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### A Case of Esophagogastric Junction Outflow Obstruction Resulting to Megaesophagus in a 37-Year-Old Male

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# A case of esophagogastric junction outflow obstruction resulting to megaesophagus in a 37-year-old male

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## Abstract

Esophagogastric junction outflow obstruction (EGJOO) is a major motility disorder characterized by an increase of > 15 mmHg in the integrated relaxation pressure (IRP) with or without problems in esophageal motility.<sup>1,2,3</sup> This presents as dysphagia, chest pain, regurgitation, heartburn, cough, and a globus sensation, with dysphagia. Severe cases may lead to a megaesophagus causing compression of mediastinal structures.<sup>6</sup> Its similarity to other esophageal disorders urge an investigation to improve its identification and treatment. This report presents a case of EGJOO in a 37-year old cachectic male patient with dysphagia and vomiting, eventually determined to be EGJOO with diagnostic manometry.

## Case Presentation

A 37-year old male with no medical history, presented to the ED with a complaint of dysphagia, nausea and vomiting, reported to have been gradually worsening over the past year. Patient initially noted having reflux symptoms. He then started having a sensation of food getting stuck in the chest. To relieve the sensation, the patient would reposition himself into a more upright or slightly leaning forward position, followed by drinking fluids. Laying flat was reported to result in worsening of symptoms. He saw his primary care doctor for the evaluation of these symptoms, and was prescribed with PPI but provided no relief. Patient has lost a significant amount of weight, estimated to be around 30-40 lbs loss.

3 days prior to presentation at the ED, patient noted symptoms progressed to complete intolerance of both solids and liquids, resulting in significant nausea and vomiting. Patient, feeling very weak, ill and diaphoretic, prompted patient to seek medical care.

Upon presentation, patient was cachectic and ill-appearing, but otherwise comfortable. Vital signs were HR of 132, BP of 119/86 mmHg, RR of 16, Temperature of 98 F (36.7 C). He weighed 54.4 kg with BMI of 16.7 kg/m<sup>2</sup>. On examination, significant for temporal wasting, dry lips, and bull-frog neck appearance. Rest of the physical examination was unremarkable.

On workup, lab work revealed CBC: WBC 23, Hgb 17.3 g/dL, Hct 51.9%, BMP: Na 152 mmol/L, BUN 76 mg/dL, Creatinine 1.50 mg/dL, AST 82 U/L, ALT 84 U/L. Initial Lactate was 2.5 mmol/L. Chest Xray demonstrated increased convex margins and large air-fluid level in the superior mediastinum favored to represent a distended esophagus. CT Chest/Abdomen/Pelvis with IV contrast was then done, revealing diffusely distended esophagus measuring 6.8 x 9cm with anterior displacement of mediastinal structures, and pneumomediastinum. The esophagus was dilated throughout with beaking of the distal portion, suspicious for underlying lesion versus achalasia.

Patient was then transferred to a higher level hospital. Endoscopic ultrasound showed markedly distended esophagus, hypertonic LES dilated to 18mm, suspect for achalasia. Manometry done showed 100% failed swallows and 100% panesophageal pressurization, consistent with Achalasia Type II.

Patient then had EGD done which showed EGJOO with large hiatal hernia and resultant megaesophagus, S/P self-expandable stent placement. Patient's diet was gradually advanced with avoidance of refeeding syndrome post-procedure and placed on PPI therapy. Surgery for the hiatal hernia was planned in the future.

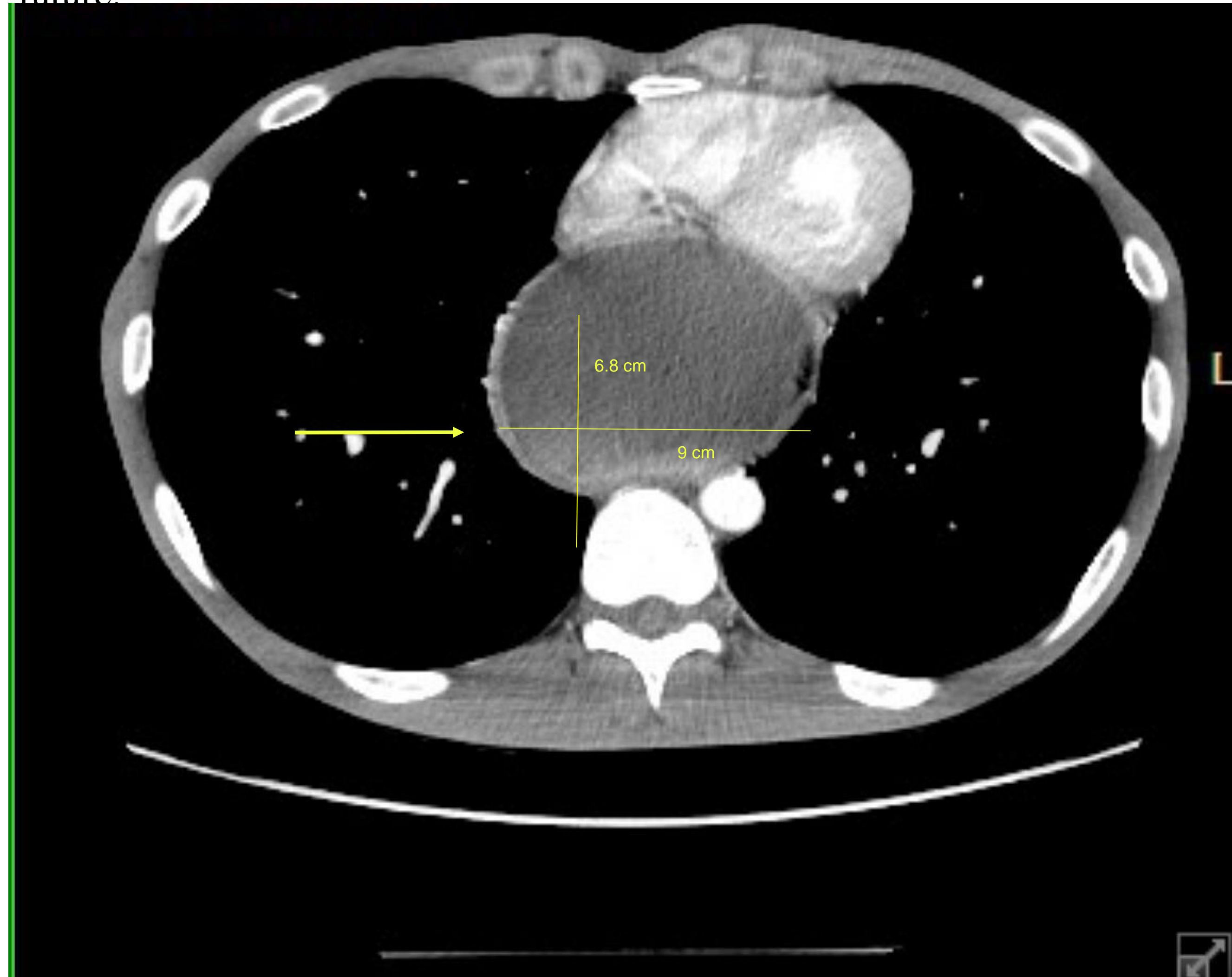


Figure 1. Computed tomography showing distended esophagus measuring 6.8 cm x 9 cm

## Discussion

Esophagogastric junction outflow obstruction (EGJOO) is characterized as a major syndrome seen with a significant rise in integrated relaxation pressure (IRP) (> 15 mmHg).<sup>1</sup> Despite characteristic elevation of the IRP, it is differentiated from achalasia as esophageal motility remains intact or weak.<sup>1,5</sup> Previous studies show that incidence of the EGJOO is rare, ranging from 1.6-11% among patients undergoing high resolution manometry.<sup>3,7</sup>

Prognosis of EGJOO is reported to be variable, with 75% of cases reporting resolution of symptoms even without treatment.<sup>5</sup> However, severe cases of EGJOO may lead to a sigmoid esophagus causing severe dysphagia and compression of mediastinal structures leading to further complications.<sup>6</sup>

Signs and symptoms of EGJOO reported across various studies include dysphagia, chest pain, regurgitation, heartburn, cough, and a globus sensation, with dysphagia cited as most common.<sup>1-3,5</sup> Its causes can be idiopathic or secondary to other conditions such as post-surgery complications, esophageal structural anomalies, malignancies, and inflammation. Secondary causes are responsible for 13-66% of EGJOO cases.<sup>5</sup> Hiatal hernia, however, accounts for about 70% of EGJOO cases secondary to an anatomical abnormality, the highest reported among structural causes.<sup>2</sup> Medications such as chronic opiate use can also be a secondary cause.<sup>1,5</sup>

While differentiated from achalasia, EGJOO can further progress to achalasia and a megaesophagus. At this stage, patients suffer from worsening dysphagia, with cardiorespiratory complications due to compression of mediastinal structures.<sup>3,6</sup> High-resolution manometry is used to identify topographic patterns and aid in the diagnosis of the disorder.<sup>5</sup> Its etiology, however, remains ambiguous as diagnosis is made mainly through manometry, thus, a need for further exploration of diagnostics for such cases.<sup>2</sup> A thorough medical history, accompanied by other imaging studies such as EGD, is also necessary to consider possible secondary causes (e.g. hiatal hernias), differentiate the condition, and to promptly treat underlying causes, as necessitated.<sup>3</sup>

Identification of underlying causes is the first step in EGJOO management, hence appropriate diagnosis is highlighted in its treatment.<sup>3</sup> For cases secondary to hiatal hernia, primary or reparative surgery may be done to manage the condition.<sup>3,4</sup> Medical management is also provided for accompanying symptoms such as reflux esophagitis through administration of proton-pump inhibitors.<sup>4-5,7</sup> For milder cases, treatment may be directed at the LES to release pressure in the area such as pneumatic dilation, PEOM, and botulinum injections.<sup>4</sup>

## Conclusion

EGJOO is a motility disorder that greatly impacts a patient's quality of life. With various etiologies, identification of underlying causes of the condition through accompanying tests and imaging for appropriate management and treatment is emphasized.

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