Diagnostic and Surgical Challenges of Pulmonary Venous Stenosis

Pulmoner Venöz Stenozun Tanısal ve Cerrahi Zorlukları

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Pulmonary venous stenosis (PVS) is a rare and progressive foetal disease[(1)]. It is categorised into congenital and acquired forms. Acquired PVS occurring due to atrial fibrillation (AF) ablation and after surgical correction of pulmonary venous return anomalies has been frequently described, but limited information is available regarding congenital PVS[(2)]. The incidence of the disease is 0.4% in patients with congenital heart disease, whereas that of acquired PVS is 10%-15% after TAPVD correction and as high as 30%-40% after AF ablation[(1,2)].

Although the specific pathophysiology of patients with normal venous return remains unknown, it is thought to develop due to myofibroblast-like proliferation of intimal cells[(3)]. PVS is emphasised in patients with congenital heart diseases such as prematurity; bronchopulmonary disease; Down syndrome; patent ductus arteriosus with left-right shunt and increased pulmonary blood flow such as in patients with atrial septal defect, ventricular septal defect and atrioventricular septal defect[(1-3)].

The current pattern of pulmonary venous flow classically can be evaluated using transthoracic echocardiography, cardiac catheterisation, computed tomography and magnetic resonance imaging[(3,4)]. In echocardiography, pulmonary venous flow without stenosis should be phasic and the gradient should not be seen. In the spectral Doppler examination, continuous turbulent flow and computation of mean gradient over 5 mmHg is significant for stenosis[(4)]. The assessment of right ventricular and pulmonary arterial pressures is also valuable. In addition, when PVS is present, it is known that the flow in lung segments will be redistributed. Therefore, it may not be possible to obtain a gradient value that is directly proportional to stenosis. Catheter angiography can be used for both diagnostic and therapeutic purposes. Stenosis can be visualised using angiography, and balloon angioplasty and/or stent implantation can be performed if appropriate. During the evaluation of PVS, which is a rare disease with poor prognosis, until the diagnosis is complete and sufficient data for intervention is obtained, continuing the examination may increase treatment effectiveness.

Treatment of PVS can be divided into four main categories, i.e., medical, catheter, surgery and transplantation, depending on the aetiology of the stenosis. The most remarkable data in medical treatment are the adjuvant chemotherapy protocol with bevacizumab and imatinib mesylate drugs, and the research is still ongoing. Catheter angiography seems advantageous because it can be used for both diagnosis and treatment. However, the results of balloon angioplasty and stent implantation in stenosis patients were worse than expected in a study, and stenosis tended to recur within a short time after stent implantation because of tissue proliferation[(4,5)].

The surgical procedures include left atrioplasty, pericardial patch plasty, unroofing and seamless repair. In the left atrioplasty technique, the appendix of the atrium is anastomosed to the after stenotic segment of the pulmonary vein and can be repaired using the appendix as a flap[(5)]. Pericardial tissue can be used to expand the stenotic segment as an autologous or xenograft after preparation. Proliferating tissue that causes stenosis can be resected at the level of the vein roof by reaching pulmonary venules through left atriotomy. For this reason, this technique is referred to as the 'unroofing' procedure. The success of all these techniques has

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been limited in accordance with histopathology of the pulmonary venous system. For this reason, using seamless technique is a controversial issue. Because there is no suture line in the pulmonary venules, it is believed that trauma and trauma-related proliferative response will be minimised using this technique. Patients in the terminal stage of pulmonary venous disease can be referred for lung transplantation depending on phrenic nerve damage, laryngeal nerve damage, the procedure applied and right ventricular function[4,5].

PVS is a progressive disease or a complication that causes irreversible injury due to diagnostic delay and misdiagnosis owing to its non-specific symptoms. Because they are rarely seen, the disease is difficult to diagnose and is progressive, with limited success rates observed with medical and surgical treatments; thus, clinical management of PVS is challenging. Although outcomes are considered to improve with specific surgical techniques and treatments, there is need for research on PVS to find advanced techniques for its management.

REFERENCES

1. Holt DB, Moller JH, Larson S, Johnson MC. Primary pulmonary vein stenosis. Am J Cardiol 2007;99:568-72.
2. Latson LA, Prieto LR. Congenital and acquired pulmonary vein stenosis. Circulation 2007;115:103-8.
3. Riedlinger WF, Juraszek AL, Jenkins KJ, Nugent AW, Balasubramanian S, Calicchio ML, et al. Pulmonary vein stenosis: expression of receptor tyrosine kinases by lesional cells. Cardiovasc Pathol 2006;15:91-9.
4. Lo Rito M, Gazzaz T, Wilder TJ, Vanderlaan RD, Van Arsdell GS, Honjo O, et al. Pulmonary vein stenosis: Severity and location predict survival after surgical repair. J Thorac Cardiovasc Surg 2015;151:657-66.
5. Seale AN, Uemura H, Webber SA, Partridge J, Roughton M, Ho SY, et al. Total anomalous pulmonary venous connection: outcome of postoperative pulmonary venous obstruction. J Thorac Cardiovasc Surg 2013;145:1255-62.