

## Key Points and Rare Presentations of Cardiac Fibroelastoma in Pediatrics: Review Article

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### ABSTRACT

**Background:** The majority of primary heart tumors are benign, but they are uncommon. The two most prevalent forms of primary cardiac tumors in adults are cardiac myxomas and papillary fibroelastomas (PFEs). A benign neoplasm is PFE. According to a single series of several patients by some authors, PFEs are the most prevalent kind of cardiac malignancy. The detection of intracardiac malignancies has resulted from the use of sophisticated imaging methods like transesophageal echocardiography (TEE).

**Objective:** This review article aimed to highlight key points and rare presentations of cardiac fibroelastoma in pediatric.

**Methods:** The terms PFE, pediatric, stroke, and cardiac tumor were used to search PubMed, Science direct and Google scholar. Additionally, the writers culled references from the pertinent literature, identifying and included just the most current or comprehensive study out of all the found studies and reviews. The search for literature was limited to English language works. Dissertations, oral presentations, conference papers, unpublished articles, and abstracts from smaller scientific investigations were excluded.

**Conclusion:** Among benign cardiac tumors, cardiac PFE (CPFE) are extremely rare in children. Although most CPFE do not cause symptoms, there is sufficient evidence to state that these tumors can cause life-threatening complications. Therefore, the diagnosis of CPFE in infants and children is of vital importance due to the high risk for embolization. Accordingly, prophylactic tumor excision with valve repair or replacement when necessary is considered to be the treatment of choice, even in asymptomatic patients.

**Keywords:** PFE, Pediatric, Stroke, Cardiac tumor.

### INTRODUCTION

A benign, pedunculated tumor with strand-like extensions emerging from the endocardium is called a cardiac PFE. These tumors can enlarge and cause significant neurologic problems, even though they are frequently found by accident. The best time to do surgery on individuals with this uncommon primary heart tumor following a cerebrovascular incident is not well-documented in pediatric literature [1].

Aortic valve prolapse may cause patients to get acute embolic strokes. When previously healthy children report with sudden stroke, a strong index of suspicion should be maintained for such malignancies notwithstanding their rarity [1].

In 1975, a myocardial infarction (MI) caused by an embolic complication of PFE was the first case of PFE that was documented. From that time on, PFEs have been linked to cardiac arrest, stroke, and vascular embolism. These so-called "benign" tumors are known to cause embolic difficulties downstream, therefore there is still much debate about whether to treat them surgically or medically [2].

### Etiology:

Due to their relative rarity, the exact cause of cardiac PFE is still mostly unclear. Since it is thought to develop in regions of valve endothelial injury, they mostly come from the valvular endocardium [3].

### Epidemiology:

Primary cardiac tumors are incredibly uncommon; fewer than 0.1% of cases have been reported in one research [3]. The actual occurrence is unclear, but it is

more prevalent among men. The aortic valve accounts for 35% to 63% of PFE cases, however other valves such as the mitral valve (9% to 55%), tricuspid valve (6%–15%), and pulmonic valve (0.5–8%) are also often affected [4-5].

### Pathophysiology:

Nonetheless, a number of explanations about the pathophysiology of PFEs have been proposed. According to one theory, these lesions are acquired entities that start off as microthrombi. On valvular surfaces, these microthrombi converge at locations where endothelial damage is minimal. These microthrombi eventually develop into excrescences and PFEs [6]. These PFEs keep becoming bigger and have the potential to embolize, which might lead to problems like stroke, MI, ventricular fibrillation, and unexpected death. The cerebral arteries are the arterial beds most frequently impacted by embolization, which can result in anything from brief ischemic episodes to stroke [7].

### Histopathology:

PFE has a gross look that is traditionally characterized as a "sea anemone" with arms that resemble fronds extending outward from a central stalk. These tumors can range in size from several centimeters to as tiny as 2 mm in vivo, with a typical size of 9 mm. PFEs are mostly composed of collagen, elastin, and reticulin on a histological level, with very little vasculature [5]. The endothelium makes up the outer layer, followed by an intermediate layer with connective

tissue rich in mucopolysaccharides and a central core made of fibrin and mucopolysaccharides [7].

**Diagnosis:**

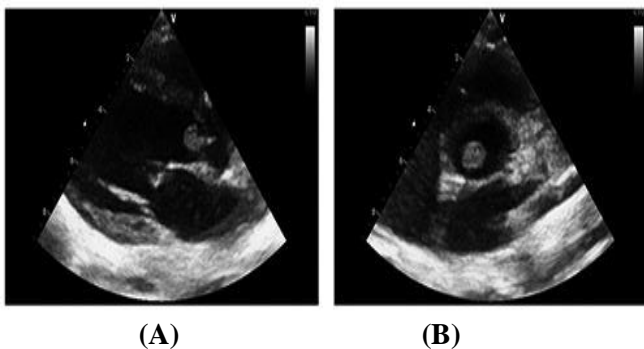
**Symptoms and physical examination:**

PFE is typically asymptomatic and unintentionally found. PFE might first present as embolization symptoms (stroke, TIA, MI) or cardiac obstruction symptoms (syncope, heart failure, sudden death). In thirty percent of cases, a stroke is the most frequent manifestation. Additionally, one-third of patients report it to be an accidental discovery. PFEs do not change the results of a physical examination, but if they are producing complications like blockage, the physical examination will reveal findings that are in line with those complications. It is highly unusual to diagnose PFE based alone on history and physical examination [8].

**Echocardiography:**

**I-Transthoracic 2D echocardiography (TTE 2D):**

Since transthoracic echocardiography (TTE) is the least intrusive method of diagnosing PFE, it should be the first line of treatment. For the identification of PFE larger than 2 mm, TTE has a sensitivity and specificity of 88.9% and 87.8%, respectively. To better see the intracardiac structures, a transesophageal echocardiography (TEE) can be done if there is a high index of suspicion for a cardioembolic etiology of stroke [9]. For TTE and TEE, the total sensitivity of PFE smaller than 2 mm is 61.9% and 76.6% respectively. Most PFEs are discovered by accident during heart surgery or echocardiography imaging. PFE sizes are typically seen to be pedunculated on echocardiography and range in size from 2 mm to 40 mm. They move independently and along the PFE's edges in a speckled pattern. PFEs can originate from either the aortic or ventricular valve surface; the lesion can develop equally on both valve surfaces. While other PFE was identified on the ventricular side of the aortic valve, it was often located on the aortic side of the aortic valve leaflets [10].



**Figure (1):** TTE parasternal short axis (A) and long axis view (B). A circular movable mass, approximately 8 mm in diameter, connected to the ventricular side of the aortic valve and protruding at the free edge of the two leaflets. No stalks could be identified [10].

**Cardiac MRI and CT:**

They could be able to provide spatial resolution that is superior to echocardiography. But these modalities, cardiac MRI in particular, are far more costly, and conducting and assessing the study calls for specific resources. Pathologic study and inspection are necessary for a clear diagnosis, much like with other solid tumors [11].

**Treatment:**

**Treatment modalities available for management of PFE:**

The two main alternatives for therapy are either careful monitoring or surgery. Unless there is a medical reason why surgery cannot be performed, surgical excision is usually the advised course of action. Reconstruction of any resulting intracardiac abnormalities and removal of all visible malignancies are the objectives of surgical therapy. PFE tumors can be precisely excised and are usually connected to a pedicle. To prevent missing several PFEs, inspections are conducted on the other heart chambers. At the time of operation, valve replacement or repair may be necessary if the PFE seriously destroys a valve. It makes sense to explore anticoagulation if the patient is symptomatic but not a good surgical candidate. This strategy is justified by the desire to avoid embolic consequences. Nevertheless, there aren't any studies to back up this approach. Surgery should be considered for asymptomatic individuals with a PFE size more than 9 mm, highly mobile masses, and independent motion as these factors are predictive of poor outcomes [4].

**Differential Diagnosis:**

Although there are many possible diagnoses for a cardiac valvular tumor, they may be divided into groups according to how the patient presents clinically. A TTE or TEE guided detection of mass is extremely indicative of endocarditis if the patient is presenting with worrying indications of infection. Take non-bacterial endocarditis into consideration if the mass is present in the clinical context of lupus or inflammatory conditions. In the differential diagnosis, a thrombus, Lambi's excrescence, or valvular calcifications should be taken into account. Additionally, leiomyosarcoma usually does not affect the aortic valve [11].

The three cardiac tumors that pediatric patients most commonly face are rhabdomyomas, fibromas, and myxomas. Rarely have reports of cardiac PFEs in children and young adults been made [12].

**Prognosis:**

The prognosis for PFE is usually favorable when the tumor is removed. There is little chance of a recurrence or metastatic spread due to their benign nature. The latter could happen if the tumor was not completely removed during the original operation [13].

### Complications:

Complications are classified according to the risk of embolism or sudden cardiac death, ventricular fibrillation and MI, or stroke. The dangers associated with intracardiac surgery on CPB include emboli, hemorrhage, arrhythmias, and infection [14].

### CONCLUSION

CPFE are quite uncommon in youngsters when it comes to benign cardiac tumors. There is enough data to conclude that these tumors can result in potentially fatal consequences, even if the majority of CPFE do not exhibit symptoms. Because of the significant risk of embolization, it is imperative that CPFE be diagnosed in babies and children. Therefore, even in individuals who are asymptomatic, preventative tumor excision combined with valve repair or replacement when needed is seen to be the best course of action.

An uncommon cause of cardioembolic events is PFE; in both younger and older patients, the cardiac source of embolism must be taken into account. For cardiac tumors that have the potential to embolic, surgery is now the only treatment option.

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