Diagnosis in a 1stdegree relative (including children)

Referral to **ICC Clinic**   
**(Urgent if arrhythmia symptoms/seizures/syncope)**

**Management**

Appropriate to underlying condition – otherwise discharge

For children consider age related penetrance prior to discharge

**Follow-up**

Local arrhythmia service  
Database link to ICC for outcomes

Paediatric follow-up appropriate to age related penetrance

Family History, 12 lead, 24hr and Exercise ECG, imaging  
 +/- provocative testing, SaECG

*Exclude drugs****\*****, metabolic and structural disease as cause of phenotype*

-ve or not done

**\*** For drugs list visit

[www.azcert.org](http://www.azcert.org) or [www.sads.org.uk/drugs\_to\_avoid.htm](file:///\\ad.ucl.ac.uk\slms\home3\rmhasdi\Downloads\www.sads.org.uk\drugs_to_avoid.htm)

and [www.brugadadrugs.org](file:///C:\Users\ebehr\Dropbox\AICC%20and%20SLICC\AICC%20meeting%202013\www.brugadadrugs.org)

**VUS** = variant of unknown significance – pathogenic effect can be assessed by segregation study of variant and phenotype

+ve

Diagnosis and risk evaluation

ECG  
Discharge

Mutation +ve

Mutation -ve

Segregation analysis in family

Cascade Screening of Family

Diagnostic evaluation - 12 lead, 24hr and Exercise ECG, imaging  
 +/- provocative testing, SaECG

VUS

Low likelihood of diagnosis

Mutation Analysis in Proband

Phenotype

Probable or definite diagnosis (e.g. LQTS, HCM)

**Management**

Confirm diagnostic status

Lifestyle advice**\***

Assess SCD risk

Develop management plan