Inversion of the Left Atrial Appendage
An Unusual Finding After Pediatric Heart Transplantation

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A n 8-year-old girl with hypoplastic left heart syndrome underwent orthotopic heart transplantation for end-stage right ventricular dysfunction, after previous surgical palliations with the modified Norwood and Glenn procedures. The donor heart was transplanted into the recipient with the use of a bicaval anastomosis. Postoperative transesophageal echocardiography was noteworthy for a prominent tubular-shaped structure within the left atrium (Figure 1A through 1C and Movies I and II in the online-only Data Supplement), in proximity to the mitral valve orifice. Doppler color-flow mapping suggested no flow within the structure. The patient’s marginal hemodynamic status after separation from cardiopulmonary bypass resulted in a decision to pursue delayed sternal closure, and the structure was not addressed at the time of transplantation.

Transcatheter echocardiography performed on the first postoperative day revealed the continued presence of this structure within the left atrium, in proximity to and directed at the mitral valve orifice (Figure 1D), with normal mitral valve function. Differential diagnostic considerations at this point included thrombus, atrial suture line, inversion of the left atrial appendage, and vegetation. Because the inversion of the left atrial appendage was felt to be the most likely cause of the these echocardiographic findings, the patient underwent direct visual inspection of the external surface of the left atrium while undergoing planned delayed sternal closure on the second postoperative day. At that time, the left atrial appendage was not visualized on the external surface of the heart and, therefore, confirmed to be inverted; manual reversion was performed without complications. Figure 2A and Movie III in the online-only Data Supplement demonstrate resolution of the inverted left atrial appendage via transesophageal echocardiography immediately after delayed manual reversion and delayed sternal closure; transthoracic imaging (Figure 2B) demonstrates continued resolution 2 days later.

Inversion of the left atrial appendage is an uncommon occurrence after heart surgery, with ≈20 cases reported in the medical literature to date. To our knowledge, this case represents the first reported occurrence of inversion of the left atrial appendage after heart transplantation. Inversion of the left atrial appendage has been reported in several clinical settings, including after a variety of operations in adults for ischemic and structural heart conditions, and surgery for congenital heart defects, as well. A postulated etiology of left atrial appendage inversion is negative intra-atrial pressure created by venting of the left atrium during the surgical procedure, or by intrapericardial pressure exceeding intra-atrial pressure in the setting of large pericardial effusion; both of these mechanisms are unlikely in the patient presented in this report. Although inversion of the left atrial appendage is regarded as a benign phenomenon, its natural history is uncertain, and spontaneous resolution has been documented.

Recognition of this uncommon clinical entity is probably the most important aspect of its clinical management. The relative rarity of left atrial appendage inversion may perplex the echocardiographer and may lead to missed or erroneous diagnoses, with potential impacts on patient morbidity if evaluations for alternative diagnoses are pursued. Inversion of the left atrial appendage can readily be diagnosed with echocardiography by the stereotypic tubular nature of the structure within the left atrium. The inverted appendage is usually directed at the mitral valve orifice and will have echocardiographic image intensity similar to the atrial wall; Doppler color-flow interrogation will not demonstrate evidence of flow within the structure, although the lack of flow does not necessarily discriminate inversion of the left atrial appendage from other diagnoses. Finally, an inability to image the left atrial appendage in its typical location should also prompt consideration of this diagnosis.

As discussed above, it is less likely that the above-mentioned mechanisms of left atrial appendage inversion could have applied in this case. An additional consideration specific to transplantation is the manipulation of the donor heart during organ retrieval, transport, and inspection before implantation. During postoperative imaging of the heart transplant recipient, echocardiographic acoustic reflections are frequently encountered within the left atrium as a manifestation of suture lines around the atrial cuff and are generally readily apparent to the echocardiographer. In most cases, these suture lines are quite pronounced and are located much more posteriorly, occupying a variable

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length within the atrial cavity. However, the unusual orientation and conical/tubular nature of the structure in the case described above clearly pointed to an alternative diagnosis. The transesophageal and transthoracic imaging contained within this report highlight the echocardiographic appearance of this finding and emphasize the need to be aware of this condition in patients after cardiac surgery, including orthotopic heart transplantation.

Disclosures

None.

References

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Movie Legend

**Movie 1.** Multiplane transesophageal echocardiogram clip from the midesophageal 0\(^0\) 4-chamber view demonstrating the inverted left atrial appendage oriented toward the mitral valve orifice. Best viewed with Windows Media Player.

**Movie 2.** Multiplane transesophageal echocardiogram clip from the midesophageal 109\(^0\) view with Doppler color flow mapping. The inverted left atrial appendage is oriented toward the mitral valve orifice, with no apparent Doppler color flow through the structure. Best viewed with Windows Media Player.

**Movie 3.** Multiplane transesophageal echocardiogram clip from the midesophageal -9\(^0\) view obtained immediately following manual reversion and delayed sternal closure demonstrates resolution of the inverted left atrial appendage. Best viewed with Windows Media Player.