Primary cardiac tumors are rare in children, with the prevalence of 0.0017–0.28% in autopsy series.\(^1\) Benign tumors account for over 90% of the total. Cardiac fibromas (CFs) represent the second most common benign cardiac tumor in the pediatric population following rhabdomyomas. Patients can be asymptomatic or present with palpitations, cardiac murmur, arrhythmias, congestive heart failure, and even sudden death according to the size and location of the tumor. Surgical resection should be considered as the best option in symptomatic patients.\(^2,3\) This study reported four surgical cases of large left ventricular fibroma in children.

Between December 2013 and April 2016, four children (two males and two females) diagnosed with primary intramural tumors arising from the left ventricle (LV) underwent a complete surgical resection. Their ages ranged from 4 to 11 years, and weight ranged from 19.9 kg to 34.0 kg. Echocardiography and cardiac magnetic resonance imaging (MRI) confirmed the diagnosis of a large cardiac mass located at the free wall of LV in all the patients. Of them, cardiac systolic and diastolic function in a patient with nonsustained ventricular tachycardia (VT) was worsening with ejection fraction (EF) of 50%. Basal characteristics of the patients are shown in Table 1.

Surgery was performed through a median sternotomy with cardiopulmonary bypass and cold cardioplegic arrest. The pericardium was opened, and a large mass was evident arising from the free wall of LV. Dissection was started by incising the epicardium which appeared to be the thinnest portion over the mass. The mass was pale, firm, and nonencapsulated, which was carefully dissected free of the surrounding myocardium using sharp dissection [Figure 1a]. Then, the mass was completely removed [Figure 1b] with or without entering the ventricular cavity, leaving a large defect in the lateral wall of LV. The residual defect together with the cut edge of LV wall was closed with a running 5-0 Prolene suture. Then, the area was covered with a piece of autologous fresh pericardium.

There were no hospital and late deaths and no major complications. Postoperative electrocardiogram of all the patients showed normal sinus rhythm and echocardiography showed normal EF with no residual mass. Histopathology confirmed the diagnosis of CF in four patients. All patients remained symptom-free without episodes of ventricular
arrhythmias (VA) during the follow-up period. The echocardiography revealed normal left ventricular size and function of each patient without tumor recurrence. The EF in the third patient [Table 1] increased from 50% to 60% after the operation and to 64% after 18 months.

Large fibromas of LV are exceptionally rare in children. CF is usually solitary, nonencapsulated, firm, nodular, and gray-white intramural tumors. They are composed of elongated fibroblasts in broad spiral bands and whirls mixed with collagen and elastin fibers. The clinical presentation of CF depends on the size and location of the tumor. Congestive heart failure and VA may be the first clinical manifestation in large fibromas. Patients with large fibromas seem to be the most concerning group, with a VT risk exceeding 50%. Diagnosis is usually established by echocardiography, CT, and MRI.

Surgery is required as an effective management strategy in the children when CF cause ventricular inflow/outflow tract obstruction, valve dysfunction, heart failure, and arrhythmia. In the present study, three patients presented with some form of VA, ranging from frequent premature ventricular beats to nonsustained VT. The onset of VA for displacement or compression of the conduction system is often life-threatening and difficult to control, which can result in sudden death. The VA was eliminated after our patients underwent complete fibroma excision, suggesting that significant debulking of the tumor mass might be sufficient to reverse the arrhythmogenic substrate.

Although the goal of surgery is as complete a resection as possible, the large tumor size in relationship to the left ventricular cavity has often been considered a limiting factor for complete excision of fibromas. Of our two patients, the size and extent of the rare giant fibromas (7 cm × 5 cm) had put them in the realm of incompletely resectable tumors, but they still underwent the complete excision of the tumors without recurrence, with resolution of symptoms and arrhythmic events. Hence, we think that the key to operation is not only carefully and completely dissect the tumor from healthy surrounding muscle without damage to other normal structures, but also to reconstruct the left ventricular wall for preventing dyskinetic motion and the formation of the ventricular aneurysm, and avoid bleeding after closure of the excision.

In conclusion, complete surgical resection should be recommended as the first-choice treatment for large left ventricular fibromas in children. The cardiac function at short- to medium-term follow-up is excellent, with a resolution of symptoms and arrhythmic events.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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