



Case Report

Isolated infundibular pulmonary stenosis. A case report

K El ouahabi*, N Erraboun, A Bouzerda, M Raissouni, I Asfalou, A Benyass, Z Lakhal, Y Moutakillah, M Ait Houssa and E Zbir

Department of Cardiology, Hôpital Militaire d'Instruction Mohamed V, Rabat, Morocco

Received: 29 May, 2020

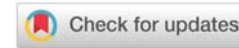
Accepted: 10 June, 2020

Published: 12 June, 2020

*Corresponding author: Kawtar El ouahabi, Department of Cardiology, Faculty of Medicine and Pharmacy, Rabat, Morocco, Tel: +212662090329; E-mail: kawtar-elou@hotmail.com

Keywords: Isolated infundibular stenosis; Fibromuscular hypertrophy; Infundibulectomy

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Abstract

Introduction: Infundibular pulmonary stenosis is one of the numerous pathologic features of the right tract (from the right ventricular outflow tract to the peripheral pulmonary arteries) occurring at the infundibular level. It is usually associated with ventricular septal defect or part of tetralogy of Fallot, but rarely isolated.

Methods and results: We report here a case of an adult with isolated infundibular stenosis, and discuss the relative frequency, clinical manifestations, the diagnosis methods, and the results of surgical correction of this lesion.

Conclusion: Infundibular pulmonic stenosis with intact ventricular septum is a rare presentation of pulmonary stenosis. Its early detection with a comprehensive examination of the right heart before right ventricular failure is rewarding since it usually can be treated well with a good guidance of surgical management, and low surgical risk.

Abbreviations

RV: Right Ventricle; ECG: Electrocardiogram; RA: Right Atrium; TTE: Transthoracic Echocardiography; IS: Infundibular Stenosis; TR: Tricuspid Regurgitation; PA: Pulmonary Artery; PV: Pulmonary Valve; RVOT: Right Ventricular Outflow Tract; PR: Pulmonary Regurgitation; MRI: Magnetic Resonance Imaging; CT: Computerized Tomography; PS: Pulmonary Stenosis; 2D: 2 Dimensions; 3D: 3 Dimensions

Introduction

Obstruction to the outflow of blood from right ventricle may occur as a result of congenital stenosis of the infundibulum which leads to RV hypertrophy. Isolated infundibular pulmonary stenosis with intact ventricular septum occurs in several patients but still an uncommon and rare cardiac abnormality [1]. Early surgical treatment corresponding to an excision of the infundibular muscle band can lead to improvement in functional class and a substantial decrease or normalization of RV pressure [2].

Case presentation

A 25 years old female with no medical history was admitted

in our department with complaints of chest heaviness and shortness of breath with two episodes of faintness for one year. On examination, the patient was of average built. A systolic thrill was found over the pulmonary area. First heart sound was normal with a splitting of second heart sound. There was a rough ejection systolic murmur best heard in pulmonary area patient bending forward (grade 5/6) without irradiation. The murmur increases immediately at the end of the Valsalva and during inspiration. Lungs were clear with no added sound. The patient was eupneic with an oxygen hemoglobin saturation of 97% at ambient air. There were no clinical signs of acute right heart failure. Other finding was an important hepatomegaly with a liver span of 16 cm.

The electrocardiogram showed a regular heart rate at 87 beats/min, a right axis deviation and RV hypertrophy with strain pattern. Chest X ray showed a cardiomegaly at the expense of the RV hypertrophy and a right atrium enlargement.

The transthoracic echocardiography showed a severe infundibular stenosis (peak pressure gradient at 95 mmHg, peak velocity at 4,9 m/s), corresponding to an infundibular muscle band, a marked concentric RV hypertrophy (maximum width 18mm) with slightly altered systolic function and a

paradoxical septal motion. Other findings comprise an RV pressure estimated at 77 mmHg, a mild to moderate tricuspid regurgitation and a dilated RA (Figures 1-4).

Cardiac MRI helped to visualize the exact location of the stenosis at the infundibulum and confirmed the TTE findings. Coronary CT angiography did not show any abnormality of coronary birth or signs of associated congenital heart disease.

The abdominal ultrasound exam showed a homogeneous liver with regular edges and dilated hepatic veins.

The patient was operated under general anesthesia. After median sternotomy and thymus dissection, pericardiotomy was done. The heart was globally dilated, mostly the right cavities with an important RV hypertrophy and an ectasic RA. The pulmonary artery looked normal while there was a manifest depression at the infundibulum. The right coronary artery was of large caliber, giving birth to an infundibular branch of medium size. This branch was sacrificed with no consequences since it was only responsible for the vascularization of the constrictive muscle band. The left anterior descending coronary artery was normal.

Cardiopulmonary bypass was established with bicaval cannulation and aortic cannulation without clamping. Vertical infundibulotomy expanded to the wall of the RV was done respecting the pulmonary valve. This one was found normal with gross hypertrophy of the RV wall and the infundibular muscle band. The stenosis admitted only a small 10 Fr suction probe. Total excision of the stenosis and the infundibular muscle bands that block the virtual right ventricular outflow tract was done. The integrity of the sub valvular tricuspid apparatus and the interventricular septum was respected. An enlargement of the RVOT with a large autologous pericardic patch sewn on the banks of the incision was done, with a good result (Figures 5-8).

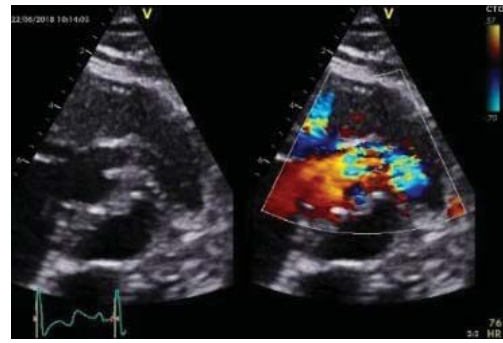


Figure 3: parasternal short view at the RVOT showing an aliasing at the infundibulum under the pulmonary valve.

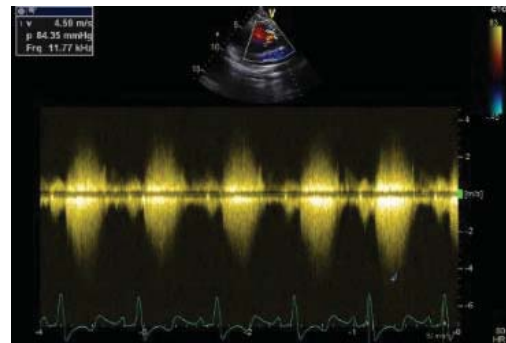


Figure 4: Continuous doppler showing a severe IS (peak pressure gradient at 95 mmHg, peak velocity at 4,9m/s).



Figure 5: First view of the heart after pericardiotomy showing an important RV hypertrophy, an ectasic RA, and a depression at the infundibulum.



Figure 6: The stenosis admitted only a small 10 Fr suction probe.



Figure 7: Infundibulectomy and reconstruction of the RVOT with a pericardic patch



Figure 1: Important RV hypertrophy shown in the parasternal long and short views.



Figure 2: Important RV hypertrophy shown in the parasternal long and short views.



Figure 8: The stenosis was due to a muscular fibrotic band.

Progressive weaning of the cardiopulmonary bypass was done under low doses of dobutamine and decannulation. After achieving hemostasis chest was closed leaving three drains (among which one was left pleural). Total bypass time 95 min.

The patient was shifted to the intensive care unit with minimum inotropic support. She was extubated on the same day, shifted to general ward on the 2nd post-operative day and discharged home on the 14th postoperative day. The post-operative TTE showed a mild pulmonary regurgitation. The patient remains asymptomatic on the subsequent follow up. She is on minimum medication and leading a normal life.

Discussion

Pulmonary stenosis can be valvular (the most common form), sub valvular (infundibular or sub infundibular) or supralvalvular. IS may occur in a variety of pathological conditions and is usually seen as part of Tetralogy of Fallot while isolated IS of primary origin is exceedingly rare with a reported incidence of 0.4% of patients with congenital heart disease [2-4].

Other congenital anomalies associated with isolated IS are described such as single coronary artery, calcific aortic stenosis, or main pulmonary artery aneurysm [5].

IS is caused by arrest of the normal involution of the bulbus cordis [4]. Two types of IS are identified: (1) Double chambered right ventricle caused by fibrous anomalous muscle bundles at the line of the junction of the RV and the infundibulum, producing a stenosis of the outflow cavity and dividing it in two. The infundibulum may be enormously dilated above the constriction [4,6]. (2) The infundibulum becomes shrunken and fusiform due to its thick muscular walls. However, in this type there can be a wide variation in the anatomy of the IS depending on the abnormal position of the bands.

Infundibular hypertrophy may be secondary and occur in various cases [6] such as PS where the reactive RV hypertrophy can cause a dynamic outflow obstruction, or in patients with hypertrophic cardiomyopathy, protrusion of right sinus of Valsalva into the RVOT, aneurysm of membranous ventricular septum, and intra or extracardiac mass lesions in the RVOT such as sarcoma [2].

IS remains well tolerated for long time. The severity of stenosis can progress with age; however, it is not well correlated to clinical manifestations. Frequently, patients with a significant stenosis are totally asymptomatic, and are referred

for advanced check up in the presence of a heart murmur, chest X-ray abnormality or electric signs of RV hypertrophy. Exertional dyspnea, fatigue and precordial pain are the most common symptoms. Exertion may provoke faintness, syncope or even death. Epigastric pain is often present.

Physical findings are systolic ejection murmur at left sternal border and RV thrill. The ejection click helps to distinguish between valvular and IS. In this case, the patient had been symptomatic with chest heaviness and shortness of breath for last 1 year and was first diagnosed in her late 25 years of age because no intracardiac shunt was present.

In patients with mild PS the ECG can be normal or show a more pronounced right-ward axis deviation which is normal in children and adolescents. In severe stenosis, ECG shows signs of RV hypertrophy (high R wave amplitude in lead V₁, deep S waves in the left precordial leads, with an R:S ratio <1 in V₆), further rightward axis deviation, or heart rhythm disorder such as atrial fibrillation or flutter.

TTE and Doppler imaging are the gold standard to detect IS and quantify its severity [6]. PV is fine and flexible. Peak and mean gradients are measured. Isolated IS is usually suspected when there is a mosaic color flow in RV outlet, flow directed away from the transducer and neither thickening nor prominent a-wave of PV. TTE makes a quantitative assessment of RV volume and function with the 2D and the 3D modes which estimates RV volume and ejection fraction, but it has limitations due to a limited acoustic window. Qualitative assessment estimates dilatation and deterioration of function over time. Routine examination includes measurements of RV inflow and outflow dimensions, assessment of the tricuspid and pulmonary valve, long and short axis dimensions, tricuspid annular plane systolic excursion, and fractional area change. The determination of the regurgitant volume in PR is important after a prior intervention.

MRI is useful for studying the anatomy of the RVOT, PA and its branches. It helps to distinguish between sub valvular, valvular and supralvalvular stenosis since it visualizes the precise location of the stenosis. Flow volumes can be accurately assessed by MRI while flow velocities are determined by TTE. This is especially useful for determination of the regurgitant volume in PR after a prior intervention. MRI is considered the gold standard for assessment and quantification of RV, mass, and function. It is not bothered by the geometric assumptions that are unavoidable in TTE. Multislice CT might be the superior diagnostic technique to study the anatomy of the RVOT and the PA tree especially, but it lacks possibilities for volumetric and functional assessment.

In this case the diagnosis of IS was made easily since all its signs were present: mosaic color flow in RV outlet, with supple, thin PV, intact ventricular septum, and no other causes of RV hypertrophy. The diagnosis was thereafter confirmed with cardiac MRI and during the surgical treatment.

The goal of treatment is to relief the RVOT obstruction, and so the pressure overloaded by an excision of the hypertrophied

muscular band. Surgical treatment is more successful when the obstruction is muscular and only involve a short segment of the outflow tract [1]. Indications for surgical intervention varies according to authors. Any obstruction in the RVOT and/ or PV with a Doppler derived peak instantaneous gradient >64 mm Hg (peak velocity >4 m/s) should be repaired, or if the stenosis is severe with gradients exceeding 100mmHg with or without symptoms such as shortness of breath, cyanosis and chest pain [1,6]. The clinical presentation of our patient and the TTE data meets the criteria described above. The procedure corresponds to an infundibulectomy associated or not to tricuspid annulorraphy or tricuspid valve replacement; or RV reconstruction according to the anatomy and the lesions found [2].

In our case, IS is of primary origin since no other causes were found during imaging evaluation and surgery exploration, the patient was operated under a cardiopulmonary bypass with a beating heart since the RV tolerates badly ischemia. We have done an excision of pulmonary infundibular muscle band with a reconstruction of the RVOT using a pericardic patch which is described in the literature [2].

Mortality rate in surgically repaired cases of IS have decreased due to improved surgical technique and post-operative care systems [2,7]. Symptomatically, patients are improved, and the exercise tolerance is increased. On physical examination, the thrill tends to disappear in all patients and the murmur to decrease in intensity. The ECG may show a significant diminution in the degree of RV hypertrophy. Postoperative PR is common and well tolerated in patients following valvulotomy for valvular pulmonary stenosis. However, it can lead to irreversible RV damage and thus should be followed carefully with regular echocardiographic imaging and ECG, checking for RV pressure, size and function, degree of PR and TR as well as the occurrence of arrhythmias [7].

Our patient showed a good response. She remains asymptomatic during the 3 and 6 months follow up after surgery while physical examination shows a slight murmur of PR with no hemodynamic repercussion in the TTE checkup.

Conclusion

Infundibular pulmonic stenosis with intact ventricular septum is a rare presentation of pulmonary stenosis. Its early detection with a comprehensive examination of the right heart before right ventricular failure is rewarding since it usually can be treated well with a good guidance of surgical management, and low surgical risk. Nevertheless, patients can develop residual lesions that require lifelong cardiological follow-up.

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Citation: El ouahabi K, Erraboun N, Bouzerda A, Raissouni M, Asfalou I, et al. (2020) Isolated infundibular pulmonary stenosis. A case report. *J Cardiovasc Med Cardiol* 7(2): 199-202. DOI: <https://dx.doi.org/10.17352/2455-2976.000137>