**MYOTONIC DYSTROPHY AND CARDIAC CONDUCTION SYSTEM ABNORMALITIES: HOW AWARENESS AFFECTS THE PATIENT OUTCOME?**

**K. Khalighi1,2**, S.B. Thapamagar1,2, N. Singh1,2

1Easton Hospital, Easton, PA, USA, 2Drexel University, Philadelphia, PA, USA

Background: Myotonic dystrophy is a multisystem disorder with multiple cardiac manifestations. Conduction system abnormality is the most common systemic manifestation which is usually progressive but can be unpredictable. The majority of patients die due to cardiac reasons and one-third of such deaths are sudden cardiac death. Case reports and discussion: In this case series, we present 7 consecutive patients with myotonic dystrophy presenting with recurrent lightheadedness, dizziness, near-syncope and fatigue. All had EKG evidence for intraventricular conduction delay but only 5 had first degree atrioventricular block. The HV intervals were greater than 72 msec in 5 patients. Four had inducible sustained ventricular tachycardia requiring immediate cardioversion and received ICD. Two underwent permanent pacemaker placement due to complete heart block and infra-hisian block. These electrophysiological findings are typical in patients with myotonic dystrophy. Five out of 7 patients were seen in last 2 years, an unusual surge of referrals attributed to an increased awareness of primary care physicians to cardiac manifestations of myotonic dystrophy. Since one third of cardiac deaths in these patients are sudden, prophylactic pacemaker placement is recommended despite minimal conduction system abnormality. However, the common practice is to identify patients with high risk conduction abnormalities by an electrophysiology study and provide prophylactic invasive strategies.

Conclusion: There is a clear need for identifying the subgroup of patients with myotonic dystrophy and to intervene for those at high-risk with prophylactic invasive strategies. Identifying such patients may be difficult unless there is increase awareness among our primary care physicians.