Technique of Coronary Sinus Plasty for Left Pulmonary Vein Stenosis

David Kalfa, MD, PhD, Wyman Lai, MD, Anne Ferris, MD, Usha Krishnan, MD, and Emile Bacha, MD

Department of Pediatric Cardiac Surgery, Morgan Stanley Children’s Hospital of New York, Columbia University, New York, New York

We describe a technique of coronary sinus plasty used to treat an infant with compression of the left pulmonary vein between the descending aorta and a dilated coronary sinus related to a persistent vena cava. The left lower pulmonary vein stenosis was relieved by repositioning the floor of the coronary sinus more anteriorly, toward the mitral annulus. The potential role played by a persistent left superior vena cava in some cases of left pulmonary vein stenosis is discussed.

Pulmonary vein stenosis (PVS) is a severe disease, whose cause and surgical management are still controversial and challenging. Many mechanisms have been proposed to explain primary or native PVS: abnormal incorporation of the common pulmonary vein (PV) into the left atrium [1], inflammatory process, and neoproliferation of myofibroblastic cells [2]. Current therapeutic options including sutureless repair, PV patch enlargement, endarterectomy, and angioplasty/stenting have not substantially improved the poor prognosis of this disease. We describe a technique of coronary sinus plasty used to treat left PVS related to compression by a dilated coronary sinus in a 1.6-year-old girl.

Technique

Through a ministernotomy, the patient underwent mild hypothermic bicaval cardiopulmonary bypass. The heart was retracted superiorly, and the severity of the left PV could be evaluated from outside the heart. In this particular anatomic form of PVS, the left PVs were in close contact with an enlarged coronary sinus related to a persistent left superior vena cava (LSVC). In this case, the left upper PV was found to be a long atretic cord. After cardioplegic arrest of the heart, the right atrium was opened and the LSVC was controlled and clamped intermittently. A vent was placed in the left atrium through the atrial septal defect (ASD) to preserve the right PV. The severity of PVS was assessed by direct inspection through the left atrium; the PVS was related to the coronary sinus, creating a bulbous roof over the vein. The left upper PV ostium was pinpoint and irretrievable.

The aim of the procedure was to anteriorly reposition the structure corresponding to the floor of the coronary sinus and the roof of the left PV. The floor of the coronary sinus was incised lengthwise parallel to the wall of the left atrium all the way until the level of the mitral valve annulus (Fig 1). This coronary sinus flap was then sutured back onto the free wall of the atrium but in a more anterior line, with use of a fine 6-0 polypropylene running suture, and thus helped to narrow the coronary sinus lumen lengthwise and enlarge the left PV ostium (Fig 2). The ASD was then closed with a 3-mm fenestrated autologous pericardial patch (Fig 3). This coronary sinus plasty could be associated to the resection of some luminal scar in the left PV lumen, if any. In this case, a minimal intrinsic luminal scar was found in the left lower PV lumen and was resected sharply.

Comment

We have successfully used this technique to treat a 1.6-year-old girl, ex-premature (birth weight 600 mg), presenting with a history of worsening shortness of breath for several months, requiring oxygen. Ultrasoundography showed left upper PV atresia, left lower PV stenosis with a mean gradient of 8 mm Hg, a predominant LSVC with an enormous coronary sinus draining into the right atrium, and a large ASD with a bidirectional shunt. The left lower PV narrowing and flow acceleration started at the level of the compression between the LSVC/coronary sinus and the descending aorta. The right pulmonary veins were normal. Echocardiography, cardiac catheterization, and ventilation/perfusion scanning demonstrated a pulmonary artery pressure above 80% of systemic, with marked favoring of the right lung (63%) over the left lung (37%), with an extreme flow deficit in the left upper lobe (2%). Magnetic resonance imaging showed a “vice effect” with compression of the left lower PV in between the descending aorta and the large coronary sinus (Fig 4). This patient
was successfully treated by a sinus coronary plasty as just described. Postoperative transesophageal echocardiography showed an enlarged left lower PV lumen with a mean gradient reduced from 8 mm Hg to 2 mm Hg. The pulmonary artery pressure decreased from 80% to 50% postoperatively. The patient was extubated on day 1 after the operation and was discharged from the hospital on day 4 with a mean gradient across the left lower PV of 1 mm Hg. At the 6-month follow-up visit, the patient was asymptomatic, required no oxygen, and had significantly decreased pulmonary hypertension and no recurrent stenosis on the left lower PV on echocardiography.

We describe here a simple and reproducible technique of coronary sinus plasty used to treat left PVS related to compression by a dilated coronary sinus. The preoperative and peroperative understanding of the mechanism of this left PVS allowed is to guide the surgical repair and relieve this unusual form of left PVS by reducing and repositioning the floor of the coronary sinus. The indications for this surgical technique are limited to this particular anatomic form of left PVS related to a dilated coronary sinus.

Numerous pathologic and surgical findings can support the hypothesis of a role played by an LSVC in the...
pathogenesis of left intra-atrial obstruction and thus the rationale for such a surgical technique. Persistent LSVC can be a congenital cause of supramitral stenosis in relation to a dilated coronary sinus [3] and frequently coexists with cor triatriatum [4]. Gharagozloo and colleagues [5] described in an autopsy series of cor triatriatum a consistent relationship of the LSVC to the insertion of the intra-atrial membrane. Ascunto and colleagues [6] reported similar cases in which a LSVC joining a dilated coronary sinus divided the left atrium into two chambers. From an embryologic point of view, a portion of the left anterior cardinal vein (future LSVC) is pulled adjacent to the posterior wall of the left atrium during the migration of the sinus venosus to separate the left atrium from the systemic veins [7]. If the left anterior cardinal vein does not obliterate, it could compress the posterior wall of the developing left atrium and thus alter the development of the pulmonary veins.

Such a hypothesis involving a persistent LSVC in the development of left intra-atrial obstruction might be extended to the pathogenesis of left pulmonary vein stenosis, all the more because cor triatriatum is considered to be related to stenosis of the common pulmonary vein. The case and surgical technique described here support this concept by demonstrating the proximity of these two structures and the feasibility of relieving the pulmonary vein stenosis by coronary sinus plasty. Although the results of this surgical technique are encouraging, long-term follow-up is needed to assess the risk of recurrent PV stenosis.

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References