ORIGINAL ARTICLE



Lambl's excrescences in children: Improved detection via transthoracic echocardiography

Amanda L. Phillips¹ | Muhammad Yasir Qureshi MBBS^{1,2} Benjamin W. Eidem MD^{1,2,3} | Frank Cetta MD^{1,2,3}

¹Division of Pediatric Cardiology and Department of Regenerative Medicine, Wanek Family Program for Hypoplastic Left Heart Syndrome, Rochester, Minnesota, USA

²Division of Pediatric Cardiology, Mayo Clinic, Rochester, Minnesota, USA

³Department of Cardiovascular Diseases, Mayo Clinic, Rochester, Minnesota, USA

Correspondence

Frank Cetta, MD, Gonda 6335, Mayo Clinic, 200 First Street – SW, Rochester, MN 55905. Email: cetta.frank@mayo.edu

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Abstract

Background: Lambl's excrescences (LE) are fibrous extensions that can be found along the lines of closure of the aortic valve. Due to improvements in ultrasound technology, LE are frequently imaged during transthoracic echocardiography (TTE) in adults.

Objective: The purpose of this study was to determine the prevalence of LE among children from two eras (2004–2006 and 2011–2012) and the effect of technological advancements on LE detection.

Methods: TTE from 700 subjects (age 18 years old or younger) were reviewed. All parasternal long and short axis images of the aortic valve were reviewed by a board certified echocardiographer, and the positive studies were then reviewed by two additional observers to confirm the presence of LE. A two-sample *t* test with 95% significance was used to analyze the presence of LE in the cohorts. Median follow-up duration was 66 months.

Results: Of the 700 subjects, 12 (1.7%) children were found to have LE. No significant difference in prevalence was found between the two eras (.9% vs. 2.6%, P = .08) and the presence of LE was not related to age (P = .36). The youngest subject with an LE was 5 months old. During long-term follow-up there were no clinical events in the 12 children identified with a LE.

Conclusions: The prevalence of LE in children is lower than that reported in adults, this supports the age-related "wear and tear" process that has been described in previous studies. LE do not require intervention or more aggressive invasive imaging in children.

KEYWORDS

children, Lambl's excrescences, transthoracic echocardiography

1 | INTRODUCTION

Lambl's excrescences (LE) were initially identified by Vilem Dusan Lambl in 1856 as filiform extensions on the ventricular surface of the aortic valve.¹ LE are currently believed to be a degenerative phenomenon without associated thromboembolic disease.² Histologic examination of these excrescences reveals 1 mm wide lesions that are usually 1–10 mm long and have a central fibroelastic core covered in a single layer of endothelium.¹ Their presence along the lines of valve closure (Figure 1), along with an age-dependent increase in prevalence, supports the "wear and tear" pathogenesis theory.³ Transesophageal echocardiography (TEE) was usually needed to image LE,³ but with advancements in ultrasound technology, LE are now frequently detected during transthoracic echocardiography (TTE).

In 1995, a retrospective TEE analysis of 1559 adults by Freedberg et al., reported a prevalence of LE of 5.5%.⁴ To date, there is no study reporting the prevalence and clinical significance of LE in children. The purpose of this study was to determine the prevalence of LE detection in a cohort of normal pediatric TTE studies performed during two eras and to determine the effect of improved imaging technology.

2 | METHODS

We reviewed 700 TTE studies in subjects \leq 18 years of age. From these, 350 consecutive studies were from 2004 to 2006 (cohort 1), and 350 consecutive studies were from 2011 to 2012 (cohort 2).

²⁵² WILEY Congenital Heart Disease

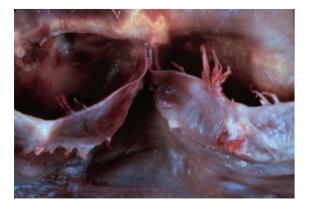


FIGURE 1 Pathologic specimen of Lambl's excrescences (LE). Note the filiform extensions (arrows) from the ventricular surface of the aortic valve cusps (with permission of Dr. William Edwards, Mayo Foundation, Rochester, MN)

Inclusion criteria included subject age < 18 years old and normal intracardiac anatomy with normal ventricular and valvular function. All parasternal long and short axis images of the aortic valve were reviewed. Indications for these studies were murmur, chest pain, syncope, and screening for inherited diseases. All studies were reviewed by a board certified pediatric/congenital echocardiographer (MYQ). The positive studies were then reviewed independently by two additional observers (BWE,FC) to confirm the presence of LE (recognized as fibrous strands <1 mm in width on the ventricular surface of the aortic cusps) (Figure 2). The prevalence of LE in the two cohorts was calculated and analyzed using a two sample *t* test with a 95% level of significance.

3 | RESULTS

Of the 700 subjects (age range: 1 day–18 years, median 10 years), 12 (1.7%) children were found to have LE (Table 1). In cohort 1: 3 (.9%) subjects had LE. In cohort 2: 9 (2.6%) subjects had LE. No significant difference in prevalence was found between the two eras (P = .08). The presence of LE was not related to age (P = .36). The mean age of

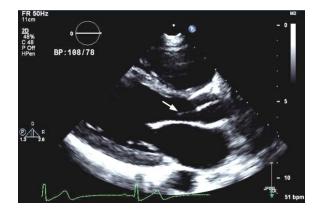


FIGURE 2 Parasternal long-axis echocardiograph demonstrating a long LE (arrow) from a 13-year-old male with an otherwise normal echocardiogram

TABLE 1 Subject demographics

Characteristics	Statistics
Total patients	700
Age at echo, median (range)	10 y (1 d-18 y)
Females	47%
Weight, median (range)	34 kg (1.3-118 kg)
Height, median (range)	140 cm (36-195 cm)
Indication for echo Murmur Screening Syncope Abnormal ECG Chest Pain Dyspnea Palpitations Abnormal chest radiograph	40% 20% 16% 12% 8% 2% 1% 1%
Final impression of echo Normal	100%
Lambl's excrescences present	2% (12)

subjects with LE was 11 ± 5.9 years (range: 5 mos-17 years) (Table 2). None of these subjects had further testing. Median follow-up was 66 months (range: 3–130 mos). No subjects with LE had a neurologic event or endocarditis during the follow-up period.

4 DISCUSSION

The prevalence of LE in children is lower (.9%–2.6%) than that reported in adults. This supports the age-related "wear and tear" process that has been described in previous studies. One pathologic study, by Magarey et al. in 1949, showed that no aortic valves in children younger than 1 year old had LE, whereas every specimen older than 60 years had LE.¹ It is important to differentiate LE from thrombus, vegetation, or papillary fibroelastoma, all of which have a risk of embolism. Historically, LE in young people have not required intervention or more aggressive invasive imaging.

A stratified study by Roldan et al. in 1997 reported a prevalence of LE of 38% in healthy patients, 47% in patients "suspected" of having thromboembolic events, and 45% in patients with confirmed embolic events. They concluded that LE are normal findings with no association with embolic disease.⁵ However, there are case reports that attempt to causally link LE to embolic events in patients undergoing surgery to excise the LE.⁶ Justification for these interventions is lacking. However, a more recent case report and review of the literature,⁷ suggested that antiplatelet agents have a therapeutic role for adults with LE and stroke. That literature review evaluated case reports of 23 patients with stroke or transient ischemic attack and LE on the aortic or mitral valves. The youngest patient was 30 years old. Approximately half of the patients had surgical excision of the LE and the others were treated medically with antiplatelet or vitamin K antagonist therapy. There was no correlation between length of the LE strand and stroke. The histologic similarities between LE and papillary fibroelastomas (fibroelastic

Age	Sex	Primary indication of echo	Final impression on echo	Lambl's excrescences noted on report	Final clinical diagnosis	Follow-up duration in months
5 m	Female	Murmur	Normal study	No	Innocent murmur	68
3 у	Female	Screening	Normal study	No	normal cardiac anatomy and function	36
4 y	Female	Murmur	Normal study	No	Innocent murmur	114
9 y	Female	Murmur	Normal study	No	Innocent murmur	3
10 y	Male	Syncope	Normal study	No	Vasovagal syncope	9
13 y	Male	Murmur	Normal study	No	Innocent murmur	130
14 y	Female	Syncope	Normal study	No	Vasovagal syncope	68
16 y	Female	Syncope	Normal study	No	Vasovagal syncope	67
16 y	Female	Abnormal ECG	Normal study	No	normal cardiac anatomy and function	26
17 y	Female	Syncope	Normal study	No	Vasovagal syncope	67
17 y	Female	Chest pain	Normal study	Yes	Musculoskeletal pain	65
17 y	Female	Abnormal ECG	Normal study	Yes	normal cardiac anatomy and function	5

and hyalinized stroma) were cited in this review. But the most efficacious therapy, if any, for LE remains controversial even in older patients with stroke or transient ischemic attack.

Improvement in ultrasound imaging technology may result in increased detection due to better visualization of LE. However, this study did not show a statistically significant difference (P = .08) in detection of LE between the two eras of echocardiographic imaging.

5 | CONCLUSIONS

LE are detected in normal, healthy children, but less frequently than in adults. With improved technology and continued advances in the imaging capabilities of ultrasound platforms, visualization of LE during TTE in otherwise healthy children may increase.

CONFLICT OF INTEREST

None.

AUTHOR CONTRIBUTIONS

Data interpretation: Phillips, Qureshi, Cetta Drafting article: Phillips, Qureshi Critical revision of article, Data collection: Qureshi, Eidem, Cetta Concept/Design: Qureshi, Cetta Statistics: Qureshi, Eidem Approval of article: Eidem, Cetta

ORCID

Muhammad Yasir Qureshi (b) http://orcid.org/0000-0002-9898-2318

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