

operation, the patient was hemodynamically stable without catecholamines. The patient received 500 mL of packed red blood cells and 600 mL of plasma.

The patient was extubated at 12 hours postoperatively. His postoperative course was complicated by bronchopneumonia and a transitory psychotic syndrome. The patient was discharged on postoperative day 20.

The histopathologic workup showed that the surgical margins were negative in the IVC (R0). In the pulmonary endarterectomy specimen, the neoplastic nature of the embolus was proven; however, margins could not be judged properly.

On follow-up visitation at 6 months, the patient was in a good clinical condition, without any complaints, and had returned to work. The follow-up computed tomography imaging showed no tumor recurrence or metastasis. The reconstructed IVC was patent.

### Comment

The IVC is subdivided into 3 segments delineated by the inflow of the hepatic and renal veins [2]. Tumors of the inferior or middle segment can be resected during clamping of the IVC and by performing the Pringle maneuver. As soon as the upper segment or the RA is involved, CPB is mandatory.

The techniques used for the resection and reconstruction of the IVC include ligation, graft replacement [3], patch cavoplasty, or primary repair [2]; however, the choice of techniques to be used is controversial. If perivascular infiltration is observed, a preferably radical approach with wide resection and ligation or replacement of the IVC is mandatory [4]. However, a predominantly intravascular growth pattern is shown in a relevant portion of leiomyosarcomas [2]. In this patient, no perivascular infiltration was seen intraoperatively or during the pathologic workup; therefore, we determined that partial resection with patch reconstruction was adequate.

To prevent thrombosis of the reconstructed IVC, the construction of a temporary arteriovenous fistula is often advised. We and others [3] think that this is not necessary in case of suprarenal cavoplasty. In our experience of more than 30 cases of cavoplasty using a native pericardial patch, we have never seen a thrombosis; in addition, the lowest rates of leg edema are reported for patch reconstruction [2]. However, we advise anticoagulation with phenprocoumon for 3 months after IVC reconstruction.

In the described case, IVC resection and repair were performed during HCA because the suprahepatic segment and the RA had to be inspected. After RA closure, we initially tried to complete the cavoplasty during CPB; however, massive backflow through the hepatic veins and collaterals was extremely hindering and led us to complete the reconstruction during HCA. For management of possible complications, especially intraoperative tumor embolization, cannulation was done before luxation of the liver. Cerebral oxygen saturation monitoring was used to prevent critical desaturation during HCA.

Although pulmonary embolectomy for acute pulmonary embolism can be done as a beating-heart procedure,

pulmonary endarterectomy is required for chronic pulmonary embolic disease, regardless of whether it is thromboembolic or neoplastic. The latter procedure usually requires HCA to facilitate a blood-free field in patients with compensatory increased blood flow in bronchial arteries and collaterals [5].

In the tumor in the pulmonary vasculature, unlike the tumor in the IVC, it was impossible to prove R0 resection by means of histopathology, whereas intraoperatively, all macroscopic parts of the tumor were removed completely. However, the patient's history and symptoms indicated a slow and less-invasive growth pattern of the tumor. Therefore, pulmonary recurrence of the tumor should be detected early by follow-up computed tomography and optionally could be treated by appropriate lung resection.

We conclude that the combined removal of a large leiomyosarcoma of the IVC and pulmonary endarterectomy for extensive intravascular neoplastic disease using the described approach of intermittent HCA is feasible and can produce a good perioperative and midterm postoperative result.

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## Development of a Subaortic Aneurysm Secondary to Disseminated Tuberculosis in a Child

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**Subvalvular aneurysms of the left ventricle are very rare and often the cause is uncertain. Most of the cases of subvalvular left ventricular aneurysms described in the**

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literature are due to congenital weakness of the fibromuscular annuli. We describe a unique case of a child with a tuberculous subaortic aneurysm observed at different stages of development by serial transthoracic echocardiography. The patient underwent successful cardiac surgery after the initial conservative treatment for tuberculosis.

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Subvalvular aneurysms of the left ventricle are very rare and often the cause is uncertain. Very little data in the literature describes subvalvular aneurysms in children and most of the data are derived from case reports. We describe a unique case of a human immunodeficiency virus negative child with a tuberculous subaortic aneurysm observed at different stages of development by serial transthoracic echocardiography. The patient underwent successful cardiac surgery after the initial conservative treatment for tuberculosis (TB).

A 3-year-old black boy who was negative for human immunodeficiency virus presented to our institution with a 1-week history of coughing, fever, night sweats, and generalized body swelling. On examination he appeared chronically ill and was underweight for his age. He had generalized lymphadenopathy in the axillary, supraclavicular, cervical, and submandibular regions. He had ascites, hepatomegaly (liver span, 12 cm), and splenomegaly (3 cm below costal margin). The boy was tachypneic with a respiratory rate of 50 breaths/min. He had intercostal and subcostal recession and audible bilateral rhonchi. His heart rate was 150 beats/min, with a gallop rhythm present and with muffled heart sounds. There were no murmurs, but he was in congestive cardiac failure. His blood pressure was 80/40 mm Hg.

A chest roentgenogram demonstrated cardiomegaly, a widened mediastinum, and paratracheal lymphadenopathy. The C-reactive protein, lactate dehydrogenase lev-

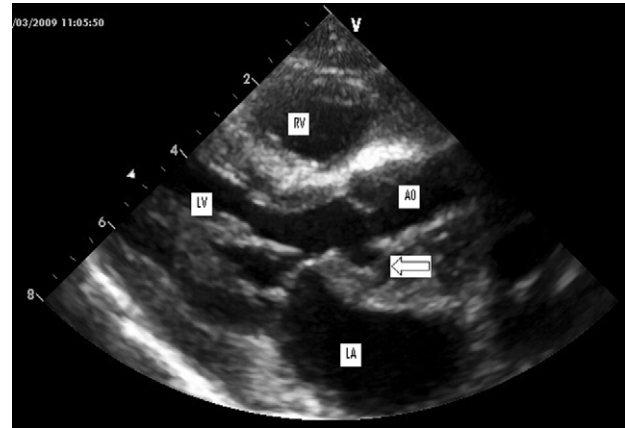


Fig 2. Repeat transthoracic echocardiogram 6 weeks after presentation. Parasternal long axis view demonstrating a small pouch in the subaortic area (arrow). (AO = aorta; LA = left atrium; LV = left ventricle; RV = right ventricle.)

els, and erythrocyte sedimentation rate were elevated. Echocardiography (Fig 1) revealed normal intracardiac anatomy with a large organized and loculated pericardial effusion. A biopsy of a supraclavicular lymph node demonstrated caseating necrosis with sites of granulomatous inflammation. Epithelioid cell granulomas and Langhans giant cells were also seen. The Ziehl Neelsen stain highlighted occasional acid and alcohol fast bacilli, which confirmed the TB. There were no features of lymphoma or malignancy present. Mycobacterium TB was also isolated on cultures of gastric aspirates. A diagnosis of disseminated TB with organized TB pericarditis was made.

The patient was treated with anti-TB drugs (isoniazid, rifampicin, pyrazinamide, and ethambutol), prednisone, and appropriate cardiac failure therapy. His clinical condition improved with resolution of the generalized edema and ascites and with a decreased size of his lymph nodes. A repeat echocardiogram (Fig 2) after 6 weeks of treatment demonstrated a small pouch (2 mm × 3 mm) in the subaortic area with mild aortic regurgitation. Cardiac surgery was not considered at this stage because of the active TB and the possibility of friable tissue. He was closely monitored while receiving TB treatment, and he remained clinically asymptomatic. Another follow-up echocardiogram (Fig 3) 3 months later demonstrated a large subaortic aneurysm just below the left coronary cusp that measured 30 mm × 24 mm and compressed the left atrium. The neck of the aneurysm measured 5 mm and a color Doppler echocardiogram demonstrated flow in and out of the aneurysm from the left ventricle. The pulsed wave spectral Doppler echocardiogram demonstrated restrictive bidirectional flow into the aneurysm with a flow velocity of 4 m/s. There was no distortion of the aortic valve and only mild aortic regurgitation was evident. These findings were confirmed on transesophageal echocardiography (Fig 4). A left ventriculogram (Fig 5) demonstrated the large aneurysm clearly, which also appeared to be compressing the left coronary artery.

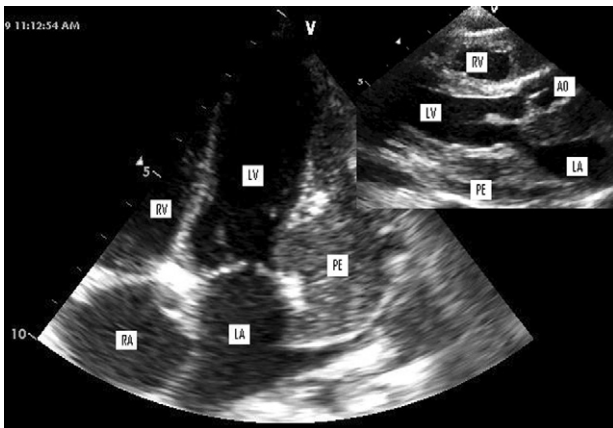


Fig 1. Apical four-chamber view and parasternal long-axis view echocardiogram (inset) demonstrating the organized loculated pericardial effusion. (AO = aorta; LA = left atrium; LV = left ventricle; PE = loculated organized pericardial effusion; RA = right atrium; RV = right ventricle.)

The boy underwent successful cardiac surgery 5 months after TB therapy was initiated. There was extensive fibrosis in the thoracic cavity and pericardial space. The aneurysm was approached through an incision in the ascending aorta and through the aortic valve. The neck of the aneurysm was sutured and closed to isolate the aneurysm, and blood was allowed to resorb spontaneously. Due to the extensive fibrosis and the posterior position of the aneurysm, evacuation was deemed too risky. The boy was doing well 4 months after the surgery and had completed the anti-TB therapy.

**Comment**

Subvalvular left ventricular aneurysms can either be subaortic or submitral. The submitral aneurysms are

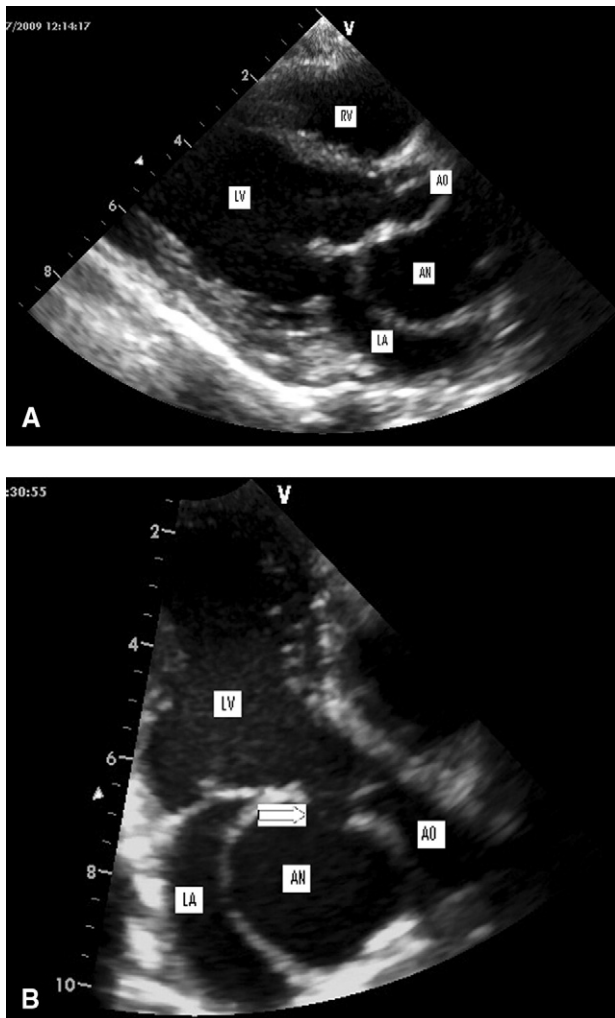


Fig 3. Repeat transthoracic echocardiogram 3 months after presentation. (A) Modified parasternal long-axis view demonstrating a large subaortic aneurysm. (B) Modified apical three-chamber view demonstrating the large subaortic aneurysm compressing the left atrium (LA). The opening of the aneurysm is seen just below the aortic valve (arrow). (AN = aneurysm; AO = aorta; LV = left ventricle; RV = right ventricle.)

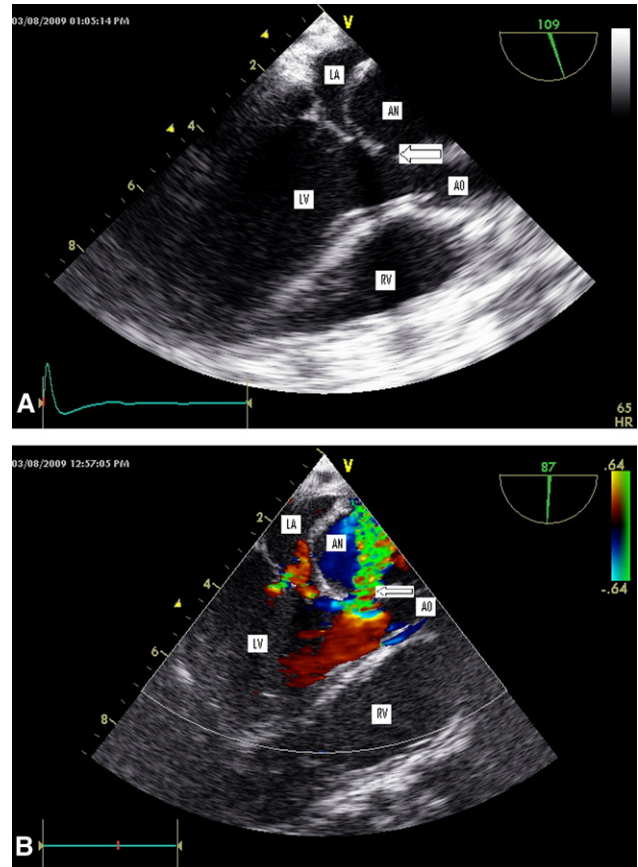


Fig 4. Transesophageal echocardiogram: (A) demonstrating the large subaortic aneurysm compressing the left atrium and narrow neck (arrow). (B) With color Doppler showing the restrictive flow through the neck of the aneurysm (arrow). (AN = aneurysm; AO = aorta; LA = left atrium; LV = left ventricle; RV = right ventricle.)

more common than the subaortic type [1]. Subaortic and submitral aneurysms arise in the fibro-muscular rings of the aortic valve and mitral valve annuli, respectively [2].

Most of the cases of subvalvular left ventricular aneurysms described in the literature are due to congenital weakness of the fibro-muscular annuli. Chesler and colleagues [2] postulate that a dehiscence of the fibro-muscular union will result in aneurysm formation. The majority of these patients are of African ancestry, although similar conditions have been found to a lesser degree in other race groups [1, 3, 4]. They have also been described as a complication in patients with infective endocarditis, tuberculosis, and syphilis [4-7]. Du Toit and colleagues [6] in their series of 12 patients found an association with rheumatic carditis in 2 patients. The causal effect of all these infections has always been speculative, as it may just reflect the high incidence of the infections in endemic areas, rather than a causal mechanism [6].

Our patient had disseminated mycobacterium TB infection with cardiac involvement. There was no aneurysm noted on the initial echocardiogram, thus excluding a congenital origin. The aneurysm was initially identified as a small pouch 6 weeks after presentation. By 4 months



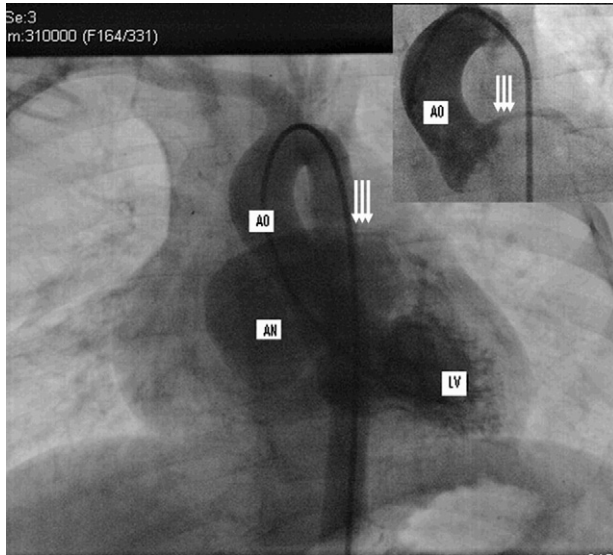


Fig 5. Left ventriculogram and ascending aortogram (inset) demonstrating the large aneurysm compressing on the left coronary artery (arrows). (AO = aorta; AN = aneurysm; LV = left ventricle.)

the small pouch had grown into a large aneurysm requiring surgical intervention.

We believe this case is the first report in which a congenital cause could be excluded by echocardiogram, and aneurysm development was linked to disseminated TB. We also believe that this is the youngest patient reported with a subaortic aneurysm secondary to TB. Disseminated TB is often associated with acquired immunodeficiency syndrome, but our patient was negative for human immunodeficiency virus and was immunocompetent. There were no vegetations identified on any of the valves to suggest infective endocarditis as the cause.

Subaortic aneurysms must be distinguished from sinus of Valsalva aneurysms. The aortic valve and the sinuses were normal, both on transesophageal echocardiogram and on angiogram, excluding a sinus of Valsalva aneurysm.

Surgical repair is the treatment of choice for large subaortic aneurysms, which are at risk of rupture, calcification, and infective endocarditis if left untreated [8]. Large aneurysms have been reported to compress the coronary arteries, causing myocardial infarction and sudden death [1, 2], or to compress the conduction system, causing heart blocks [9]. The ventriculogram in our patient demonstrated compression of the left coronary artery, although the electrocardiogram did not reveal any ischemic changes.

Transthoracic and transesophageal echocardiography played a pivotal role in identifying, monitoring, and planning surgical intervention in our patient.

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## Fenestrated Exclusion of the Right Ventricle for Tricuspid Atresia and Absent Pulmonary Valve

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The case of a boy who had an absent pulmonary valve, tricuspid atresia, intact ventricular septum, coronary-to-right-ventricular fistula, and dysplasia of the right ventricular free wall is described. At the bidirectional cavopulmonary shunt procedure, the right ventricular free wall was opened and two major fistula orifices to the cavity were closed with sutures. A fenestrated circular patch was placed in the main pulmonary artery and the right ventricular free wall was plicated. The patient then underwent completion for total cavopulmonary connection. Follow-up catheterization showed that the pulmonary artery was partially excluded with minimal pressure wave conduction from the right ventricle, which significantly shrank. This new approach seems to be effective and reproducible in this particular situation.

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**A**b absence of the pulmonary valve occurs infrequently in association with tricuspid atresia, intact ventricular septum, and dysplasia of the right ventricular free wall. The right ventricle is nonfunctional and potentially hazardous for Fontan completion because of nonphysiologic backward flow in the systemic vein, and surgical management is challenging and controversial [1]. In the following Fontan case, fenestrated exclusion of the pathologic right ventricle (RV) is described.

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