In Essence

- Myotonic dystrophy type 1 (DM1) is a progressive systemic disease that causes left ventricular dysfunction and conduction disturbances within the His-Purkinje system due to myocyte hypertrophy, fibrosis, focal fatty infiltration.
- We describe a case of reverse-selective His-bundle pacing in a patient with DM1 and prior history of LV dysfunction.

Case Presentation

A 19 year-old, 50kg male with DM1 Presented for Pacemaker Implantation

Cardiac History:
- Atrial fibrillation and flutter
- Symptomatic bradycardia with Mobitz Type II
- Progression to high grade 2:1 heart block and symptomatic sinus node dysfunction
- History of left ventricle dysfunction (LVEF of 45% on ACE-inhibitor)

Decision-Making:
Given progression of His bundle loss in up to 60% of patients with DM1, but young age and need history of LV dysfunction, reverse-selective His bundle pacing was performed.

Procedure:
Baseline EP study demonstrated an initial H-V measured at 71 ms.

His-bundle lead placement with reverse-selective His-bundle capture was performed by utilizing 3-dimensional mapping prior and choosing distal His location utilizing a 3830 lead.

Background

DM1 Background:
- Prevalence in general population
  - 1 per 8000 people
- Conduction disturbances
  - Affects ~30–75% of DM1 patients
- Mortality
  - ~7x that of the general population
  - Typically occurring in the 4th-5th decade of life

His-bundle Pacing Background:
- Long-term right ventricular pacing is associated with left ventricular dysfunction and cardiomyopathy in up to 13% of the cases
- His-bundle pacing achieves physiological pacing and avoids marked ventricular de-synchrony, reduce the risk of pacemaker induced cardiomyopathy, and reverse pacing-induced cardiomyopathy

Take Home Points

His-bundle pacing is beneficial for Patients with DM1 by:
- Providing Physiologic Pacing
- Decreasing the risk of cardiomyopathy and reversing pacer-induced cardiomyopathy
- Providing the ability to modify and transition to non-selective pacing in the event of further cardiac conduction dysfunction secondary to focal fatty infiltration