The role of the arrhythmia nurse in families with inherited cardiac conditions

A family case study

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“when sudden cardiac death occurs, NHS service have systems in place to identify family members at risk and provide personally tailored, sensitive and expert support, diagnosis, treatment, information and advice to close relatives”, (DOH 2005).
In 2006 the Liverpool Heart and Chest employed 2 HRNS, now a team of 4.

These posts were introduced following the launch of Chapter 8 of the National Service Framework with a main focus to providing a fair and high quality service “for arrhythmias and better prevention of sudden cardiac death”, (DOH 2005). It incorporated three quality requirements and 20 markers of good practice.
Nationally and Locally

- Nationally – arrhythmia care co-ordinators and arrhythmia nurse specialists are now relatively well established.
- BHF have funded over 30 posts.
- In Liverpool: LHCH the only hospital to have a team of specialist nurses dedicated to the care of arrhythmia patients.
- Locally: unaware of any community based arrhythmia care teams.
What do we do?

* Outpatient clinics – Heart failure, arrhythmia, support the device clinics
* Telephone information and support line
* Inpatient review
* Offer one to one support/advice and education
* Cardioversion service
* Facilitate and assist in provocation testing

**Nursing team profile**

- ICC and sudden cardiac death
- Heart failure management
- 4 specialist nurses and full time Secretarial support
- ICD, pacing care And support
- Arrhythmia care and support

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*Telephone information and support line*
*Inpatient review*
*Offer one to one support/advice and education*
*Cardioversion service*
*Facilitate and assist in provocation testing*
The ICC service focuses on inherited cardiac conditions associated with a risk of sudden cardiac death. These principally include the inherited channelopathies (long QT, Brugada syndrome, CPVT, etc.) and the familial cardiomyopathies (hypertrophic, arrhythmogenic right ventricular and dilated).
Case Study

• Referred in by the Coroner’s Officer
• Female age 21, fit and well
• Asymptomatic
• Sudden unexplained death of son aged 5
• Normal post mortem
Adrenaline challenge
Base line ECG
Adrenaline Challenge

End of stage 1
0.05mcg/kg
Adrenaline Challenge

End stage 2
0.10mcg/kg
Adrenaline Challenge
• Family screening

• Referral to Genetics

• On going review and support
Case Study

- Referred from DGH for ICD implant
- Diagnosed long QT syndrome (type 2)
- Genotyped KCNH2
Copy of family pedigree on WB

LONG QT (KCNH2) = 3

WB aged 22
Age 3 years.
Ish 1 mesh.
• ICD explanted following lead fracture and inappropriate shocks
• Stopped taking beta blocker
• Housing issues
• Passed away aged 22yrs
The Patient Journey

The Heart rhythm should take the journey with them, offering advice and support throughout.
Wilson (2009) believed that “since the publication of the NHS Plan (DOH 2000), healthcare professionals have been encouraged to ensure that ‘the patient remains at the centre of everything we do’. In essence, all patient processes should be developed around the needs of the patient, not for the ease of the staff that provide the care”. A similar statement had been addressed in a later document from the DOH: High Quality for all (DOH 2008), whereby it suggested that organisations should cover safety, effectiveness (including timeliness of provision) and innovation.
Would the patient get from A to B without us

Undoubtedly YES!!!!!

• However......
• Named point of contact to provide precise and expert information, support, empathy
• Improves the overall patient experience
Any Questions

Questions are guaranteed
In life answers aren’t
What do we do

• Meet with individual
• Reassure and discuss tests required
• Provide contact details
• Refer or involve other organisations CRY, SADs, STAR, HRUK
• Take detailed family tree
• Liaise with genetic team
• Support both the patient and family
• Provide written information
A Provocation test is whereby a drug either Adrenaline or Ajmaline (in some units they use flecainide) and in some cases both drugs are required, they are infused intravenously in accordance with the trust protocol. The trust has two protocols which were devised by the electrophysiology department with some input from the heart rhythm nurse specialists, achieved prior to the nurse specialists overseeing the test. If both are required then Adrenaline is employed first.

Adrenaline tends to be infused over 25 minutes with a gradual increase through up titration of the infusion every five minutes with half an hour cardiac monitoring following completion of the test. As opposed to Ajmaline whereby it is administered over a ten minute period with an hour monitoring afterwards, while some units will give this as a bolus.

There are two protocols: the ‘shimizu protocol’ and the ‘Mayo protocol’ that have performed studies on individuals in respect to adrenaline infusion and the identification of concealed LQTS. Vyas & Ackerman (2006) revealed that both models ‘expose a degree of accuracy’ and that it is a useful diagnostic tool in those suspected of concealed LQTS. However, Ackerman (2008) points out that the Mayo protocol demonstrates a 75% predictive value.

Risk Stratification

“Genetic testing is often useful in probands with a clinical diagnosis of LQTS to provide more accurate risk stratification and to guide therapeutic strategies.”

“It has been shown that the interplay between genetic defect, QT duration, and gender may provide an algorithm for risk stratification.”

Family Testing

“Genetic analysis is very important for identifying all mutation carriers within an LQTS family.”
Arrhythmogenic Triggers Differ by LQTS Subtype

- Knowing a patient’s LQTS subtype will help to determine appropriate lifestyle modifications, decreasing the risk for cardiac events. 

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References


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