prostheses to this population. Early results with the Freestyle are encouraging [3].

In this very challenging case, we implanted a stentless porcine xenograft in the SSV position in the youngest patient reported to date. The patient developed a fibrin sheath in the left ventricular outflow tract and Freestyle valve 7 months after initial implantation. The precise mechanism of fibrin deposition is unknown, although it may be related to turbulence from the ventricular septal fenestration, decreased leaflet excursion and leaflet washing in a small patient, or clopidogrel resistance in an infant [10]. A similar observation of fibrin-like deposition has been reported by Ohnaka and colleagues with a bioprosthetic valve 27 months after initial implantation in an adult patient [11].

The woven polyester cuff of the Freestyle is of variable height (Fig 1). Therefore, the prostheses must be rotated to facilitate coronary button implantation. For typical coronary anatomy, this involves a 180° rotation of the prosthesis to match the least-tall cuff with the posterior coronary. In most cases, the patient's left coronary button is implanted into the porcine root noncoronary sinus, and the patient's right coronary button is implanted anteriorly in the location of the porcine coronary.

In very small patients, the right pulmonary artery and left coronary artery must be mobilized to allow for unobstructed left coronary button implantation. Left ventricular outflow enlargement may be necessary as 19 mm is the smallest Freestyle currently available.

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Birt-Hogg-Dubé Syndrome in a Patient Presenting With Familial Spontaneous Pneumothorax

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Birt-Hogg-Dubé (BHD) syndrome is a recently discovered autosomal-dominant disease caused by a mutation in the folliculin gene. We report a patient with familial spontaneous pneumothorax who was found to have BHD syndrome. Patients with a personal and family history of pneumothoraces and computed tomographic (CT) findings of multiple pulmonary cysts should alert the thoracic surgeon to this syndrome; additional evaluation and testing may be warranted.

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Birt-Hogg-Dubé (BHD) is a genetic syndrome characterized by skin lesions, renal tumors, and recurrent pneumothoraces. Patients with a family history of pneumothoraces and suggestive CT findings should be referred for genetic testing and offered screening for the detection of renal neoplasms.

A 28-year-old man presented to our emergency department with a 1-day history of progressive dyspnea and a chest roentgenogram that demonstrated bilateral pneumothoraces (Fig 1). The patient was a nonsmoker and had no significant past medical history. However, his family history was notable for recurrent pneumothoraces affecting both his mother and his maternal grandmother.

A CT scan of the chest demonstrated several parenchymal cysts, which raised concern for BHD syndrome (Fig 2). The pneumothoraces were evacuated with bilateral pigtail catheters, and the patient was discharged home the following day. Further testing was conducted on an outpatient basis. His α_1-antitrypsin serum level was normal, as were anti-RNP, Ro, and La antibody levels. Abnormal sequence analysis of the folliculin (*FLCN*) gene was consistent with BHD syndrome. Given the association of BHD with renal neoplasms, the patient underwent renal ultrasonography, which was unremarkable.

Comment

BHD is an autosomal-dominant genetic syndrome that was first described in 1977 [1]. The initial report of the

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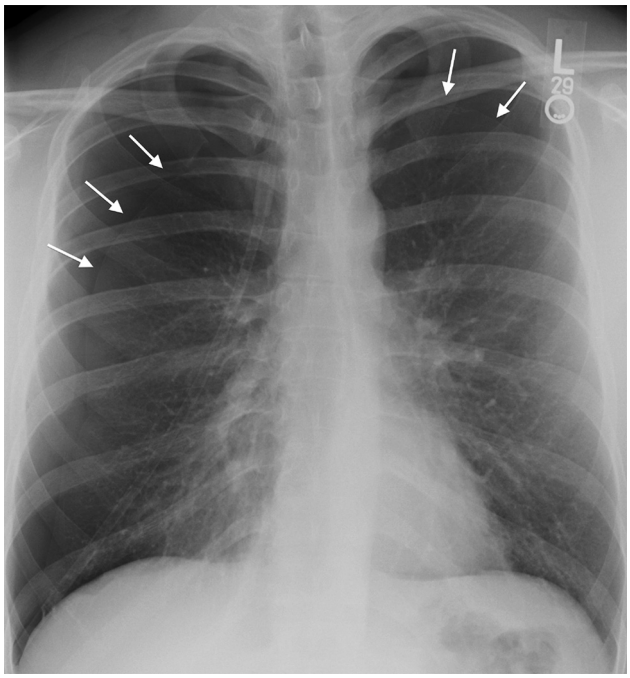

Fig 1. Chest roentgenogram revealing bilateral pneumothoraces (arrows).

syndrome described the classic dermatologic findings, such as fibrofolliculomas and trichodiscomas. These lesions usually manifest as small dome-shaped papules on the nose and cheeks. In 2001, the genetic locus of BHD syndrome was localized to chromosome 17, and further analysis identified a novel gene, *FLCN*, which encodes for the folliculin protein. Although folliculin has no known function, subsequent studies suggest it may play a role as a tumor suppressor.

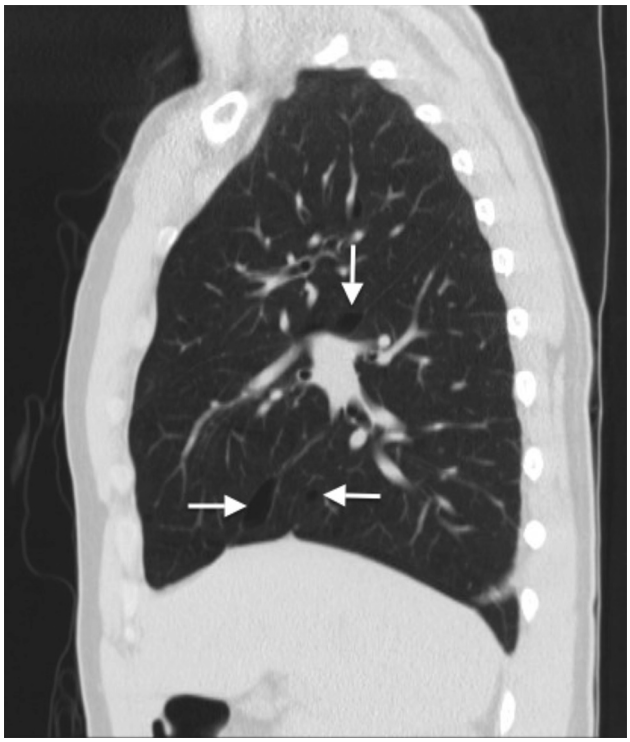

Fig 2. Sagittal computed tomographic (CT) image demonstrating numerous parenchymal cysts (arrows).

In addition to skin lesions, BHD syndrome is also associated with pulmonary cysts, recurrent pneumothoraces, and renal neoplasms. Up to 80% of patients with BHD syndrome will have multiple pulmonary cysts on CT scans [2]. These are typically small thin-walled cysts, are adjacent to the fissures, and often involve the lower lobes. Therefore, it is important to consider both α_1-antitrypsin deficiency and BHD syndrome in the differential diagnosis. These cysts are prone to rupture, and consequently a pneumothorax may be the initial presentation of this disease. Indeed, the risk of pneumothorax is estimated to be 50 times higher in patients with BHD syndrome. Recurrent pneumothoraces are common and have been reported in children as young as 7 years of age [3].

The most concerning feature of BHD syndrome is its association with renal cancer. The risk of renal cancer is estimated to be 7 times higher for those affected with BHD [4]. Additional studies of BHD families have calculated the prevalence of renal tumors to be 27%, with a median age of onset of 50 years [5]. A variety of histologic subtypes may develop, including clear cell, oncocytic, and papillary. Multifocal or bilateral tumors have been described, and patients have been diagnosed with metastatic disease as young as 27 years of age. In addition to renal cancer, it has also been suggested that colorectal polyps and invasive colon cancer are more prevalent in patients with BHD syndrome, although this has not been clearly established [6].

Although BHD may be readily diagnosed with sequence analysis of the *FLCN* gene, the diagnosis can be easily overlooked. The presentation of the condition is known to be highly variable. The classic skin manifestations may be absent, and renal tumors may present decades after a pneumothorax. For these reasons, it is important that thoracic surgeons have a high index of suspicion for this syndrome in the appropriate context. It is suggested that patients with recurrent pneumothoraces or a family history of pneumothorax undergo CT imaging of the chest [3]. The finding of multiple pulmonary cysts should prompt a referral for genetic testing. Other syndromes such as tuberous sclerosis may also present with skin and pulmonary findings and should be considered.

Establishing a diagnosis of BHD is important for several reasons. First, the diagnosis may impact the management of the pneumothorax. Because patients have multiple pulmonary cysts, it is unlikely that an apical wedge resection will be therapeutic. Simple drainage of the pneumothorax, perhaps with pleurodesis should be considered. Second, these patients should be advised to undergo regular surveillance for renal cancer. Although there are no established guidelines, it has been suggested that affected

individuals undergo annual magnetic resonance imaging examinations [7]. Surveillance may begin as early as 20 years of age and should continue for the lifetime of the individual. Finally, family members of the patient should be counseled and offered genetic screening.

In summary, thoracic surgeons should have a high index of suspicion for BHD syndrome. The initial presentation is often a pneumothorax, and unless further imaging is pursued, the condition may be misdiagnosed as a spontaneous primary pneumothorax. Sequence analysis of the *FLCN* gene is diagnostic, and those diagnosed with the syndrome should be referred for counseling and radiographic surveillance of the kidneys.

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Reoperative Lung Transplantation for Donor-Derived Pulmonary Mucormycosis

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A 64-year-old male with end-stage lung disease underwent right orthotopic lung transplantation. After doing well initially, he developed acute hypoxemic respiratory failure with allograft pneumonia. Donor operative cultures demonstrated mold of the *Mucor* species, which were corroborated by donor endobronchial cultures obtained near the right mainstem bronchial anastomosis. The patient was treated with reoperative bilateral orthotopic lung transplantation in combination with antifungal agents. The operation was performed successfully, using lungs donated after cardiac death and treated with ex vivo lung perfusion. The patient has recovered well, remaining on room air with good allograft function, without evidence of fungal disease.

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Pulmonary mucormycosis has an extraordinarily high mortality. The extant literature suggests that a combined strategy of extirpation and antifungal therapies yields the best possible outcomes. However, little is known about mucormycosis in lung allografts and whether pneumonectomy for extirpation with retransplantation is efficacious. Donor organ management can also be challenging in the setting of reoperative lung transplantation. Strategies to expand the donor organ pool may be required for these higher-risk patients to ensure timely retransplantation. Such strategies include the use of organs obtained in the setting of donation after cardiac death and screening for prospective allograft function using ex vivo lung perfusion.

A 64-year-old man with end-stage restrictive lung disease secondary to idiopathic pulmonary fibrosis and no other noteworthy medical or surgical history was referred to the University of Maryland Medical Center for lung transplantation evaluation. At referral, the patient required noninvasive support via a high-flow supplemental O_2 system with a fraction of inspired O_2 of 60%, with declining activity tolerance. The remainder of his pre–lung transplant evaluation was unremarkable, and he was deemed a suitable prospective lung transplant recipient.

Given the patient's disease process, age, and rapidly declining functional capacity, he was listed for single lung transplantation. A suitable donor lung with excellent gas exchange and mechanics was obtained from a healthy male donor; the history was noteworthy for the donor being found with his face in soil. However, donor bronchoscopy was satisfactory, with minimal secretions present. The patient underwent right orthotopic lung transplantation. Cardiopulmonary bypass, via cannulation of the ascending thoracic aorta and right atrium, was required because of exacerbated hypoxemia upon institution of single lung ventilation. The operation was technically satisfactory, with a total organ ischemic time less than 3 hours, and excellent completion arterial blood gases, pulmonary circulatory hemodynamics, transesophageal echocardiography, and flexible fiberoptic bronchoscopy. The patient was extubated within 24 hours following the operation, with 3 L of supplemental O_2 via nasal cannula within 48 hours and without any inotropic

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