Version 1, 24 September 2019



#### **Arrhythmia Syndromes**

#### Selection Criteria – Arrhythmia Syndromes

Exclusion criteria:

- Deceased individuals
- NGS-based test performed from 2013 onwards
- Individuals from families with a known disease-causing variant
- Unascertained sudden death

For BrS, MUST meet diagnostic criteria for dx AND must be able to tick at least 1 box from at least 2 of the following categories: Age criteria, FHx criteria, Clinical criteria. PLEASE NOTE: FHx can be considered standalone if confirmed, i.e. if diagnostic criteria AND family history criteria are met, the patient is eligible.

For LQTS, MUST meet diagnostic criteria for dx:

- LQTS risk score  $\geq$  3.5 in the absence of a secondary cause for QT prolongation and/or a QTc interval  $\geq$  500 ms in repeated 12-lead ECG and in the absence of a secondary cause for QT prolongation.

For CPVT, MUST meet diagnostic criteria and  $dx \le 40$ yo:

- Structurally normal heart, normal ECG & unexplained catecholamine-induced bidirectional VT or polymorphic ventricular beats or VT in an individual < 40 or relative of a CPVT case with normal heart who manifests exercise induced PVCs or bidirectional polymorphic VT.

exercise induced PVCs or bidirectional polymorphic VT.
Diagnosis
BrS (go to i.) LQTS (go to ii.) CPVT (go to iii.)
i. BrS
Diagnostic criteria:
(MUST meet diagnostic criteria for dx)
HRS/EHRA/APHRS Expert Consensus Statement at any age
Family history criteria:
(If family history AND diagnostic criteria are met, the patient is eligible)
<ul> <li>≥ 1 first or second degree relative with documented BrS</li> <li>≥ 1 first or second degree relative with sudden death before 50 (must attempt to obtain PM report, death certificate and/or medical records to exclude other causes of death)</li> </ul>
Clinical criteria:
Spontaneous type 1 pattern Previous OHCA Induced type 1 pattern (OHCA = out-of-hospital cardiac arrest)
ii. LQTS
Diagnostic criteria: (MUST meet diagnostic criteria for dx)
<ul> <li>LQTS risk score ≥ 3.5 in the absence of a secondary cause for QT prolongation</li> <li>QTc interval ≥ 500ms in repeated 12-lead ECG and in the absence of a secondary cause for QT prolongation</li> </ul>
iii. CPVT
Diagnostic criteria:
(MUST meet diagnostic criteria for dx)



or polymorphic ventricular premature beats or VT in an	ned exercise or catecholamine-induced bidirectional VT individual ≤ 40 yo, OR fests exercise induced PVCs or bidirectional polymorphic
VT	rests exercise induced 1 ves of bidirectional polymorphic
Age criteria:	
Must be dx ≤ 40yo	
Clinical History	
Height (cm)	Weight (kg)
Calculated total Body Surface Area (m2)	
Date of diagnosis: (If unsure of the exact date please set to January 1 of the year the event took place and set the "Exact date" option to "No".)	Is this an exact date? Yes No
Symptoms at diagnosis: Yes No Shortness of breath	Heart failure
Chest pain Pre-syncope Syncope Palpitations  Number of other additional symptoms:	Heart failure OHCA Seizure(s) Epilepsy Other
, ,	<u> </u>
First additional symptom:  *SNOMED CT Capture Field  First additional symptom description (if code not	
found):	
Second additional symptom: *SNOMED CT Capture Field	
Second additional symptom description (if code not found):	
Asymptomatic list	☐ Diagnosed on family screening ☐ Incidental diagnosis; details:
Other Conditions	
HTN Diabetes CAD Cancer Syndrome diagnosis Metabolic conditions Neuromuscular conditions	☐ AF ☐ Previous VT ☐ Conduction system abnormalities ☐ Ventricular arrhythmias ☐ OHCA (other than at presentation) ☐ Other (Do not include the principal diagnosis)
HTN  First LITN data of diagnosis	
First HTN date of diagnosis	
Is this an exact date?  Diabetes	Yes No Date not known
Date Diabetes diagnosed	
Is this an exact date?	Yes No Date not known
CAD	



Number of coronary artery disease (CAD) conditions:	
First CAD type:  *SNOMED CT Capture Field	
First CAD type description (if code not found):	
First CAD date of diagnosis:	
Is this an exact date?	Yes No Date not known
Second CAD type:	
*SNOMED CT Capture Field	
Second CAD type description (if code not found):	
Second CAD date of diagnosis:	
Is this an exact date?	Yes No Date not known
Cancer	
Number of cancers:	
First cancertype:	
*SNOMED CT Capture Field	
First cancer type description (if code not found):	
First cancer date of diagnosis:	
Is this an exact date?	Yes No Date not known
Second cancertype:	
*SNOMED CT Capture Field	
Second cancer type description (if code not found):	
Second cancer date of diagnosis:	
Is this an exact date?	Yes No Date not known
Chemotherapy drugs:	Yes No
Syndrome Diagnosis	
Syndrome diagnosis: *OMIM Capture Field	
Syndrome diagnosis (if code not found):	
Syndrome diagnosis date of diagnosis:	
Is this an exact date?	Yes No Date not known
Metabolic Conditions	res No Date not known
Metabolic conditions:	
*OMIM Capture Field	
Metabolic conditions (if code not found):	
Metabolic conditions date of diagnosis:	
Is this an exact date?	Yes No Date not known
Neuromuscular Conditions	
Neuromuscular conditions:	
*OMIM Capture Field	
Neuromuscular conditions (if code not found):	



Neuromuscular conditions date of diagnosis:	
Is this an exact date?	Yes No Date not known
AF	
AF condition	
*SNOMED CT Capture Field	
AF conditions (if code not found):	
AF date of diagnosis:	
Is this an exact date?	Yes No Date not known
Previous VT	
Previous VT:	
*SNOMED CT Capture Field	
Previous VT (if code not found):	
Previous VT date of diagnosis:	
Is than an exact date?	Yes No Date not known
Conduction System Abnormalities	
Conduction system abnormalities type:	
*SNOMED CT Capture Field	
Conduction system abnormality type description (if	
code not found):	
Conduction system abnormalities diagnosis date	
Is this an exact date?	Yes No Date not known
Ventricular Arrhythmias	
Ventricular arrhythmia type:	
Ventricular arrhythmia type: *SNOMED CT Capture Field	
Ventricular arrhythmia type: *SNOMED CT Capture Field  Ventricular arrhythmia type description (if code not	
Ventricular arrhythmia type: *SNOMED CT Capture Field  Ventricular arrhythmia type description (if code not found):	
Ventricular arrhythmia type: *SNOMED CT Capture Field  Ventricular arrhythmia type description (if code not found):  Ventricular arrhythmia date of diagnosis:	
Ventricular arrhythmia type: *SNOMED CT Capture Field  Ventricular arrhythmia type description (if code not found):  Ventricular arrhythmia date of diagnosis:  Is this an exact date?	Yes No Date not known
Ventricular arrhythmia type: *SNOMED CT Capture Field  Ventricular arrhythmia type description (if code not found):  Ventricular arrhythmia date of diagnosis:  Is this an exact date?  Out of hospital cardiac arrest (OHCA)	
Ventricular arrhythmia type: *SNOMED CT Capture Field  Ventricular arrhythmia type description (if code not found):  Ventricular arrhythmia date of diagnosis:  Is this an exact date?  Out of hospital cardiac arrest (OHCA)  OHCA	Yes No Date not known  Yes No
Ventricular arrhythmia type: *SNOMED CT Capture Field  Ventricular arrhythmia type description (if code not found):  Ventricular arrhythmia date of diagnosis:  Is this an exact date?  Out of hospital cardiac arrest (OHCA)  OHCA  OHCA date:	
Ventricular arrhythmia type: *SNOMED CT Capture Field  Ventricular arrhythmia type description (if code not found):  Ventricular arrhythmia date of diagnosis:  Is this an exact date?  Out of hospital cardiac arrest (OHCA)  OHCA	
Ventricular arrhythmia type: *SNOMED CT Capture Field  Ventricular arrhythmia type description (if code not found):  Ventricular arrhythmia date of diagnosis:  Is this an exact date?  Out of hospital cardiac arrest (OHCA)  OHCA  OHCA date:	Yes No
Ventricular arrhythmia type: *SNOMED CT Capture Field  Ventricular arrhythmia type description (if code not found):  Ventricular arrhythmia date of diagnosis:  Is this an exact date?  Out of hospital cardiac arrest (OHCA)  OHCA  OHCA date:  Other  Number of other additional conditions:	Yes No
Ventricular arrhythmia type: *SNOMED CT Capture Field  Ventricular arrhythmia type description (if code not found):  Ventricular arrhythmia date of diagnosis:  Is this an exact date?  Out of hospital cardiac arrest (OHCA)  OHCA  OHCA  OHCA date:  Other  Number of other additional conditions:  First additional condition:	Yes No
Ventricular arrhythmia type: *SNOMED CT Capture Field  Ventricular arrhythmia type description (if code not found):  Ventricular arrhythmia date of diagnosis:  Is this an exact date?  Out of hospital cardiac arrest (OHCA)  OHCA  OHCA  OHCA date:  Other  Number of other additional conditions:  First additional condition: *SNOMED CT Capture Field	Yes No
Ventricular arrhythmia type: *SNOMED CT Capture Field  Ventricular arrhythmia type description (if code not found):  Ventricular arrhythmia date of diagnosis: Is this an exact date?  Out of hospital cardiac arrest (OHCA)  OHCA  OHCA  OHCA date:  Other  Number of other additional conditions:  First additional condition: *SNOMED CT Capture Field  First additional condition description (if code not	Yes No
Ventricular arrhythmia type: *SNOMED CT Capture Field  Ventricular arrhythmia type description (if code not found):  Ventricular arrhythmia date of diagnosis:  Is this an exact date?  Out of hospital cardiac arrest (OHCA)  OHCA  OHCA  OHCA date:  Other  Number of other additional conditions:  *SNOMED CT Capture Field  First additional condition description (if code not found):	Yes No
Ventricular arrhythmia type: *SNOMED CT Capture Field  Ventricular arrhythmia type description (if code not found):  Ventricular arrhythmia date of diagnosis:  Is this an exact date?  Out of hospital cardiac arrest (OHCA)  OHCA  OHCA  OHCA date:  Other  Number of other additional conditions:  First additional condition: *SNOMED CT Capture Field  First additional condition description (if code not found):  First additional condition date of diagnosis:	Yes   No
Ventricular arrhythmia type: *SNOMED CT Capture Field  Ventricular arrhythmia type description (if code not found):  Ventricular arrhythmia date of diagnosis:  Is this an exact date?  Out of hospital cardiac arrest (OHCA)  OHCA  OHCA  OHCA date:  Other  Number of other additional conditions:  First additional condition: *SNOMED CT Capture Field  First additional condition description (if code not found):  First additional condition date of diagnosis:  Is this an exact date?	Yes No
Ventricular arrhythmia type: *SNOMED CT Capture Field  Ventricular arrhythmia type description (if code not found):  Ventricular arrhythmia date of diagnosis:  Is this an exact date?  Out of hospital cardiac arrest (OHCA)  OHCA  OHCA date:  Other  Number of other additional conditions:  First additional condition: *SNOMED CT Capture Field  First additional condition description (if code not found):  First additional condition date of diagnosis:	Yes   No



Second additional condition description (if code not found):	
Second additional condition date of diagnosis:	
Is this an exact date?	Yes No Date not known
Echo Parameters at Diagnosis	
Echo performed?	<ul><li>Not performed</li><li>Normal</li><li>□ Abnormal</li><li>□ Unknown</li></ul>
Describe abnormalities	
Electrophysiological Parameters	
Resting QTc at diagnosis:  NB: ensure this measurement is off QT prolonging drugs and not immediately post arrest.	(msec)
Exercise stress test (EST) performed?	Yes No Unknown
EST type	☐ Bruce ☐ Sprint
EST result	Normal Abnormal QTc in recovery Blood pressure drop during exercise VT Ventricular ectopic beats Other arrhythmia
Longest QTc in recovery:	(msec)
VT:	<ul><li>☐ Monomorphic</li><li>☐ Polymorphic</li><li>☐ Bidirectional</li></ul>
Ventricular ectopic beats morphology:	☐ Monomorphic ☐ Polymorphic
Ventricular ectopic beats distributional pattern:	Single Couplets Triplets
Ventricular ectopic beats, when?	At rest During exercise During max exercise In recovery
Other arrhythmia:	
Holter monitor:	



Normal Normal	Ventricular ectopic beats - monomorphic
Unknown	Ventricular ectopic beats - polymorphic
Atrial tachycardia	Ventricular ectopic beats - couplets
Afib	Ventricular ectopic beats - triplets
Torsades	Sustained VT
AV Block II - intermittent	Non sustained VT
AV Block II - persistent	AVRT/AVNRT
AV Block III - intermittent	Abnormal pauses for age
AV Block III - persistent	Significant Bradycardia
Brugada pattern	Other; please specify
Ajmaline challenge performed?	Not performed
	Positive
	Negative
	Unknown
Brugada pattern type:	Type 1
	Type 2
	Type 3
Flecainide challenge performed?	☐ Not performed
	Positive
	Negative
	Unknown
Brugada pattern type:	Type 1
	Type 2
	Type 3
Adrenaline challenge performed?	Not performed
	Positive
	☐ Negative
	Unknown
Longest QTc:	
Intervention Procedures	
PPM:	Yes No
Date implanted:	
	(If unsure of the exact date please set to January 1 of
	the year the event took place and set the "Exact date"
	option to "No".)
Is this an exact date?	Yes No
ICD:	Yes No
	(If the patient has had multiple ICDs, this refers to the
	first one)
Date implanted:	•
F 1 1 1 1 1 1 1 1 1 1 1 1 1 1 1 1 1 1 1	(If unsure of the exact date please set to January 1 of
	the year the event took place and set the "Exact date"
	option to "No".)
Is this an exact date?	Yes No
Primary or secondary prevention	Primary Secondary
Cardiac transplant:	Yes Listed No
Carulac Hallsblatt.	



Date inserted:	
	(If unsure of the exact date please set to January 1 of
	the year the event took place and set the "Exact date"
	option to "No".)
Is this an exact date?	Yes No
Date listed:	
	(If unsure of the exact date please set to January 1 of
	the year the event took place and set the "Exact date"
	option to "No".)
Is this an exact date?	☐ Yes ☐ No
LVAD:	Yes No N/A
Date inserted:	
	(If unsure of the exact date please set to January 1 of
	the year the event took place and set the "Exact date"
1.11.2	option to "No".)
Is this an exact date?	Yes No
Uploads	
Echo at diagnosis or representative echo	
Number of ECGs at diagnosis or representative ECGs	
First ECG	
First ECG lead placement	Standard
C	Elevated
Second ECG	
Second ECG lead placement	Standard
Third FCC	Elevated
Third ECG	Chandand
Third ECG lead placement	Standard
Faunth FCC	Elevated
Fourth ECC lead alexander	Chandand
Fourth ECG lead placement	Standard
Fifth FCC	Elevated
Fifth ECG	Chandard
Fifth ECG lead placement	Standard
CMDI at diagnosis or representative CMDI	Elevated
CMRI at diagnosis or representative CMRI	
Ajmaline challenge report	
Flecainide challenge report	
Adrenaline challenge report	
Operation report	