ACTA FACULTATIS
MEDICAE NAISSENSIS

UDC: 616.12-007
DOI: 10.5937/afmnai40-37783

*Case report*

# Incidental Finding of a Rare Left Atrial Appendage Anomaly: A Case Report

Feridoun Sabzi, Aghigh Heydari, Mohammad Rouzbahani, Atefeh Asadmobini

*Cardiovascular Research Center, Kermanshah University of Medical Sciences, Kermanshah, Iran*

**SUMMARY**

**Introduction. Congenital hypoplasia of the left atrial appendage (LAA) with stenotic ostium in an abnormal position is an extremely rare entity. We report an exceptional case of hypoplastic LAA with ostial stenosis in an abnormal location in the patient with mitral valve stenosis that was mistaken in the transesophageal echocardiography (TEE) for an intraluminal thrombus.**
**Case report. A 43-year-old woman was admitted to our center with dyspnea on exertion. TEE revealed the presence of severe mitral stenosis and clot in the LAA. The patient underwent open cardiac surgery by cardiopulmonary bypass. Intraoperative inspection of the left atrium revealed hypoplasia of the LAA with stenotic and slit-like ostium. There was also a malposition of the LAA that was placed in a paravalvular location. The ostium of the LAA was not round and wide but narrowed and slit-like.**
**Conclusion. The authors believe that this anomaly may have some physiologic consequences in patients with mild structural failure. Indeed, this anomaly may aggravate the severity of structural failure by pressure burden on the atrial wall and valve.**

*Keywords*: **congenital heart disease, left atrium, atrial appendage, cardiovascular abnormalities**

Corresponding author:
**Atefeh Asadmobini**
e-mail: a.asadmobini@gmail.com

## INTRODUCTION

Left atrial appendage (LAA) is a pedunculated, muscular protrusion of the left atrium originating from the adjacent ostium of the left upper pulmonary vein. Its origin is inferolateral from the left atrial wall and lies near the atrioventricular groove, where the proximal segment of the main circumflex artery gives the lateral branches. It usually has a wide ostium that narrows to its distal end, which is converted to a long tubular or hooked-like structure. In opposition to the ostial portion that has a wide mouth, its trunk has a narrow junction with the pectinate muscle. A numbers of studies that have been performed for a description of the left atrial anatomy are scanty, but numerous articles about the anatomy of the right atrial appendage have been observed in the medical literature (1). Because of ignorance of the LAA in the anatomic study and a few reported anatomic varieties of appendage, our case report describes one of the exceptionally rare anomalies. With the improvement of imaging modalities, transesophageal echocardiography (TEE) has revolutionized the visualization of small structures of the heart such as the LAA, which was difficult to describe by the old imaging technique. The importance of LAA anatomy is related to its role in possible thromboemboly during atrial fibrillation (AF) cardioversion. A few cases of atresia have been reported in patients that were prepared for AF cardioversion. Therefore, the position, size, and numbers of the pectinate muscle have a vital role in possible future thrombus formation in the stagnation of blood in the left atrium (2). Herein we report an exceptional case of hypoplastic LAA with ostial stenosis in an abnormal location in the patient with mitral valve stenosis that was mistaken in the TEE for an intraluminal thrombus.

## CASE REPORT

A 43-year-old woman was referred to our center complaining of a two-month history of breathlessness on exertion. On physical examination, vital signs such as blood pressure (110/70 mmHg), heart rate (90 beats per minute), and respiratory rate (23 breaths per minute) were unremarkable. Her rhythm was irregular. Auscultation of the thorax showed fine crepitation in the lower zones of the lung but no wheeze or rhonchi was heard. Cardiac auscultation showed a diastolic rumble with an opening snap. ECG exhibits atrial fibrillation with a rapid ventricular response. A TTE showed a normal left ventricular function with preserved ejection fraction, severe mitral stenosis, and a moderately dilated left atrium. TTE also revealed the presence of a dense echogenic mass within the left atrial appendage compatible with a thrombus. The ostium of the left atrial appendage or blood flow between the appendage and the left atrial cavity was not studied by TEE. The patient was scheduled for surgery with midline sternotomy. After aortic and bi-cava cannulation, the

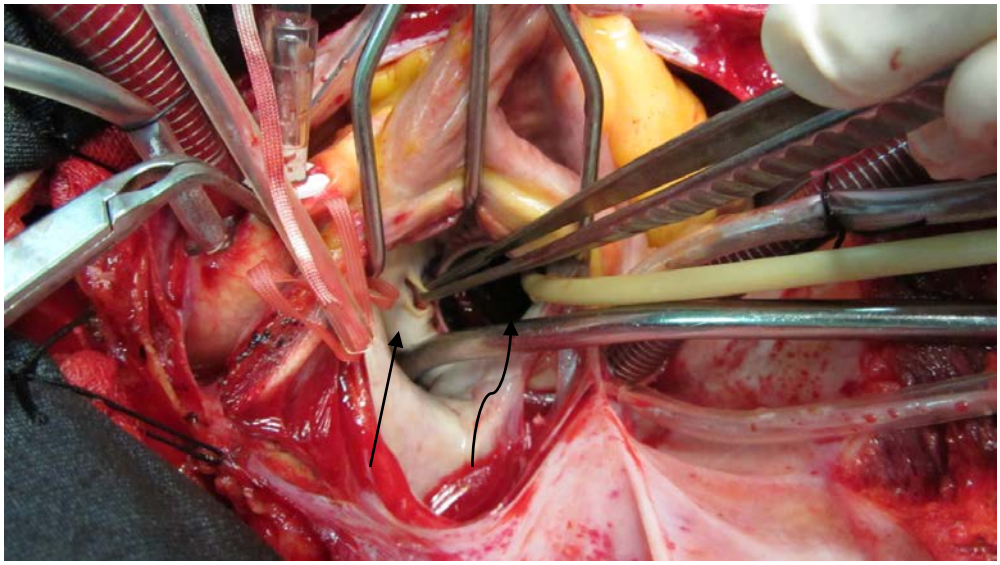

**Figure 1**. *The stenotic ostium of hypoplastic left atrium (vertical arrow) as well as the prosthetic mitral valve (the curved arrow)*

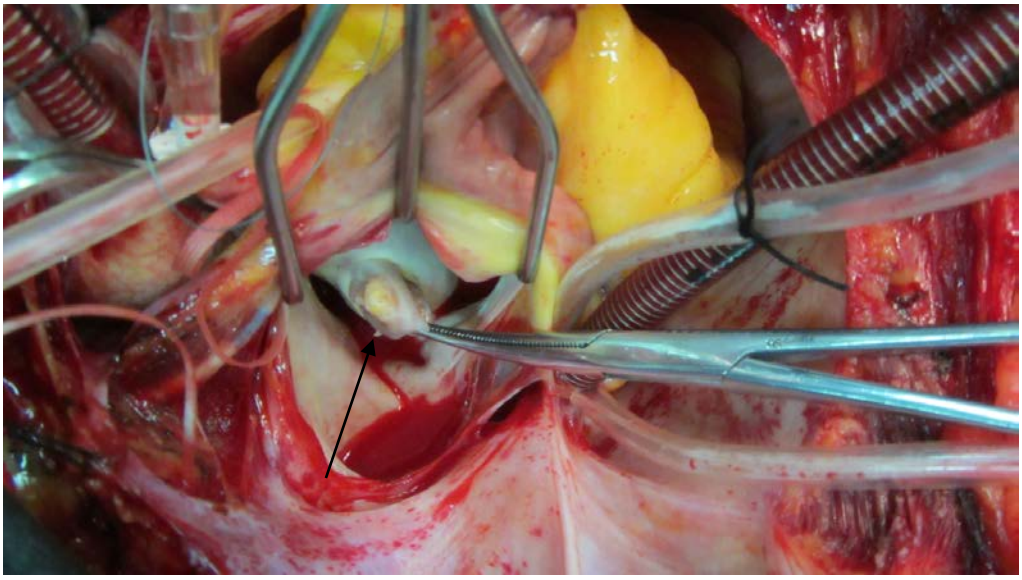

**Figure 2.** *An inverted hypoplastic left atrial appendage (the black arrow)*

ascending aorta was cross-clamped and cardioplegia was delivered into the aortic root. With the cardioplegic arrest, an interatrial groove was incised, and access to the left atrium cavity was provided by upward traction. On exploration of the left atrial cavity, LAA was hypoplastic. Its ostium was stenotic and slit-like covered by endocardium and located in an abnormal location near the mitral valve ring (Figure 1, 2). It seems that the stenotic and slit-like ostium of the thick wall appendage prevents the diastolic filling, so adherence of appendage walls to each other formed a muscle mass that is interpreted in TTE as an intrathrombus. Inversion of the appendage revealed its hypoplastic nature and absence of usual thick pectinate muscles as observed in normal LAA. The patient underwent mitral replacement with a prosthetic mitral valve (29 number, carbomedic, sorine group). The LAA was left intact and the patient was weaned from a cardiopulmonary bypass. The postoperative period was uneventful and discharged on the 7th day of the operation with aspirin and warfarin medications.

## DISCUSSION

LAA is a blind secular and hooked-like structure originating from the upper corner of the left atrium (LA) and presents in early fetal life as the primitive bud that leads to a future appendage. Although it is known as a simple tubular structure with an undefined physiologic effect, it has been associated with some serious complication that relates to its complex anatomic and histologic characteristics of this structure (3). During the third week of fetal growth, a pair of endocardial cushion tissue appears on each side of the fetus in the midline due to the rapid development of endocardial cushion cells derived from primitive cardiac structure. In the third week of fetal life, an embryonic primitive bud comes out from the left upper portion of the left atrium that later gives rise to LAA. This muscular structure seems to have an important physiologic effect. Some authors believe that this finger-like structure with a wide ostium functions as a decompression reservoir that reduces the left atrial pressure during forceful systolic contraction and in other phases of increased left atrial pressure (4). This physiologic effect is provided by the anatomic characteristics of LAA ostium. The ostium regulates the volume and speed of blood flow to LAA during the systolic function of LA. This regulation of blood flow circulation in LA may prevent the backflow knock of blood column to pulmonary veins and pulmonary circulation. On the other hand in some cases, atresia of LAA may be associated with early structural failure of the mitral valve in patients that have a mild structural malformation in their mitral valve apparatus. It seems that the shock absorber effect of LAA may prevent the hemodynamic knock effect of the rush of blood flow on the mitral valve with mild structural failure. We report this case to intensify the association of this rare congenital malformation of unknown physiolo-

gical sequels with the concomitant mitral valve disease that warrans to be identified in future studies. In a case report, a patient with a narrowed LAA ostium was detected that had hemodynamically important stenosis across the LAA ostium to the main LA (5). Although one case of iatrogenic stenosis of the left atrial appendage ostium has been reported following an incomplete ligation by surgery during mitral valve replacement, congenital stenosis of LAA ostium diagnosed by echocardiography for detection of another pathology is a very rare event. This phenomenon was reported by Ha et al. in a case of congenital LAA stenosis (6). In a case study by Coughlan, a subject with a transient ischemic attack was described, having idiopathic atrial appendage stenosis (7). The author postulated that the anomaly highlighted a congenital malformation that constituted a rare type of cor triatriatum, in which fibromuscular membranous septum divided the primitive left atrium higher than the atrial appendage location into two accessory chambers. In two separate studies by Zhang and Collier, LAA atresia was reported in two patients without any associated cardiac pathology. The AF was absent in all of these four cases. However, our patient presented with AF that may be caused by concomitant mitral valve pathology (8, 9). Other specific features of our case that distinguish it from previously reported cases of LAA refer to the concomitant presence of malposition and hypoplasia of LA with stenotic LA ostium. This complex malformation was not detected preoperatively by TTE and the absence of blood flow in the hypoplastic and thick lumen was interpreted as a left atrial clot which is a common finding in MS (10). However, a hypothesis for possible clinical sequelae of this type of anomaly is not clearly defined by the presence of a small number of cases that have been reported in the medical literature so far. However, clot formation in two types of malformation isolated stenosis of the ostium and LAA hypoplasia with ostial stenosis is theoretically possible because of the relative stasis that existed behind the stenosis (11). It is also a possible risk that the high-velocity blood jet may knock the wall of the left atrium causing fibrosis and thrombus formation.

## CONCLUSION

Congenital hypoplasia of LAA with stenotic ostium and presence in an abnormal location is an extremely rare cardiac anomaly. No cases have been reported so far. The authors believe that this anomaly may have some physiologic consequences in patients with mild structural failure. Indeed, this anomaly may aggravate the severity of structural failure by pressure burden on the atrial wall and valve.

### Acknowledgement

The authors appreciate the experts of Research Development Center of Imam Ali and Ayatollah Taleghani Hospital in Kermanshah University of Medical Science.

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Article info
Received: May 10, 2022
Accepted: March 17, 2023
Online first: May, 5, 2023

# Slučajni nalaz retke anomalije aurikule leve pretkomore: prikaz slučaja

Feridoun Sabzi, Aghigh Heydari, Mohammad Rouzbahani, Atefeh Asadmobini

*Kardiovaskularni istraživački centar, Univerzitet medicinskih nauka u Kermanšahu, Kermanšah, Iran*

## SAŽETAK

**Uvod. Kongenitalna hipoplazija aurikule leve pretkomore sa stenotičnim uščem u abnormalnoj poziciji je ekstremno retka pojava. Predstavljamo redak slučaj hipoplastične aurikule leve pretkomore sa stenozom ušća u abnormalnoj poziciji kod bolesnice sa stenozom mitralne valvule, koja je tokom transezofagijalne ehokardiografije (TEE) greškom protumačena kao intraluminalni tromb.**
**Prikaz slučaja. Četrdesettrogodišnja žena primljena je u naš centar sa dispnejom pri naporu. Transezofagijalna ehokardiografija (TEE) je pokazala prisustvo ozbiljne mitralne stenoze, kao i ugrušak u aurikuli leve pretkomore. Bolesnica je podvrgnuta otvorenoj operaciji srca ugradnjom kardiopulmonalnog bajpasa. Intraoperativna inspekcija leve pretkomore je ukazala na hipoplaziju aurikule leve pretkomore sa stenotičnim uščem nalik na prorez. Takođe, primećen je nepravilan položaj aurikule leve pretkomore koja se nalazila paravalvularno. Ušće aurikule leve pretkomore nije bilo okruglo i široko, već suženo, poput proreza.**
**Zaključak. Autori smatraju da ova anomalija može imati neke fiziološke posledice kod bolesnika sa blagom strukturalnom insuficijencijom. Ova anomalija može pogoršati ozbiljnost strukturalne insuficijencije pritiskom na zid pretkomore i valvulu.**

*Ključne reči*: **kongenitalna srčana bolest, leva pretkomora, aurikula pretkomore, kardiovaskularne abnormalnosti**