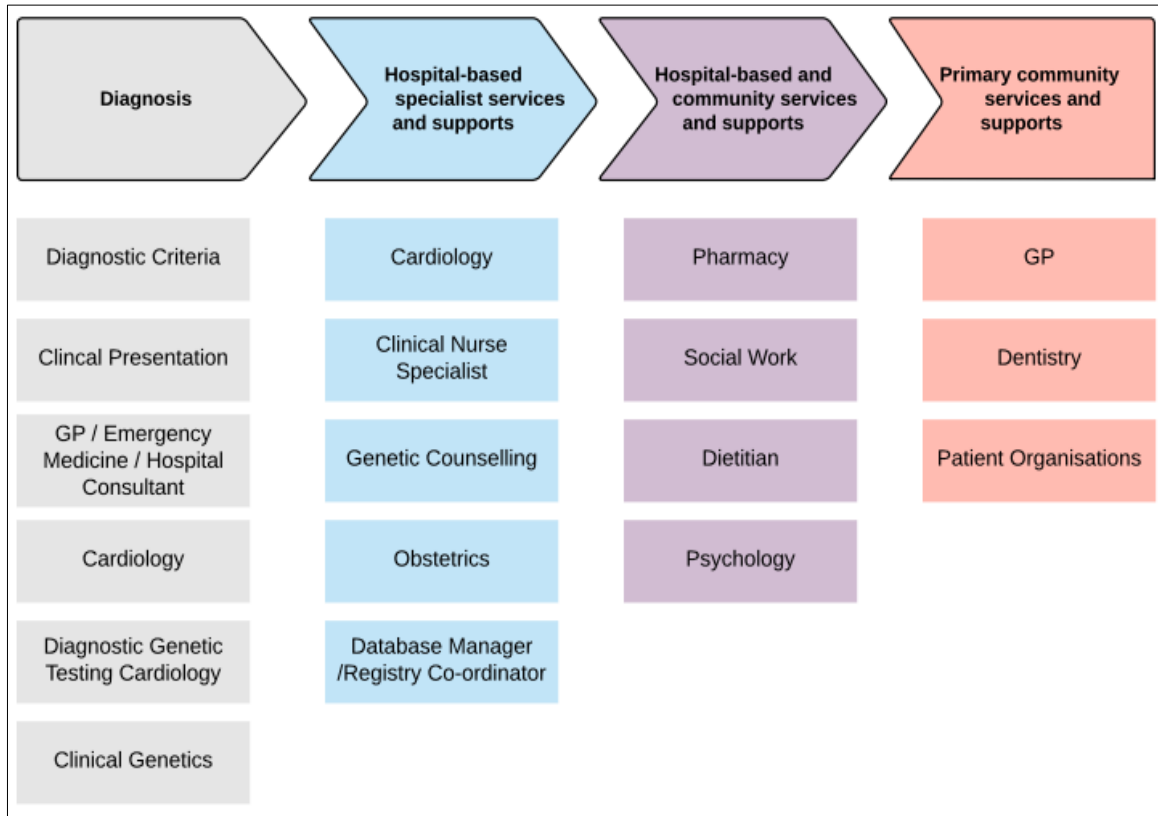




Long QT Syndrome (LQTS)

Adult Care Pathway

Long QT Syndrome (LQTS) Adult Care Pathway



Clinical Characteristics:

Orphacode: 768

Disease Definition: Congenital Long QT Syndrome (LQTS) is a hereditary cardiac disease characterized by a prolongation of the QT interval at basal ECG and by a high risk of life-threatening arrhythmias

Diagnosis	Diagnostic Criteria	Long QT diagnosis based on Schwartz criteria; QTc > 480ms = 3 points, 460-470ms = 2 pts, > 450ms (Males) = 1 pt, Torsades = 2 pts, notched T waves in 3 leads = 1 pt, resting HR < 2nd centile for age = 0.5 pt, Syncope with stress = 2pts (without stress = 1 point), congenital deafness = 0.5 pts, FHx LQTS = 1 pt, SADS in 1st deg family member = 0.5 pt. < 1 point = low, 2-3 points = intermed and > 4 pts = high probability of LQTS
Diagnosis	Clinical Presentation	Syncope which may lead to cardiac arrest and death (most commonly preteen years to 20s); ECG abnormalities, prolongation of QT interval and T wave abnormalities
Diagnosis	GP / Emergency Medicine / Hospital Consultant	Referral criteria: Family history of Long QT syndrome or Family history of sudden cardiac death (SCD) due to sudden arrhythmic death syndrome (SADS) in first degree relative or History of exertional syncope with prolonged QT on ECG or Documented PMVT Exertional ventricular polymorphic ectopy Asymptomatic QTc > 470ms with a normal QRS duration in the absence of other high-risk features Refer to family screening clinic – CRYP centre (cardiac risk in young persons), Tallaght University Hospital, Dublin 24 Or Family Heart Screening Clinic, Heart House, Mater University Hospital, Dublin 7 Or Children to Inherited Cardiac Conditions Clinic, CHI-Crumlin, Dublin 12
Diagnosis	Cardiology	Assessment: Full history Cardiac examination Detailed family pedigree Schedule investigations (urgent if syncope, seizure, sudden death in 1st degree relative): 12 lead ECG 15 lead Brugada ECG, 24-hour Holter / QT analysis TMET with stand-up QT measurement and recovery to < 100/min with post-exercise QT Consider Adrenaline challenge (0.05-0.2mcg/kg/min)
Diagnosis	Diagnostic Genetic Testing Cardiology	Core LQTS panel in probable or definite LQTS patients Consider LQTS panel in patients with repeated ECGs confirming isolated QTc > 470ms

Diagnosis	Clinical Genetics	<p>Predictive genetic testing for known familial Long QT pathogenic variant</p> <p>Consider phenotypic features indicative of rare syndromic forms</p>
Hospital-based specialist services and supports	Cardiology	<p>Commence beta-blocker (e.g. Nadolol or twice daily long acting Propranolol) and titrate dose</p> <p>Consider intracardiac device (ICD) if QTc > 500ms, Syncope or VT on BBL or 1st degree relative with SCD</p> <p>Consider sympathectomy if unable to take BBL or ICD shock</p> <p>Advise patient and family on avoidance of stimulants and exercise, sports to moderate or high level</p> <p>Children - advise schools on AED defibrillator</p> <p>Pregnancy - liaise with high-risk Obstetrics team, ECG and holter throughout pregnancy, beta-blocker dose adjustment.</p> <p>Advise first degree adult relatives to seek cardiology clinical screening for Long QT via GP referral</p> <p>Arrange clinical cardiology screening for children of / child siblings of individuals with Long QT</p>
Hospital-based specialist services and supports	Clinical Nurse Specialist	<p>Co-ordinate SADS proband post-mortem report, pathology results, medical records, tissue / blood from SADS biobank, newborn-screening blood spot, consent family for molecular autopsy</p> <p>Co-ordinate patient and family investigations</p> <p>Point of contact</p> <p>Patient and family education, advice & support</p> <p>Advise patient and family on avoidance of stimulants and exercise, sports to moderate or high level</p> <p>Give patient and family list of QT prolonging medications to avoid, link to credible meds website and app www.crediblemeds.org</p> <p>Advise on self-care for sick days to avoid dehydration and electrolyte imbalance</p> <p>Link patient and family to patient organisations</p> <p>Liaise with health care professionals</p> <p>Transition planning</p>
Hospital-based specialist services and supports	Genetic Counselling	<p>Review genetic report; recurrence risk, identification of at-risk relatives; cascade testing; link family to patient organisations.</p> <p>Refer patients with Long QT for cardiology screening and request cardiologist to see first-degree relatives for phenotypic cardiology assessment.</p> <p>Advise first degree adult relatives to seek cardiology clinical screening for Long QT via GP referral and provide family with information letter to support this</p> <p>Refer children of / child siblings of affected individual for clinical cardiology screening for Long QT</p>

Hospital-based specialist services and supports	Obstetrics	High risk pregnancy; close liaison with cardiology specialist centre throughout pregnancy and post-partum
Hospital-based specialist services and supports	Database Manager/Registry Co-ordinator	Create and maintain database of patients attending service Record patient biographical, clinical and research data Audit and quality improvement Ensure minimum data set standards for ERN registry
Hospital-based and community services and supports	Pharmacy	Awareness and avoidance of QT-prolonging drugs – www.crediblemeds.org Hospital Pharmacy – expert medication advice Community Pharmacy –advise patients and family about over-the-counter medication and GP prescribing advice
Hospital-based and community services and supports	Social Work	Psychosocial support: Assess social and family supports, safeguarding Link with community supports as required e.g., GP, Public Health Nurse, Primary Care SW, Local authority SW, Mental Health SW, Disability SW, TUSLA Offer 1-1 counselling or GP referral to Counselling in primary care (CIPC) www.hse.ie/eng/services/list/4/mental-health-services/counsellingpc/ Financial support (as required): Patient advocacy, support applications for Medical card, Disability allowance, Supplementary Welfare allowance, Exceptional Needs payment, Long-term illness card, direct to Citizens' information www.citizensinformation.ie/ Housing and/or mobility issues: Advocacy and support Home Care Packages: if issues with activities of daily living - arrange application for inpatients, liaise with Public Health Nurse to arrange for out-patients Respite Care: liaise with public health nurse or community disability services to arrange Employment issues: Link to Intreo public employment and EmployAbility services www.gov.ie/en/campaigns/fb84c0-intreo/ www.gov.ie/en/service/8578c4-access-the-employability-service/
Hospital-based and community services and supports	Dietetics	Nutrition assessment to ensure nutritional adequacy and optimal weight

services and supports		<p>Provide dietary advice to help lower risk of arrhythmic events e.g. Mediterranean diet, avoid large meals, electrolyte balance</p> <p>Weight and dietary management support (primary care)</p>
Hospital-based and community services and supports	Psychology	<p>Person and family-centred support for chronic condition at diagnosis, transition and as required – coping skills, depression and anxiety management, bereavement, adherence to lifestyle adjustments, compliance with medication and follow-up care</p>
Primary / community services and supports	GP	<p>Liaise with cardiac specialist centre</p> <p>Beta-blocker prescribing</p> <p>Awareness and avoidance of QT-prolonging drugs for management of coexisting conditions - https://crediblemeds.org</p> <p>Advise patients on self-care for sick days to avoid dehydration and electrolyte imbalance</p> <p>Counselling in Primary Care for mild to moderate psychological issues - anxiety, loss, coping</p> <p>www.hse.ie/eng/services/list/4/mental-health-services/counsellingpc</p> <p>Management of inter-current conditions; co-ordination of local services and supports; refer/communicate across services</p>
Primary / community services and supports	Dentistry	<p>Awareness and avoidance of QT-prolonging drugs – www.crediblemeds.org</p>
Primary / community services and supports	Patient Organisations	<p>Advocacy, support & information - Irish Heart Foundation (www.irishheart.ie)</p> <p>Long QT syndrome support group (www.irishheart.ie/get-support/support-groups/long-qt-syndrome-support-group/)</p> <p>Cardiac Risk in the Young (CRY) charity (www.cry.ie)</p> <p>CRY Charity: + 353 1 414 2235</p> <p>CRY Helpline: + 353 1 839 5438</p> <p>Email: familysupport@cry.ie</p> <p>Rare Ireland Family Support Network (www.rareireland.ie)</p> <p>Rare Diseases Ireland (www.rdi.ie)</p>

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