INVESTIGATION OF UNEXPLAINED CARDIAC ARREST

Background
- Acute Life Threatening Events (ALTE) in infancy rarely fall into this category, where airway, respiratory and neurological issues predominate. This guideline should be used where a malignant arrhythmia is documented. Malignant arrhythmia in the very young is less likely to be familial, but rather due to a de novo mutation.
- Maintain higher level of suspicion if ALTE occurs outside the usual age for sudden infant death syndrome (i.e. under 2 weeks and over 6 months) and if sudden collapse occurs whilst awake.
- Family investigations may not be necessary if a non-heritable diagnosis has been made. However, there is often a considerable time in which there is doubt, particularly during cerebral cooling, or after amiodarone when the QT interval may be very long. Furthermore it is likely that many family members will be present and available for testing; it can be difficult weeks later to achieve this.

Who requires Detailed Cardiac Investigation
- Documented unconscious VF or VT
- Any child who needed and responded to CPR
- Near drowning in a strong swimmer
- Seizure needing CPR or with documented VT/VF

Assessment:

Initial Assessment
- A paediatric cardiologist, ideally an electrophysiologist, should be involved early.
- Review and copy ambulance/defibrillator rhythm strips and get a clear history on the resuscitation from those who gave it.

History
- Document a clear history on presentation - activity at the time, and arrhythmic triggers including recent illness, fever (a trigger in Brugada syndrome), medications, drugs.
- Previous history of syncope or seizure?
- Any previous cardiac tests done?

Family History
- Draw up a family tree asking specifically for history of sudden death in young people, seizures (long QT and other channelopathies are commonly mistaken for epilepsy), syncope, and mention specifically the inherited heart diseases.
Investigations:

Cardiac Tests

- Obtain a number of 12 lead ECGs, at least daily, document temperature, electrolytes and QT prolonging medication at the time, write these on the ECG. (Hypothermia, low K+, Mg++, Ca++, amiodarone prolong the QT interval, fever may unmask the Brugada sign). Look particular for pre-excitation, the Brugada sign, repolarisation abnormalities, ischaemia, measure the QT interval in at least V5 and Lead II.

- Echocardiogram. Look carefully for coronary arterial anomaly and cardiomyopathy in particular.

- Consider endomyocardial biopsy if history, echocardiogram and ECG are suspicious for myocarditis. Cardiac MRI may also be helpful.

Blood Tests

- DNA extraction and storage (EDTA sample):
  - Genetic testing is guided by other investigations.
  - Contact the Cardiac Inherited Disease Group (CIDG) coordinator during daylight hours (cidgadmin@adhb.govt.nz or phone 23634 or discuss with Jon Skinner) if a familial cause is suspected or not excluded. Do not contact the CIDG if a non familial condition (such as myocarditis) has been made.
  - The coordinator will obtain family consent for DNA storage and possible testing, and help coordinate family investigations.

- Save blood for possible toxicology and metabolic screen and virology.

Family Investigations

- Explain to the family about inherited disease as a possible cause.
- Obtain expert review of a 12 lead ECG on both parents and all siblings. Further tests may be needed depending on detailed findings in the presenting child.
Provocative Tests
The family may find this stressful and they will need a clear explanation and guidance. It is wise to obtain the family’s consent.

These tests should be performed in ICU with the patient attached to a 12 lead ECG as well as ICU ECG and blood pressure monitoring. A cardiologist and intensivist should be present, with resuscitation equipment and drugs readily available.

Adrenaline
This test is important if an event has occurred with exercise (especially swimming) or excitement. Used if an exercise test cannot be done, this will unmask Catecholamine Precipitated Ventricular Tachycardia (CPVT) through induction of ventricular extra beats, bidirectional or polymorphic VT, and long QT syndrome since the QT interval fails to shorten and prolongs after the infusion is stopped. Adrenaline is given centrally at 0.05mcg/kg/min increasing in 5 minute intervals through 0.1 and 0.2 mcg/kg/min. 12 lead ECGs are taken at baseline and at the end of each stage. A QTc over 0.5 seconds is considered to be diagnostic of long QT syndrome; expert review is essential.

Ajmaline
This test is especially important in unmasking cardiac events which occur at sleep/rest or during fever. Unmasks the Brugada signature in those with the syndrome. Sensitivity of the test is improved by moving leads V4-6 (which are not needed) and placing them one intercostal space above V1-V3 and rename as V1-3 upper. Infuse 1mg/kg ajmaline over ten minutes; stop early if positive to minimise risk of inducing VF.

Adenosine
By blocking AV nodal conduction transiently this will unmask concealed pre-excitation due to an accessory pathway. Give 300mcg/kg/min by rapid bolus into a central vein. If heart block or pre-excitation is not seen, give a larger dose until it is.
Appendix

Measuring the Qtc

Brugada Signature

Wolf-Parkinson-White

Lown-Ganong-Levine