Rare Case of Congenital Absence of Left Atrial Appendage and AFIB.

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The left atrial appendage (LAA) is a finger-like muscular part of the left atrium with a narrow neck where blood can collect and form clots leading to stroke and other embolic phenomena. Congenital absence of the left atrial appendage is extremely rare [1, 2] and only 6 cases have been reported in the literature, limiting our understanding of its clinical significance, especially in patients with atrial fibrillation requiring anticoagulation.

An 84 year old Caucasian male with new onset atrial fibrillation presented for TEE guided cardioversion. He did not have any history of Transient Ischemic Attack (TIA) or stroke. Initially managed with rate control strategy; he then developed dyspnea on exertion and was scheduled for elective cardioversion. His TTE showed normal ejection fraction with mild left ventricular hypertrophy and mildly dilated left atrium. A TEE was performed and no LAA could be identified with multiple views. (Fig A) His surgical history was reviewed, and he had no history of surgical or percutaneous LAA ligation. A CT of the heart confirmed the absence of the LAA (Fig B and C). The patient was cardioverted to normal sinus rhythm.

The Significance of congenital absence of the LAA remains uncertain and there are no guidelines or recommendations about long term anticoagulation in these rare cases. Based on the limited data available from LAA closure and effect of LAA morphology on stroke risk, it appears that with absence of the LAA, risk of thrombosis and AF associated stroke may be significantly less and CHADS2-VASC2 scoring system may not apply to this particular population. Risk of stroke in these patients may be equivalent to those undergoing LAA closure.

Based on recent data from LAA closure studies [3, 4], one can postulate that congenital absence portends a lower risk of intra atrial thrombosis and stroke in patient with AF and long term anticoagulation may be reconsidered. Of course this hypothesis needs to be confirmed with further studies and follow up of such patients. In our case, multimodality imaging was vital in confirming the diagnosis.

References: