identify the apex of the triangle of Koch by annotating the following structures on a CARTO-Sound 3-D electroanatomic map: the tricuspid annulus, the Os of the coronary sinus, and the tendon of Todaro. Using this CARTO-Sound slow pathway map as a reference, safe and effective ablation sites were consistently found. Ablation was performed with an irrigated ablation catheter at sites with a 0.25-1:1 A:V ratio on a 3.5 mm ablation tip within the region demarcated by CARTO-Sound. In all six patients, sustained slow junctional rhythm was observed during ablation, and patients demonstrated no slow pathway conduction and were non-inducible for AVNRT following ablation (20-30W; 30 seconds; 3-6 lesions).
Results: Successful identification of the slow pathway was accomplished using ICE and the CARTO-Sound 3-D mapping module. Safe and effective ablation was performed using this technique, as heart block was not observed during ablation and no recurrence of SVT was observed in any of the patients.
Conclusion: Successful identification of the slow pathway was accomplished using ICE and the CARTO-Sound 3-D mapping module. Safe and effective ablation was performed using this technique, as heart block was not observed during ablation and no recurrence of SVT was observed in any of the patients.

B-PO02-014

PLN VARIANTS IN PATIENTS WITH SUDDEN CARDIAC ARREST/DEATH WITHOUT DILATED OR ARRHYTHMOGENIC CARDIOMYOPATHY: A CASE SERIES

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Background: Phospholamban, encoded by the *PLN* gene, is a regulator of intracellular Ca^{2+}. Pathogenic *PLN* variants cause cardiac remodeling and early death. It is plausible that *PLN* variants confer a pro-arrhythmic phenotype without overt structural remodeling. To date, most *PLN* variants associated with sudden cardiac arrest/death (SCA/D) are accompanied by structural cardiomyopathies. The prevalence of *PLN* variants in autopsy-negative SCA/D cases without overt cardiomyopathy is unclear.
Objective: We present a case series of patients with *PLN* gene variants identified due to SCA/D without overt dilated or arrhythmogenic cardiomyopathies. We describe *PLN* gene variants presenting with primarily arrhythmic phenotypes. The *PLN* gene should be strongly considered in genetic testing strategies for SCA/D.
Methods: Cases were ascertained through professional collaboration among cardiovascular genetic counselors. We prioritized cases with *PLN* variants presenting without recognized cardiomyopathy phenotypes and/or in autopsy-negative SCA/D.
Results: We describe five cases with pathogenic *PLN* variants. Two are previously reported (p.Arg14del, p.Leu39Ter), and one is novel (p.Gln22LeufsX19). There were 3 pediatric cases and 2 adult cases, and 2/3 pediatric cases presented with SCD and negative autopsies. The other pediatric case survived SCA, but follow-up cardiac investigations were normal. Four of five cases had uninformative cardiac phenotypes, though one case had some evidence of cardiac hypertrophy at autopsy. Interestingly, 4/5 cases had SCA/D occur during rest, and 1/5 cases occurred with some activity. Last, one adult with SCD had a history atrial fibrillation, though no other cases had known arrhythmias. Table 1 summarizes these findings for comparison and contrast.
Conclusion: With a role in Ca^{2+} regulation, it is plausible that *PLN* variants confer a pro-arrhythmic substrate and increased of SCA/D in the absence of cardiomyopathy. We raise awareness for the possible role of *PLN* variants in phenotype- and autopsy-negative SCA/D and encourage further investigation. This may help guide genetic counseling for *PLN* variants found in patients at autopsy and/or in patients who have not yet developed cardiomyopathy.

B-PO02-015

ENHANCING MUTANT I_{KS}CHANNEL ACTIVITY BY ALTERING ENDOGENOUS PIP_2 LEVELS AND ITS INTERACTION WITH PKA SIGNALLING PATHWAY

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Background: Phosphatidylinositol-4,5-biphosphate (PIP_2) is implicated in the regulation and modulation of the I_{KS} channel.
Objective: N/A
Methods: We initially transfected Human Embryonic Kidney (HEK) cells with a KCNQ1 gene, along with KCNE1 to form the wild type (WT) I_{KS} channel. The cells were also transfected with a constitutively active PI(4)P 5-kinase(PIP5K), which increases endogenous levels of PIP_2. To ensure the enzyme remains localised at the plasma membrane we attached it to CFP-FKBP and we co-transfected the cells with cherry tagged Lyn11-FRB construct that tethers to the plasma membrane.
Results: We substituted serine with alanine at site 27 and 92(S27A/S92A) to generate a mutant known to disrupt cAMP mediated upregulation, there was a statistical increase in current density when co-expressed with CF-PIP5K. We then substituted serine with aspartic acid (S27D/S92D) to create a Phosphomimetic mutation, this mutant reproduces the effects of sympathetic mediated augmentation of I_{KS} channel. Increased PIP2 levels failed to increase current density in S27D/S92D mutant, implying the channel is at its maximum activity and hence we failed to observe any further modulation.
In the presence CF-PIP5K, whole cell voltage clamp recordings demonstrated a statistically significant increase in WT channel activity (at +80mV, $p<0.001$), when compared to unaltered PIP_2 conditions. Heterozygous Serine566phe and Phe340del mutants had statistically significant reduction in current density compared to wild type in basal conditions. When these mutants were expressed with the active CF-PIP5K, Serine566phe and Phe340 had an increase current. We then proceeded to interrogate how PIP_2 interacts with sympathetic signaling system. Pseudojanin (PJ) causes depletion of PIP_2 hence perturbing channel activity. When PJ was expressed with KCNQ1 and KCNE1 we observed an 80% reduction in channel activity at +80mV. When we perfused these cells with isoprenaline the channel activity was restored to normal.
Conclusion: Here we illustrate how increasing PIP_2 levels can revive I_{KS} channel activity in mutant genotype therefore supporting evidence of its capabilities as a potential therapeutic tool. This modulation is independent of the PKA-cAMP system.

B-PO02-016

BUCCAL MUCOSA CELLS AS A DIAGNOSTIC TOOL IN PATIENTS WITH ARRHYTHMOGENIC CARDIOMYOPATHY

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Background: Arrhythmogenic cardiomyopathy (ACM) is predominantly caused by mutations in genes encoding desmosomal proteins (such as *plakophilin-2*), however also mutations in non-desmosomal proteins like phospholamban (*PLN*) are found. Previous research showed that plakoglobin protein levels and localization in cardiac tissue of ACM patients is disturbed and this could be an additional tool to discriminate