

# The Use of Contrast Echocardiography in the Diagnosis of an Unusual Cause of Congestive Heart Failure: Achalasia

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*Extrinsic compression of the left atrium is a potentially life-threatening but unusual cause of congestive heart failure. Achalasia is a motility disorder characterized by impaired relaxation of the lower esophageal sphincter and dilation of the distal two-thirds of the esophagus. We report only the third known case in the world literature of massive left atrial compression by a dilated esophagus in a patient with achalasia. The use of contrast echocardiography with perflutren protein-type A microspheres allowed for differentiation between a compressive vascular structure and the esophagus. This resulted in prompt treatment leading to hemodynamic stability after nasogastric decompression and Botulinum toxin injection at the gastroesophageal junction. (ECHOCARDIOGRAPHY, Volume 21, February 2004)*

*achalasia, contrast, echocardiography, left atrium, compression*

An 81-year-old female with a past medical history significant for coronary artery disease, hypertension, and Parkinson's disease was admitted to a community hospital for complaints of substernal chest pain and dysphagia. She also complained of progressive shortness of breath and was subsequently intubated for respiratory failure. Physical exam revealed hypotension (blood pressure 85/55 mmHg), regular pulse at 90 to 100 beats per minute, bilateral jugular venous distension, and pulmonary rales.

Chest X ray revealed a tubular structure in the upper mediastinum with an air-fluid level, pulmonary vascular congestion, and a right pleural effusion. Congestive heart failure was the working diagnosis.

A transthoracic echocardiogram revealed an echogenic tubular mass extrinsically compressing the left atrium with the major and minor diameters of 58 and 52 mm, respectively. The remaining left atrial cavity was crescentic in shape. It covered a surface area of approximately 30 mm<sup>2</sup> in the apical 4-chamber view at

the end of ventricular systole. In addition, there was minimally detectable color flow within it (Fig. 1).

Because of its anatomic location and appearance, its differential diagnosis included a dilated and extended esophagus or a tortuous and dilated descending thoracic aorta. A contrast injection of 0.5 ml of perflutren protein-type A microspheres (OPTISON®, Amersham Health, St. Louis, MO) into a peripheral vein at a rate of 1 ml per second was performed.

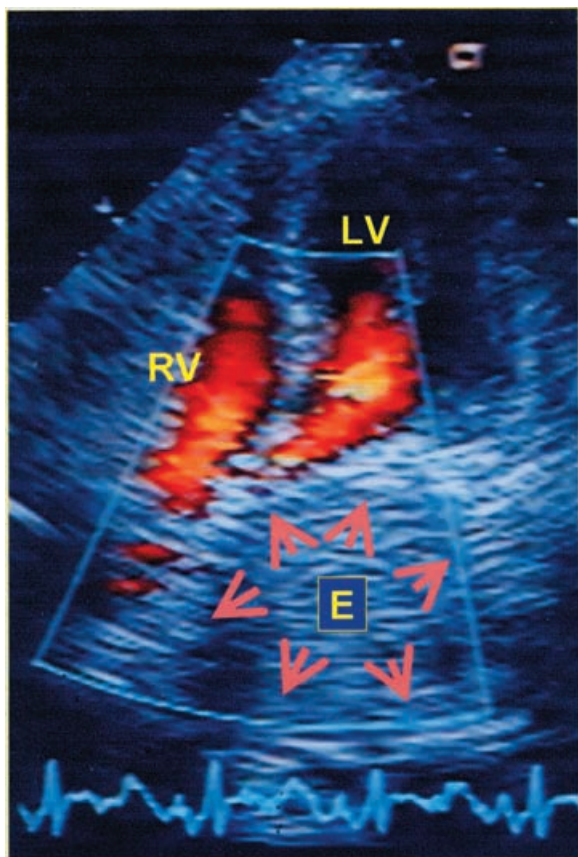
All four cardiac chambers were opacified but the left atrium appeared as an opacified crescentic sliver. However, the mass extrinsic to the left atrium was not opacified suggesting that this was not a vascular structure.

Left ventricular wall motion and ejection fraction were normal. Mild left ventricular hypertrophy was present. The mitral inflow pattern was consistent with the patient's age and revealed no evidence of mitral inflow obstruction (peak early diastolic wave velocity 77 cm/s; deceleration time of the early wave 180 ms, peak atrial wave velocity 98 cm/s; isovolumic relaxation time 67 ms). Pulmonary hypertension was present with the pulmonary artery systolic pressure estimated at 40 mmHg above the right atrial pressure using the peak velocity of the tricuspid regurgitation jet.

Hence, in the presence of normal left ventricular systolic function and no significant

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**Figure 1.** Due to extrinsic compression by a massively dilated esophagus (arrows radiating from E), left atrium is reduced to a minute crescentic sliver having a minimal color Doppler flow on this transthoracic echocardiogram. The portion of the esophagus compressing the left atrium had major and minor diameters of 58 and 52 mm, respectively. The remaining left atrial cavity covered a surface area of approximately 30 mm<sup>2</sup>. RV, right ventricle; LV, left ventricle.

left ventricular diastolic dysfunction, intrinsically abnormal myocardial function was not the likely cause of her congestive heart failure.

Echocardiographic imaging after oral ingestion of a carbonated drink to identify the upper gastrointestinal compressing structure could not be performed in our patient since she was unable to swallow liquids.

Subsequently, a CT scan of the chest confirmed atrial compression by a massively dilated esophagus measuring 56 × 50 mm in diameter and having an air-fluid level within it. There were also bilateral pleural effusions consistent with progressive heart failure (Fig. 2).

The patient was immediately treated with nasogastric decompression and had marked clinical improvement with resolution of hypotension (blood pressure 125/65). Follow-up transthoracic echocardiogram revealed a more patent left atrium (its surface area in the apical 4-chamber view increased to approximately 60 mm<sup>2</sup>).

Esophagogastroduodenoscopy was then performed with the injection of Botulinum toxin at the gastroesophageal junction with significant improvement in the patient's ability to swallow. Repeat echocardiogram 1 week after Botulinum toxin injection revealed a left atrial surface area in the apical 4-chamber view of approximately 80 mm<sup>2</sup> and resolution of the compressive effects from the dilated esophagus. The major and minor diameters of the tubular structure decreased to 38 mm and 28 mm, respectively (Fig. 3).

She was discharged to a rehabilitation facility in stable condition and was doing well 9 months posttherapy.

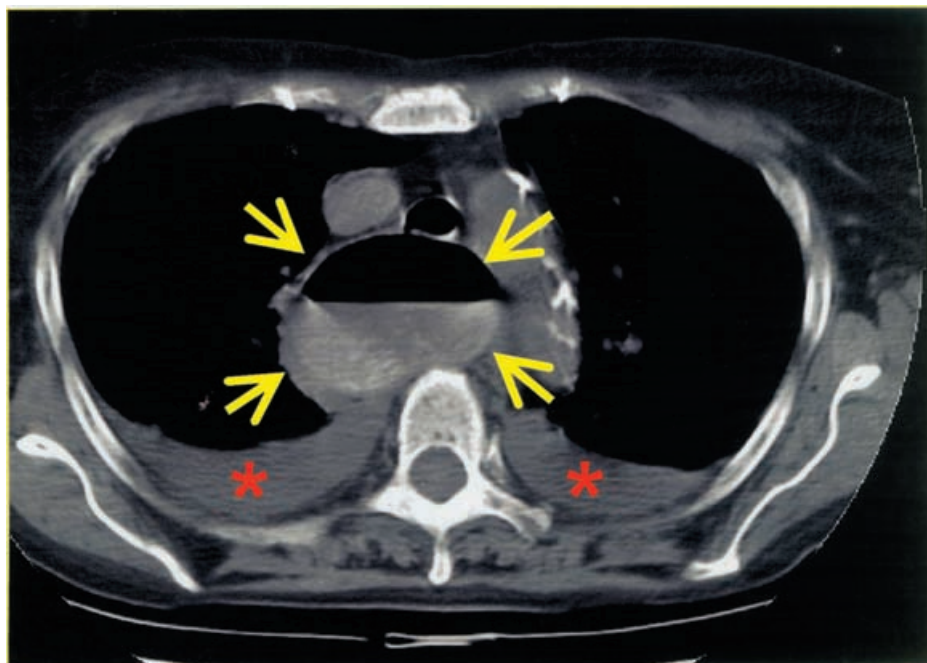
## Discussion

This is the first reported case of contrast echocardiography as a method of differentiating a compressive vascular structure (i.e., aorta) from an upper gastrointestinal structure (i.e., esophagus). In addition, it is a third reported case of left atrial compression by a dilated esophagus due to achalasia.

Our patient presented with an unusual cause of pulmonary edema because of extrinsic compression of the left atrium. A possible mechanism for the observed hemodynamic compromise could be impaired left atrial filling, as a result of the compression, with a concomitant rise in pulmonary venous pressure. A functional tamponade due to direct impairment of ventricular filling has also been described.<sup>1</sup>

Extrinsic compression of the left atrium is an unusual cause of congestive heart failure. Other mediastinal structures, which have been described to cause left atrial compression, include bronchogenic cysts,<sup>2</sup> lymphoma,<sup>3</sup> carcinoma,<sup>4</sup> thymoma,<sup>5</sup> diaphragmatic hernia,<sup>6</sup> and, commonly, aortic aneurysms.<sup>7</sup>

Left atrial compression by upper gastrointestinal structures (stomach, esophagus, or both) is uncommon. Achalasia is a motility disorder characterized by impaired relaxation of the lower esophageal sphincter and loss of peristalsis in the distal two-thirds of the esophagus.



**Figure 2.** Chest CT, which was obtained after oral contrast ingestion, reveals an air-fluid level in a massively dilated esophagus (arrows) and bilateral pleural effusions (asterisks). The esophagus measured 56 × 50 mm.

Achalasia is a motor disorder of the smooth muscle of the lower esophageal sphincter. The underlying abnormality is loss of intramural neurons. Primary achalasia (idiopathic) is the most common form found in the United States while secondary achalasia can be caused by gastric carcinoma, lymphoma, or Chagas' disease, etc.

Achalasia is suspected in a patient with dysphagia and typical radiographic features such as the absence of the gastric air bubble or the presence of an air-fluid level in the mediastinum representing retained fluid in the esophagus. Although manometry confirms the diagnosis of achalasia, upper endoscopy is required to exclude other diseases that may mimic achalasia and particularly to screen for adenocarcinoma of the stomach, which is the most common neoplasm associated with an achalasia-like presentation. Treatments with smooth-muscle relaxants, Botulinum toxin injection, dilation, and myotomy are directed at palliation of symptoms and prevention of complications.

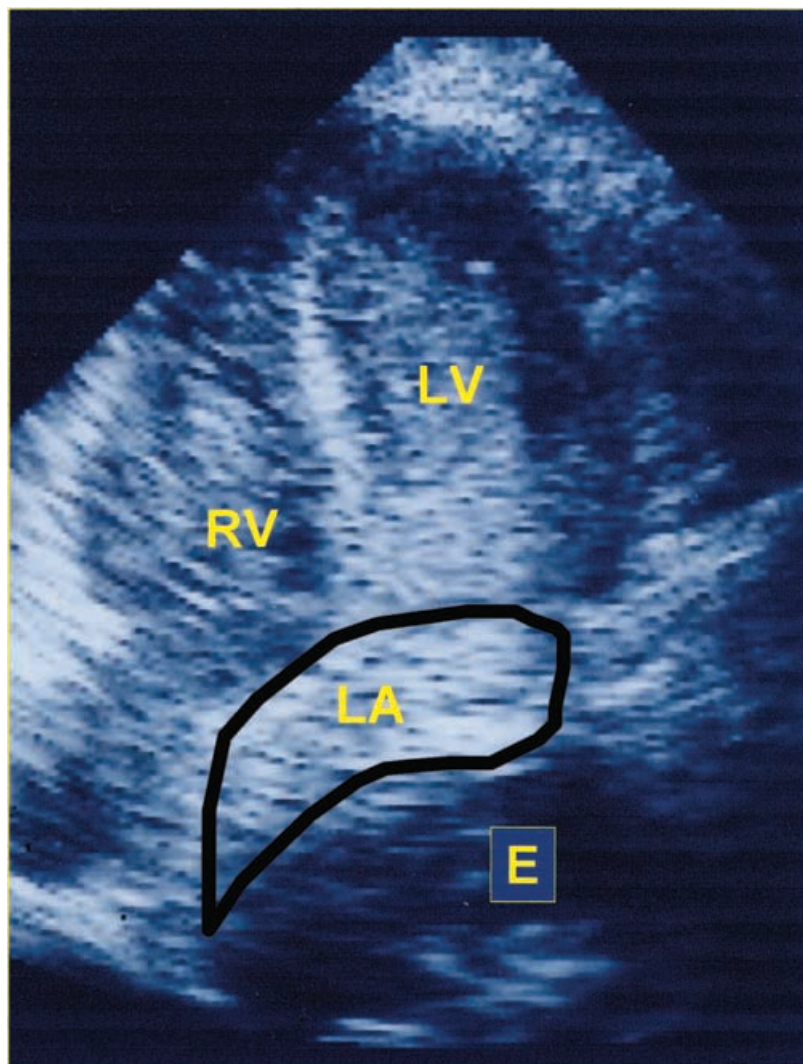
There are two previous cases reported of decompensated achalasia resulting in left atrial compression as seen on two-dimensional echocardiography.<sup>8,9</sup> In one case, the patient

had no symptoms of cardiac compromise, but the second case described a case similar to ours in that the patient had significant signs of congestive heart failure.

Previous use of echocardiography in this situation relied on identifying the echocardiographic appearance of air contrast in the esophagus during the ingestion of liquids containing carbon dioxide.<sup>10</sup> In cases of hiatal hernia or an empty esophagus, carbonated liquids would appear in the dilated structure.

We believe that in the setting of achalasia, the use of echo contrast may be a more reliable method than an oral ingestion of liquids because severe long-standing achalasia may be associated with both dysphagia to liquids and retained food in the esophagus, thereby, limiting the passage of ingested liquids.

In conclusion, this is a rare and unique cause of congestive heart failure due to extrinsic compression of the left atrium by a dilated esophagus. Left atrial compression is associated with severe hemodynamic compromise and a quick and accurate diagnosis is of critical importance. Contrast echocardiography in a patient with either known or suspected achalasia can readily make the diagnosis where rapid treatment will lead to hemodynamic stability.



**Figure 3.** Following nasogastric decompression and Botulinum toxin treatment, the esophagus (E) decreased in size and measured approximately  $28 \times 38$  mm. The left atrium (LA) partially reexpanded. Optison® contrast opacifies the left atrium but not the esophagus on this transthoracic echocardiogram. RV, right ventricle; LV, left ventricle.

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### References

1. Raza ST, Mukherjee SK, Danias PG, et al: Hemodynamically significant extrinsic left atrial compression by gastric structures in the mediastinum. *Ann Intern Med* 1995;123:114–116.
2. Volpi A, Cavalli A, Maggioni AP, et al: Left atrial compression by a mediastinal bronchogenic cyst presenting with paroxysmal atrial fibrillation. *Thorax* 1988;43:216–217.
3. Iwase M, Nagura E, Miyahara T, et al: Malignant lymphoma compressing the heart and causing acute left sided heart failure. *Am Heart J* 1990;9:68–70.
4. DeLuca A, Daniels S, Pathak N: Pulmonary edema due to extreme left atrial compression. *N J Med* 1991;88:37–38.
5. Canedo M, Otken L, Stefadouras MA: Echocardiographic features of cardiac compression by a thymoma simulating cardiac tamponade and obstruction of the superior vena cava. *Br Heart J* 1977;39:1038–1042.
6. Baerman JM, Hogan L, Swiryn S: Diaphragmatic hernia producing symptoms and signs of a left atrial mass. *Am Heart J* 1988;116:198–200.
7. Breall JA, Goldberger AL, Warren SE, et al: Posterior mediastinal masses: Rare causes of cardiac compression. *Am Heart J* 1992;124:523–526.
8. Mates M, Veselka J, Belohlavek J: Esophageal achalasia compressing the heart diagnosed by echocardiography. *Int J Cardiol* 1998;66:225–227.
9. Yilmaz MB, Arat N, Biyikoglu SF, et al: Extrinsic left atrial compression in a patient with achalasia. *Int J Cardiol* 2002;85:301–303.
10. D'Cruz IA, Feghali N, Gross CM: Echocardiographic manifestations of mediastinal masses compressing or encroaching on the heart. *Echocardiography* 1994;11:523–533.