mean LVEF improved from 37,3 to 44,1% and QRS duration from 172,9 to 136,8ms. Pts with less than 50% LVEF had a response rate of 75,5% (increase in LVEF from 4-30%), and 30% of patients increase their LEVF between 10-30% (deemed super responders), only one pt decrease 10% her LVEF (a His failure implant).

CONCLUSION: Cardiac resynchronisation is feasible with less leads needed, potentially with longer duration of batteries and lesser cost for the health system. Response rate is similar to CRT historical controls but super responders are twice to three times higher tan historical controls.

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DEVELOPMENT OF A DIAGNOSTIC SCREENING ALGORITHM FOR THE IDENTIFICATION OF TRANSTHYRETIN AMYLOID CARDIOMYOPATHY IN HIGH-RISK PATIENT POPULATIONS

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BACKGROUND: Transthyretin amyloid cardiomyopathy (ATTR-CM) is an underdiagnosed disease, resulting in heart failure, arrhythmias, and valvular disease in a sizable (but still undetermined) proportion of older patients. Disease modifying therapy exists but is more effective in earlier-stage disease. Effective screening methods are needed to identify ATTR-CM at a treatable stage and improve outcomes for this complex population.

METHODS AND RESULTS: We designed a pragmatic screening study to identify patients in high-risk settings, including heart failure (HFC), atrial fibrillation (AFC), and transcatheter valve (TAVR) clinics. Patients were included if they were >60 years old, met one of several broad screening criteria, and their treating physician felt that an ATTR evaluation was appropriate. Patients with a clear alternate diagnosis or known diagnosis of amyloidosis were excluded. Patients were screened with nuclear scintigraphy (PYP). Patients also underwent blood and urine screens for monoclonal protein. Baseline demographic, clinical, laboratory, and imaging data were collected into a prospective observational registry. Patients are followed for 3 years. To date, 97 patients have been fully screened, 57% of these in HFC. The mean age of participants is 78 years; 54% are male. 20 patients (21%) have been diagnosed with ATTR-CM, including 5 (5%) who had a monoclonal protein identified. Of screening criteria used, the positive predictive value (PPV) for ATTR-CM ranged from 6-36%. 12/33 participants (PPV 36%) with moderate-severe diastolic dysfunction (DD) had ATTR-CM, 20/66 with age >70 years (PPV 30%), 19/72 with heart failure with preserved ejection fraction (HFpEF, PPV 26%), 15/59 with left ventricular septal thickness >/=12 mm (IVSD12, PPV 25%), 19/79 with natriuretic peptide values disproportionately elevated for NYHA class (BNP, PPV 24%), 2/13 with LVEF < 40% and normal ventricular dimensions (HFrEF, PPV 15%), and 1/16 with severe low-flow low-gradient aortic stenosis (AS, PPV 6%). Conversely, the negative predictive value was highest for age >70 (NPV 100%), followed by HFpEF (NPV 96%), BNP (NPV 94%), DD (NPV 87%), IVSD12 (NPV 86%), HFrEF (NPV 79%), and AS (NPV 76%). Combining screening criteria yielded improved test characteristics. A combination of [age >70 and DD] had PPV 52% and NPV 89%, while a combination of [age >70 and HFpEF] had PPV 34% and NPV 98%.

CONCLUSION: Broad screening criteria applied to high-risk patient populations yield a large number of new ATTR-CM diagnoses. Further refinement of these criteria should lead to even greater diagnostic yield while minimizing unneeded testing. Efficient diagnosis of ATTR-CM may allow earlier diagnosis and more effective therapeutic interventions.

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EARLY DETECTION OF POLYNEUROPATHY IN PATIENTS WITH HEREDITARY TRANSTHYRETIN AMYLOIDOSIS CARDIOMYOPATHY

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BACKGROUND: Hereditary Transthyretin Amyloidosis (hATTR) is a disorder that can present with cardiac, neurological, or mixed signs and symptoms. Patients with hATTR cardiomyopathy (CM), can show early neurological signs which lead to the eventual onset of neurological symptoms. Early detection of polyneuropathy among patients with hATTR-CM can identify candidates for new disease modifying therapies that could significantly reduce mortality and morbidity in these complex patients. The purpose of this study is to examine the role of systematic neurological examinations in patients with hATTR-CM.

METHODS AND RESULTS: Twenty-eight patients with hATTR-CM who did not have any neurological symptoms were enrolled in this prospective study from the Amyloidosis Clinic at the University Health Network. All patients (average age 65.71± 14.23 years; 39.3% males; 60.7% females) were referred to a neurology program specialized in hATTR polyneuropathy (PN). Comprehensive neurological assessments were conducted including a physical examination and nerve conduction studies (NCS). NCS was performed on the upper and lower extremities. Twelve (42.9%) patients did not display any neurological abnormalities on the physical examination, but they had abnormalities on the NCS suggesting the presence of TTR-PN. The abnormalities on the NCS were diffuse sensory greater than motor polyneuropathy. Ten (35.7%) patients displayed clinical and NCS abnormalities. Six (21.4%) patients did not display any clinical or NCS abnormalities. Based on the systematic neurological assessment, twenty-two patients in our study were identified as