


Median Arcuate Ligament Syndrome Presenting as Hyperadrenergic POTS.



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- POTS is a symptom complex rather than a disease entity itself, with underlying **heterogeneous pathophysiologies**, such as:
 - Neuropathic
 - Mast Cell activation
 - Autoimmune

Index Case

- 16 years old female with Hyperadrenergic POTS
- Persistent Gastrointestinal symptoms including postprandial abdominal pain, nausea, vomiting and weight loss.
- Abdominal Bruit

Index Case

Born at 39 wks. GA, Birth weight: 7lbs 5oz, Apgar's 8/9

Frequent severe gastric reflux and high pitch cry

At 11 years old: long bout of vomiting, weight loss and significant abdominal pain for several weeks.

Abdominal CT suggestive of mesenteric adenitis

13 ½ yr. old, 5 days episodes of vomiting, Missed 3 + weeks of school primarily due to night-time vomiting and lethargy during the day.

Frequent to ER visits due to abdominal pain, persistent vomiting, and shortness of breath. Sharp abdominal pains, knife like, below the xiphoid process, and radiates throughout the ULQ and underneath rib cage. Some nights she can't get off the bathroom floor, pain at 10/10

Daytime brings intense abdominal pains, pains are almost always present, and intensify with eating.

Postprandial discomfort and sense of extreme fullness.

Tried liquid diet x10, no improvement.

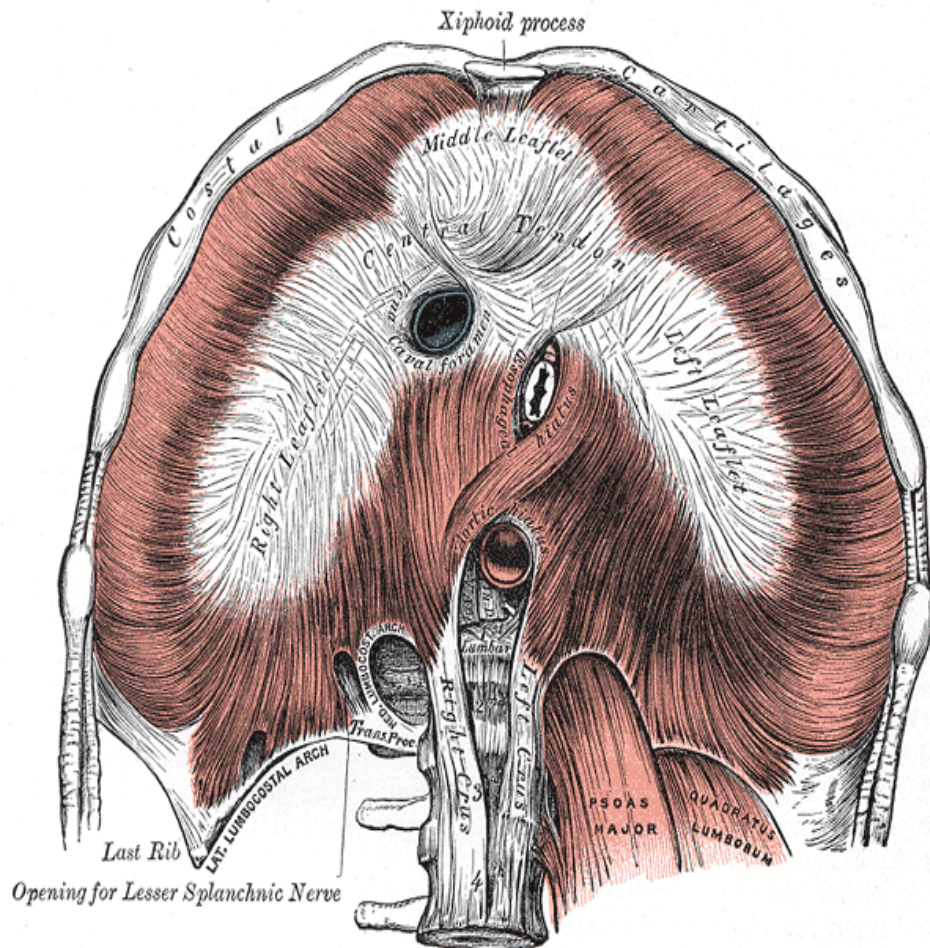
No Relief from any medication



Median Arcuate Ligament

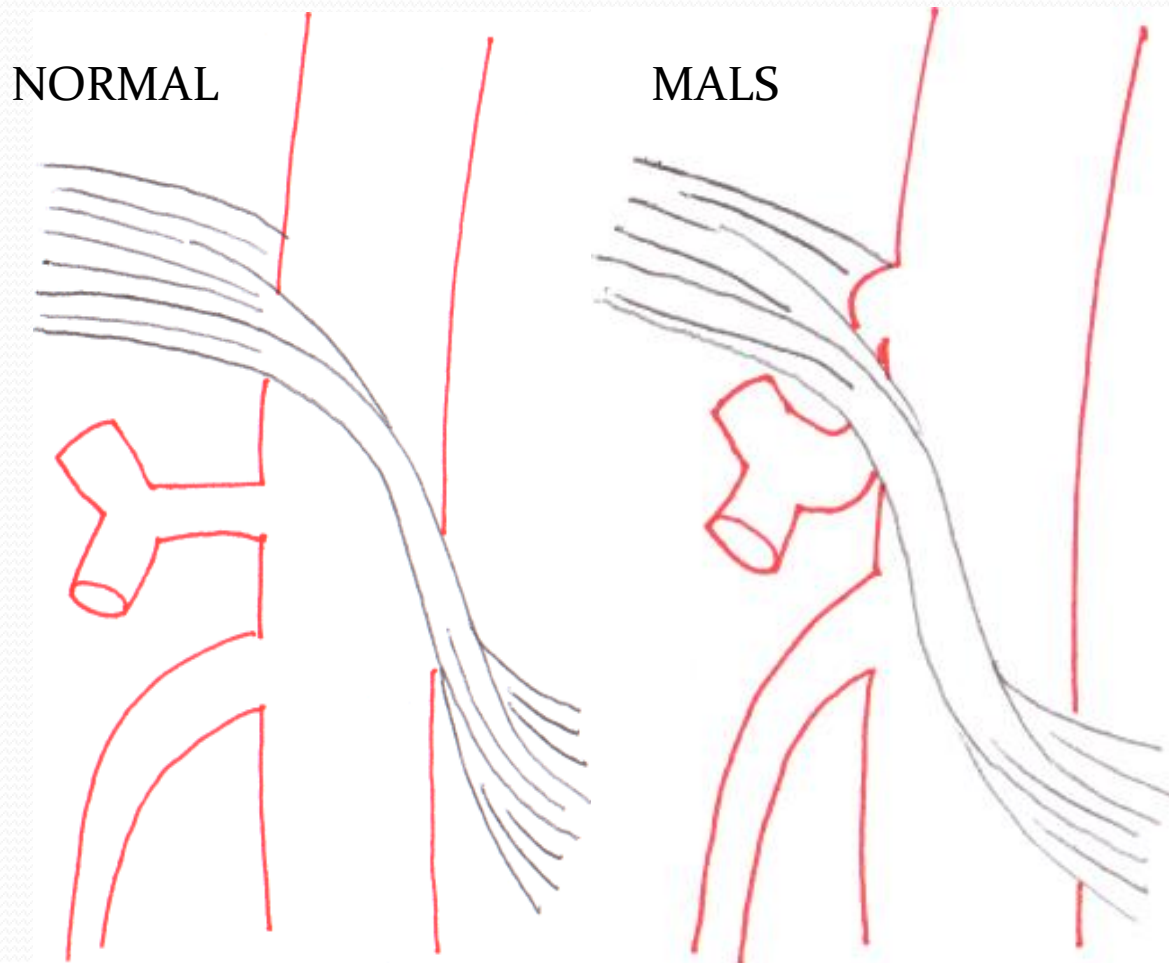
A ligament formed at the base of the diaphragm where the left and right diaphragmatic crura join near the 12th thoracic vertebra.

This fibrous arch forms the anterior aspect of the aortic hiatus



Median Arcuate Ligament Syndrome(MALS)

The median arcuate ligament encroaches on the celiac artery and celiac ganglia



Study Population

- **Patients referred to CHI Dysautonomia Clinic**
 - **February 2013 through August 2014**

- **Met criteria for POTS:**

POTS was defined as a HR increase of ≥ 30 bpm within 10 minutes of upright tilt in the absence of hypotension

- **Persistent Gastrointestinal Symptoms despite extensive GI laboratory and Endoscopic Work up.**

Study Protocol

- **Retrospective chart review**
 - **Demographics**
 - **Previous Testing**
 - **Medications**
 - **Clinical Course**
- **Tilt Table Test**
- **Valsalva's Maneuver**
- **Celiac and Superior Mesenteric Artery Duplex Testing**
- **Vascular Abdominal CT-Angiogram.**
- **Review of Surgical Laparoscopic outcome**
- **Quality of life (QOL) was determined by pre- and postsurgical administration of PedsQL™ questionnaire**

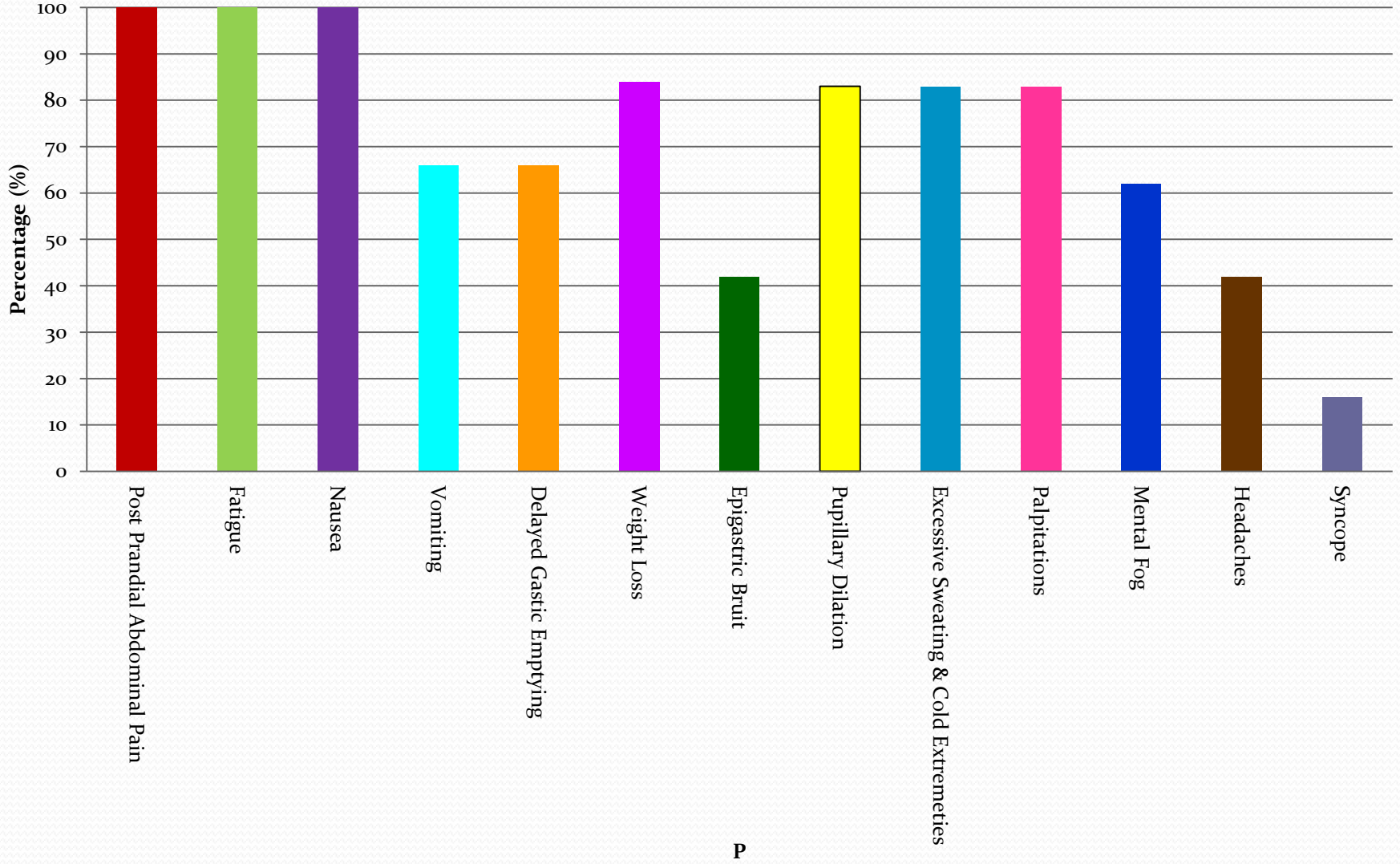
Prior Work up and Diagnostic Studies

- 1. Complete blood count with differential, platelet count
- 2. ESR, C-reactive protein
- 3. Amylase, lipase
- 4. Comprehensive metabolic panel (including liver function tests)
- 6. Thyroid function tests
- 7. Celiac Disease workup
- 8. UGI
- 9. Upper endoscopy with biopsy
- 10. Colonoscopy
- 11. Abdominal ultrasound
- 12. Abdominal CT-scan
- 13. Urinalysis,
- 14. pregnancy test

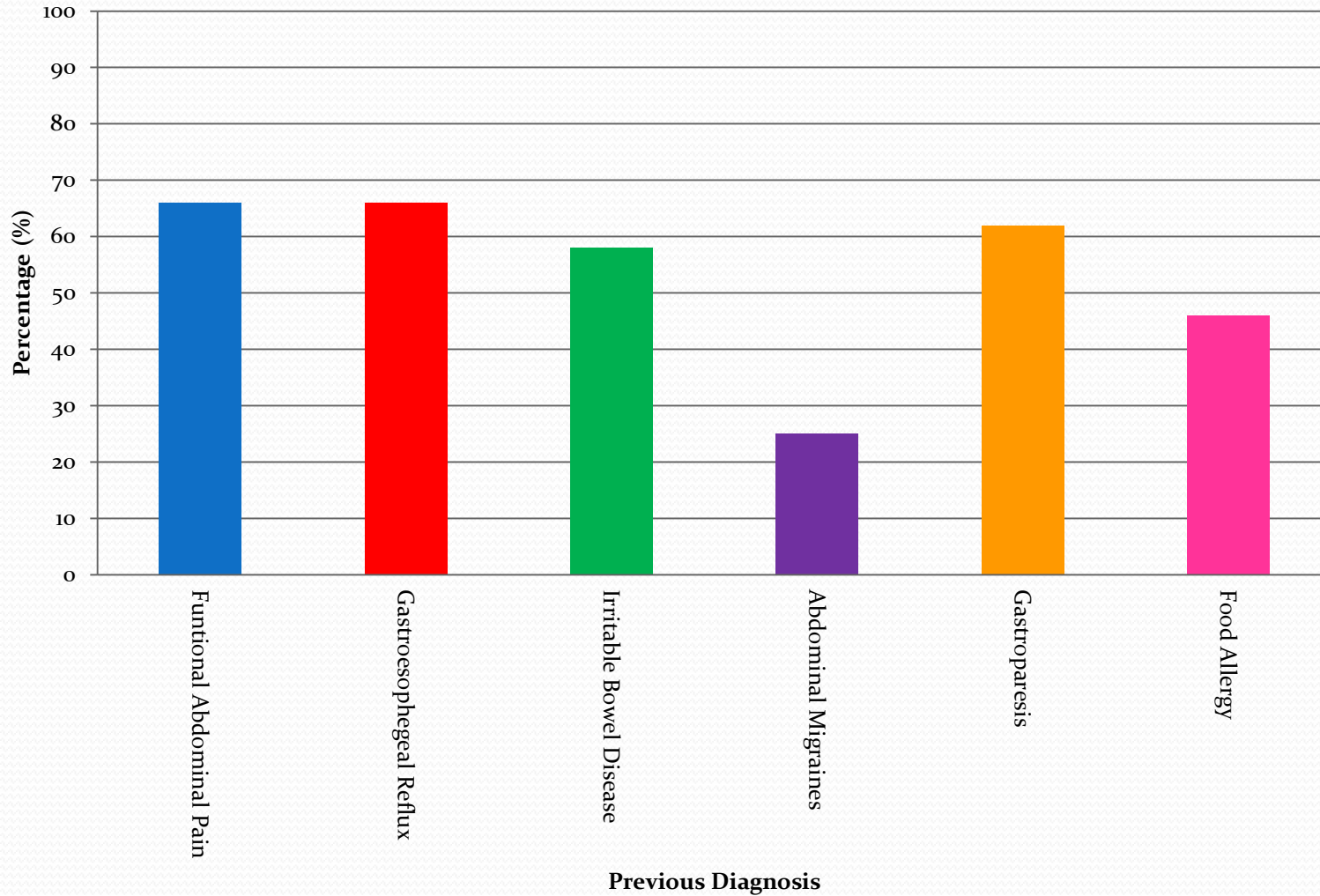
Demographics

Subjects (n)	24
Gender (Percent Female)	21/3(79⁰%)
Median Age (years)	16.2
BMI	20 ± 4
Race (Percent Caucasian)	100⁰%
Years of symptoms @ presentation	4.1 ± 3.2
State of Residence	Virginia 6, Maryland 4, West Virginia 2, Pennsylvania 3, New Jersey 4, Florida 2, Alabama 2, Ohio 1.

Presenting Symptom % Distributions



Previous Diagnoses %



Tilt Table Results

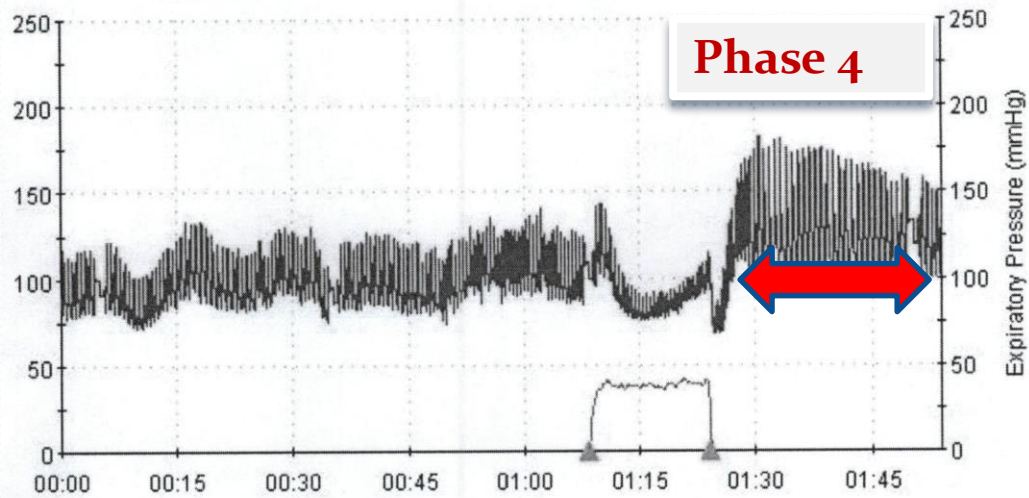
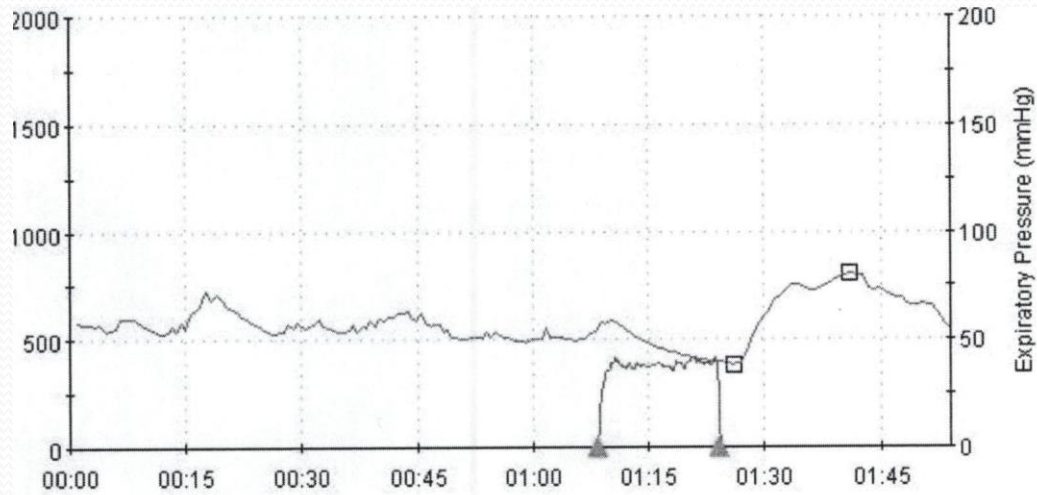
	MALS	Control	P-Value
Supine HR	90±5	71±4	0.05
HR@10 Minute tilt	138±4	89±5	
Orthostatic HR change	48±4	18±4	0.01
Supine Systolic BP	115±4	98±6	0.05
Systolic BP@10 Minute tilt	128±4	102±5	0.05
Orthostatic Systolic BP change	13	4	0.01

Valsalva Findings

SBP at the end of late phase II of the Valsalva maneuver was 132 ± 5 versus 110 ± 9 in controls; $P=0.05$),

SBP overshoot in phase IV (55 ± 6 versus 15 ± 3 mm Hg in controls; $P < 0.05$),

these findings were consistent with Hyperadrenergic state.



Vascular Duplex Examination

Equipment:

Acuson Sequoia 512 9 (Acuson Corp, Mountainview, CA) ultrasound scanner with linear array 4-7 MHz or 5-10 MHz transducers

Protocol:

The Celiac and SMA were examined in the supine position. The Peak systolic velocity (PSV) and end diastolic velocity (EDV) were measured from the proximal, mid and distal arterial segments

Same Measurements were repeated at deep inspiration and at end expiration.

Measurements were suggestive of flow-reducing stenosis:

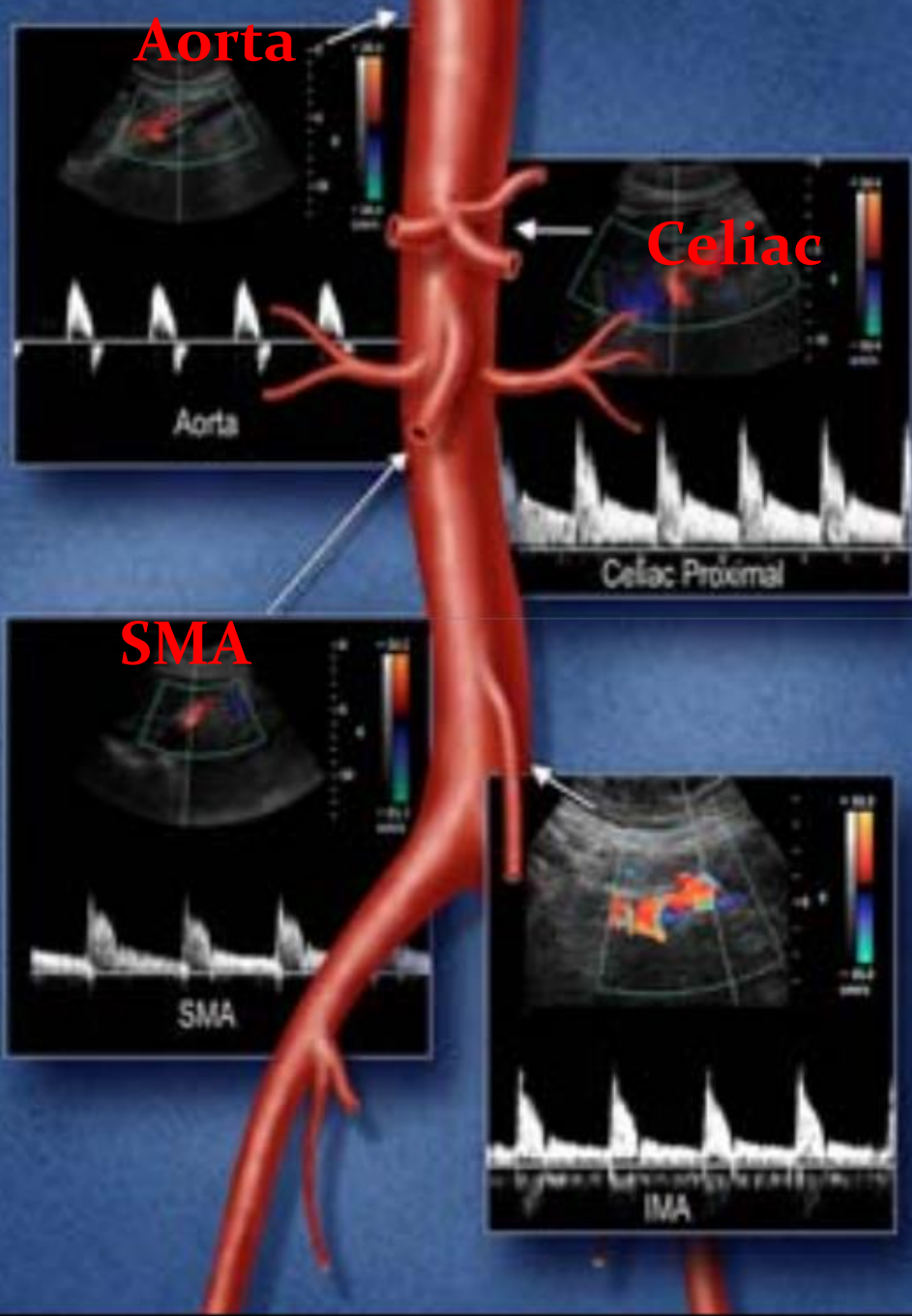
PSV > than 300 cm/sec

EDV > 55 cm/sec suggested a flow-reducing stenosis

Post-stenotic Color Doppler turbulence

Spectral broadening of the Pulsed Doppler waveform

A Decrease in PSV with deep inspiration and increase with expiration was suggestive of MALS.



Aorta

Celiac

SMA

IMA

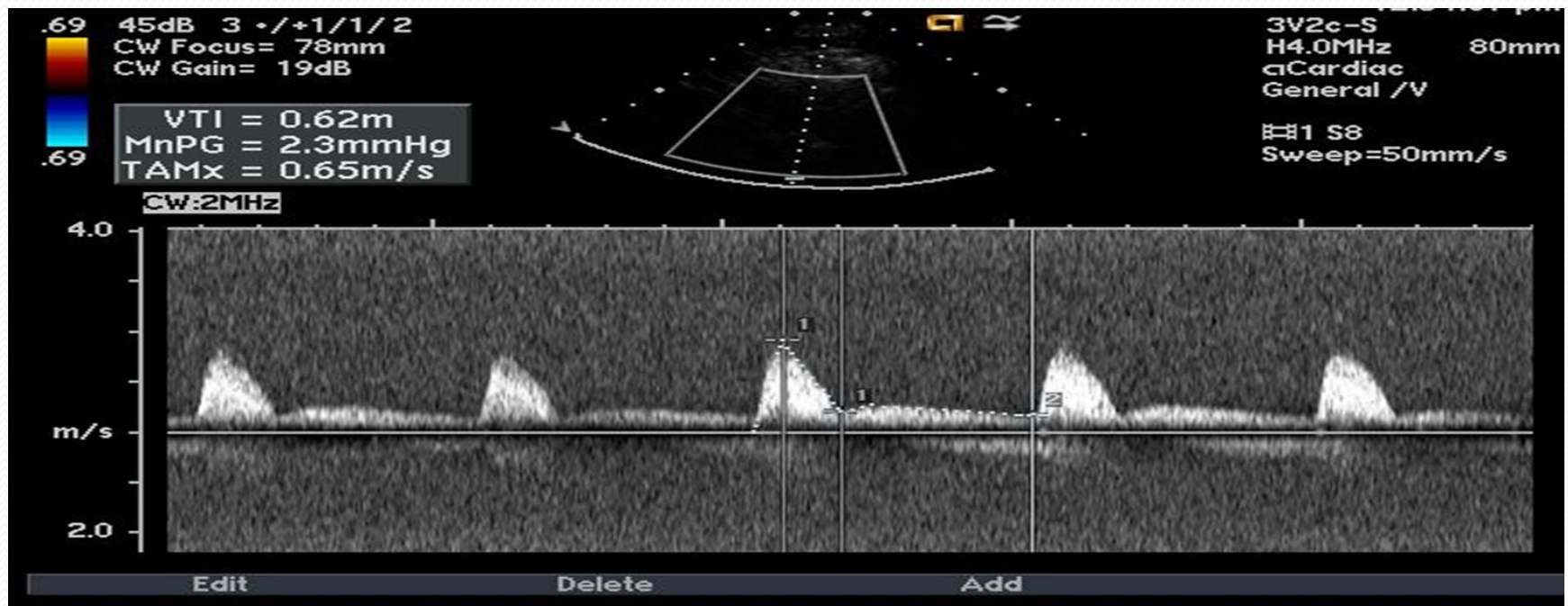
Normal Duplex Velocity Measurements

Site	Peak Systolic Velocity (PSV) cm/s	End Diastolic Velocity (EDV) cm/s	Turbulence
Abdominal Aorta	80 +/- 20	0.0	None
Celiac	101 +/- 3.5	33 +/- 3.4	Trivial
Superior Mesenteric (SMA)	113 +/- 3.9	15 +/- 1.1	Trivial

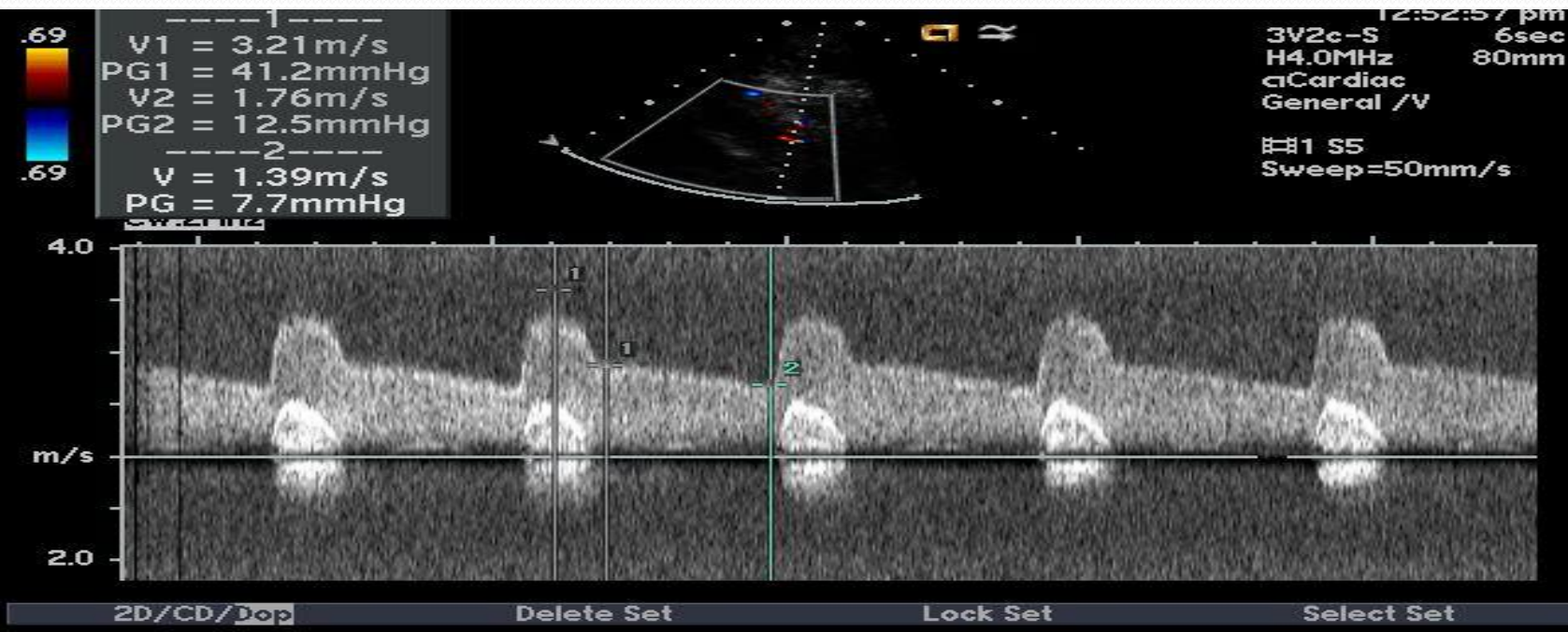
Duplex Celiac Measurements Pre and Post Operatively

	Mean PSV Deep inspiration	Mean PSV End Expiration	Mean EDV Deep Inspiration	Mean EDV End Expiration	
Pre- operative	190±18	486±26	32±4	138±12	
Post- operative	178±12	220±18	26±6	34±8	
Change	12±15	266±22	6±4	104±4	

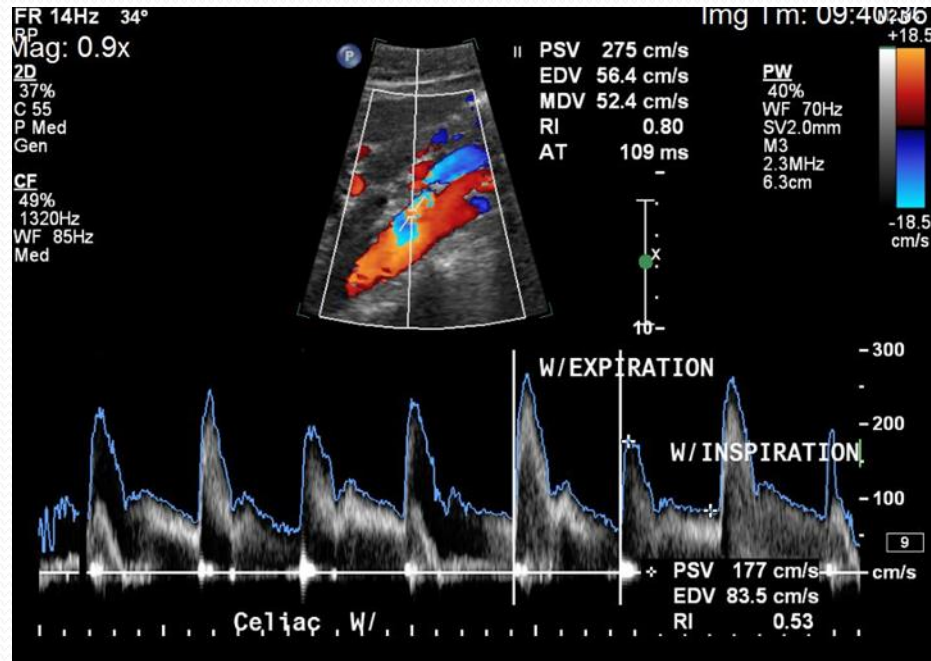
Aortic Doppler Flow Velocity



Celiac Artery Doppler Flow Velocity



Accentuated Celiac Flow Restriction at End Expiration



90% Celiac Artery Stenosis



Abdominal CT-Angiogram

Fish Hook Configuration



Operative Technique

- Laparoscopic abdominal approach
- Electrocautery is used to open the diaphragmatic crura directly onto the abdominal aorta.
- The muscle fibers are dissected and divided with cautery in a stepwise manner until the adventitia of the aorta is exposed.
- Dissection proceeds distally until the origin of the celiac artery is identified.
- The ganglionic nerve fibers of the celiac plexus overlying the celiac artery are also divided by hook electrocautery

Intraoperative Ultrasound

- A laparoscopic ultrasound probe is used to measure flow in centimeters per sec (cm/sec), within the aorta and celiac artery looking closely at the area of the narrowing.
- Typically flows within the proximal celiac artery are well above 300 cm/sec in all patients.
- Resection of the median arcuate ligament is performed until normal celiac artery flow velocity is documented to be around the aortic flow.

Surgical Results

- There were no deaths
- One complication resulting from inadvertent celiac artery injury requiring conversion to open laparotomy and surgical repair of the celiac artery tear.

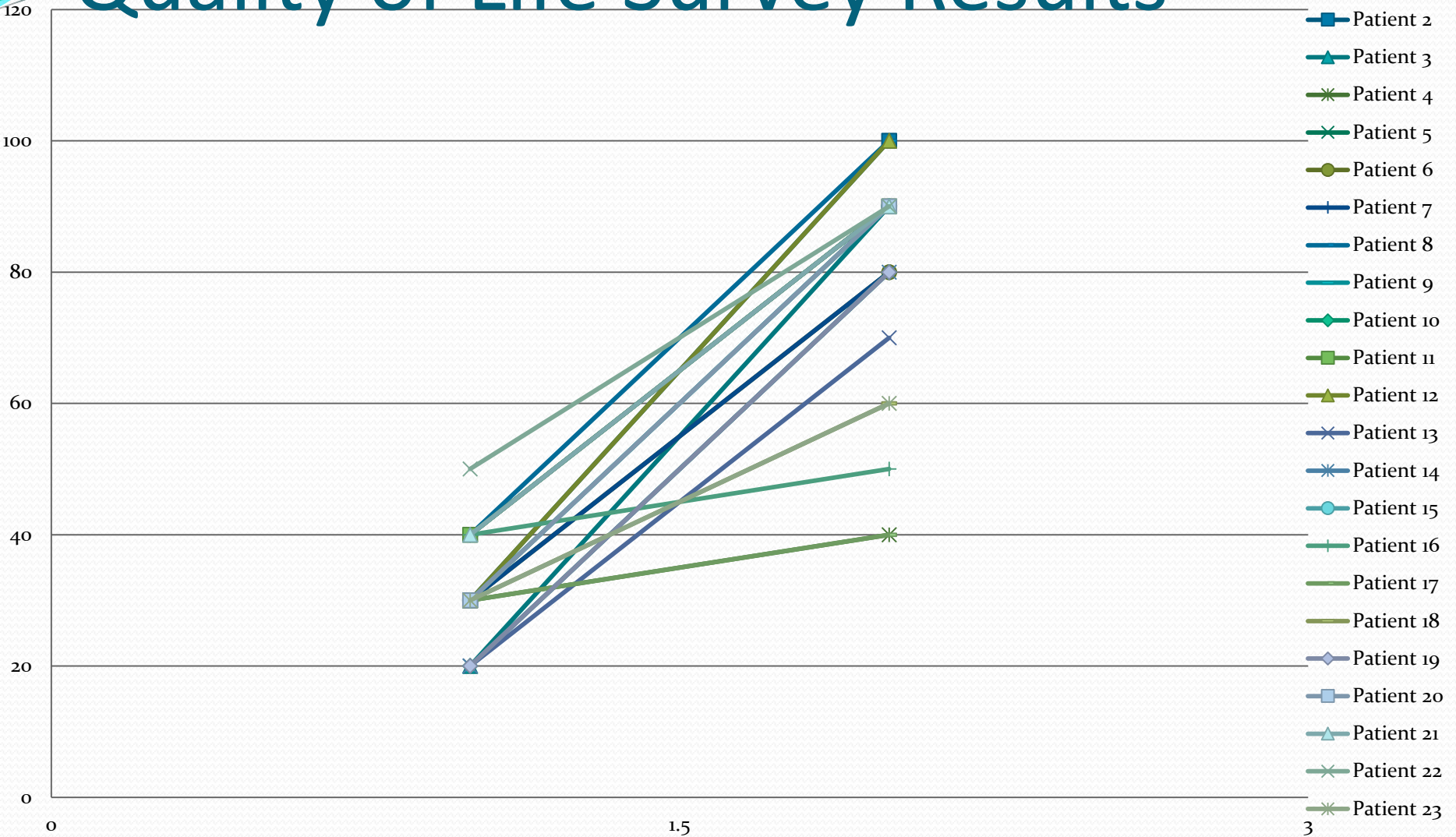
Quality of life (QOL)

- Scores were based on physical, emotional, social and school functioning and converted to a 0–100 scale with higher scores indicating better QOL
- Patients who did not return the survey, received a phone call and follow-up e-mails that included the QOL survey.

Quality Of Life Survey Results

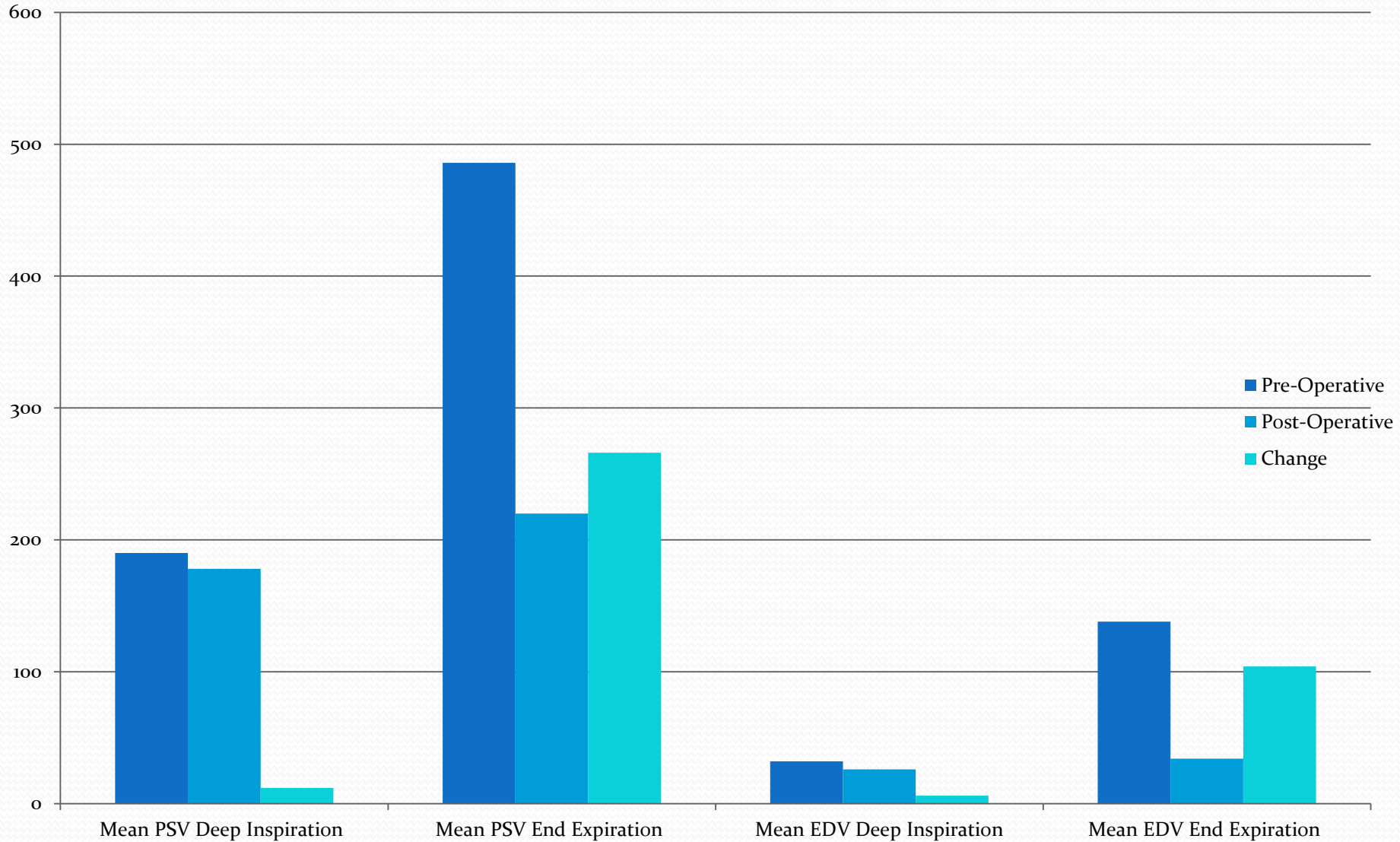
- The average total preoperative score was 43.2 and
- the average total postoperative score was 84.6.
- Median follow up of 9.2 months (2.5–22 months)

Quality of Life Survey Results



Pre-Op VS. Post Op

Duplex Follow Up Results



Incidence of MALS

32% in patients with POTS
and persistent
Gastrointestinal symptoms

Pathognomonic Sign

Epigastric Bruit

@

End Expiration

Celiac Neuropathy?

- Direct Catecholamine Release
- Regional Complex Pain Syndrome
- Disturbed Adrenal-Cortical Interactions
- Disturbed Ovarian Steroidogenesis

