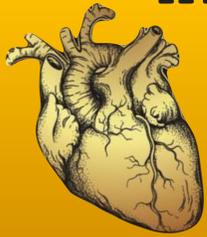


# His Bundle Pacing in Patient with Myotonic Dystrophy Type 1

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## 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

## 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

## 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.

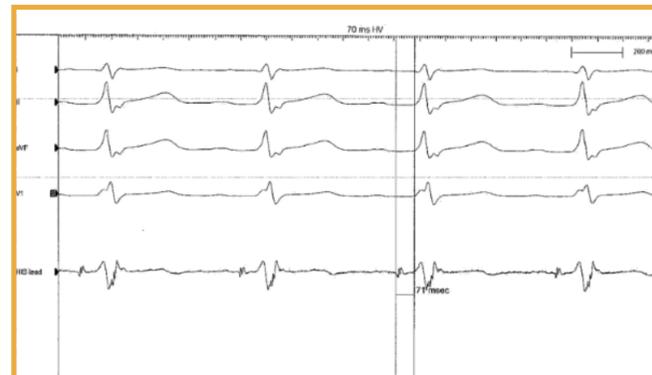


Fig 1. Initial H-V Measurement

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.

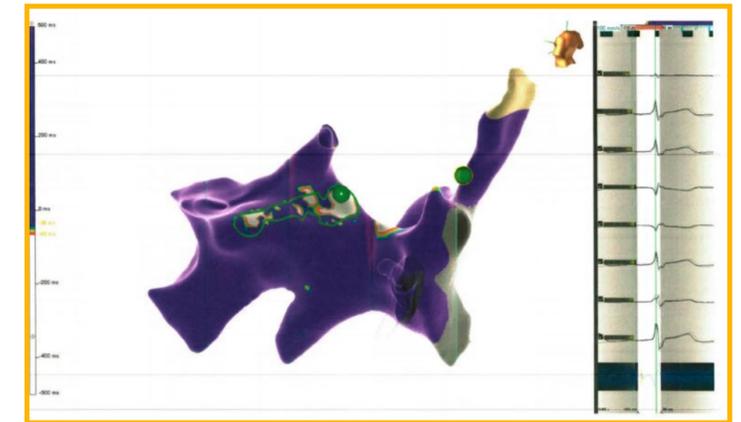


Fig 2. 3-dimensional mapping of right atrium and ventricle in left posterior oblique view demonstrating atrial and ventricular leads (Green Markers). Ventricular lead on His bundle location.



Fig 3. His bundle signal on permanent ventricular lead at end of case.

	Non-Selective Threshold	His Threshold	Sensing	Impedance
At Implantation	2V @ 0.4ms	1V @ 0.4ms	2.5 mV	460
3 Month Follow-up	2V @ 0.4ms	1V @ 0.4ms	3.5 mV	437

Table 1. Pacemaker parameters at implantation and 3 month followup

## 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