

ECHO ROUNDS Section Editor: Edmund Kenneth Kerut, M.D. _____

Bronchogenic Cyst with Extrinsic Pulmonary Vein and Left Atrial Compression Presenting as Exertional Dyspnea

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(*ECHOCARDIOGRAPHY, Volume 24, February 2007*)

bronchogenic cyst, echocardiography, computed tomography, extrinsic pulmonary vein compression, TEE

A previously healthy 52-year-old male presented with a several week history of progressive exertional dyspnea. Physical examination and electrocardiogram were unremarkable. A transthoracic echocardiogram (TTE) was performed (Fig. 1). Initially it was thought that a membrane was within a large left atrium (LA). To further delineate the LA and its anatomy, peripheral injection of sonicated contrast (Optison, Amersham Health, Inc., Princeton, NJ) was performed (Fig. 2). A “contrast-free” cystic appearing structure became readily apparent. Computed tomography (CT) of the chest likewise demonstrated a cystic structure adjacent to the LA (Fig. 3). Transesophageal echocardiography (TEE) was then performed. In addition to the cystic mass (Fig. 4), the flow velocity profile in the left upper pulmonary vein (LUPV) was consistent with extrinsic pulmonary vein compression (Fig. 5). The patient subsequently underwent surgical resection of the mass, and had an uneventful recovery. The chief complaint of progressive exertional dyspnea resolved. Histopathology identified ciliated stratified epithelium, consistent with a bronchogenic cyst.

A bronchogenic cyst is a congenital lesion that is a remnant from abnormal budding of the embryonic foregut. These cysts are usually single,

but multiple cysts may exist, and range in size up to 5 cm in diameter. They may or may not be connected to bronchi or bronchioles. These cysts are well marginated and lined by ciliated, mucus-secreting respiratory epithelium. If not connected to a bronchiole, they are filled with mucinous material, but may become infected, leading to suppuration.¹

Most cases are either asymptomatic or present with respiratory symptoms.² Patient presentation may range from respiratory distress at the time of birth, to those that are

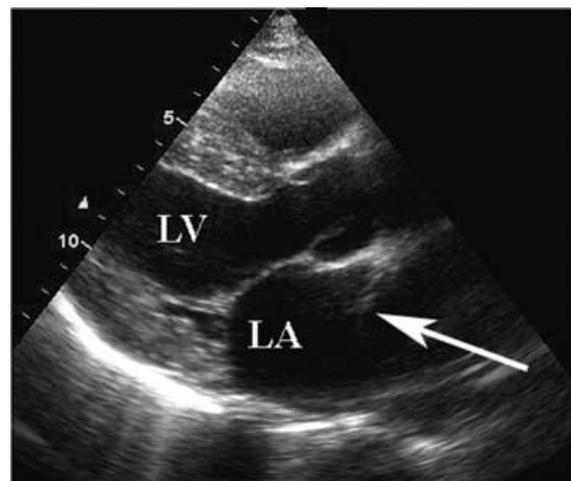


Figure 1. Parasternal long-axis image suggested a large sized left atrium (LA), but there was a suggestion of a “membrane” within its cavity (arrow). LV = left ventricle.

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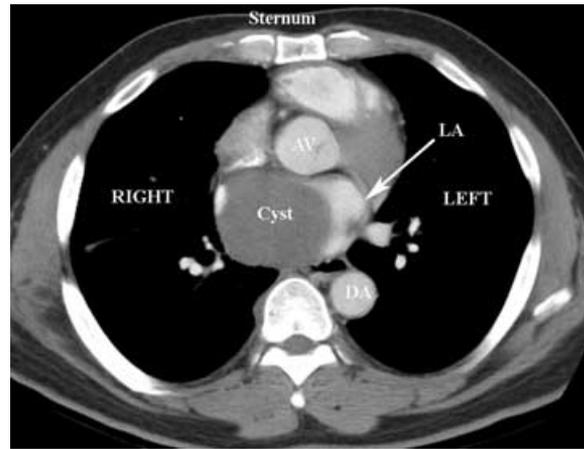
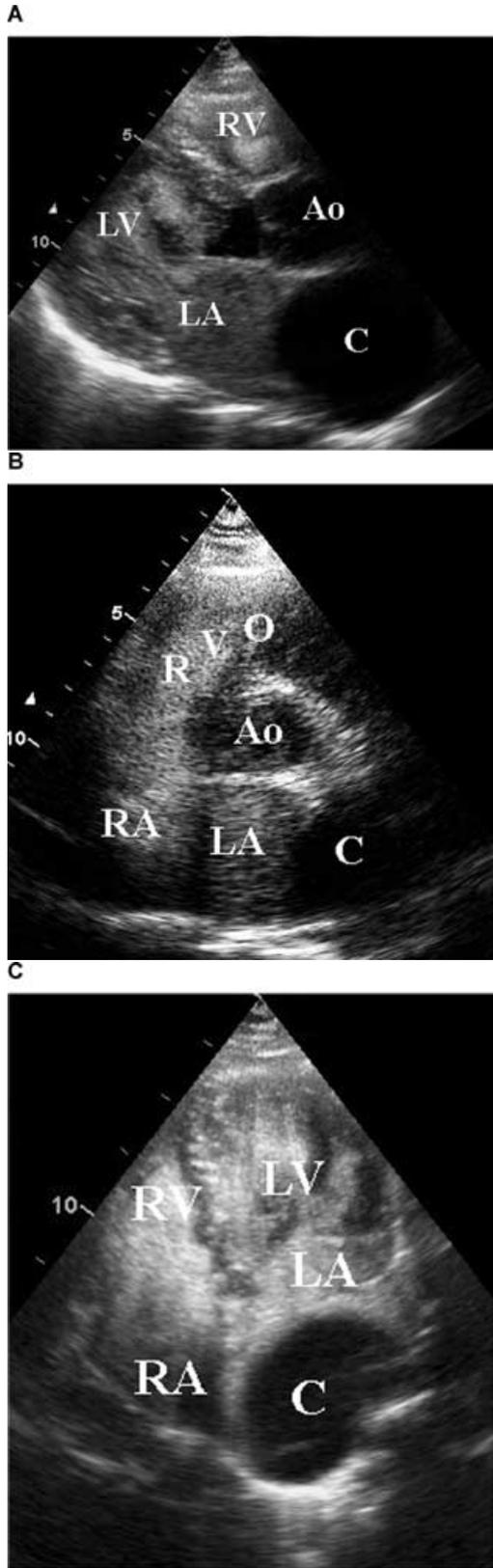


Figure 3. Cross-sectional computed tomography (CT) with intravenous contrast at the level of the aortic valve and proximal ascending aorta (AV). The nonopacified mediastinal cystic structure (Cyst) is noted to compress the left atrium (LA). DA = descending aorta; LEFT = left lung; RIGHT = right lung; Sternum = sternum.



Figure 4. Transesophageal echocardiography (TEE) within the mid-upper esophagus in a nearly vertical plane (100°) revealed a cystic mass (C) that appeared to contain a “gelatinous” material. The cyst was adjacent to the superior vena cava (SVC) and cranial to the left atrium.

Figure 2. Transthoracic echocardiography (TTE) with peripheral venous injection of Optison. (A) Parasternal long-axis demonstrated a “contrast-free” cystic (C) appearing structure adjacent to the left atrium (LA), and posterior to the proximal ascending aorta (Ao). LV = left ventricle; RV = right ventricle. (B) Parasternal short-axis again demonstrated a cystic structure (C) adjacent to the left atrium (LA). Ao = proximal ascending aorta, RA = right atrium; RVO = right ventricular outflow tract. (C) Apical imaging. C = cystic structure; LA = left atrium; LV = left ventricle; RA = right ventricle; RV = right ventricle.

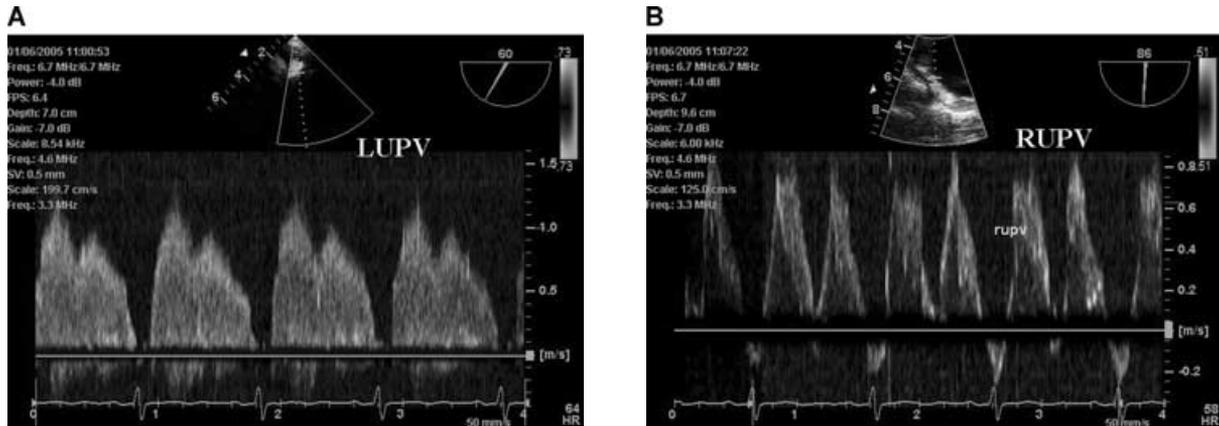


Figure 5. Transesophageal echocardiography (TEE) with pulsed-wave Doppler of the pulmonary veins. Sampling was performed within 1 cm of the orifice of the pulmonary vein evaluated. (A) Left upper pulmonary vein flow demonstrated a relatively high velocity (1 m/sec), with a prolonged deceleration slope of both systolic and diastolic waves. In addition, the atrial reversal was not present. These findings are consistent with extrinsic compression of that pulmonary vein. (B) Right upper pulmonary vein flow (RUPV) revealed flow velocities in the range normally found.

asymptomatic throughout life. Cough appears to be a common presenting symptom; but hemoptysis, pneumothorax, esophageal compression, infected cyst, and postobstructive pneumonia have also been noted.³ In addition, chest pain simulating aortic dissection,⁴ and also transformation to carcinoma has been reported.⁵ It is presumed that this patient's presentation was at least partly related to extrinsic compression of the LUPV, and also LA compression, leading to progressive exertional dyspnea.

By CT, a bronchogenic cyst is a single smooth mass with a uniform attenuation value.⁶ By echocardiography, bronchogenic cysts have been described as homogeneous masses with an echo-reflectance similar to blood. Doppler color flow distinguishes the cystic structure lacking blood flow from a vascular abnormality.⁷

This case had several echocardiographic findings that may serve as "clues" for diagnosis of bronchogenic cyst, namely:

1. Sonicated contrast (Optison) helped differentiate the cyst as a nonblood-filled structure extrinsic to the heart.
2. The cyst wall appeared thin.
3. The material within the cyst, especially as noted by TEE, appeared homogeneous and "gelatinous."

4. Although not specific for cysts, LA and pulmonary vein extrinsic compression probably were related to the patient's presenting symptoms of progressive exertional dyspnea.

The following supplementary material is available for this article online: Movie clips: figures 1, 2a, 2b, 2c, 4a, 4b, 5.

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