Table 1. Safety Outcomes* for outpatient intravenous diuresis visits in patients with cardiac amyloidosis

Safety Outcome, n (%)	All cardiac amyloidosis IV Diuresis Visits (N=56)	AL-CA visits (N=26)	ATTR-CA visits (N=30)
Asymptomatic hypotension	8 (14)	4 (15)	4 (13)
Symptomatic hypotension	0	0	0
Mild acute kidney injury	2 (4)	0 (0)	2 (7)
Severe acute kidney injury	0	0	0
Mild hypokalemia	9 (16)	3 (12)	6 (20)
Severe hypokalemia	3 (5)	0 (0)	3 (10)

*Safety outcome definitions: Symptomatic hypotension = any decrease in SBP or DBP with associated lightheadedness, dizziness, pre-syncope, or syncope; Asymptomatic hypotension = ≥20 mmHg decrease in SBP or ≥10 mmHg decrease in MAP post-diuresis; Mild acute kidney injury = ≥ 20% decrease in post-diuresis eGFR; Severe acute kidney injury = ≥2.0 times increase in post-diuresis creatinine; Mild hypokalemia: K+ ≤ 3.5 mmol/L post-diuresis; Severe hypokalemia: K+ ≤ 3.0 mmol/L post-diuresis.

Abbreviations: AL-CA=light chain cardiac amyloidosis; ATTR-CA=transthyretin cardiac amyloidosis; IV=intravenous

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Outcomes After Bariatric Surgery In Patients With Ventricular Assist Devices: Systematic Review And Individual Participant Data Meta-analysis

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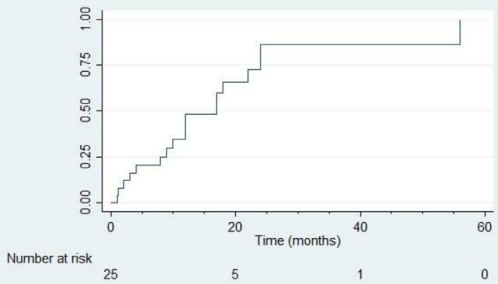
Background: Body mass index (BMI) ≥ 35 kg/m^2 is a major contraindication for heart transplantation (HT). Consequently, many patients who would otherwise be good HT candidates require ventricular assist devices (VADs) as destination therapy. Furthermore, some HT candidates who initially had a BMI < 35 kg/m^2 but gained weight during VAD support may lose their candidacy. All evidence for a potential role of bariatric surgery (BS) in obese VAD patients comes from small cohorts and case reports which makes its interpretation challenging. **Methods:** A systematic search was performed in ClinicalTrials.gov, Cochrane, Embase and PubMed. Other sources screened included Google Scholar, meeting proceedings, journal sites, and references cited in included studies. Selected subjects were obese, adult patients with VADs who underwent BS and had follow-up BMI data. **Results:** Twelve references with 29 patients (age 41.9 [± 12.2] years, 63.6% male) were included. The baseline BMI was 45.2 (± 6.7) kg/m^2. Most patients (82.8%) underwent sleeve gastrectomy while 17.2% received Roux-en-Y gastric bypass. Median follow-up was 24 (12-30) months. Eleven (39.3%) patients had postoperative adverse events after BS. Among the 23 patients with documented listing status (listed vs. not listed for HT) after BS, 78.3% lost enough weight and were listed for HT. Thirteen patients underwent HT 14.4 (± 7) months after BS, and three patients had myocardial recovery with VAD explantation after weight loss. Twenty-two of 28 (78.6%) patients achieved the composite outcome of BMI < 35 kg/m^2/OHT/listing for OHT/myocardial recovery at 11 (3-17) months. Figure 1 shows a Kaplan-Meier curve for the composite outcome. There were no deaths during the HT-free 1-year follow-up. **Conclusions:** In VAD patients who are not HT candidates due to obesity, BS may allow for enough weight loss to improve their candidacy for HT or even achieve myocardial recovery. Although there were no deaths reported during HT-free 1-year follow-up, there was a high rate of adverse events during the postoperative period. However, the rate of these events may decrease as programs become more experienced with BS in VAD patients.

Figure 1. Kaplan-Meier estimate of the composite outcome



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Survey Conducted At An Academic Medical Center Revealed Knowledge Gaps of Transthyretin Amyloidosis Cardiomyopathy Predominantly In Primary Care Providers

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Introduction: Transthyretin amyloidosis (ATTR) is a progressive, fatal disease caused by the formation of amyloid fibrils that misfold and accumulate in various tissue in the body, including the heart, leading to a form of heart failure called transthyretin amyloidosis cardiomyopathy (ATTR-CM). ATTR-CM consists of two subgroups: a wild-type (ATTR-wt) or an inherited mutant autosomal dominant gene (hATTR). ATTR-wt and hATTR have an estimated median survival 3.6 years and 2.5 years after diagnosis, respectively. The diagnosis of ATTR-CM is often delayed or missed, and one potential factor playing in role in this may be lack of provider awareness and knowledge of this disease. The objective of this analysis was to assess ATTR-CM knowledge of healthcare providers (HCPs) at a large academic medical center. The hypothesis of this analysis is that there is lack of foundational knowledge regarding ATTR-CM among HCPs. **Methods and Results:** This analysis utilized a cross-sectional observational design in which HCPs were asked to voluntarily complete an electronic questionnaire. This questionnaire collected demographic characteristics, captured awareness of ATTR-CM, confidence in managing patients with ATTR-CM, and desire for additional education. Forty-six HCPs completed the survey. All respondents identified themselves as physicians, with the majority practicing in internal medicine or family medicine (43.75%). Approximately 2/3 of the HCPs had been in practice ten years or less. Most respondents reported that less than half of their current patient population have heart failure (87.5%). All respondents stated some level of awareness to ATTR-CM. However, the majority (84.79%) were not confident differentiating between hATTR and ATTR-wt, and 73.91% were not familiar with the various mutations in hereditary ATTR-CM. The survey revealed areas of knowledge gaps with majority indicating not being confident with genetic testing to determine if ATTR-CM is hereditary (73.3%), identifying the soft tissue signs and symptoms of ATTR-CM (67.39%) and differentiating between ATTR-CM and light chain amyloidosis (63.04%). The majority (67.39%) indicated a desire for further education. **Conclusions:** Major knowledge gaps were identified in this survey indicating further education is needed to increase clinician suspicion of ATTR-CM, so patients, who may require further testing to make an accurate diagnosis, can be identified.

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Improvement of Cardiac Structure And Function In Hereditary Transthyretin Amyloidosis Cardiomyopathy With Inotersen: A Case Report

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Introduction: Hereditary transthyretin amyloidosis (hATTR) is a progressive, fatal disease caused by systemic deposition of misfolded transthyretin (TTR) fibrils resulting in cardiomyopathy (CM) and polyneuropathy (PN). Treatment options include stabilizers (tafamidis) to prevent TTR tetramer dissociation, silencers (inotersen, patisiran) that inhibit hepatic *TTR* synthesis, or liver transplantation. Silencers have favorable outcomes in PN, with limited data on CM. This case highlights improvement in hATTR-CM in a patient treated with inotersen. **Patient Case:** A 68 year old female with a family history of hATTR presented for evaluation as a heterozygous carrier of the p.T80A (T60A) *TTR* mutation. She experienced refractory nausea, vomiting, diarrhea alternating with constipation, weight loss (10 kg) over the preceding 6 months, and 2 months of new onset exertional dyspnea and was in NYHA Class 2 heart failure. ECHO showed severe concentric hypertrophy (septal and posterior walls, 2.1 cm and 1.9 cm), moderate reduction in left ventricular systolic function with a left ventricular ejection fraction (LVEF) of 32%, and moderate pericardial effusion. BNP and troponin I levels were 532 pg/mL and 0.05 ng/mL. Diffuse 4-chamber delayed gadolinium enhancement was noted on cardiac MRI (A). Nerve conduction studies confirmed sensorimotor neuropathy with an estimated neuropathy impairment score of 10. Furosemide and spironolactone resulted in a 6.4 kg diuresis with improved dyspnea. Right heart catheterization showed right atrial pressure of 6 mm Hg, pulmonary artery pressure of 39/16 mm Hg, and pulmonary capillary wedge pressure of 10 mm, with a cardiac index of 2. Episodic orthostatic hypotension precluded initiation of beta blockers and vasodilators. She was initiated on inotersen for symptoms of neuropathy. After 6 months, LVEF increased to 45% and diastolic function improved from stage 2 to 1. The pericardial effusion resolved. Six minute walk distance showed an increasing trend over 10 months (B). BNP levels were 108 pg/mL after 1 year on inotersen, and modified BMI stabilized. **Conclusion:** Inotersen was associated with improvements in cardiac function in this patient with hATTR-CM who could not be treated with guideline-directed heart failure therapy. For hATTR-CM with no convincing benefit from conventional therapy beyond symptom control, inotersen may offer a treatment option to alter the natural history of this fatal disease.