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.

References

Birt-Hogg-Dubé Syndrome in a Patient Presenting With Familial Spontaneous Pneumothorax
Andrew Auerbach, MD, David H. Roberts, MD, Sidhu P. Gangadharan, MD, and Michael S. Kent, MD
Division of Thoracic Surgery and Interventional Pulmonology and Division of Pulmonary, Critical Care and Sleep Medicine, Beth Israel Deaconess Medical Center, Harvard Medical School, Boston, Massachusetts

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.


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 pulmonary opacification (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 α1-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 syndrome was in 1977, and a few years later, the gene responsible for BHD was identified in 2007 [2]. The authors emphasize the importance of genetic testing and screening for renal neoplasms in patients with BHD, as this condition is associated with a high risk of renal cell carcinoma. The case presented highlights the potential for recurrent pneumothoraces in patients with BHD, which underscores the need for early recognition and appropriate management.

Accepted for publication Sept 5, 2013.

Address correspondence to Dr Kent, 185 Pilgrim Rd, Deaconess 201, Boston, MA 02215; e-mail: mkent@bidmc.harvard.edu.

© 2014 by The Society of Thoracic Surgeons
Published by Elsevier Inc
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.

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 \( \alpha_1 \)-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...
Reoperative Lung Transplantation for Donor-Derived Pulmonary Mucormycosis

Keshava Rajagopal, MD, PhD, A. Claire Watkins, MD, Marc Gibber, MD, Zachary N. Kon, MD, Pablo G. Sanchez, MD, PhD, Aldo T. Iacono, MD, and Bartley P. Griffith, MD

Division of Cardiac Surgery, Department of Surgery and Division of Pulmonary/Critical Care Medicine, Department of Medicine, University of Maryland Medical Center and School of Medicine, Baltimore, Maryland

A 64-year-old male with end-stage lung disease underwent right orthotopic lung transplantation. After doing...