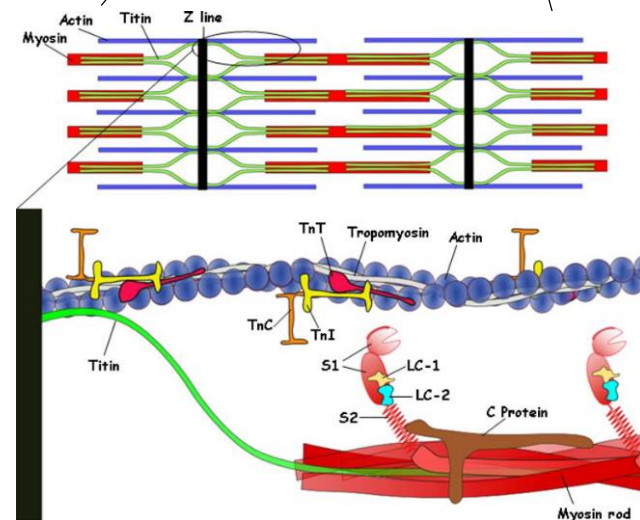
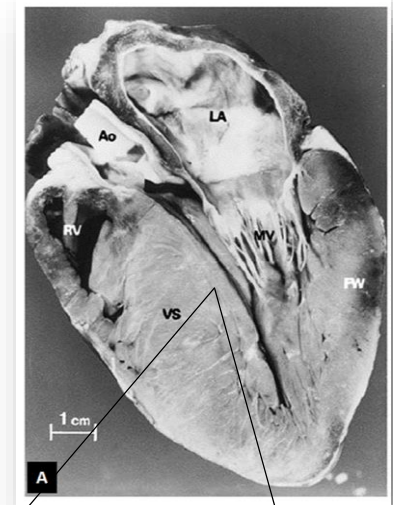
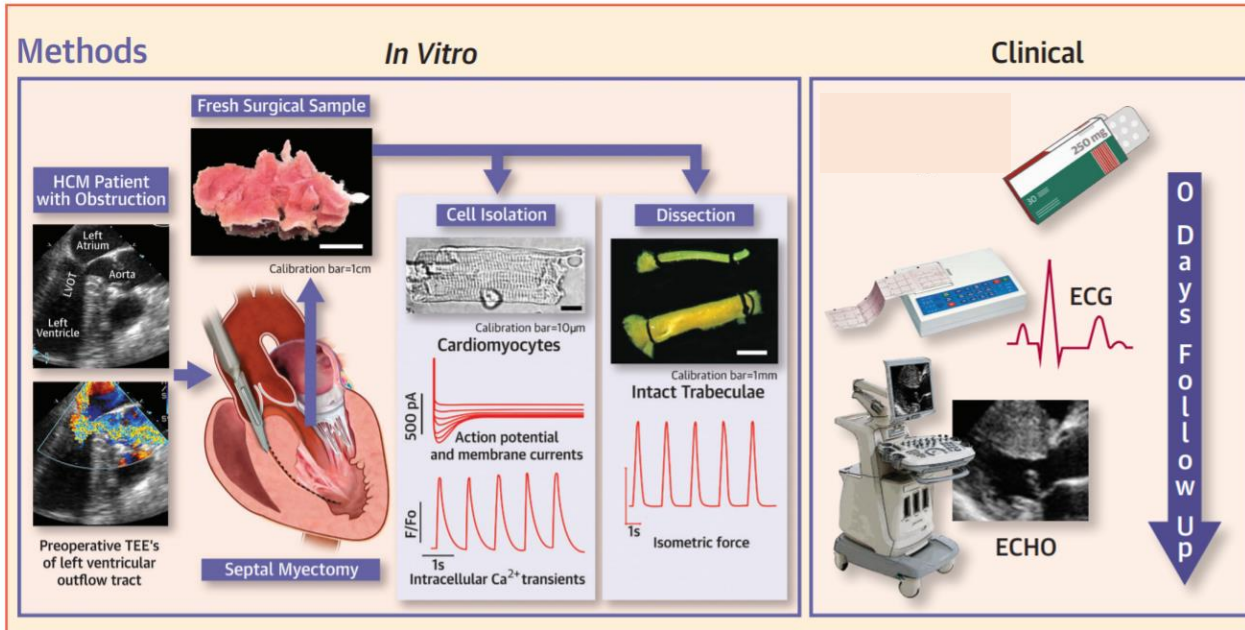


Denothe Unità 10

Hypertrophic Cardiomyopathy HCM



Physiology

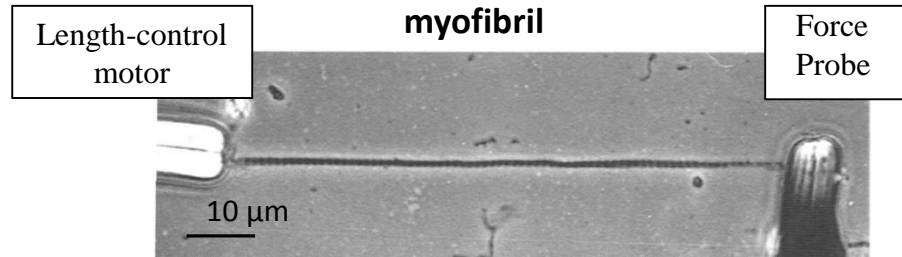
Pharmacology

Cardiology

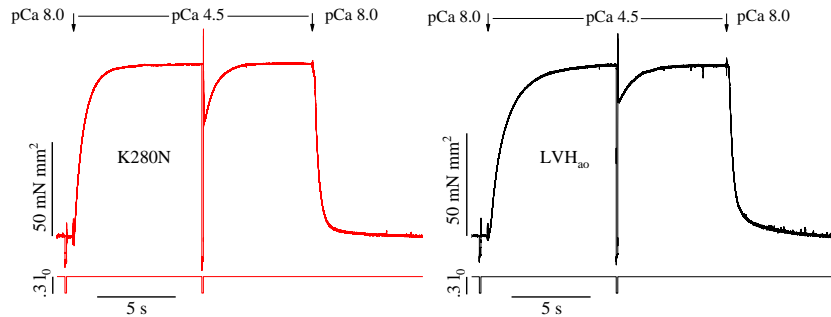
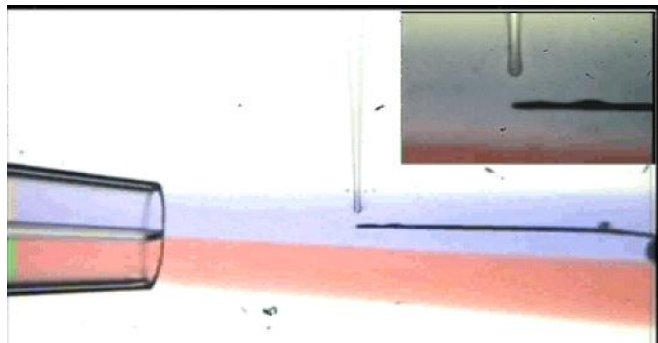
LENS
Morpho-functional
imaging

Nutrition

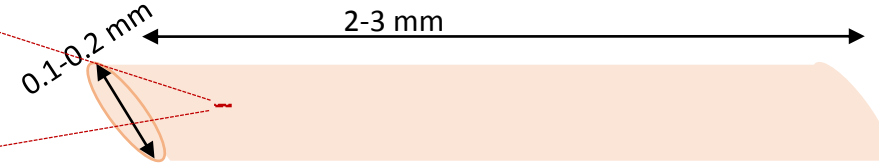
Single myofibril mechanics



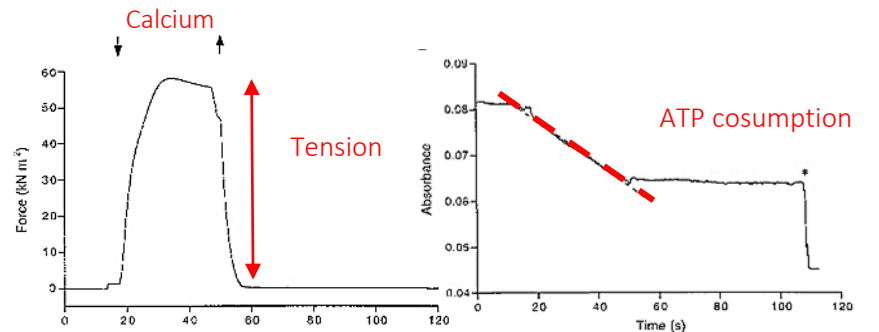
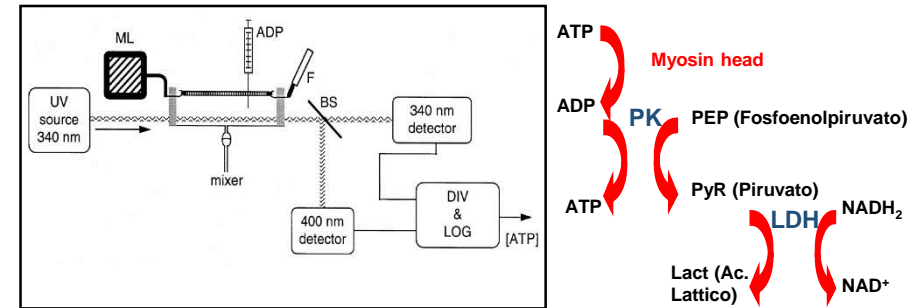
Fast solution switching technique



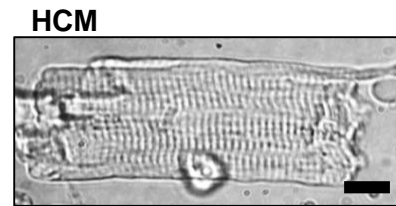
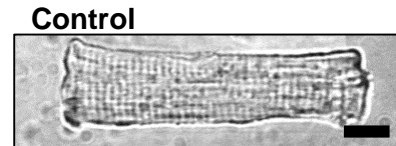
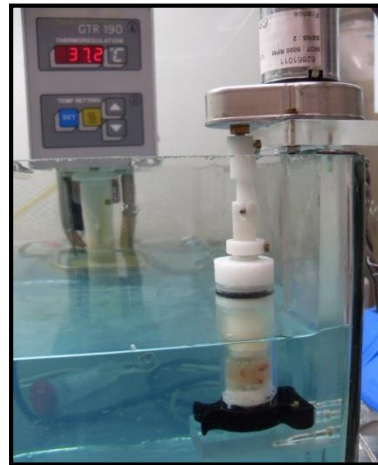
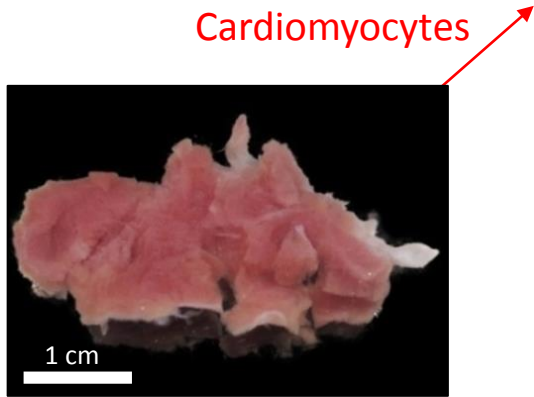
Myocardial energetics



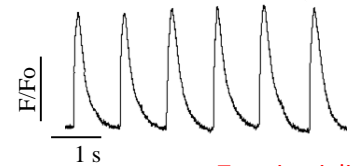
ATPase activity measured by an enzymatic assay



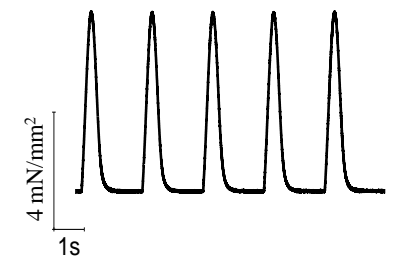
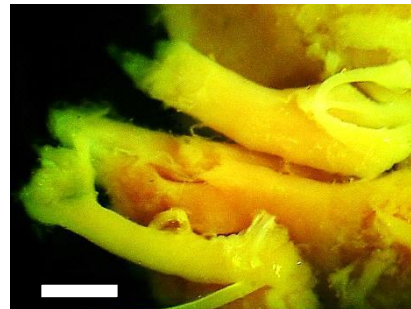
Secondary changes : electro-mechanical remodeling in HCM



Control HCM

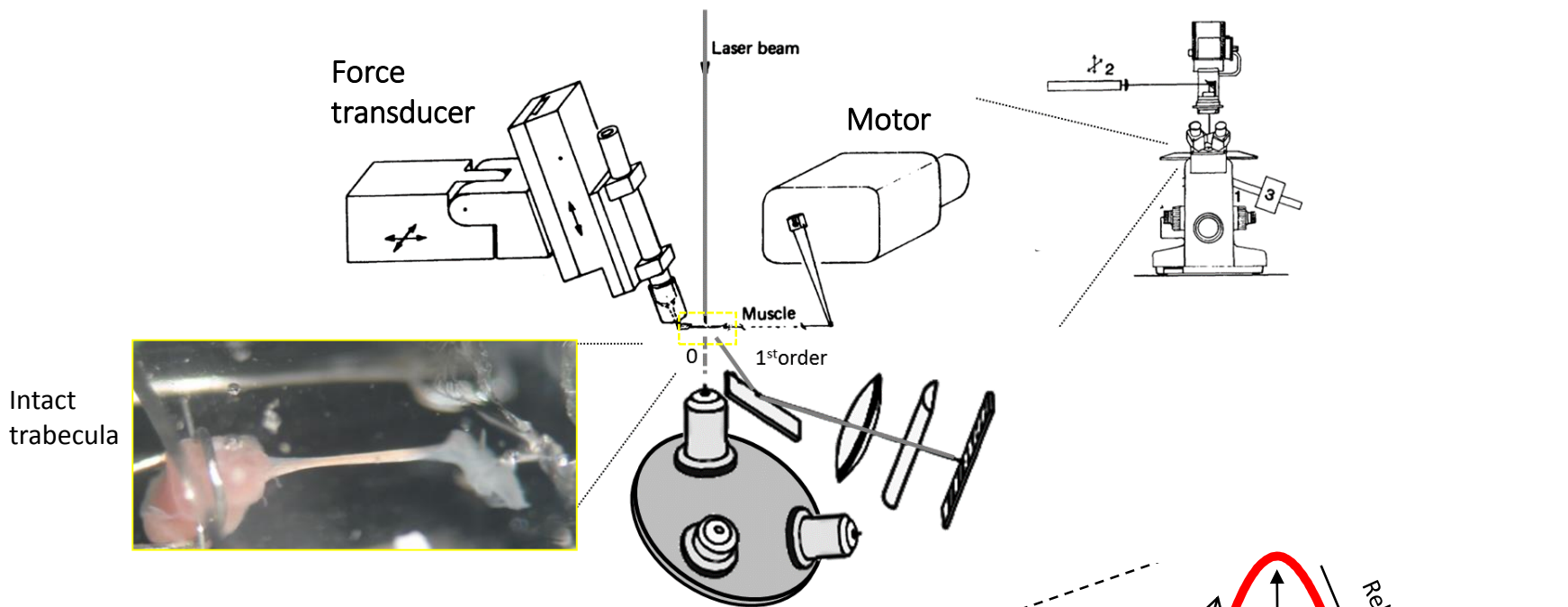


Transienti di Ca^{2+}

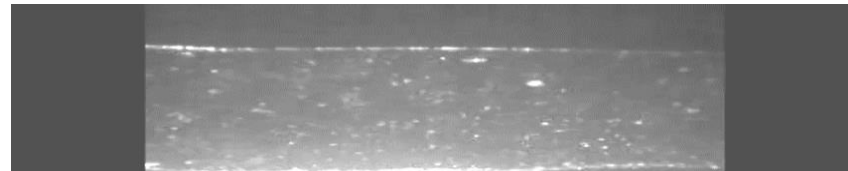
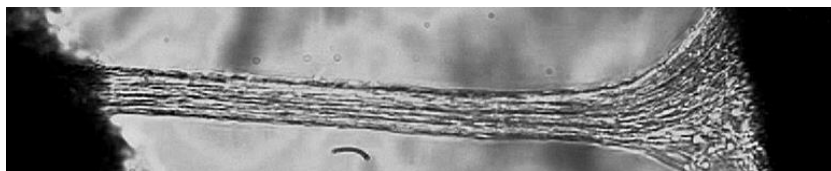
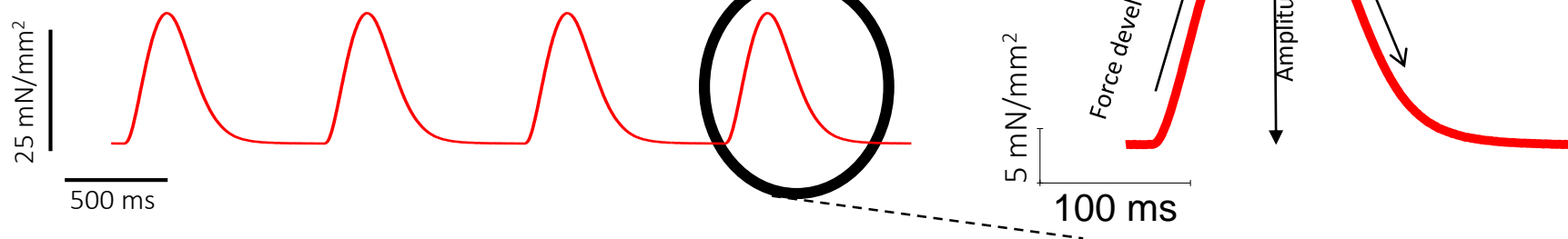


Forza isometrica

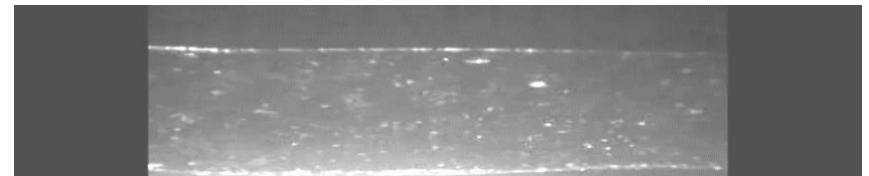
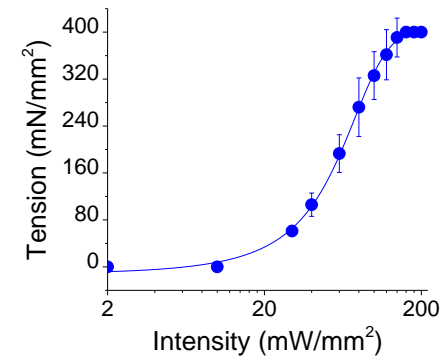
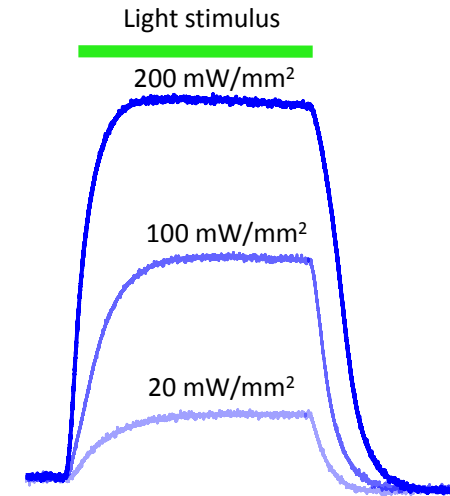
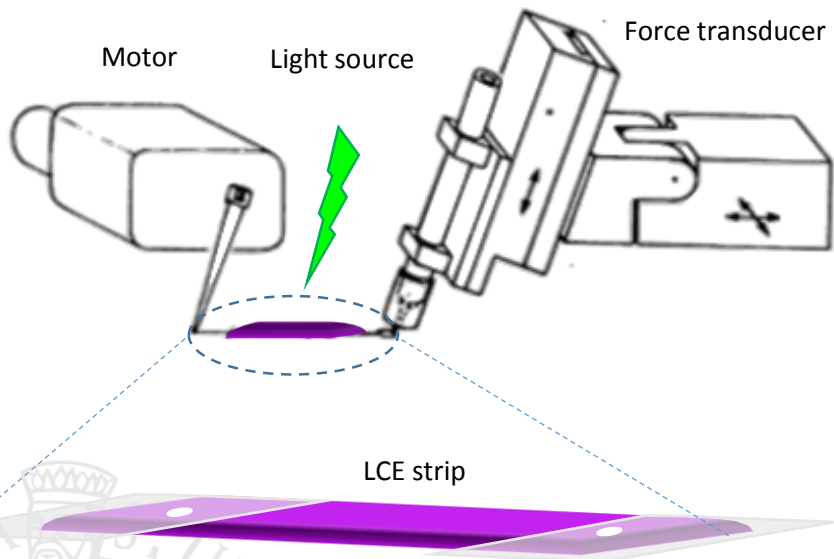
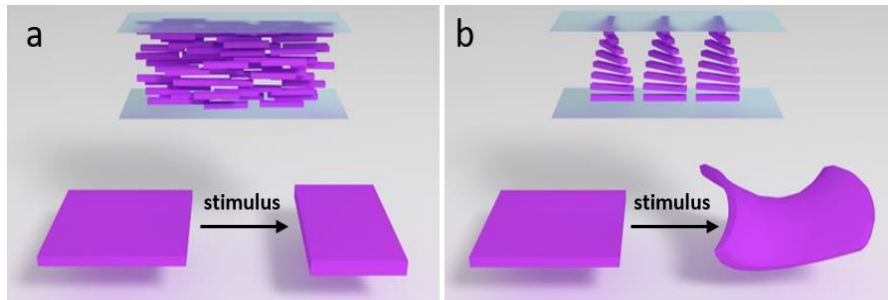
Apparatus for mechanical experiments



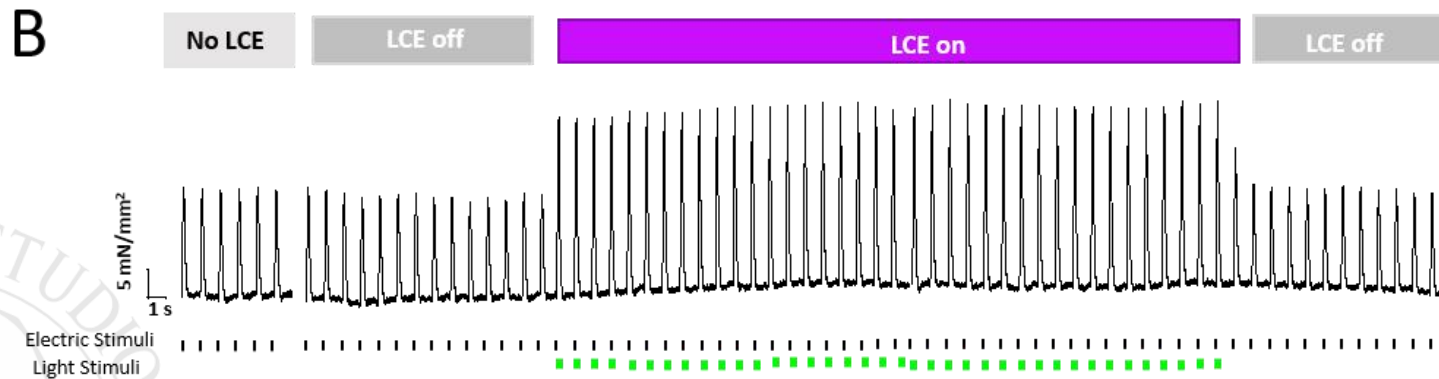
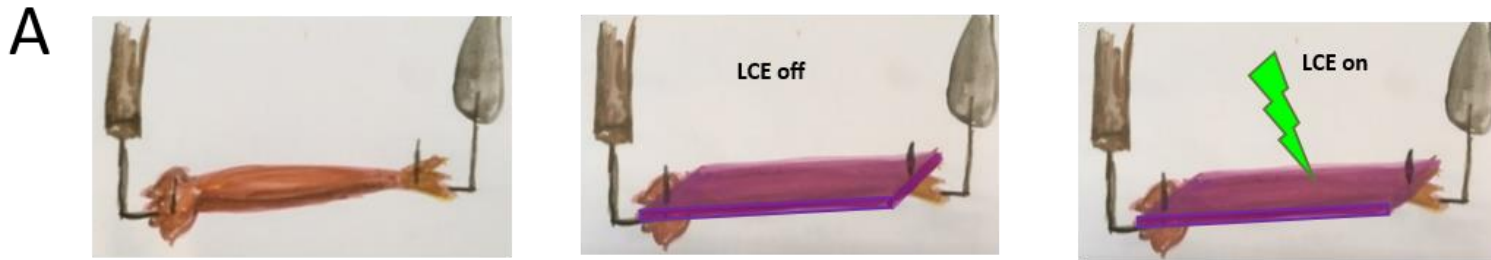
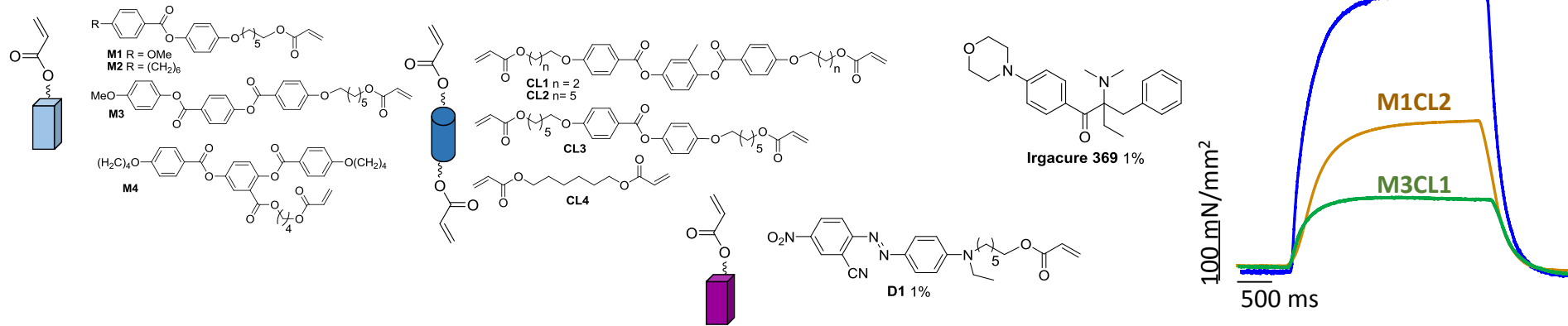
Force Recordings



Mechanical characterization of Liquid Cristal Elastomers



Mechanical characterization of LCEs



Limitations of biophysical studies on human fresh tissue

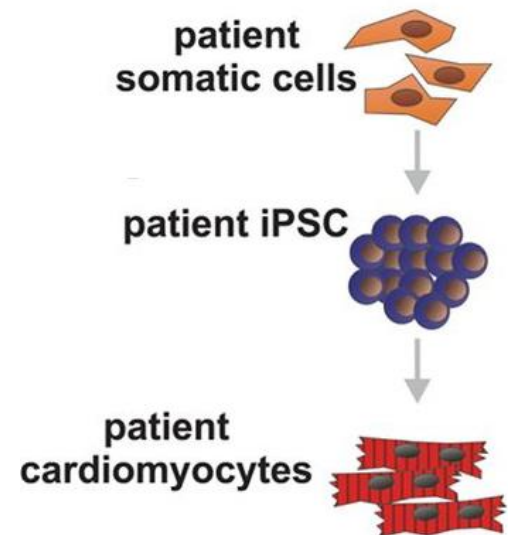
- Limited number of samples (10-12/year)
 - Mutations-specific mechanisms?
- Advanced disease stages

Alternatives:

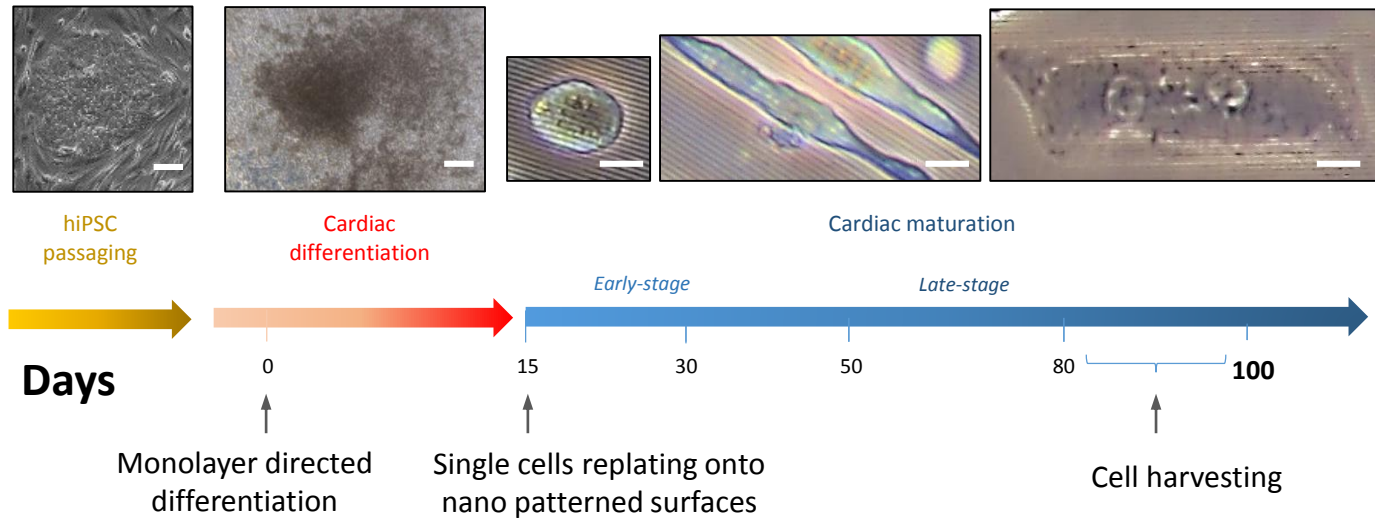
1. Mouse models



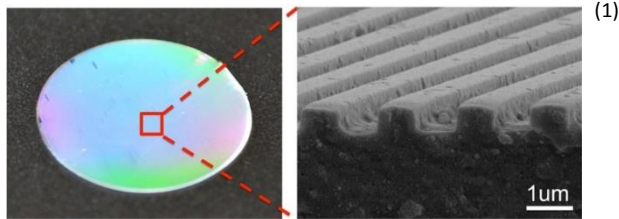
2. Human iPS-derived cardiomyocytes



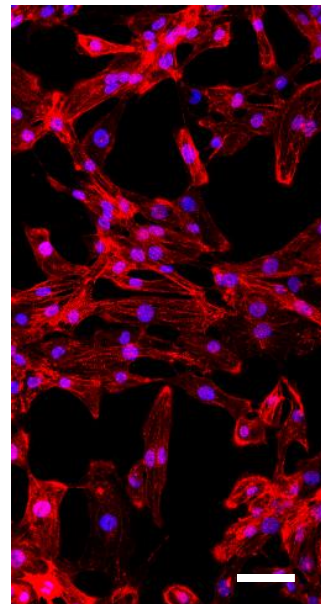
Improved cardiac maturation: combining nanopatterns and long-term culture



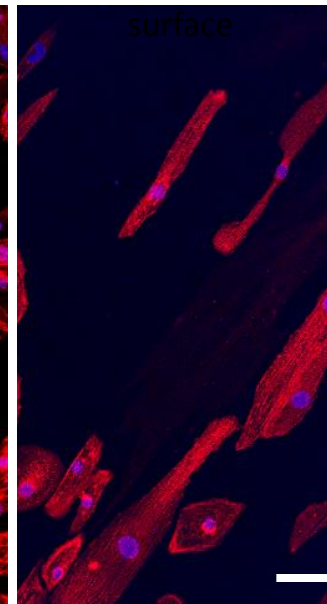
Nanopatterned surfaces mimic extracellular matrix



Flat surface



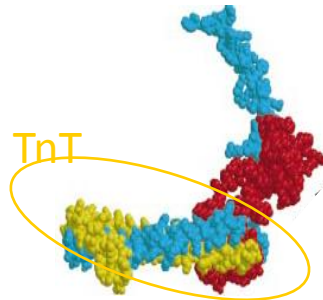
Nanopatterned surface



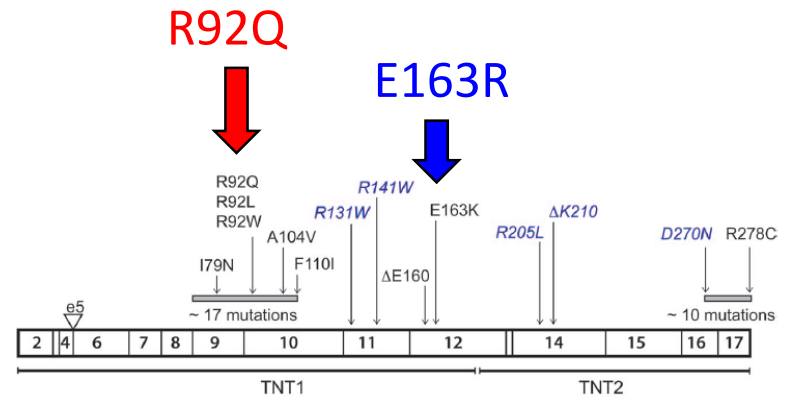
(1) Macadangdang J., J Vis Exp., 2014



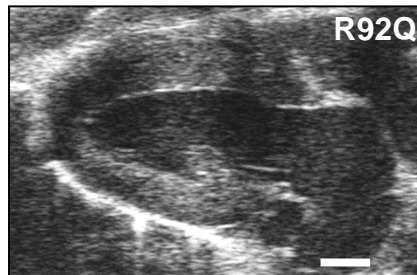
~ 30 mice per group,
8-10 months



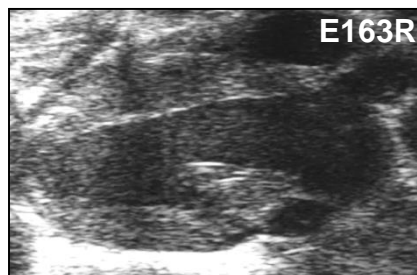
Troponin complex



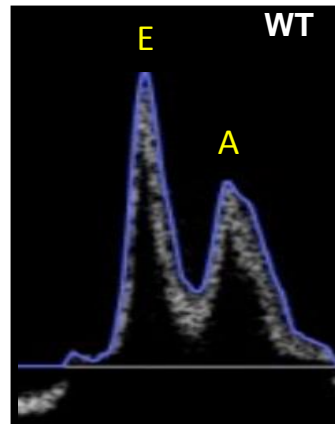
WT



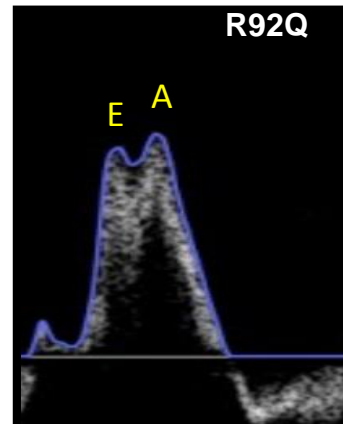
R92Q



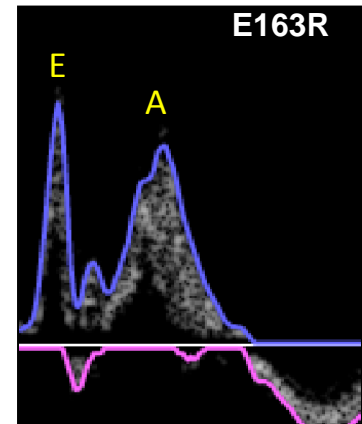
E163R



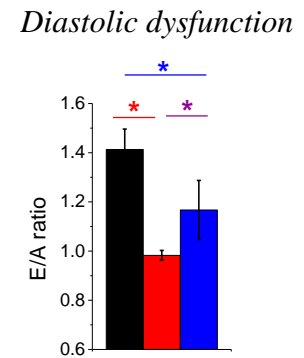
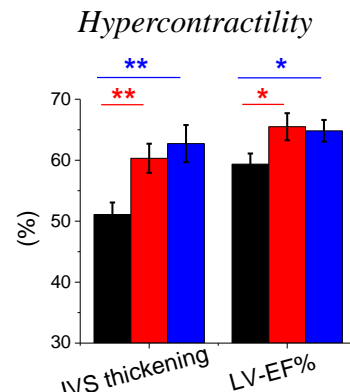
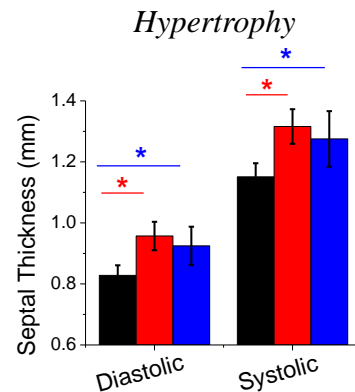
WT



R92Q

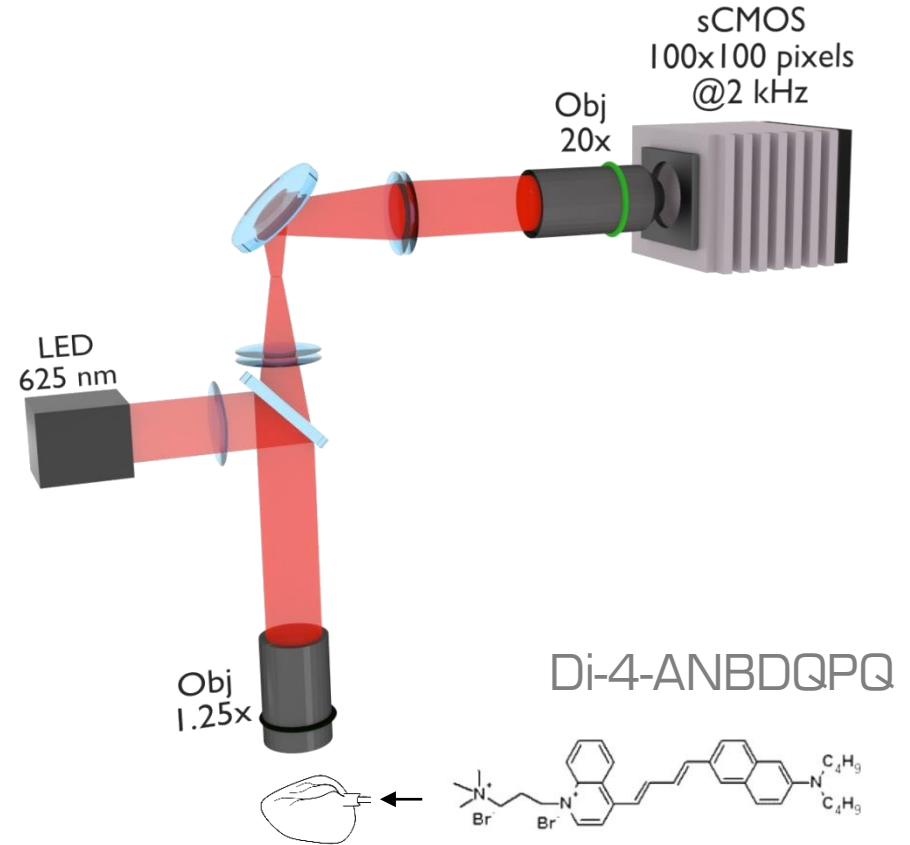
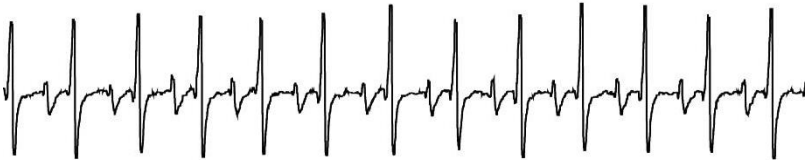
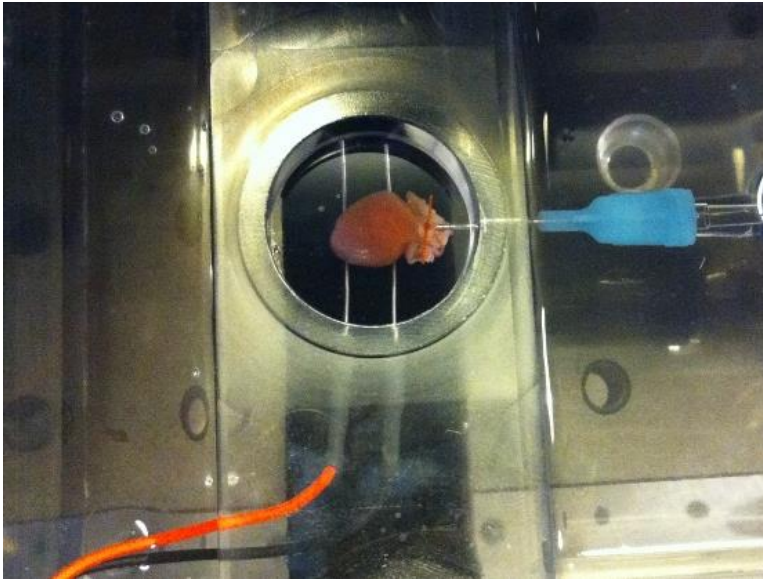


E163R

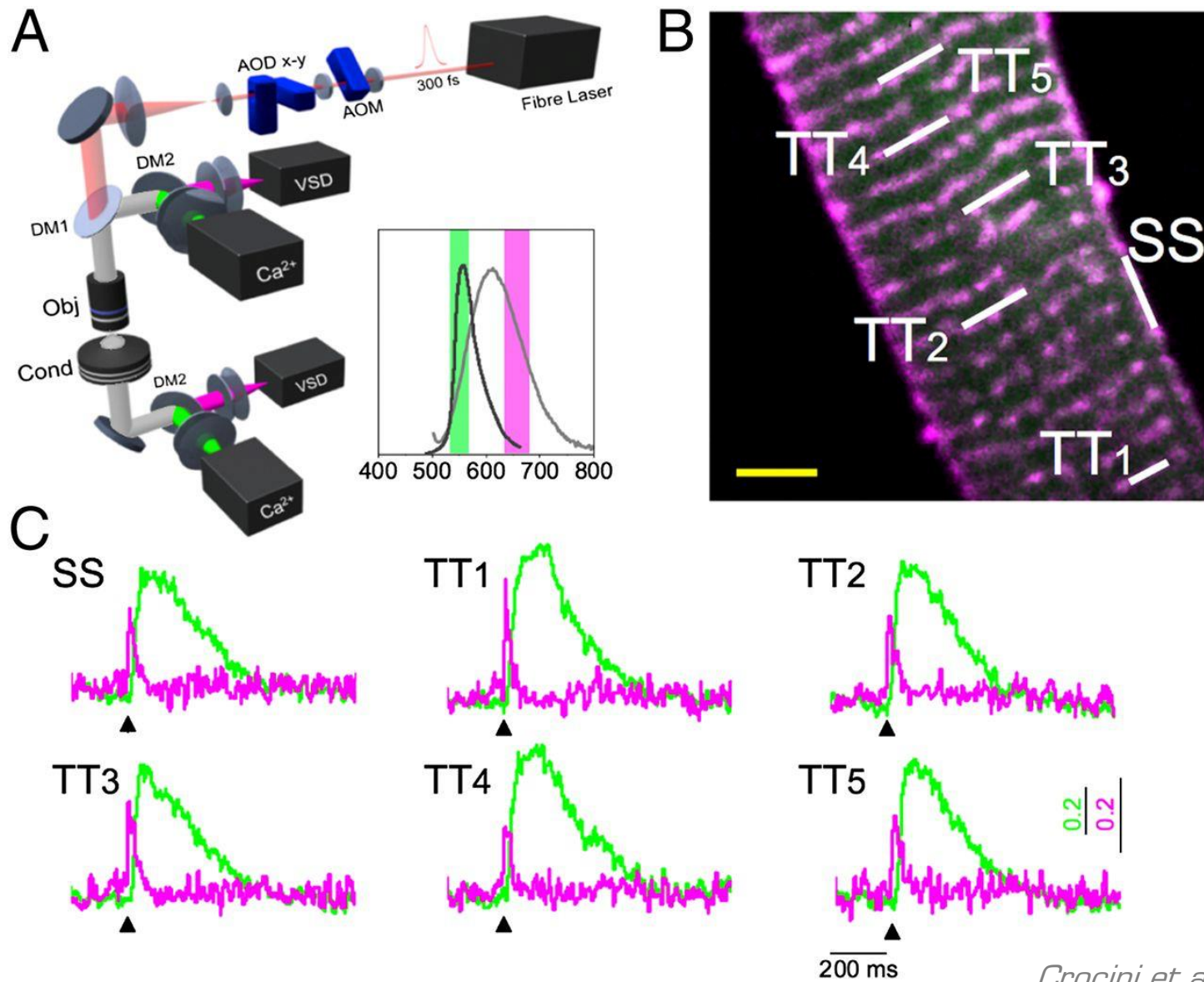


Ferrantini et al. JAHA 2017

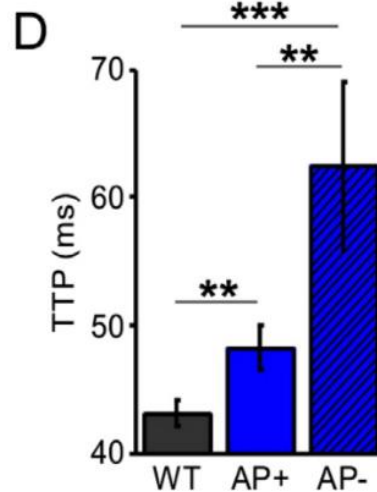
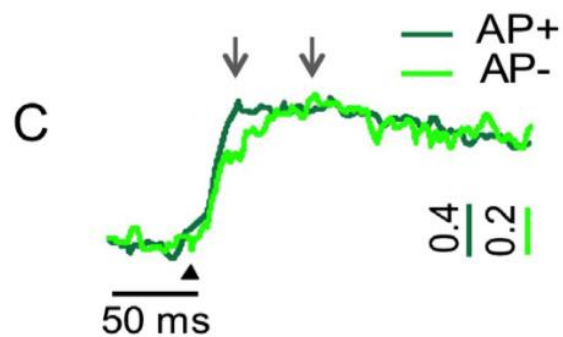
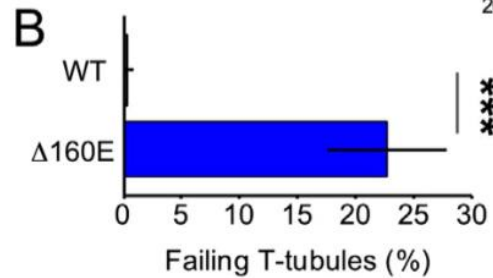
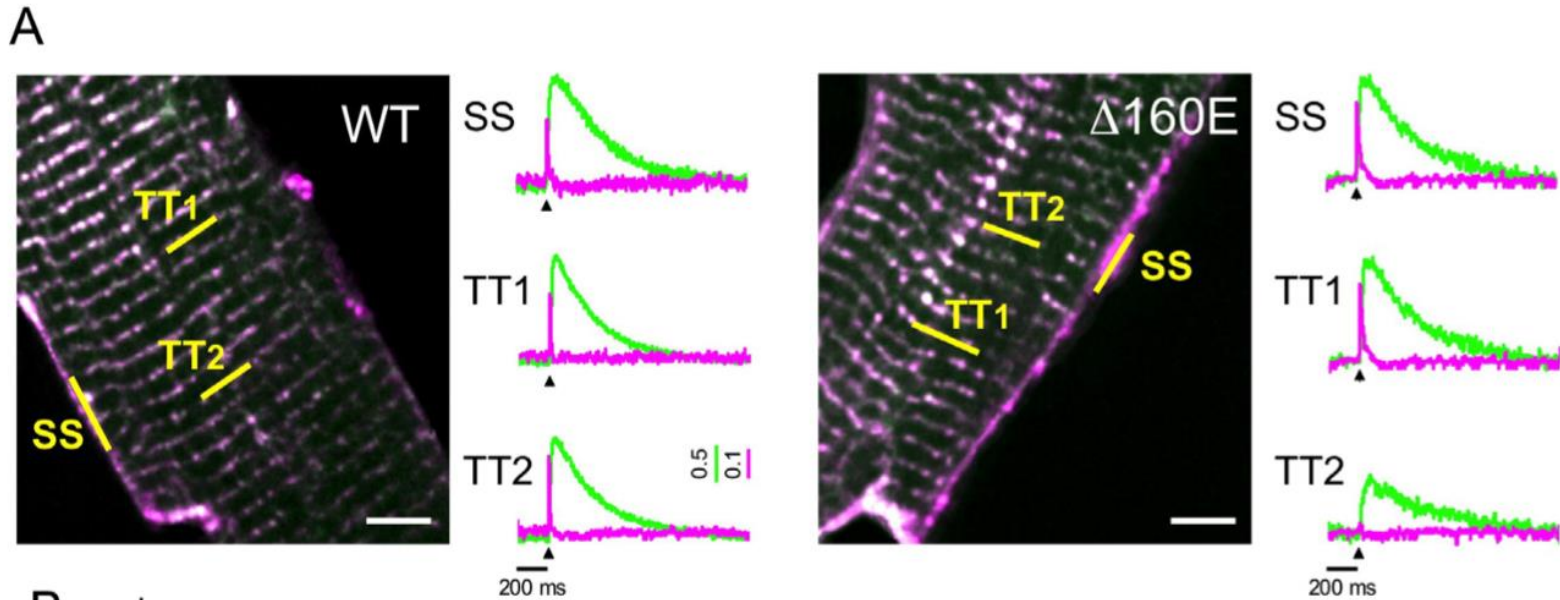
Optical mapping



Simultaneous multisite voltage and Ca^{2+} recording



T-tubular electrical defects in HCM mouse model



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