

Left atrial anomalous fibromuscular bands: a systematic review of case reports

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Medical Studies 2026; 42 (1): 7–11

DOI: <https://doi.org/10.5114/ms.2026.160485>

Key words: left atrial, anomalous fibromuscular band, string-like structure, mitral valve prolapse.

Abstract

Introduction: Left atrial fibromuscular band (LAMB) is an infrequently described anomaly of the heart.

Methods: Our knowledge about it is still limited and comes solely from case reports.

Results: Prevalence of LAMB is nowadays estimated as 0.1% in the general population.

Conclusions: The majority of all cases appears to be completely asymptomatic and the diagnosis is made accidentally during imaging of the heart (echocardiography or computed tomography). Conversely, there is a group of patients in whom the bands are attached to mitral valve leaflet triggering mitral valve prolapse and severe regurgitation that often demand surgical repair. Finally, the LAMB appears to be a possible cause of a cryptogenic stroke. However, this issue remains unclear because there is not enough evidence until now.

Introduction

Anomalous muscular bands localized in the right atrium and left ventricle have been precisely described in many previous reports. Conversely, those found in the left atrium (LA) seem to be substantially less frequent and their exact clinical significance remains unclear. Along with the ongoing development of imaging techniques as well as an increasing number of the visual examinations of the heart, a growing number of case reports on left atrial (fibro)muscular band (LAMB) has been published recently.

The aim of this report is to summarize a contemporary knowledge about the anomalous left atrial muscular band: its prevalence, location, impact on a heart dysfunction and possible methods of treatment.

Previously, many authors used various terms for describing these structures e.g. left atrial anomalous muscular (LAMB), string-like structure, fibromuscular cord, accessory or aberrant chordae (tendineae). In this text we will use the abbreviation LAMB for all cases irrespective of the initial term used by the case report authors.

Methods

The inclusion criterium for the review was the presence of an additional band in a left atrium confirmed at least in one imaging technique. The following were excluded from further analysis – cases with concomitant abnormal structures in the left ventricle or other morphological defects of the heart like cor triatriatum etc. Finally, only the adult patients were qualified

for this review due the fact that pediatric cases differ much with respect to their symptomatology and baseline characteristics [1].

The data were obtained from the PubMed database. Initially 136 reports published in the last two decades (2003 to 2023) were extracted. The vast majority of them fulfill the above exclusion criteria and finally only 32 case reports (that cover 40 patients with abnormal band structures in the left atrium irrespective of a clinical context were included in the review).

The vast majority of the reports consist of a single case report and only the analysis from Italy covers 6 patients [2] (Table 1). Almost all reported cases came from a limited list of countries like Japan [1, 3–8], Italy [2, 9–12], Turkey [13–16] and the United States of America [17–25].

Results

The prevalence of LAMB is currently estimated as about 0.1% in the general population. These data come from a large cohort of 1040 patients qualified for catheter ablation of atrial fibrillation. A cardiac computed tomography (CT) revealed only 1 patient with a string-like structure in the left atrium [1]. The latest retrospective analysis of Italian patients suggests that the prevalence of LAMB is even lower. They described only 6 cases of left atrial muscular bands out of 30000 echocardiographic examinations, which accounts for prevalence of about 0.02% in the general population. However, the authors claimed that many cases were not been detected by transthoracic echocardiography [2].

Table 1. List of case studies on left atrial muscular bands (LAMB)

Ref.	Year of publ.	No. of patients	Country	Diagnosis based on	Age	Location of the band in the LA	Pathology	Surgical treatment
[1]	2016	1	Japan	CT	ND	IAS to LA (RSPV area)	AF	None
[2]	2023	6	Italy	TTE/TEE	71, 84, 18, 36, 77, 79	Various: From IAS (2x) or from ant-sup wall of LA (4x) to AMVL (3x)	Endocarditis susp (4x), Mild MR (1x), TEE pre EPS (1x)	None
[3]	2019	1	Japan	TEE/CT	54	IAS to LA (post. wall)	AF	None
[4]	2018	1	Japan	ECHO/CT	79	IAS to LA (RSPV area)	Mitral valve stenosis	MVR but no LAMB
[5]	2015	1	Japan	CT	65	IAS to LA (roof)	AF	None
[6]	2020	1	Japan	TEE	62	IAS to AMVL	AF, Severe MR	Surgery
[7]	2021	1	Japan	TEE/CT	62	IAS to LA (roof, RIPV area)	AF	None
[8]	2015	1	Japan	TEE/CT	72	IAS to LA (post. wall)	AF	None
[9]	2020	1	Italy	TEE/CT	49	IAS to LA (LIPV area)	AF	None
[10]	2010	1	Italy	Sur	58	IAS to AMVL	MVP/severe MR	Surgery
[11]	2018	1	Italy	TEE	85	LA (ant. wall) to AMVL	Severe MR	Surgery
[12]	2016	1	Italy	CT	30, 67	IAS to LA (post. wall)	Palpitation/stroke	None
[13]	2003	1	Turkey	TTE/TEE/MRI	43	IAS to AMVL	MVP/severe MR	Surgery – only LAMB
[14]	2009	2	Turkey	TTE/TEE	34, 56	LA	Stroke	None
[15]	2012	1	Turkey	TTE/TEE/CT	32	IAS to AMVL	Palpation	None
[16]	2003	1	Turkey	TTE/TEE	43	IAS to AMVL	Severe MR	Surgery – only LAMB
[17]	2019	1	USA	TTE/TEE	55	IAS to AMVL	MVP/severe MR	Surgery
[18]	2018	2	USA	TTE/TEE	22, 56	IAS to LAA/AMVL	Sepsis/stroke	None
[19]	2015	1	USA	Sur	82	LA (ant-lat wall) to AMVL	AF/moderate MR	Surgery
[20]	2008	1	USA	TEE	72	LA (dome) to AMVL	MVP/severe MR	Surgery
[21]	2002	1	USA	TEE	20	IAS to AMLV	MVP/severe MR	Surgery
[22]	2016	1	USA	TTE/TEE	39	LA (dome) to AMVL	Mild MR	None
[23]	2012	1	USA	TTE/TEE	27	LA (dome) to AMVL	Severe MR	Surgery
[24]	2016	1	USA	TEE	ND	IAS to AMVL	Severe MR	Surgery
[25]	2010	1	USA	TEE	72	Dome of LA to AMVL	Severe MR	Surgery
[26]	2019	1	Czech Republic	TEE/MRI	67	IAS to LA (roof, RSPV area)	AF/susp. foreign body	Surgery – only LAMB
[27]	2017	1	South Korea	TTE/TEE	76	IAS	AF	None
[28]	2014	2	RSA	TTE	29, 43	LA	TIA	None
[29]	2011	1	UK	TEE	75	LA (roof) to AMVL	Moderate MR	None
[30]	2008	1	UK	TTE/TEE	33	IAS to LV (lat. wall)	Severe MR	Surgery
[31]	2014	1	South Korea	TEE	67	IAS to AMVL	Severe MR	Surgery
[32]	2011	1	UK	TTE	49	IAS to AMVL	Severe MR	Surgery

TTE – transthoracic echocardiography, TEE – transesophageal echocardiography, CT – computed tomography, MRI – magnetic resonance imaging, Sur – surgical (intraoperative), LA – left atrial/atrium, LV – left ventricle, IAS – interatrial septum, AMVL – anterior mitral valve leaflet, LIPV – left inferior pulmonary vein, RSPV – right superior pulmonary vein, RIPV – right inferior pulmonary vein, AF – atrial fibrillation, MVP – mitral valve prolapse, MR – mitral valve regurgitation, LAMB – left atrial muscular band, TIA – transient ischemic attack, MVR – mitral valve replacement, EPS – electrophysiologic study, ND – no data.

In our analysis the patient age ranged from 18 to 85, with 16 patients aged below fifty and 8 individuals over seventy 5 years old (Table 1). In the literature, there are numerous case reports of LAMB diagnosed in pediatric patients as well.

In the vast majority of cases the LAMB originated from a left atrial septum (IAS), in the area of a fossa ovalis. Only in few cases it originated from different parts of the LA like a dome [20, 22, 23] or anterior superior wall of the left atrium [2, 11]. The distal part of the LAMB is often attached to an anterior leaflet of a mitral valve (AMVL) [2, 10, 13, 15–18, 20–23, 25] or a roof of the LA (often nearby a pulmonary vein orifice) [1, 4, 7–9, 12, 26] (Table 1).

The LAMB diagnoses were made based on different imaging techniques. The predominant method of detection was echocardiography, both transthoracic (TTE) and transesophageal examination (TEE). The latter was the most common method of the final confirmation of the LAMB as well as its precise location in the heart (Figures 1 and 2 – cases from our archive from the Świętokrzyskie Cardiology Centre in Kielce, Poland). Only in few cases, CT imaging was the first diagnostic method for LAMB detection [1, 6, 13]. More often multimodality imaging, i.e. echocardiography along with computed tomography were used in the diagnostic pathway [3, 4, 7–9, 15]. Substantially less frequently magnetic resonance imaging (MRI) was utilised [13, 26]. Only in 2 cases anomalous fibromuscular bands were initially diagnosed intra-surgically during the mitral valve repair procedure [10, 19] (Table 1).

Discussion

Based on data from the previous case reports there are many different possible clinical consequences of LAMB: from a completely uneventful presence of string-like structures in the left atrium, through the increased risk of embolic events up to a severe dysfunction of a mitral valve causing heart failure symptoms.

What seems to be a frequent clinical scenario lately is that LAMBs are often accidentally encountered in imaging techniques before the invasive procedures in the left atrial/atrium, especially an ablation of atrial fibrillation. What is important, the LAMB did not prevent a successful procedure (pulmonary vein isolation – PVI) in the left atrium despite the fact that they are commonly attached to the part of the left atrium nearby a pulmonary vein orifice [1, 2, 7, 8]. Similarly, also in patients qualified for an electric cardioversion due to persistent atrial fibrillation asymptomatic LAMBs were observed [3, 27]. Their presence did not alter primary intention to treat. A really interesting case of suspected foreign body presence in the left atrium after catheter ablation was reported by Skala *et al.* Due to the unspecified character of a string-like structure in the left atrium potential risk of embolism and a cardiac surgery was performed and chord-like material was excised from LA [26].

Anomalous fibromuscular band may imitate an endocarditis especially when fever or sepsis of unidentified origin is present. These cases appear to be exceptionally confusing for differential diagnosis and demand the highest proficiency from the physician analyzing echocardiography [2, 18]. Moreover, in some cases more than one additional structure may be observed – Pizzuti *et al.* present a patient with two filamentous structures in the left atrial/atrium [2].

Additionally, the LAMB is associated with a possible risk of embolism episodes. There are case reports of the unexplained stroke with the presence of the string-like structure in the left atrium [12, 14, 18, 28]. A cryptogenic stroke is an extremely challenging diagnosis of exclusion (there are many other possible underlying causes of central nervous system ischemic episodes). To our knowledge, the exact significance of the LAMB as a potential cause of embolism is still uncertain. Moreover, LAMB is often associated with a patent foramen ovale (PFO) or a mitral valve prolapse (MVP) what makes this issue even more unclear [28].

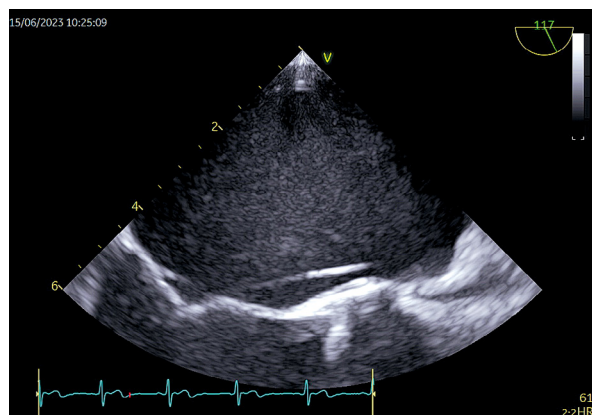


Figure 1. LAMB in transesophageal echocardiographic examination (TEE)

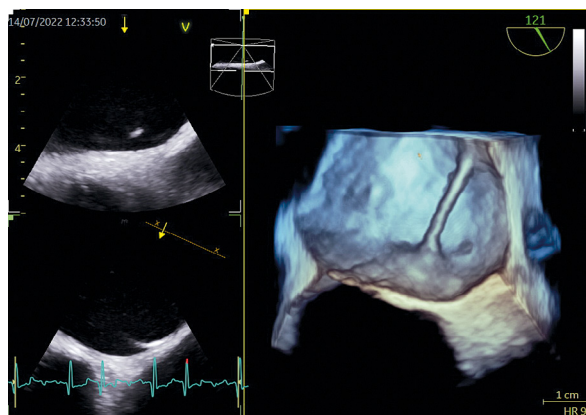


Figure 2. LAMB in three-dimensional (3D) presentation in transesophageal echocardiographic examination (TEE)

In many cases a severe mitral valve dysfunction is observed. In these individuals a specific mitral valve prolapse is present causing intensive heart failure symptoms. Very often these patients are finally qualified for surgical repair of a mitral valve. In our analysis only 4 patients with a mitral valve regurgitation (classified as mild to moderate) were treated conservatively [2, 15, 22, 29]. Interestingly, in some other cases an isolated removal of the LAMB restored a normal function of the mitral valve so that any other intervention was not necessary [13, 16, 26]. A large group of patients with the LAMB and a concomitant severe mitral valve regurgitation underwent a valve surgery. In all reported cases a successful procedure of valvuloplasty was performed with preservation of a native mitral valve [6, 10, 17, 19–21, 23–25, 30–32] (Table 1).

Conclusions

Additional left atrial fibromuscular band is a rare inborn anomaly of the heart. Its clinical consequences mainly depend on the presence of mitral valve prolapse – in these cases a surgical treatment for restoration of a normal mitral valve function is necessary. Conversely, very often the LAMB seems to be a benign asymptomatic entity with no further risk for the patient. What is important, it does not prevent invasive procedures on the heart. Along with an increasing number of heart imaging techniques we expect a rise in incoming cases of LAMB.

Funding

No external funding.

Ethical approval

Not applicable.

Conflict of interest

The authors declare no conflict of interest.

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Received: 7.10.2024

Accepted: 28.04.2025

Online publication: 25.03.2026