac disease was present in <1% of patients presenting with ALTE (although cardiac abnormalities were found in 4% overall). The only clinical predictor of cardiac disease was prematurity. Does this mean we do not need to do ECGs? Of those with significant findings, ventricular pre-excitation and cardiomyopathy were the diagnoses, and these were identifiable on ECGs at the time of the event.

It is also worth considering: **Cardiac channel mutations in SIDS: a population-based molecular autopsy study, Ackermann et al, Circulation 2002.**
When you use molecular testing 5% of children screened post-mortem for genetic mutations have genes associated with LQTS.
It therefore follows that if an ECG is to be done as routine (based on local guidelines) or if the circumstances of the individual patient means an ECG is indicated we should review the ECG as a whole, but specifically document the presence or absence of ventricular pre-excitation and additionally QRS/T wave changes that could represent either a cardiomyopathy or LQTS.

ADVANCED CASE 1

A 12 year old girl was heard to be coughing in the middle of the night.
Her mother went in to check and found her daughter coughing and distressed.
A few seconds later she fell forwards into her mother's arms and became lifeless.
Mum (first aid trained) gave back blows and started CPR. Paramedics attended within 7 min, this was her 3-lead ECG:



Following a single DC shock she reverted into sinus rhythm at home and was brought to hospital.

What are your initial actions on her arrival?

A child coming in from the community with a history of having a DC shock should trigger a cardiac arrest call (regardless of current clinical condition).
As she is now back in normal sinus rhythm the usual APLS structured approach to assessing her should be undertaken ensuring that the correct people are present should a further episode occur.

This child has had an out of hospital arrest and following stabilisation, subsequent care should be in an intensive care unit with access to cardiology service or designated CICU.

Priorities to discuss:

1. CVS stabilisation (including intubation if required)
a. Choice of anaesthetic agents should be discussed with a team with appropriate experience, depending on your situation, this may be the local intensive care or transport team. www.cats.nhs.uk/wp-content/uploads/guideline-intubation.pdf
2. Electrolyte management
3. Is a transfer necessary, if so, what arrangements need to be initiated?

Are there any specific conditions that might be implicated?

Based on the information from the rhythm strip and the response to DC cardioversion, this child has had a VF arrest. This aetiology in children is rare and when it does occur, is usually a degeneration of another malignant arrhythmia (such as VT). **Paediatric survivors of out-of hospital VF: etiologies and outcomes, Silka et al, Heart Rhythm 2018**: Survival rates for paediatric out of hospital arrest are 12-29%, however, in cases where there was a sustained return of circulation, more than 80% had an underlying cardiac aetiology identified. So what were these causes?

1. Primary electrical disease 33%
2. Cardiomyopathy 27%
3. Congenital heart disease 11%
4. Other cardiac 13%

89% of paediatric patients in this study survived to hospital discharge with 38% having a normal neurological outcome.

In this particular case we can think about the most likely cardiac causes of her arrest using the above statistics combined with the situation in which she had her arrest. Given that she is 12, a congenital cardiac condition seems unlikely with no prior history. This leaves two main groups:

Primary electrical disease – a rhythm disturbance caused by an underlying (genetically inherited) predisposition. This group of conditions can be called channelopathies and patients typically have a structurally normal heart. The most common are LQTS (multiple different subtypes) and Brugada syndrome. Certain subtypes of channelopathies are more likely to present during sleep.

Cardiomyopathies – this is a structural change in the heart which compromises function and can predispose to arrhythmias. Hypertrophic and dilated cardiomyopathies would be the most commonly encountered in paediatrics.

This girl needs a full cardiac workup, including an echo, which is unlikely to be done immediately in the ED. A full clinical examination, CXR and 12- lead ECG might provide some clues to whether this is primary electrical (J point changes, QTc, T wave morphology changes) or a cardiomyopathy (shifted apex, abnormal heart sounds, large cardiac shadow on CXR, large QRS complexes on ECG).

If we think carefully about the information we have:

- VF arrest
- No preceding symptoms
- Onset of symptoms at night (rest)

This makes a primary electrical disorder that presents at rest most likely here.

How might these present in a less catastrophic way to the ED?
Referring back to Paediatric survivors of out-of hospital VF: etiologies and outcomes, Silka et al, Heart Rhythm 2018, prior to the arrest, 26% of patients had pre-existing cardiac symptoms.
Signs and symptoms of channelopathies can start to appear in childhood, typically in the teenage years. These include recurrent syncope and in particular, syncopal events in unusual situations such as during exercise and when lying down. It is also important to be aware of the family inheritance pattern here. Most are autosomal dominant (although phenotypic features are widely variable) so asking a multi-generational family tree could reveal pertinent information.

Depending on the experience of the learning group, there is also a window here for a discussion about the subtypes of primary electrical disorder and typical presentation/ECG changes.

LQTS:
Congenital problem affecting ion channels within the heart. This can cause either a loss or gain of function in potassium/sodium channels depending on what the precise genetic abnormality is. There are many different types with identifiable mutations and some patients (around 30%) who have diagnostic investigations with no currently known genetic mutation identified. The majority of patients fall into the first three groups.

	Trigger	ECG	Channel affected	% events during sleep
LQTS 1	Exercise	Broad-based T wave	Potassium loss of function	3
LQTS 2	Emotion/loud noise	Bifid T wave	Potassium loss of function	29
LQTS 3	Sleep	Long isoelectric ST segment	Sodium gain of function	39

Brugada syndrome:
Congenital sodium channel mutation causing incomplete right bundle branch block and anterior precordial ST elevation. Autosomal dominant, however, males more likely to have phenotypical symptoms/ECG changes.

CPVT:
Catecholaminergic polymorphic VT. This is a rare condition where intrinsic catecholamine release related to stress/emotion causes runs of ventricular tachycardia. Children will often present with dizziness/syncope/presyncope during exercise.

What do you anticipate the ongoing plans to be for this patient and the family as a whole?

These children will need a full cardiac workup, depending on the echo results this will be extended to include exercise and pharmacological stress testing. From ED, we need to consider where the best place is for ongoing care and how she will safely get there. For most children, a transfer will be necessary.

As she has had an out of hospital arrest, the likely path would be (following cardiac work up) to be offered an implantable defibrillator (ICD).

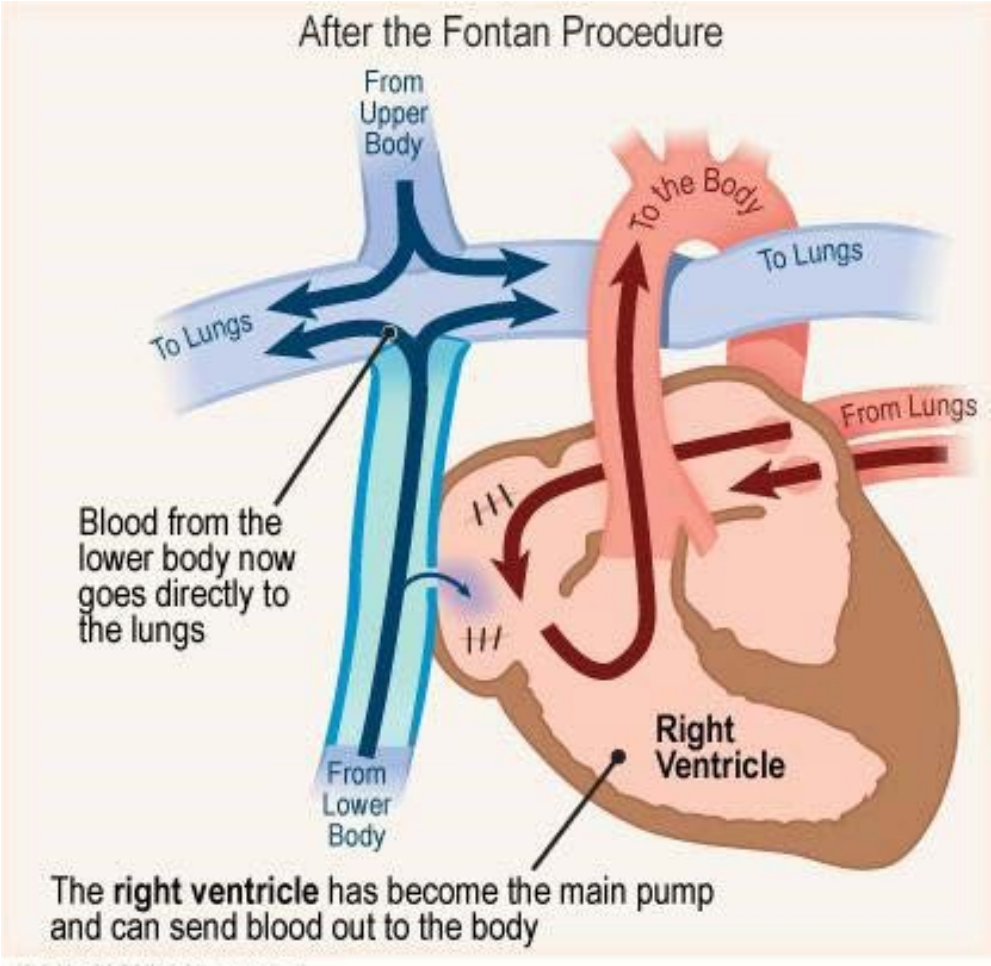
The likely diagnosis here is of a channelopathy and this has implications for the rest of the family. As the majority are autosomal dominant at an appropriate juncture, all first degree relatives should be referred for screening.
This can cause some amount of concern within the family and genetic counselling is appropriate, especially where further children are planned.

ADVANCED CASE 2

A 16 year old boy with a completed Fontan circulation for a congenital diagnosis of hypoplastic left heart syndrome. He is complaining of intermittent butterflies in his tummy. Mum has brought him to the ED as she has been unable to contact his specialist team and he was feeling dizzy whilst walking. There is no history of arrhythmias since discharge from hospital. You do not have access to his inpatient notes from the time of repair as this was done over a decade ago.
He is haemodynamically stable at triage.

What rhythm possibilities does this case suggest?

The Fontan circulation was an operation initially developed around 50 years ago to treat tricuspid atresia. It is now used to treat a wide variety of congenital cardiac lesions where achieving a dual ventricle circulation is impossible. Due to ongoing improvements in patient selection and the procedure itself, 80% of Fontan patients now survive to 30 years. The increasing longevity of this repair means that children survive into adulthood and long term complications are becoming more evident.



There are different potential surgical techniques that can be employed in a staged Fontan operation and the various suture lines involved can interfere with electrical conduction pathways. Below are the types of arrhythmia and their respective incidence ranges (for those that survive beyond 5 years) which are highly variable as it depends on the duration of follow up and the initial surgical approach used. Information on complications following Fontan has been brought together by: **Management of the Child and Adult with Fontan Circulation, Rychik et al, Ciculation 2019.** Numbers specific to arrhythmias are:
Sinus node dysfunction 15-60%
Supraventricular tachycardia 5-80%
Ventricular tachycardia 2-10%

What are the medical priorities for this child?

This is a complex case, and that is important to acknowledge, but the holistic care of this child relies on doing the basics well and involving the correct subspecialists at the right time. What you would do as a non cardiologist for the patient described above is in fact, very similar across all of the complex congenital heart conditions. The European Society of Cardiology released a position statement, Arrhythmias in Congenital Heart Disease, Hernandez-Madrid et al, Europace 2018 suggesting the following priorities:

1. Assess the clinical impact currently
- History & knowledge of anatomy where possible
- Haemodynamic status

In ED this is covered by an ABC assessment and the decision about whether immediate DC cardioversion is required.

2. Optimise haemodynamic status

3. Blood work up to include; infection markers, electrolytes (with replacement where necessary), TFTs

4. Urgent collaboration with the cardiac centre
- Information about how to optimise patient
- Pharmacological agents that have worked previously
- Transfer for ongoing management and detailed cardiac work up

It is also worth mentioning here that many of our traditionally used anti-arrhythmic agents have negative inotropic effects and can therefore confer significant disadvantage in those who already have ventricular dysfunction.
In a haemodynamically stable patient, pharmacological management should be discussed with the tertiary cardiac team.

QUIZ QUESTIONS: (10 MINUTES)

Question 1.

Supraventricular tachycardia. A child presents to emergency with a narrow complex tachycardia without CVS compromise that does not respond to vagal manoeuvres but does respond quickly to the first dose of adenosine. What age of child is least likely to require long term therapy or interventional treatment but still subsequently have long term resolution of symptoms?

A: 6 weeks old

B: 5 years old

C: 10 years old

D: 16 years old tomorrow (we will have to refer to adult cardiology anyway!)

Answer:

The majority of typical SVT, by which we mean narrow complex tachycardia responding to vagal manoeuvres and/or adenosine, in childhood is mediated by an accessory pathway. The presence of these pathways is documented inutero and they usually regress by around 20 weeks gestation. It has been inferred that the persistence of inutero pathways is the substrate for SVT presentations during childhood (including in those born prematurely).

For children presenting with SVT early, there is certainly a good chance of spontaneous regression of the pathway and therefore long term medical therapy or interventional treatment may not be necessary. Depending on what you read and the exact definition applied to 'recurrence' of SVT somewhere between 30-50% of infants will not require therapy beyond 12-18 months.
Those children that present later, and certainly those who represent with SVT in later childhood after a documented period of SVT during their infancy are much more likely to need medical or interventional therapies. In the older age groups medical therapies are increasingly being replaced by catheter ablations due to improving safety profile and reducing complications of this procedure.
A catheter ablation provides the potential for a 'cure' without the need for ongoing medical therapy.

Question 2.

BRUE. A child presents at 8 weeks old following a BRUE. Which of the following puts them in a higher risk category for a cardiac diagnosis?

A. Bruising to the upper arms seen during examination

B. Vomited milk and coughed immediately before the event

C. Recurrent UTIs

D. Mycoplasma pneumonia

Answer:

BRUE or brief resolved unexplained event is an event occurring in a child under the age of 1yr lasting less than one minute with one or more of; cyanosis/pallor, absent decreased or irregular breathing, change in tone, altered level of responsiveness. Formally, the term BRUE should only be applied where reasonable consideration has been given to whether there is an alternative cause or possible diagnosis, the list of possibilities is wide but for simplicity could be broken down by system not forgetting that inflicted injuries need to be considered in childhood. Bruising is common in childhood for both accidental and non-accidental reasons. The exception to this statement is non mobile children. Accidental bruising in non-mobile children is rare (0-1.3%) and therefore should be taken as a potential serious indicator of child abuse in this situation. The presence of vomiting immediately prior to symptoms suggests a gastrointestinal cause, possibly with stimulation of the vagal nerve. Parental reports of unexplained events need to be recorded accurately and should be addressed, but risk stratification should be based on a clinician determined objective review of the event. The appearance of 'dead' to each individual will vary based on their prior experience so exactly what is meant needs exploration. In this situation the risk specific to a cardiac diagnosis is raised due to the family history. This child has a first degree relative with a minimally explained medical history who has died suddenly and unexpectedly at an early age. The majority of inherited cardiovascular disorders are genetically inherited in an autosomal dominant fashion so this child would need further investigation into their cardiac status, both through a more detailed review of the father's medical history if available and direct testing of the child.

Question 3.

Corrected congenital heart disease. You encounter a child who had a complete repair of Tetralogy of Fallot during their first year of life in another country. They are now 12, in the UK and have no formal cardiac follow up. They present with the feeling of palpitations which occur intermittently throughout the school day but rarely during holidays. What is the likely cause of these palpitations?

A. SVT

B. Non sustained VT

C. Sinus tachycardia

D. All of the above

Answer:

The outcomes for congenital heart disease have improved and increasingly children are surviving longer following surgical procedures. This has even resulted in a new subspecialty for our colleagues in adult medicine (Adult Congenital Heart Disease/ACHD). As long term survival is evolving there are useful consensus documents to rely upon both for adults and children; **Pharmacological and non-pharmacological therapy for arrhythmias in the pediatric population: EHRA and AEPC-Arrhythmia Working Group joint consensus statement.** Children, young people and adults who have ever had cardiac intervention are higher risk for the development of arrhythmias than the general population.

The highest risk is in the immediate postoperative phase which remains the domain of cardiac intensive care units. Development of an arrhythmia substrate following cardiac surgery is dependent on a number of factors including but not limited to; pre-surgical cardiac status, age at surgery, site of scarring and development of fibrotic tissue and ventricular dilatation (which can change over time). Post repair of ToF both supraventricular and ventricular arrhythmias have been reported. Sudden cardiac death occurs in 1-3% of repaired ToF patients, the presumed mechanism being a ventricular arrhythmia. In addition to all of this we need to remember that this is an adolescent, in a new country, likely to have recently moved schools with a significant cardiac diagnosis, she can also develop sinus tachycardia related to extra cardiac reasons or non cardiac stimuli in the same way as any other adolescent. The answer, therefore, is d – this child has clear potential substrate for a cardiac arrhythmia and should be worked up and referred with this in mind but there needs to be holistic oversight of the other possibilities relevant to adolescents as well.

Take home tips

1. Haemodynamically stable SVT can be initially managed non pharmacologically, modifications to the Valsalva improve it effectivity

2. Infants who present with BRUE should be risk stratified.
ECGs should be specifically screens for ventricular pre-excitation, hypertrophic changes and changes in the T wave/QTc

3. Over 80% of out of hospital VF arrest have a cardiac aetiology, a full work up is necessary and often also extends to familial screening

4. Arrhythmias are a late complication of congenital heart disease seen more due to extended life expectancy in this group of patients

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