Background Channelopathies are the leading cause of sudden cardiac death in the young (<35 years of age) in Ireland. Implantable cardioverter defibrillators (ICDs) are necessary for high-risk cases. It is recommended that these often-challenging conditions are managed at subspecialist inherited cardiac conditions centres (ICC) to optimise lifestyle measures, direct pharmacological and procedural interventions, and ensure optimal ICD programming. However, there is a paucity of outcome data in terms of recurrence of ventricular arrhythmias, and the interventions utilised.

Aims To examine ICDs programming and interventions post therapy in cases of Long QT Syndrome (LQTS) and Brugada Syndrome (BrS) that are attending a high-volume specialist ICC in Ireland.

Methods Retrospective analysis of a single centre ICC clinic’s device data to identify LQTS and BrS cases since Jan 2007 to Jan 2022. Local electronic records and the national cloud-based program Heart Rhythm Ireland were utilised.

Results A total of 22 patients were identified. Demographic data and diagnoses as per table 1 below. A transvenous ICD was utilized in 15 (68%), and subcutaneous ICD in 7 (32%). A total 7/22 (32%) had experienced tachycardia therapies. Appropriate therapies were experienced by 3 patients, two of had a recurrent episode. Of the 4 inappropriate aetiologies; 2 sinus tachycardia, 1 supraventricular tachycardia, 1 T wave oversensing. All 4 had transvenous ICD systems. Mean time from implant to first therapy was 1084 days (range 125–3433). Specialist ICC clinic interventions to reduce the risk of further therapy included reinforcement of lifestyle measures (5/7), medication optimisation (6/7), sympathectomy (1/7) and BrS epicardial ablation (1/7). Device programming was optimised in 5/7, to prolong the time to therapy in order to reduce risk of inappropriate defibrillation and to promote spontaneous reversion of polymorphic VT. No further ICD therapies were documented since last treatment optimisation, during a mean follow up of 678 days (range 196–4759).

Conclusion Approximately one in three channelopathy cases with an ICD experienced ICD defibrillation, the majority of which were inappropriate. Following interventions in a specialised clinic, recurrence of ventricular arrhythmia in this high-risk population appears to be low.