

## Ventricular Septal Defect Closure with Perforated Patch in Large Ventricular Septal Defect with Severe Pulmonary Hypertension and Non Reactive Oxygen Test

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### Abstract

Management of ventricular septal defect (VSD) with severe pulmonary hypertension (PH) had not been extensively studied and is still challenging. The closure of VSD in patients with high pulmonary vascular resistance (PVR) and severe PH is highly risk procedure. If high PVR and severe PH still persist after closure procedure, the patients have poor prognosis. We reported a 24-year-old woman whom was diagnosed with large VSD, bidirectional shunt with L to R dominance, dilatation of left ventricle, and mild to moderate mitral regurgitation, mild tricuspid regurgitation and severe PH. Right heart catheterization showed pre-oxygen test: mean aorta pressure 85 mmHg, mean pulmonary artery pressure 65 mmHg, flow ratio 5,4, PVR 2,3 WU and pulmonary vascular resistance index (PVRI) 3,22 WU/m<sup>2</sup>. The results of post-oxygen test: mean aorta pressure 83 mmHg, mean pulmonary artery pressure 63 mmHg, flow Ratio 2,2, PVR 0,3 WU and PVRI 0,42 WU/m<sup>2</sup>. Patient had been performed VSD closure with perforated patch 3 mm. Three month evaluation by echocardiography showed residual VSD 3 mm, L to R shunt, moderate tricuspid regurgitation and mild PH (TVG 36 mmHg). In Baumgartner criteria of VSD operability, this patient was not operable because the ratio of mean pulmonary and systemic circulation more than 2/3, but in Lopez criteria, patient is operable because PVRI below 6 WU/m<sup>2</sup>. Patient with high and moderate PH and PVR which is still operable, VSD can be closed partially. Partially VSD closure can be performed by transcatheter procedure after PH decrease and stable.

Keyword: VSD closure- perforated patch – PH severe

### Background

The management of ventricular septal defect (VSD) with severe pulmonary hypertension (PH) had not yet extensively investigated.<sup>1</sup> The closure procedure in VSD patients with increasing pulmonary vascular resistance (PVR) with severe PH is a risky procedure. The persistence of high PVR and severe PH after the closure procedure is the sign of a bad prognosis.<sup>1</sup> In a retrospective study, patient with congenital heart disease and severe PH post operatively had worse prognosis than patient with non-operative congenital heart disease.<sup>2</sup> In this case we present and discuss a patient with large VSD, severe PH and non reactive oxygen test in whom the VSD closure was performed.

### Case

A woman 24 years old came to our hospital with a chief complaint of dyspnea. She felt dyspnea on effort since 10 months before admission, there was neither orthopnea nor paroxysmal nocturnal dyspnea. Patient had been diagnosed as large VSD since April 2014 and has been planned to have VSD closure.

Her physical examination demonstrated a good general condition and compos mentis. Blood pressure was 100/60 mmHg, heart rate was 82 bpm, respiratory rate was 22 x/minute, temperature was 36,5°C and peripheral oxygen saturation was 97%. No increased jugular venous pressure. Lung examination was normal. There was cardiomegaly with ictus cordis at intercostal space V left mid clavícula line. In auscultation, S1 was normal and S2 was louder, with holosystolic murmur grade 3/6 with punctum maximum at intercostal space III-IV left parasternal border. Abdominal and extremity examination were normal

Laboratory results showed hemoglobin 13,1 g/dL, hematokrit 39%, leukocytes 7,94 x 10<sup>3</sup>/μL, segment 59,6 %, limphocytes 31,6 %, monocytes 6,9%, eosinophil 1,8%, basophil 0,1%. Blood chemistry showed BUN 11 mg/dL, creatinine 0,89 mg/dL, SGOT 21 U/L, SGPT 18 U/L, natrium 138 mEq/L, potassium 3,8 mEq/L, and chloride 102 mEq/L. An electrocardiography examination showed sinus rhythm, heart rate 76 bpm and normoaxis (figure 1).

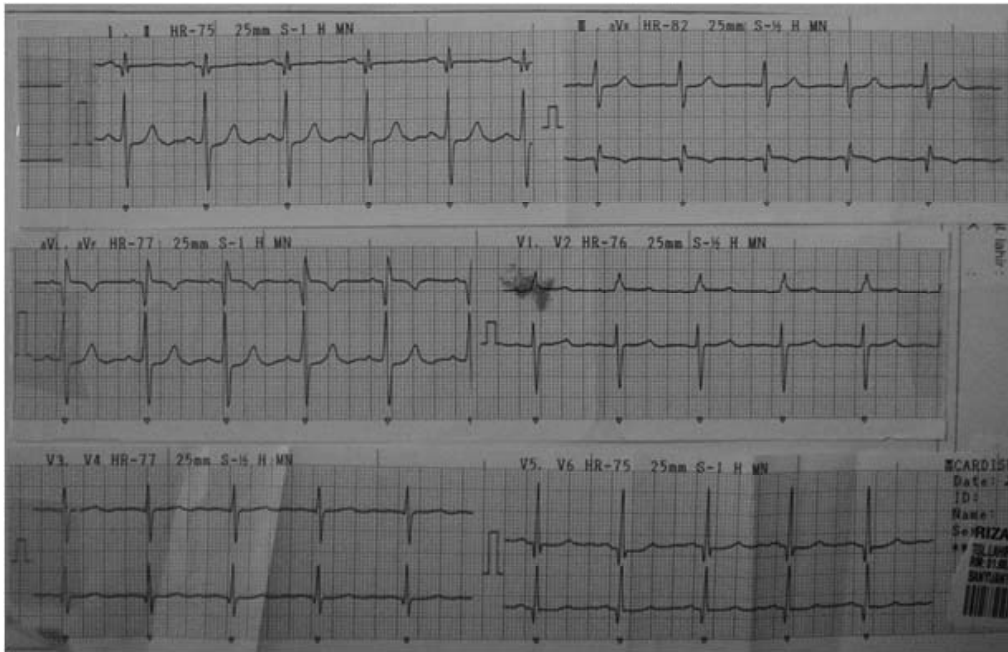


Figure 1. Electrocardiogram showed sinus rhythm, heart rate 76 bpm and normoaxis



Figure 2. Cardiomegaly with CTR > 0,5 and protruding pulmonary segment

Chest X-ray examination in showed cardiomegaly (CTR > 0,5) and a protuding of pulmonary segment indicating the presence of pulmonary hypertension (figure 2).

Echocardiography examination results showed a large VSD with diameter 17-20 mm, bidirectional shunt with L-to-R shunt dominance, dilatation of left ventricle with LV diameter 55 mm, ejection fraction 70%, diastolic dysfunction

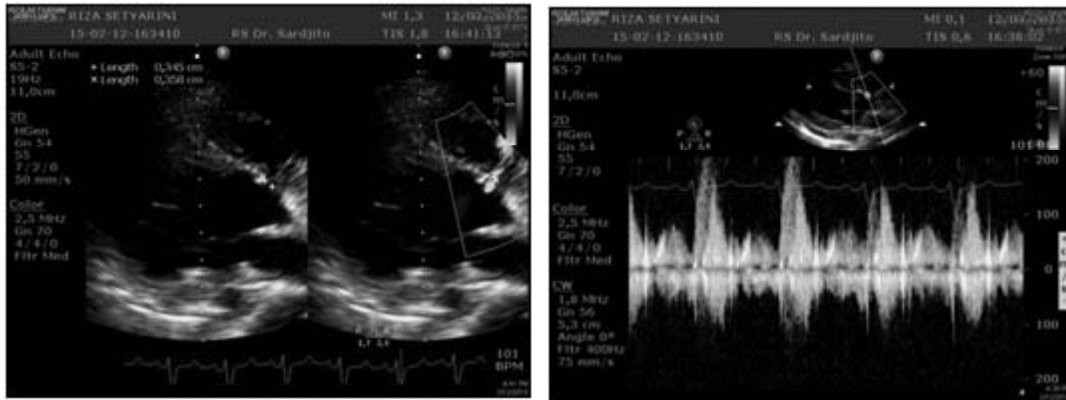


Figure 3. Echocardiogram in post-operation showed residual VSD 3 mm, L to R shunt

relaxation type, normal right ventricle function, mild to moderate mitral regurgitation, mild tricuspid regurgitation and severe PH (TVG was 74 mmHg). Right heart catheterization results showed: pre-oxygen test mean aorta pressure was 85 mmHg, mean pulmonary artery pressure was 65 mmHg, flow ratio was 5,4, PVR was 2,3 WU and PVRI was 3,22 WU/m<sup>2</sup>. Post oxygen test: mean aorta pressure was 83 mmHg, mean pulmonary artery pressure was 63 mmHg, flow ratio was 22, PVR was 0,3 WU and PVRI was 0,42 WU/m<sup>2</sup>.

The patient had been done VSD closure in February 2015 with perforated patch 3 mm. The evaluation by echocardiography done in April 2015 showed residual VSD with 3 mm diameter, L to R shunt, normal cardiac chambers, ejection fraction 63%, uncoordinated motion, normal diastolic LV function and normal right ventricle function, moderate tricuspid regurgitation and mild PH (TVG 36 mmHg) (figure 3). The patient was hospitalized within 8 days and discharge in stable condition. The peripheral oxygen saturation was 97%. The medication was sildenafil 3 x 20 mg.

#### Discussion

Pulmonary hypertension can occur in congenital heart disease because of increasing pulmonary blood flow due to L-to-R shunt through the defect. An L-to-R shunt will increase arterial pulmonary pressure and cause a stress to the vessel wall, circumferential dilatation of wall vessel and endothelial dysfunction.<sup>3</sup> There is a changing expression of vasoactive mediator such as endothelin-1, prostacyclin and nitric oxide. Pulmonary vasoconstriction will occur and persist. The increased expression of vascular endothelin-1 and fibroblast growth factors cause remodeling (hypertrophy and proliferation) of vascular smooth

muscle cells and increasing of intracellular matrix deposition. This remodelling will cause increasing of PVR and subsequent right ventricle pressure.<sup>3</sup>

Patient with moderate to large size congenital heart defect can suffers from PH but not pulmonary vascular disease (PVD) because there is no extensive and permanent remodeling in pulmonary vessel walls. Patient with severe PH and high pulmonary flow and normal PVR, may be considered to have VSD closure because the lesion in vessel walls is not yet extensive and may still be reversible. High PVR indicated that such patients have suffered from extensive and irreversible PVD.<sup>4</sup> The closure of VSD in patient with moderate to large congenital defect, L to R shunt and moderate to high PVR is very high risk procedure because the extent of lesion in pulmonary vessels is unknown and there is no guarantee for good long term outcome.<sup>4</sup> If PVR is still high in post operation, the patient's prognosis is worse.<sup>4</sup> A study in England reported that children whom suffered from severe PH post operatively had worse prognosis than patients with Eisenmenger syndrome.<sup>2</sup>

When the right heart catheterization was done, a vasoreactive test was usually performed. The gold standard of vasoreactivity test was using inhaled nitric oxide (iNO) not only oxygen.<sup>5</sup> In iNOP test 1 (inhaled nitric oxide as a preoperative test 1) study showed that by using oxygen and iNOP (100% oxygen and 80 ppm iNOP) gave 97% and 90% sensitivity and accuracy, respectively, to predict the operability in patient with congenital heart disease and PH, whereas oxygen only (100% oxygen) gave 64% sensitivity and 68% accuracy.<sup>6</sup>

Currently, there is no consensus about hemodynamic criteria to determine operability in

Table 1. Hemodynamic criteria in the operability of VSD<sup>9</sup>

Indications	Class <sup>a</sup>	Level <sup>b</sup>
Patients with symptoms that can be attributed to L-R shunting through the (residual) VSD and who have no severe pulmonary vascular disease (see below) should undergo surgical VSD closure	I	C
Asymptomatic patients with evidence of LV volume overload attributable to the VSD should undergo surgical VSD closure	I	C
Patients with a history of IE should be considered for surgical VSD closure	IIa	C
Patients with VSD-associated prolapse of an aortic valve cusp causing progressive AR should be considered for surgery	IIa	C
Patients with VSD and PAH should be considered for surgery when there is still net L-R shunt ( $Q_p:Q_s > 1.5$ ) present and PAP or PVR are $< 2/3$ of systemic values (baseline or when challenged with vasodilators, preferably nitric oxide, or after targeted PAH therapy)	IIa	C
Surgery must be avoided in Eisenmenger VSD and when exercise-induced desaturation is present	III	C
If the VSD is small, not subarterial, does not lead to LV volume overload or pulmonary hypertension, and if there is no history of IE, surgery should be avoided	III	C

<sup>a</sup>Class of recommendation.  
<sup>b</sup>Level of evidence.  
 AR = aortic regurgitation; IE = infective endocarditis; L-R shunt = left-to-right shunt; LV = left ventricle; PAH = pulmonary arterial hypertension; PVR = pulmonary vascular resistance;  $Q_p:Q_s$  = pulmonary to systemic flow ratio; VSD = ventricular septal defect.

congenital heart disease.<sup>7</sup> There is a retrospective study showed that 38 patients with VSD and severe PH and high PVR ( $>6$  WU) was closed, resulting in 8 patients died because of severe PH post operation and 30 patients (79%) had good outcome post operation.<sup>8</sup> It showed that PVR threshold in operable patient must be determined precisely, if the threshold too low it will exclude many patients whom actually may still be operable.<sup>8</sup> Baumgartner et al. (2009)<sup>9</sup> give operability hemodynamic criteria in VSD with PH according to European Society of Cardiology guideline, which is as follows:

Lopes et al. (2009) suggested a hemodynamic criteria for assessment of operability in patients with congenital heart disease and pulmonary artery hypertension. Table 2 showed the criteria suggested by Lopes et al. (2009).<sup>10</sup>

Patients with high and moderate PH and PVR which is still operable in the criteria by Lopes et al. (2009)<sup>10</sup>, VSD can be closed partially, so if severe PH still persist post-operatively, right ventricle can be decompressed.<sup>11</sup> According to Beghetti (2012) and Myers (2014), partially VSD closure can be closed by transcatheter procedure after PH decrease and stable.<sup>4,7</sup>

In this case, the patient had been performed partially VSD closure, by making 3-3,5 mm hole in the patch, so it allows right ventricle to decompress if PH still persist post operation. Subsequently, this residual VSD can be closed by transcatheter procedure. There are several case reports about transcatheter closure in patient that had residual VSD closure, such as by Ramakhrisnan et al. (2011)<sup>12</sup> who reported residual VSD closure by

Table 2. Hemodynamic criteria for operability of congenital heart defect with PH<sup>10</sup>

Condition	Hemodynamics
Baseline	PVR <6WU/m <sup>2</sup> , PVR:SVR ratio <0.3
Acute vasodilator challenge (O <sub>2</sub> , NO)	Baseline PVR 6–9WU/m <sup>2</sup> , PVR:SVR 0.3–0.5
	• Decrease in PVR index of 20%
	• PVR:SVR decrease of 20%
	• Final PVR index <6WU/m <sup>2</sup>
	• Final PVR:SVR <0.3

Criteria suggestive of a favorable outcome after operation with biventricular circulation.<sup>11</sup>

PAH-CHD, pulmonary arterial hypertension associated with congenital heart disease; NO, nitric oxide; PVR, pulmonary vascular resistance; SVR, systemic vascular resistance.

ADO II in baby and Catfield *et al.* (2005)<sup>13</sup> who reported transcatheter VSD closure in VSD patient post operatively with patch dehiscence.

### Conclusion

We reported a 24-year-old woman large VSD and severe PH. The patient had been done VSD closure with perforated patch 3-3,5 mm. In post operative echocardiogram showed residual VSD 3 mm, L to R shunt with improvement of PH from 79 mmHg to 36 mmHg. A VSD closure in this patient was performed by perforated patch, to anticipate if PH persist post operatively, and be closed by transcatheter procedure if PH improve and stable in the follow up.

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