A 14-year-old adolescent man from the Republic of Congo, with no personal medical history, presented with progressive shortness of breath and exercise limitation. On clinical examination, he had a 3/6 systolic murmur and regular cardiac rhythm, and cyanotic fingers and clubbing, as well. Pulmonary auscultation was normal. No recent episode of fever or shivering or peripheral sign of endocarditis were reported. Pulse oxymetry showed oxygen saturation at 64%. The diagnosis of Tetralogy of Fallot was subsequently established. Laboratory results showed a normal white blood cell count with no abnormalities.

The initial echocardiogram showed a large perimembranous septal defect of 1.5-cm diameter, an aortic override, a predominant right-to-left ventricular shunt, a severe infundibular pulmonary stenosis, and right ventricular hypertrophy. Left ventricular diameter was 39 mm at end-diastole, and the left ventricular ejection fraction was normal. A 4-mm mobile element was unexpectedly visualized at the level of the pulmonary valve.

An ECG-gated multidetector pulmonary computed tomography angiogram with iodinated contrast injection was then performed to explore this unusual finding. Preoperative transesophageal echocardiography was not considered as appropriate for this young, poorly cooperating adolescent patient. It revealed a 28×14 mm hypodense intraluminal filling defect attached to the pulmonary valve (Figures 1 and 2). This mass, suggesting a thrombus or vegetation, nearly occluded the pulmonary trunk. In addition, multiple bilateral pulmonary emboli were seen with images suggestive of mycotic aneurysms (Figure 3). The right inferior lobar artery was occluded. There were also pulmonary nodules in relation to distal pulmonary arteries with peripheral ground-glass opacities, indicative of peripheral pulmonary abscesses (Figure 4). Bronchial arterial collaterals were well developed.

Pulmonary arterial pressure measurement by catheter was avoided because of the major risk of material migration. Surgery was indicated to remove and analyze the mass from the pulmonary arterial trunk, and, depending on the resultant pulmonary arterial pressure, to repair the Tetralogy of Fallot. Intraoperative transesophageal echocardiography was performed to confirm the diagnosis (Movie I in the online-only Data Supplement). The intervention was performed through a median sternotomy, and pulmonary arterial pressure was isosystolic (105/64) by direct measurement. Hypothermic cardiopulmonary bypass with antegrade cold blood cardioplegia was used. Incision of the pulmonary artery showed the mass, which seemed thrombotic, attached to a bicuspid pulmonary valve. All of the abnormal material was excised from the main pulmonary artery without removing the pulmonary valve and sent for pathological and bacterial examination (Figure 5).

After closure of the main pulmonary artery and weaning from cardiopulmonary bypass, the pulmonary arterial pressure remained isosystolic (108/67). Infundibular pulmonary stenosis protected this patient from pulmonary overcirculation as we can see in Eisenmenger syndrome, but multiple distal thromboemboli led to irreversible secondary pulmonary
arterial hypertension, and thus curative treatment of the Tetralogy of Fallot could not reasonably be performed. However, the postoperative course was uncomplicated, with the use of nitric oxide, sildenafil, and norepinephrine for 48 hours.

Pathological and bacterial analysis of the mass showed the presence of a recent fibrinocruoric thrombus with numerous colonies of Gram-positive coccoidal bacteria. Abiotrophia defectiva, a nutritionally variant streptococcus, was identified with complementary tests; therefore, antimicrobial therapy with Ampicillin was used for 6 weeks to treat this endocarditis. Anticoagulants were started immediately after the intervention and continued on a long-term basis because of the major risk of lethal recurrent massive thromboemboli and despite the risk of intracranial bleeding facilitated by the Tetralogy of Fallot.

The patient was discharged from the hospital at day 5 after surgical intervention. At 3 months after surgery, there had been no worsening of his symptoms. Pulmonary arterial thromboendarterectomy was not considered because of the septic nature of the distal lesions of the pulmonary arteries. The only surgical therapy feasible for this patient would have been a cardiopulmonary transplantation, which was not chosen at this time because of social considerations.

Tetralogy of Fallot was not diagnosed in this patient in his early years, and he was recently sent to France from Central Africa by the Chain of Hope because of the late appearance of clinical signs. Although no clinical sign of infection was noticed initially, computed tomography angiography images...
were able to demonstrate severe signs of pulmonary infection with multiple pulmonary mycotic aneurysms, pulmonary peripheral abscesses, and pulmonary arterial occlusions. Those lesions were secondary to pulmonary valve endocarditis, a rare complication of Tetralogy of Fallot. Multiple septic emboli from the initial septic thrombus were responsible for distal pulmonary occlusions; therefore, severe secondary isosystemic pulmonary hypertension precluded surgical curative treatment.

Disclosures
None.

References
Septic Pulmonary Thromboemboli in an Adolescent With Tetralogy of Fallot
Patrick Farahmand, Alban Redheuil, Sylvain Chauvaud, Jérôme Jouan, Amine Jemel and Jean-Noël Fabiani

Circulation. 2011;123:2164-2166
doi: 10.1161/CIRCULATIONAHA.110.991257
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2011 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/123/19/2164

Data Supplement (unedited) at:
http://circ.ahajournals.org/content/suppl/2011/05/16/123.19.2164.DC1

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/