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A Woman with Double Chamber Right Ventricle

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ABSTRACT

An additional membrane or muscle band inside right ventricle divides it into two chambers, the proximal and distal one. It is a rare congenital malformation which makes up approximately 0.5-1 per cent of all congenital heart defects (CHD). The double chambers right ventricle (DCRV) most often present in children but rarely in adults. Transesophageal Echocardiography (TOE) is the most effective tool in diagnosing a DCRV. A 36 years old woman, diagnosed with large ventricular septal defect (VSD) by transthoracic echocardiography (TTE), underwent a TOE to complement the preparation of surgical closure. The TOE finding instead revealed a septum or membrane that divide the right ventricle into two separate chambers, later confirmed by a cardiac computer tomographic scan (Cardiac CT). The patient underwent successful surgical excision of the membrane with VSD closure to alleviate symptoms and improve the quality of life. We present this case in order to emphasize the rarity and management of this congenital heart disease.

INTISARI

Sebuah membran atau otot tambahan di dalam ventrikel kanan membagi menjadi dua ruang, yaitu proksimal dan distal. Kelainan bawaan ini termasuk langka, yang membentuk sekitar 0,5-1 persen dari semua penyakit jantung bawaan. Double Chamber Right Ventricle (DCRV) paling sering ditemukan pada anak-anak tetapi jarang pada orang dewasa. Ekokardiografi transesofageal (TOE) adalah alat yang paling efektif dalam mendiagnosis DCRV. Seorang wanita berusia 36 tahun, didiagnosis dengan defek septum ventrikel besar (VSD) oleh ekokardiografi transtorakal (TTE), menjalani TOE untuk melengkapi persiapan penutupan VSD secara bedah. Temuan TOE menunjukkan terdapat septum atau membran yang membagi ventrikel kanan menjadi dua ruang terpisah, yang kemudian dikonfirmasi dengan pemindaian tomografi komputer jantung (CT scan jantung). Pasien berhasil menjalani operasi eksisi membran dengan penutupan VSD untuk mengurangi gejala dan meningkatkan kualitas hidup. Kami menyajikan kasus ini untuk menunjukkan kelangkaan dan pengelolaan terhadap penyakit jantung bawaan ini.

Introduction

Double-chambered right ventricle (DCRV) is an anomaly in the right ventricle that divides the structure of the right ventricle into two parts namely the proximal and distal parts.[1] The proximal part has a higher pressure than the distal part with a pressure difference of more than 20 mmHg.[2] DCRV can be caused by moderator band anomaly and hypertrophy of the right ventricular muscle trabecular tissue. This disorder is often associated with other congenital anomalies such as ventricular septal defect, tetralogy of Fallot (ToF), Ebstein anomaly, and double outlet right ventricle (DORV). In moderator band anomalies, DCRV can be classified as a very rare congenital anomaly, occurring in approximately 0.5-1 percent of patients with congenital heart abnormalities, but can also be classified as acquired anomalies in progressive hypertrophy in the supraventricular crest of right ventricle.[3]

Most DCRV patients are diagnosed in childhood to adolescence before the age of 20 years. Diagnosis of DCRV patients in adulthood is more difficult and is often diagnosed as pulmonary stenosis depending on the severity of right ventricular outflow tract (RVOT) obstruction. Patients can feel asymptomatic or with

symptoms of shortness when on the move, angina, syncope, or dizziness. Physical examination usually shows systolic heart noise in the lower heart border area. The most effective investigation in diagnosing DCRV is transesophageal echocardiography (TOE).[2] This case report will discuss the diagnosis and management of patients with DCRV.

Case Presentation

A 36-year-old woman who has been diagnosed with a ventricular septal defect (VSD) accompanied by an infundibular type pulmonary stenosis goes to a cardiac clinic for routine control. The patient was diagnosed with VSD and pulmonary stenosis when giving birth to a child 7 years before. When pregnant at the time, the patient complained of heavy shortness of breath when she was active and there was noise. Patients performed echocardiography with VSD results and infundibular pulmonary stenosis. The patient successfully delivered spontaneously with shortens second stage labor and then routine control in cardiac clinic without therapy.

Patients come without significant complaints, but feel shortness of breath when on the move moderate-heavy. Other complaints such as sleeping with more than one pillow, orthopnea, swelling in the extremities, and chest pain are not present. Physical examination showed that the patient's condition was compost mentis, blood pressure 131/83 mmHg, pulse rate 84 times per minute, respiration rate 16 times per minute, and temperature of 36.8 degrees Celsius. A physical examination of the heart revealed heart sounds 1 and 2 within normal limits, there was a pansystolic noise 3/6 with a maximum punctum in the intercostal spaces of 3-4 left sternal borders without cardiomegaly. The patient is then referred for echocardiography examination before the surgical conference with the possibility of closing the ventricular septal defect and resection of infundibular pulmonary stenosis.

Transthoracal echocardiography examination showed the results of perimembranous outlet (PMO) VSD with left-to-right shunts of 0.7-0.8 cm in diameter, pressure gradient of 100 mmHg, visible bulkhead at 1/3 distal RV, turbulence (+) with pressure gradient of 130 mmHg between the two walls, left ventricular ejection fraction good 58%, normal left ventricular diastolic function, decreased ventricular systolic function (tricuspid annular plane systolic excursion, TAPSE 15 mm), moderate tricuspid regurgitation with tricuspid valve gradient (TVG) 107 mmHg , high probability of pulmonary hypertension with tricuspid regurgitation velocity (TRV) 5.16 m / s, mild aortic regurgitation, and RV dilatation. Because of the suspicion of a divider dividing the right ventricle into the proximal and distal parts, the patient underwent a trans esophageal echocardiography (TOE) procedure.

Transesophageal echocardiography examination showed VSD PMO size 0.5-0.7 cm, left to right shunt with a pressure gradient of 100 mmHg. The pseudoseptum is visible at 1/3 distal right ventricle (RV) with a pressure between 126 mmHg. Figure 1 showed the TOE result. There are no abnormalities in the structure of the pulmonary valve and its surroundings so that the infundibular type pulmonary stenosis can be removed. The patient was then diagnosed as VSD PMO, left-to-right shunt, suspected double-chambered right ventricle (DCRV) with high probability of pulmonary hypertension. The patient is then planned for a multi-slice computer tomography (MSCT) of the heart and catheterization of the tapping of the heart in preparation for the surgical conference.

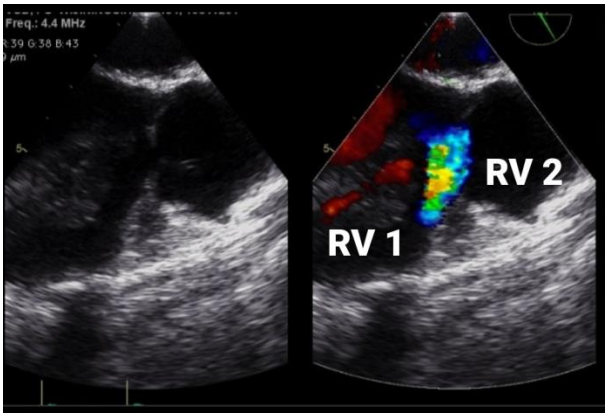

Figure 1. The TOE finding of RV 1 and RV 2

Cardiac MSCT was done with the results of site solitus, no persistent left superior vena cava (PLSVC), innominata vein, AV-VA concordance, VSD malalignment with a diameter of 4.9-9.6 mm, RV divided by pseudoseptum which becomes the first RV and the second RV. The second RV is more anterior and apical than the first RV. Mean pulmonary artery (MPA) diameter is 16.7 mm, proximal right pulmonary artery (RPA) 14.7 mm, distal 13.6 mm, proximal left pulmonary artery (LPA) 12.2 mm, distal 11.4 mm. The diameter of the descending aorta is 10.9 mm, with the conclusion: malalignment VSD, RV is divided by the pseudoseptum into two parts, with no patent ductus arteriosus (PDA) and major aorto pulmonary collateral arteries (MAPCA) found. Figure 2 showed the result of cardiac MSCT.

Patients underwent right and left heart catheterization. The result was an innominate vein, the catheter could cross from the RV to the left ventricle (LV), LV graphy appeared VSD diameter 7.13 mm, pressure at Ao 105/32 (80)mmHg , RA 7 / 4 (3)mmHG, PA 27/7 (16) mmHg, RV1 101 / -5 (17) mmHg, RV2 25 / -2 (7) mmHg, LV 104/0 (8) mmHg; saturation (%) superior vena cava (SVC) 60.4, inferior vena cava (IVC) 70.1, right atrium (RA) 74.5, pulmonary artery (PA) 81, Aorta 97, LV 97; flow ratio 2.07, pulmonary artery resistant index (PARI) 2.12, Qp / Qs 1.56, CO 3.74, CI 2.85 with conclusion: VSD shunt left to right, high flow low resistant with double chamber right ventricle (DCRV), no pulmonary hypertension. Figure 3 and 4 showed the heart catheterization result.

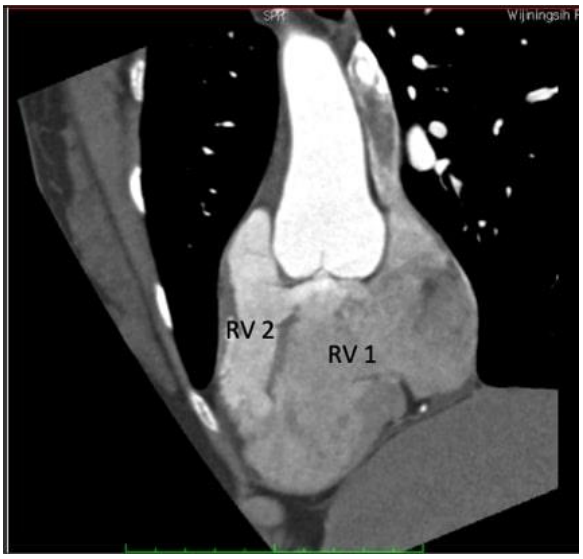

Figure 2. Cardiac MSCT scan finding of RV 1 and RV 2 separated by a septum

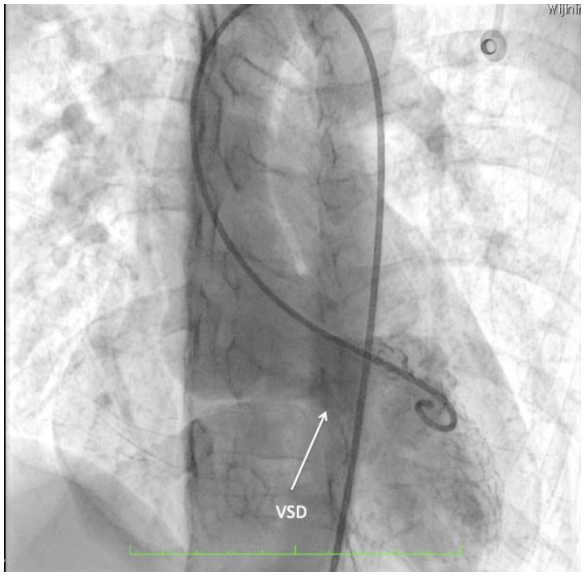

Figure 3. Right heart catheterization showing the VSD

The patient underwent surgery with VSD closure and resection of the septum without complications. Echocardiography parameter became normal albeit still had some septal tissue left in order to limit sudden surge of blood flow into pulmonary artery. Follow up indicate patient has no symptom associated with the surgery.

Discussion

The right ventricle consists of three different anatomical regions: the inlet, the trabecular apex, and the outlet. RV inlets consist of and are limited by tricuspid valves. The tricuspid valve is composed of three cuspi, anterosuperior, inferior and septal. These three cusps are bound by the chordae tendinea which are anchored by the papillary muscles to the walls of the myocardial trabecula in addition to the septal cusps which are bound directly to the interventricular septum by the septal papillary muscles. The trabecular portion of the ventricles extends to the apex which has thinner walls and is prone to injury by heart catheters and pacemaker electrodes.[4] Outlets are the parts of the RV that are not composed by the trabecular muscles. The surface of this section is smoother than other RV parts.

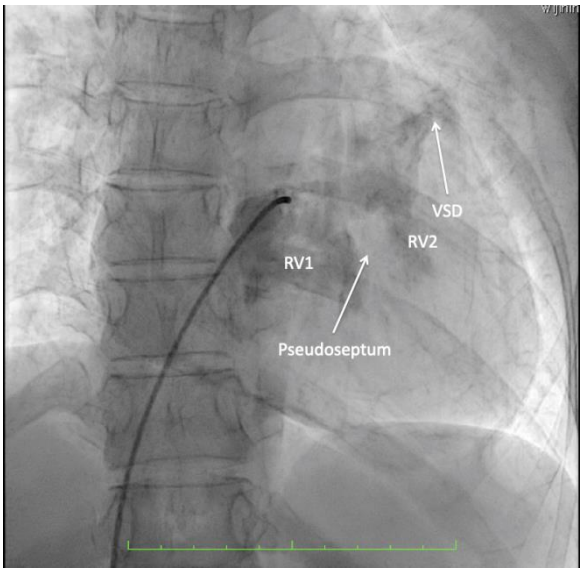

Figure 4. Right heart catheterization showing the DCRV

Double Chamber Right Ventricle is a congenital heart disorder caused by the muscle anomaly that divides RV. DCRV has two types namely DCRV type 1 and DCRV type 2. DCRV type 1 is a muscle tissue disorder that crosses RV while type 2 DCRV is parietal muscle hypertrophy and RV septal.[5] DCRV may also occur from moderator bands that come from a superior location and are almost separated from the septum wall.[6]

Usually DCRV does not stand alone. More than 90% of cases are related to congenital VSD abnormalities, especially the membrane type. In this patient the congenital abnormality accompanying DCRV is VSD PMO. Other congenital abnormalities that often coincide with DCRV include pulmonary valve stenosis, PAPVD, transposition of large arteries, PLSVC, double outlet right ventricle (DORV), tetralogy of Fallot, and Ebstein anomalies. The tendency of DCRV to associate with other congenital cardiac abnormalities indicates a strong correlation with abnormalities during fetal development (Hoffmann et al., 2004). DCRV is quite often misdiagnosed and remains asymptomatic until adulthood. In this patient there were no complaints until during pregnancy the systolic noise was typical of VSD, and after echocardiography was diagnosed as VSD with infundibular pulmonary stenosis. Muscle tissue abnormalities in RV may be quite common and only in the presence of congenital heart abnormalities that cause hypertrophy in these tissues causes obstruction of RV outflow.[7]

Patients with DCRV have similar clinical conditions with several congenital abnormalities. Some of the conditions that are often found are dyspnea, syncope, stable and unstable angina, exercise intolerance, and asymptomatic.[8] Physical examination that may be found is systolic noise VSD and RV heaving if there is prominent right ventricular hypertrophy.[9]

The most effective diagnostic tool for detecting DCRV is TOE.[2] In the echocardiographic context, obstruction is defined depending on location and appearance. The location of the lesion is described as "high" if it is located close to the pulmonary valve and "low" if it is localized at the apex. Diffuse lesions are triangular in shape with a long attachment and discrete lesions have a short attachment. The TOE approach with Doppler provides a visualization of the degree of stenosis between the two chambers. The use of continuous Doppler can calculate intraventricular pressure gradients that reduce the need for cardiac catheterization. On echocardiography, fibrosis and hypertrophy are more visible during systole because muscle tissue contracts and thickens.

Cardiac catheterization and angiography can be used to confirm the diagnosis. It can also be done if there is no TOE probe available. The use of cardiac catheterization can assess intraventricular gradient more accurately than continuous Doppler because the use of TOE uses the Bernoulli equation which is simplified by eliminating the velocity equation in the proximal ventricular section. Another differential diagnosis of DCRV associated with VSD is Tetralogy of Fallot and VSD with PS because the picture is similar on echocardiography.[3]

The most effective DCRV management is operative by removing hypertrophic tissue. With anatomic locations that may be close to the tricuspid valve, tissue resection must pay attention to the attachment of the saved papillary muscle to maintain valve function. Difficulties in visualizing intraoperative muscular abnormalities can occur if the location is "low" in the right ventricle.[10]

The conduction block is the most commonly acquired morbidity after DCRV resection. Post-surgery found 46% of patients experienced incomplete right bundle branch block (RBBB). This can occur because the atrioventricular pathway runs along the septomarginal trabeculae and the moderator band to the right ventricular apex and the anterior papillary muscle. Complete resection and a portion of the band moderator will cause iatrogenic heart block.[1]

Obstructive tissue resection is done in conjunction with VSD closure. In these patients planned closure of VSD by resection of the intraventricular septum. The location of the VSD also determines the difficulty of closing the defect. The location of the defect at the apex has more difficult access if there is obstruction of thickening of the moderator band.[6]

Indications of operative intervention in patients with DCRV are the presence of symptoms, associated anomalies that require intervention, aortic regurgitation to any degree, and significant RV obstruction that allows patients to experience symptomatic future. DCRV operative correction has good functional and long-term hemodynamics.[12]

DCRV is generally a progressive cardiac disorder so that surgical intervention before the appearance of symptoms is more beneficial. The groups of patients at high risk for developing symptoms are patients with VSD, increased flow to the pulmonary arteries, and patients with right heart failure.[1] In these patients no significant symptoms appear, but patients with concomitant VSD and aortic regurgitation at any degree in these patients are indicative of VSD closure and intraventricular septal resection.

An operative approach to dealing with DCRV is more developed today. In short, such as transventricular, transatrial, or transatrial-transventricular combination. In the era before the 2000s, transventricular was the most popular method, whereas after that transatrial and transatrial-transventricular combinations were more dominant. Transventricular is still used as the most frequent method in pediatric patients. Transventricular methods especially right ventriculotomy have several advantages, providing an accurate assessment of the RV obstructive elements and visualization of ventricular septal defect boundaries that are difficult to identify via transatrial. RV dysfunction and lethal ventricular arrhythmias are rarely encountered in DCRV operations, in contrast to long-term outcomes in patients with tetralogy of Fallot. The most important thing is to avoid damage to the tricuspid valve component in any approach.[14]

The patient underwent surgery with VSD closure and resection of the septum without complications. Echocardiography parameter became normal albeit still had some septal tissue left in order to limit sudden surge of blood flow into pulmonary artery. Follow up indicate patient has no symptom associated with the surgery.

Conclusion

36-year-old female patient diagnosed with DCRV with TOE. DCRV is a congenital abnormality that divides RV into two parts, high pressure proximal and low pressure distal. DCRV can be caused by muscle tissue anomalies, muscle trabecular hypertrophy, or moderator band abnormalities. This congenital abnormality often coincides with other cardiac disorders such as VSD and subaortic stenosis. Because of its rare occurrence, DCRV is often diagnosed as other diseases such as TOF and VSD with PS. The most effective management is operative septal tissue excision and VSD closure.

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