treta conduit is manufactured by glutaraldehyde fixation of a segment of bovine jugular vein that contains a natural trileaflet valve. The Contegra graft is currently approved as a humanitarian-use device for reconstruction of the right ventricular outflow tract in patients younger than 19 years with congenital cardiac malformations. Biologic tissue grafts are believed to be more resistant to infection than nonbiologic grafts because they allow a greater degree of antibiotic penetrance and the ability of host cellular defenses to incorporate into graft material. Increased resistance of biologic tissue grafts to infection makes them an attractive option in patients with intracardiac or mediastinal infection.

When available, appropriately sized aortic and pulmonary homografts are excellent alternatives for replacing an infected PTFE graft. However, the use of homografts in this clinical situation was limited by the relative scarcity of small homografts. In contrast, Contegra grafts are readily available and appear to have reintervention profiles comparable with homografts [1]. The use of a downsized Contegra graft has not been reported, but studies that describe the use of downsized homografts in infants and children indicate that the patency rate of downsized conduits is similar to standard homograft conduits of similar size [2,3]. Although this patient experienced multifocal thrombosis that necessitated stenting of the downsized Contegra graft, use of the modified conduit enabled clearance of purulent mediastinitis. In the setting of multifocal thrombosis, it is unclear whether the leaflets within the conduit contributed to thrombotic obstruction of the graft. It is possible that excision of the leaflets at the time of implantation could have prevented this complication. The surgically downsized Contegra bovine jugular graft appears to be a valuable alternative to replacing an infected PTFE graft in an infected space.

We are reporting a rare case of papillary fibroelastoma of the tricuspid valve in an 8-year-old child who presented with pulmonary embolism. Echocardiography was instrumental in determining the source of the pulmonary embolism, but not in delineating between tumor and thrombus. Successful surgical resection of the mass was accomplished and good outcome was attained despite the delay in diagnosis and failure of medical management. A high index of suspicion for tumor involving the tricuspid valve is emphasized despite its rarity in children.


Cardiac papillary fibroelastoma (PFE) is a rare benign cardiac tumor, with an incidence of 0.33% in autopsy series [1]. These tumors are the third most common benign cardiac tumor, with main involvement of the valvular heart structure, predominantly the left-sided cardiac valves [2, 3]. Primary cardiac tumors in children are rare, and there are only sporadic cases reported. Papillary fibroelastoma of tricuspid valve is even more rare in pediatric patients, and only a handful of cases have been reported in the literature [4–6]. We are reporting an unusual case of PFE of the tricuspid valve in an 8-year-old boy who presented with chest pain and shortness of breath as a result of pulmonary embolism (PE), and the clinical management.

This 8-year-old boy presented with a history of worsening chest pain and shortness of breath for several months, with no preceding factors. As part of his extensive workup, he underwent chest roentgenography, which demonstrated normal cardiopulmonary structures and vascularity with no infiltrates, effusion, or bony fractures or mass. He subsequently underwent cardiac computed tomography angiography (CTA), which was diagnostic for bilateral PE of lower lobe segments. His lower extremities duplex study ruled out deep venous thrombosis, despite which he was treated with anticoagulation therapy in the form of intravenous heparin infusion owing to the presence of PE. He also had extensive hematologic studies for infection, malignancy, and hemophilic factor abnormalities, which all turned out to be normal. He had a transthoracic echocardiogram to further investigate the source of his PE as well as for the clinical presence of his heart murmur. His echocardiogram demonstrated a mass on the anterior leaflet of the tricuspid valve, with estimated dimensions of 2.5 cm × 1.5 cm and moderate central regurgitation with no other cardiac defects (Fig 1). The mass was followed up with daily transthoracic echocardiograms for its resolution while the patient continued therapeutic heparin treatment for presumed thrombus on the tricuspid valve. Thrombolysis with a single dose of recombinant tissue plasminogen activator to dissolve the presumed thrombus was tried and failed.

### References


### Papillary Fibroelastoma of Tricuspid Valve in a Pediatric Patient

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Pediatric cardiothoracic surgery was consulted for surgical resection of the mass given patient’s persistent symptoms and the lack of resolution of the presumed thrombus with medical management. He was taken to the operating room and placed on mild hypothermic cardiopulmonary bypass through a median sternotomy. After cardioplegic arrest of the heart, the tricuspid valve was explored through a right atriotomy approach. Inspection of the tricuspid valve revealed the presence of a single lobulated light-gray mass on the anterior leaflet of the tricuspid valve and foreshortened septal leaflet (Fig 2). The mass was excised in its entirety with a great portion of the involved anterior leaflet of the tricuspid valve and foreshortened septal leaflet (Fig 2). The mass was replaced with a 25-mm bioprosthetic valve (Carpentier-Edwards Perimount Magna Valve; Edwards Lifesciences, Irvine, CA) after failed attempts to repair the valve. The frozen section revealed spindle cell neoplasm, and the final pathology results were consistent with papillary fibroelastoma (Fig 3). The patient had an uneventful postoperative course and was shortly discharged home on a regimen of aspirin and warfarin anticoagulation based on current anticoagulation treatment recommendations for bioprosthetic valves. He was followed up on numerous occasions after discharge, with resolution of his symptoms and PE on repeat CTA.

Comment

Papillary fibroelastoma is an uncommon primary cardiac tumor [1]. It accounts for the most common valvular heart tumor in adults, with an incidence of 7.9%, predominantly involving the left-sided valvular structure [2, 3]. It is an exceedingly rare occurrence on the tricuspid valve, and only a few anecdotal pediatric cases have been reported in the literature [4–9]. Despite the benign nature of the tumor, serious clinical complications, such as myocardial infarction, stroke, and sudden death, have been reported.

Papillary fibroelastoma used to be an incidental finding at autopsy until the advent of echocardiography. Typical echocardiographic features of the tumor include a round, oval, or irregular appearance, with well-demarcated borders and a homogeneous texture with almost half containing a small mobile stalk [10, 14]. On gross anatomic examination, PFE resembles a sea anemone, consisting of numerous fingerlike fronds radiating from a stalk. Microscopically, each frond consists of a collagenous and elastic core, surrounded by a mucopolysaccharide matrix, and covered by endothelial cells [10, 11].

The pathogenesis of PFE remains under discussion, but several possible explanations have been reported, including previous mechanical damage to the endothelium, iatrogenic factors, organizing thrombi, and a latent infectious mechanism due to cytomegalovirus [12, 13]. The occurrence in neonates and infants supports the hypotheses of hamartomatous origin or congenital abnormalities [12].

Pulmonary embolism is not an uncommon occurrence in adulthood but is a rare event in childhood. It is mostly...
caused by venous thromboembolism as the result of multiple potential risk factors such as immobility, central venous catheter, recent surgery, congenital heart disease, inherited or acquired thrombophilic abnormalities, malignancy, infection, nephrotic syndrome, etc. The most common imaging modalities used in the diagnosis of PE is ventilation-perfusion scan and computed tomography pulmonary angiography. Alternative imaging techniques, namely, cardiac CTA, conventional pulmonary angiography, and transthoracic/transesophageal echocardiography are sometimes used to make the diagnosis of PE in concert with Doppler ultrasonography for deep venous thrombosis. The D dimer levels as well as thrombophilia screening are done in addition to other potential culprits for venous thromboembolism as part of extensive workup for PE. Anticoagulation therapy in the form of intravenous unfractionated heparin or low molecular weight heparin on initial presentation and diagnosis of PE with transition to warfarin for a median period of 3 months is the conventional treatment for uncomplicated PE. Systemic thrombolysis with recombinant tissue plasminogen activator is only needed for the patients who present in cardiorespiratory distress. Emergent surgical thromboembolectomy is occasionally performed after failed thrombolysis in patients with cardiorespiratory distress. Repeat imaging is required before, during, and at the end of the treatment period to follow up on the resolution or progression of the PE [15].

This particular patient presented with persistent chronic pleuritic pain and shortness of breath with no preceding factors such as trauma, infection, or hematologic abnormalities hinting at his symptoms. The CTA demonstrated subsegmental pulmonary embolism, obviating the need for pulmonary angiography or ventilation-perfusion scan. The echocardiogram was helpful in determining the source of embolization, but was misleading toward thrombus formation, subjecting the patient to the potential risks of heparin anticoagulation therapy and thrombolysis with recombinant tissue plasminogen activator. Intraoperative transesophageal echocardiography also portrayed the same findings and definition as did preoperative transthoracic echocardiograms with respect to structural distinction between a mass versus thrombus. In retrospect, cardiac magnetic resonance imaging would have potentially delineated the nature and consistency of the tricuspid valve mass early in presentation, thus avoiding risky exposure to anticoagulation therapy and thrombolysis and leaning toward earlier surgical resection.

Tricuspid valve repair in the form of leaflet augmentation with autologous pericardium or other patch material with added anuloplasty is feasible and has favorable mid to long-term durability. In this particular case, attempts to repair the tricuspid valve were unsuccessful owing to extensive resection of the anterior leaflet involved with the mass and foreshortened septal leaflet, which prohibited valve repair, hence leading to valve replacement with a bioprosthetic valve. The downside is that this patient will potentially require that the bioprosthetic valve be replaced in the future as the result of leaflets’ degeneration despite implantation of a large-size valve. Nevertheless, having a competent bioprosthetic valve will preserve his normal cardiac geometry and function in contrast to his own native incompetent or stenotic repaired valve.

In conclusion, this is a rare case of papillary fibroelastoma of the tricuspid valve in a pediatric patient with pulmonary embolization and associated symptoms. Transthoracic echocardiography was useful in determining the source of the embolization, but not the nature of the mass. A high index of suspicion for valvular cardiac tumor in children, such as papillary fibroelastoma, is needed for more definitive imaging studies and earlier surgical treatment. Although this is a benign tumor with rare chance of recurrence, frequent follow-up with echocardiographic imaging is warranted.

References