

Anomalous Chord of the Left Atrium without Involvement of the Mitral Valve

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An anomalous chord from the left side of the atrial septum to the left atrial free wall was incidentally noted on transthoracic echocardiography in a 14-year-old boy with vasovagal syncope. Previously reported cases of anomalous chords in the left atrium were associated with the mitral valve leaflets in all but two cases. This is the first reported case of an echocardiographic diagnosis of anomalous left atrial chord without insertion of the chord into the mitral valve. (Echocardiography 2012;29:E159-E162)

Key words: pediatric cardiology, anomalous chord, left atrium

Clinical History:

A 14-year-old boy without a significant medical history presented to an emergency department following a seizure associated with vasovagal syncope. All neurological studies for syncope and seizure workup were negative. A cardiac evaluation for syncope was performed as well.

A 15-lead ECG demonstrated sinus bradycardia at a rate of 47 beats per minute. Intervals and forces were normal. A Holter monitor revealed normal sinus rhythm with sinus bradycardia. The transthoracic echocardiogram (TTE) (Phillips iE33, Andover, MA, USA) demonstrated normal intracardiac anatomy with normal ventricular dimensions and function with no intracardiac shunts. A likely artifact with the appearance of parallel linear densities crossing the mid-left atrium (LA) was appreciated (Fig. 1 and movie clip 1). Doppler interrogation of the mitral valve revealed no obstruction to flow. The patient's history revealed no prior central line placements and no intracardiac interventions. The differential diagnosis list included a foreign body, possibly a catheter, within the LA; a foreign body in a nearby vessel, such as the thoracic descending aorta, with an artifact within the LA; a reverberation artifact within the LA due to normal cardiovascular structures; an anomalously coursing vessel, possibly a coronary artery, with an artifact within the LA; and a variant of cor triatriatum sinister or supravulvar mitral ring. A possible chord was considered, but the presence of parallel linear densities coursing for some distance was thought to be more likely either a nonbiological structure or an imaging artifact.

Additional workup included a chest x-ray, which was normal with no radiopaque foreign bodies visualized. A chest CT with angiography also showed a nonmetallic linear filling defect within the LA. The thoracic descending aorta appeared unobstructed with no evidence of foreign body or artifact within it (Fig. 2).

A transesophageal echocardiogram (TEE) and catheterization were done simultaneously with the potential for removal of a foreign body or mass. The TEE was performed using an adult omniplane multifrequency probe (Siemens, Sequoia C512, Mountain View, CA, USA). The TEE revealed a linear structure extending from the inferoposterior aspect of the atrial septum to the midposterior left atrial wall just medial to the left lower pulmonary vein entrance (Fig. 3 and movie clip 2). The linear structure measured approximately 3.5–4 cm in length and 0.2 × 0.2 mm in diameter with slight tapering toward the atrial septal end. There appeared to be "tenting" of the atrial septum into the LA at the connection point of the linear structure. Although there was an echolucency in the middle (similar to a catheter), the structure reverberated with cardiac motion. The mitral valve and its chordal attachments were normal. The pulmonary venous drainage was unobstructed. There was a patent foramen ovale (PFO) with left to right shunting. The findings were most consistent with an anomalous left atrial chord that was not associated with the mitral valve.

The catheterization procedure demonstrated no abnormal opacities within the heart by cine biplane imaging. Angiographic injections in the aortic root revealed normal coronary artery origins and proximal courses. A catheter was manipulated across a PFO to the LA and into the left lower pulmonary vein within contact with the structure. Angiography in the LA revealed no

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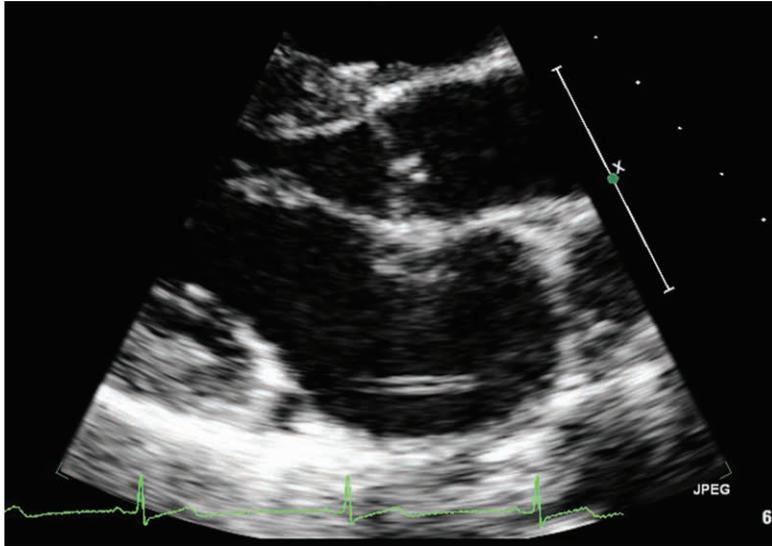


Figure 1. Parasternal long-axis view demonstrating parallel linear densities in the mid-left atrium.

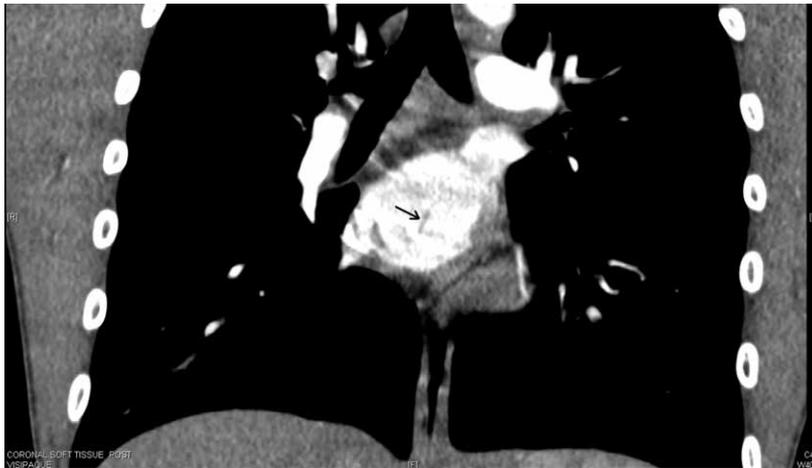


Figure 2. CT angiogram of the chest, postcontrast, demonstrating a non-metallic linear filling defect noted within the left atrium. This filling defect is not visualized on precontrast images, likely corresponding to a foreign body.

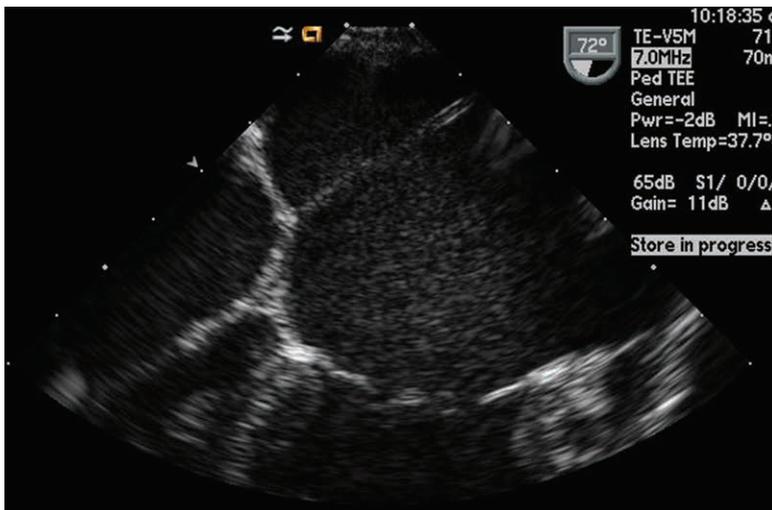


Figure 3. Midesophageal four-chamber view focused on the left atrium demonstrating a crossing chord with connection to the atrial septum. Mild “tenting” of septum primum is seen and subtle tapering of the chord at its insertion site into the atrial septum is noted.

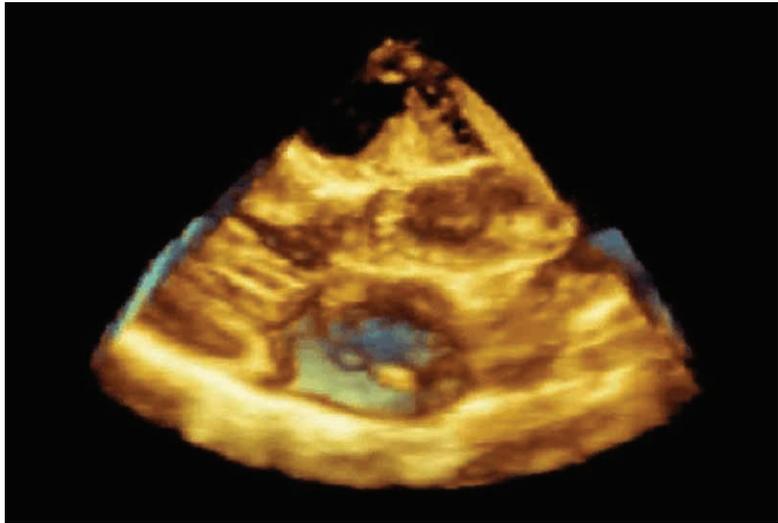


Figure 4. Transthoracic 3D echocardiographic view of the crossing left atrial chord without connection to the mitral valve.

filling defects or outlining of a foreign body with contrast.

On follow-up, a full-volume three-dimensional (3D) echocardiogram (Phillips iE33; X5-1 transducer) of the LA was performed to ascertain the exact origin and insertion of the anomalous chord, as well as to determine its course, size and shape in more detail (Fig. 4 and movie clip 3). The images showed that the chord indeed originated from the inferoposterior aspect of the atrial septum with insertion to the middle aspect of the left atrial free wall. Again, there was no connection to the mitral valve apparatus. On 3D imaging, the chord was linear in appearance and echodense with the same density as the atrial wall and septal tissue, consistent with the diagnosis of an anomalous chord of the left atrium. There was no obstruction to blood flow. No intervention was required, and the family was reassured that this was a rare but benign congenital anomaly.

Discussion:

Anomalous bands in the heart are rare. In all but two of the 21 reported cases of chords within the LA, the attachments were from the area of the foramen ovale to the anterior or posterior mitral valve papillary muscles or cusps.¹ All but one of the 21 cases was diagnosed on gross inspection of the heart during routine autopsy. Taglieri et al.² recently reported a TTE finding of anomalous LA chord connecting to the anterior mitral valve leaflet resulting in severe regurgitation.

To our knowledge, this is the first reported case of an echocardiographic diagnosis of anomalous chord crossing the left atrium without association with the mitral valve. McNamara et al.³ describe a similar anomaly during routine autopsy of the heart of an adult patient who died of noncardiac pathology. They noted a smooth round chord extending across the LA, from atrial septum to

the free wall of the LA. Histologically, the chord was composed of cardiac muscle, smoothly covered by endocardium. The location and size of the chord in our patient was similar to that described by this group.

The significance of these bands is uncertain. The embryologic origin is not well understood, though it has been theorized that these bands are part of the septum primum with deviation and extension to the left side. In our patient, its insertion near the origin of the left lower pulmonary vein begs the possibility that the developmental origin of these LA chords is similar to that of a supra-valvular mitral ring.

This is an interesting case of anomalous LA chord without involvement of the mitral valve diagnosed without surgery or autopsy. The finding of an anomalous left atrial chord is likely a congenital abnormality of no clinical significance. It is, however, important to recognize that these anomalous chords may rarely be present on routine echocardiographic evaluation, and misinterpretation of normal variants will impact management and clinical consequences.⁴ When seen, the hemodynamic significance of the chord should be fully assessed echocardiographically for attachments to mitral valve with associated valve dysfunction and for obstruction to flow within the LA.

References

1. Knoblich R, Ducey EF: Anomalous fibromuscular cord of the left atrium. Review of literature and report of a case. *Arch Pathol* 1962;73:86-88.
2. Taglieri C, Botta L, Roghi A, et al: Unusual insertion of a mitral chord causing severe valve regurgitation. *Eur J Cardiothorac Surg* 2010;38:387.
3. McNamara WL, Baker LA: Asymptomatic congenital anomaly of the heart - congenital muscular cord bridging walls of auricle above center of mitral valve. *Am Heart J* 1947;34:288-290.
4. George A, Parameswaran A, Nekkanti R, et al: Normal anatomic variants on transthoracic echocardiogram. *Echocardiography* 2009;26(9):1109-1117.

Supporting Information

Additional Supporting Information may be found in the online version of this article:

Movie clips for Figures 1–3.

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