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Indication of Interventional Procedure for Isolated Syncopal Pulmonary Stenosis in Adolescents: Case Reports and Revue of Literature

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ABSTRACT

Pulmonic stenosis refers to a dynamic or fixed anatomic obstruction to flow from the right ventricle to the pulmonary arterial vasculature. Although most commonly diagnosed and treated in the pediatric population, some patients with severe, isolated PS may be diagnosed for the first time as adults, and represents approximately 10% of all congenital heart disease. We report the cases of three adolescents, who presented to the ER after having several episodes of syncopes during exercise. The TTE showed a dome-shaped pulmonary valve, with a peak pulmonary velocity between 5,3 and 6,8 m/s, with a right ventricular hypertrophy and dilatation. We proceed to the balloon valvuloplasty and used balloon dilatation catheter until the waisting disappeared. An echocardiography was performed the day after the procedure, showing a drop peak pulmonary velocity and decreased right ventricular dilatation.

In older children and adults, percutaneous pulmonary valvuloplasty is the first line treatment for valvar pulmonary stenosis irrespective of symptoms. Catheter-based balloon valvotomy is recommended for patients with non-dysplastic valvular PS and with peripheral PS. Surgery is recommended for patients with sub-infundibular or infundibular PS and hypoplastic pulmonary annulus, with dysplastic pulmonary valves, and for patients with associated lesions which need a surgical approach, such as severe pulmonary regurgitation or severe tricuspide regurgitation.

Simple balloon dilatation can provide a definitive solution and avoid the need for surgery.

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KEYWORDS: Pulmonary stenosis; Adult Congenital Heart Disease; Interventional Cardiology

INTRODUCTION

Pulmonary stenosis refers to a dynamic or fixed anatomic obstruction to flow from the right ventricle to the pulmonary arterial vasculature, which leads to right ventricle pressure overload that in turn causes increased contractility and dilation and results in increased wall stress and compensatory right ventricle hypertrophy. Although most commonly diagnosed and treated in the pediatric population, some patients with severe, isolated pulmonary stenosis may be diagnosed for the first time as adults. Isolated valvular pulmonary stenosis comprises approximately 10% of all congenital heart disease.

CASE REPORT

We report the case of a 15, 17 and 18 years old adolescents, who presented to the ER after having several episodes of syncope during exercise. At the physical exam there was a systolic precordial murmur with a crescendodecrescendo quality heard loudest at the left upper sternal border without radiation to the carotid arteries and without hepatosplenomegaly or subcutaneous edema. Electrocardiogram revealed in our three patients right bundle branch block and right axis deviation consistent with right ventricular hypertrophy. The transthoracic echocardiography showed a dome-shaped pulmonary valve, with a peak pulmonary velocity between 5, 3 and 6,8 m/s, and a peak pulmonary gradient between 112 and 185 mmHg, with a right ventricular hypertrophy, dilatation and dysfunction, and moderate tricuspid regurgitation. We proceed to the pulmonary balloon dilatation and started with the puncture site which was the right femoral vein. Heart rate, blood pressure, respirations, and pulse oximetry were continuously monitored throughout the procedure.

A 7 Fr sheath was inserted into the vein and used a 7 Fr Berman angiographic catheter to acquire hemodynamic and angiographic data. The pressure of the right atrium was

around 15 mmHg, and the right ventricle peak systolic pressure was between 95 to 129 mmHg in our three patients, which was ⩾ 75% of the peak systolic systemic pressure, and therefore significant. We did an exchange of catheter to the right coronary artery catheter after leaving the guidewire in the left pulmonary artery, to obtain the mean pulmonary valvar gradients, which were around 13 mmHg. We choose balloon dilatation catheter Tyshak II and MedZ with a length between 3 and 4 cm, and the diameter was chosen based on the patients annulus, and inflated at 5 atm, two times each, during 10 secondes, until the waitline disappeared, and performed additional balloon inflation after, to ensure adequate valvuloplasty. We finished with the measurement of pressures in the right ventricule which were between 55 to 88 mmHg in our three patients and gradient across the pulmonary valve so that the guidewire is left in its place across the pulmonary valve while the results of valvuloplasty are evaluated. Then we placed again the Berman angiographic catheter in the right ventricular apex and an angiogram was performed (Figure 1) An echo was performed the day after the procedure, showing a drop of mean peak to peak pulmonary gradient to 25 and 65 mmHg with maximum velocity of 2,5 and 3,8 m/s, a reduction of the right ventricle dilatation and therefore decreased tricuspid regurgitation.

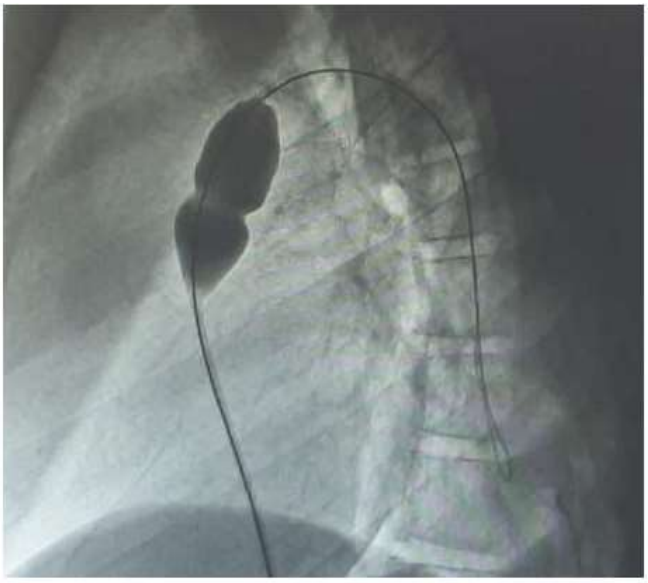

Figure 1: RV angiography showing pulmonary dilation balloon during inflation showing the waistline

DISCUSSION

Pulmonary stenosis, or more generally, right ventricular outflow track (RVOT) obstruction, is an anatomic blockage to right ventricular output. It's most commonly the result of a congenital heart defect, associated with Alagille syndrome, Rubella syndrome, foetal endocarditis …. Valvar pulmonary stenosis was first described in 1761 by Morgagni in his classic text 'De Sedibus et Causis Morborum'. He reported a patient with pulmonary stenosis and atrial septal defect who had been cyanosed during life [1]. Just over two hundred years late Kan and colleagues reported the first case of percutaneous balloon pulmonary valvuloplasty in the New England Journal of Medicine revolutionizing the management of this common congenital condition [2]. Beyond the neonatal period, most cases of valvar pulmonary stenosis are asymptomatic as the right ventricle and atrium compensate to maintain resting cardiac output. Typically symptoms, when they do emerge, are exertional and include breathlessness and fatigue. Rarely chestpain, syncope, and sudden death occur. [3] Echocardiography permits prior visualization of the nature of the valvar stenosis and assessment of the peak instantaneous systolic gradient. 36 mmHg or less is considered mild, 36 to 64 mmHg moderate, and greater than 64 mmHg severe. [4]

Indications for balloon pulmonary valvuloplasty and surgery in adults are a valvar pulmonary stenosis with an echocardiographically assessed transvalvar gradient of > 64 mmHg. If balloon valvulopalsty is ineffective, surgery should be considered in asymptomatic patients with a transvalvar gradient gradient of >80mmHg. Balloon valvuloplasty can be considered in patients with a transvalvar gradient of > 64mmHg and - symptoms related to valvar pulmonary stenosis - impaired right ventricular function - double-chambered right ventricle - relevant arrhythmias - right to left intracardiac shunting. In older children and adults, percutaneous pulmonary valvuloplasty is the first line treatment for isolated valvar pulmonary stenosis in all those with an invasive gradient across the pulmonary valve exceeding 40mmHg, irrespective of symptoms. [6] Catheter-based balloon valvotomy is recommended for patients with nondysplastic valvular PS and with peripheral PS (often with stent implantation). Surgery is recommended for patients with sub-infundibular or infundibular PS and hypoplastic pulmonary annulus, with dysplastic pulmonary valves, and for patients with associated lesions which need a surgical approach, such as severe pulmonary regurgitation or severe tricuspide regurgitation [5] Transcatheter therapy of valvar pulmonary stenosis is one of the first, if not the first, catheter interventions which have facilitated the application of catheter interventional technology to children, so that many children have benefited from less invasive treatment for structural congenital heart defects.

CONCLUSION

The interventional treatment of isolated pulmonary valve stenosis is one of the most rewarding procedures and the first treatment in line. Even in critical neonatal pulmonary stenosis, simple balloon dilatation can provide a definitive solution and avoid the need for surgery. Excellent results can be obtained if the procedure is performed with care by the interventionalist.

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