# A REVIEW OF PULMONARY ATRESIA WITH VENTRICULAR SEPTAL DEFECT TREATMENT IN HANOI HEART HOSPITAL

Nguyen Sinh Hien[1], Nguyen Minh Ngoc[1]

**ABSTRACT**

***Objective:*** *Evaluation of short-term outcome after treatment of pulmonary atresia with ventricular septal defect in Hanoi Heart Hospital.*

***Methods:*** *Patient records of 188 patients with PA-VSD treated in Hanoi Heart Hospital from 01/2005 to 10/2016, were retrospectively analysed in this study.*

***Results:*** *Mean age 3.2 ± 1.5 (0.16 to 25 years); Classification: Type I: 65 patients (34.6%), type II: 56 (29.8%), type III: 47 (25%), type IV: 20 (10.6%); Treatment: Complete repair: 106 (56.4%), MAPCAs unifocalization with RV–PA shunt (Sano shunt): 16 (8.5%), MAPCAs unifocalization with BT shunt 13 (6.9%), BT shunt only 53 (28.2%). Early death: 8/188 (4.3%).*

***Conclusions:*** *Correct diagnostic and surgery indication help improve the outcome of PA-VSD patients.*

***Key word:*** *Pulmonary atresia, ventricular septal defect.*

## I. INTRODUCTION

Pulmonary atresia with ventricular septal defect (PA-VSD) is a cyanotic congenital heart defect with anatomical diversity: the size of central pulmonary arteries varies from diminutive/absent to reasonable, no connection between right ventricle and pulmonary arteries, and pulmonary blood supply is derived either from native pulmonary arteries or major aortopulmonary collateral arteries (MAPCAs), or both. The others anomalies are similar to Tetralogy of Fallot: large VSD, overriding aorta, and RV hypertrophy. If not treated, patients with PA-VSD have less than 50% chance of being alive at 1 year and 15% at 30 years [1]. Common causes of death are: heart failure, cerebrovascular accident,...

Diagnostic of PA-VSD and indication of treatment are obtained by clinical findings, echocardiography, catheterization, multisection computed-tomography (MSCT). [2]

Treatment of PA-VSD consists of medication and surgery. The MAPCAs unifocalization and complete repair surgery have been performed since the 1980s but the results remains unclear. Common complications of surgery are: congestive heart failure, PA hypertension,...

In Vietnam, there are few studies on PA-VSD surgery. This study aims to evaluate the early results of PA-VSD repairs in Hanoi Heart Hospital, and hopefully our experiences may help improving the success in treatment of this congenital defect.

## II. MATERIALS AND METHODS

This study comprised 188 PA-VSD patients treated in Hanoi Heart Hospital from 01/2005 to 07/2016. The patients who have other combining

1. Hanoi Heart Hospital

**Corresponding author**: Nguyen sinh Hien
Email: nguyensinhhien@gmail.com;
**Received:** 21/4/2017; **Revised:** 25/5/2017;
**Accepted:** 19/6/2017

defects such as atrioventricular septal defects, anormaly of pulmonary venous return,... are excluded.

Patients' preoperative, intra-operative and post-operative records are collected and reviewed. Data are analysed by SPSS software. Comparisons are statistically significant when $p < 0.05$. Early-death means death within 30 days postoperative.

## III. RESULTS

Table 1: Preoperative characteristics (n = 188)

| | n (percentage) |
|---|---|
| Male | 103 (54.8%) |
| Female | 85 (45.2%) |
| Age | 3.2 ±1.5 years |
| Cyanotic | 188 (100%) |
| Syncope | 45 (23.9%) |
| Cerebrovascular accident | 7 (3.7%) |
| Cerebral abcess | 3 (1.6%) |
| Hct (%) | 60.2± 18.5 (41-80.5) |
| Hb (g/l) | 176.5± 25.6 (122-251) |

Table 2: Operation Technique (n = 188)

| | **Complete repair** | **BT shunt** | **Unifocalization and repair** | **Unifocalization and BT shunt** | **Unifocalization and RV-PA shunt** | **Total** |
|---|---|---|---|---|---|---|
| **Type I** | 54 | 11 | 0 | 0 | 0 | 65 |
| **Type II** | 39 | 17 | 0 | 0 | 0 | 56 |
| **Type III** | 5 | 25 | 7 | 7 | 3 | 47 |
| **Type IV** | 0 | 0 | 1 | 6 | 13 | 20 |
| **Total** | 98 | 53 | 8 | 13 | 16 | 188 |
| **Percentage** | 52.1% | 28.2% | 4.3% | 6.9% | 8.5% | |

Complete repair was performed in 106 patients (56.4%). Mean number of MAPCAs in unifocalizated patients: 4.1± 0.8 (Min:2 – Max: 6). Mean number of anastomosis in unifocalizated patients: 2.7± 0.7 (Min:1 – Max:5)

Table 3: Oxygen saturation in patients with MAPCAs unifocalization

| | Preoperative saturation | Postoperative saturation |
|---|---|---|
| Unifocalization and BT shunt | 35.5 ± 3.4 (36-58) | 75.6 ± 6.6 (74-92) |
| Unifocalization and RV-PA (Sano) shunt | 35.4 ± 3.2 (35-60) | 72.5 ± 7.5 (70-90) |

Table 4: Complication and death (n = 188)

| Complication | n | Percentage |
|---|---|---|
| Low cardiac output | 26 | 13.8 |
| Bleeding | 4 | 2.1 |
| Pericardial effusion | 10 | 5.3 |
| Sternal and wound infection | 5 | 2.7 |
| Renal failure | 3 | 1.6 |
| Reoperation | 6 | 3.3 |
| Death | 8 | 4.3 |

Mean time of intubation (in hour): 32 ± 26.4 (Min: 2 – Max: 168). Mean time of hospital admission (in day): 12.5 ± 4.5 (Min: 5 – Max: 60).

Table 5: Correlation of technique, low cardiac output syndrome and death

| **Technique** | **n** | **Death** | **Low cardiac output** |
|---|---|---|---|
| Complete repair of type I | 54 | 1 | 3 |
| Complete repair of type II | 39 | 1 | 4 |
| Complete repair of type III | 5 | 1 | 3 |
| Unifocalization and complete repair | 8 | 1 | 3 |
| Unifocalization and BT shunt | 13 | 1 | 3 |
| Unifocalization and RV-PA shunt | 16 | 2 | 5 |
| BT shunt only | 53 | 1 | 5 |
| Total | 188 | 8 | 26 |

Table 6: Characteristic of complete repair patients (n = 106)

| | n | Percentage |
|---|---|---|
| RV- PA gradient (mmHg)<br>< 40<br>≥ 40 | <br>102<br>4 | <br>96.2<br>3.8 |
| RV pressure / LV pressure<br>< 0.7<br>≥ 0.7 | <br>96<br>10 | <br>90.6<br>9.4 |
| Aortic cross-clamp time<br>CPB time | 90.2 ± 30.5 (42-180)<br>114 ± 35.5 (55-255) | |

Characteristics affect death rate:

- CPB time > 120 minutes (p = 0.003)
- RV/LV pressure ≥ 0,7 (p = 0.002)

Table 7: Postoperative echocardiography in complete repair patients (n=102)

| | n | Percentage |
|---|---|---|
| Pulmonary valve regurgitation<br>Mild<br>Moderate - Severe | <br>85<br>17 | <br>83.3<br>16.7 |
| Persistent VSD | 2 | 1.96 |
| RV-PA gradient > 40 mmHg | 2 | 1.96 |

*Death rate within 12 months*: 1/180 (0.56%). The patient was a 3-years old PA-VSD type III child who had been performed a complete repair.

## IV. DISCUSSION

### 4.1. Diagnostic

Theoretically, when the MAPCAs are adequate, cyanotic symptom is mild or absent [2]. But in our study all of the patients were cyanotic in different levels, which may be explained by late admission. The mean age was 3.2 years (youngest: 3 months and oldest: 25 years). Some patients had large but locally stenotic MAPCAs, which reduced pulmonary blood flow but also protected lung parenchyma. Cyanotic symptom was often the cause of hospital admission, but sometimes the patients presented the complications such as cyanotic syncope (23.9%), cereborvascular accident (3.7%), and cerebral abcess (1.6%). Echocardiography was performed in all patients. The anomaly images were: absent of RV-PA connection, diminution or absence of pulmonary trunk and main branchs, ventricular septal defect, overidding aorta, and RV hypertrophy. Some authors considered PA-VSD as a severe form of Tetralogy of Fallot [1]. In early time, a lot of patients were diagnosed only with echocardiography, which caused ignorance of defects. The consequences are intraoperative difficulty, postoperative PA hypertension and heart failure,... Therefore, the combination of echo, cardiac catheterization an MSCT is greatly important in PA-VSD diagnostic. It allows effective evaluation of cardiac lesions, diminutive level of pulmonary arteries, pulmonary blood flow distribution, calculation of McGoon index, Nakata index, Z value,... and helps correct the surgery indication (complete repair of temporary surgery).

### 4.2. Indication of surgery

In early time, with the limitations of image diagnostics (catheterization, MSCT), we performed BT shunt in patients with small pulmonary artery (Z < -2, based on echo and intraoperative evaluation). One-stage repair was performed on type I, type II patients with adequate pulmonary artery. Other studies demonstrate differents index for surgery indication: one-stage repair in patients with McGoon index greater than 1.2, Nakata index greater than 150mm/m$^2$, or total neopulmonary artery index (TNPAI) greater than 200mm/m$^2$[2]. At least 14/20 lobe division of lungs must have proper blood flow after unifocalization process to guarantee the ability of oxygen exchange [6], therefore, we did not perform one-stage repair in all patients. The percentage of complete repair in this study is 56.4%, primarily in type I and type II patients. In present time, with adequacy of image disagnostics, we could perform complete repair in even type III (25.5%) or type IV (5%) patients. The one-stage repair percentage of Mee et al. was 51.7% [5], of Davies et al. was 73% [4], and of Carrilo et al. was 80% [6].

According to Gupta et al., unifocalization surgery should be performed in 1-3 months of age if pulmonary blood flow totally depends on MAPCAs, in 3-6 months of age if patients have stenotic MAPCAs, and in 6-8 months of age if MAPCAs are diminutive/absent and pulmonary flow is provided mostly through the PDA. Other studies demonstrates that obstructive pulmonary

arterial disease may present in 4-week-old babies. In this study, most of the patients were lately hospitalized, which reduced the surgery results. To improve the outcome, early diagnostic should be performed, even in prenatal period.

**4.3. Technique of operation**

Surgery incision should be lateral thoracotomy or median sternotomy when performing MAPCAs. According to D.Balaguru, lateral thoracotomy should be performed for unifocalization surgery, and complete repair through median sternotomy could be performed several months later (VSD closure and reconstruction of RV-PA conduct). In our centre, we only use median sternotomy for unifocalization of MAPCAs. The incision allows full exposure of thoracic aorta and MAPCAs, and complete repair can be performed immediately if necessary. We always try to exposure and unifocalizate all MAPCAs which had been discovered by catheterization and MSCT, excluding too small MAPCAs (diameter less than 2mm), because those would be early obstructive. Mean number of MAPCAs in our study was 4, and mean number of anastomosis was 2.7 (table 2).

We prefer to perform all possible anastomosis before ligation of MAPCAs to reduce bypass time, and CPB should only be initialized when SpO2 was less than 50%. Hence, sometimes unifocalization surgery could be performed without CPB. Pericardial patch enlargement is recommended if pulmonary artery stenosis exists.

An important question: should BT shunt or RV-PA (Sano) shunt be made after unifocalization? According to table 3, the oxygen saturation is higher in BT shunt patients than in RV-PA shunt patients. In early time we preferred RV-PA shunt but some patients had low saturation postoperatively (less than 60%) and we had to leave 1-2 MAPCA (would be ligated with coil after). Thus we changed to perform BT shunt to acquire better saturation. This may be explained by low oxygen exchange in several region of lung which was caused by late admission.

The size of BT shunt conduct was determined according to table 8.

Table 8: Conduct size by patient's weight

| Weight (kg) | Size of the Goretex Conduct |
|---|---|
| 3,2-4 | 4 |
| >4-6 | 5 |
| >6-10 | 6 |
| >10 | 7 |

When performing RV–PA shunt, the conduction size should be equal to 2/3 of the theoretic pulmonary trunk size. The conduction may be homograft or biological valve tube. We used pericardial patch to construct the conduction in early time, but the pulmonary valve was often insufficient after 3-6 months

**4.4. Outcome**

Most of the patients who had been performed a complete repair have significant improvement in clinical symptoms and grow-up. Cyanotic symptom was reduced in MAPCAs and BT shunt patients. The most common complication was low cardiac output syndrome (13.8%), especially on patient with MAPCAs unifocalization. Early death rate was 3.8% in complete repair group, 10.3% in unifocalization group, 1.9% in BT shunt group, and total rate was 4.3%. The factors affecting death rate were long CPB time (more than 120 minutes) and RV/LV pressure ratio greater than 0.7 [1], [6], There was one death after 1 years of operation which was caused by persistent postoperative PA hypertension. Recent studies demonstrates similar rate. Death rate was 3.3% according to Mee et al.[5] and 5.7% according to Carrilo et al. According to Davies et al., death rate after unifocalization was 6%[4]. The death rate after complete repair in J.Souquet's study was 8% [7] .

**V. CONCLUSION**

PA-VSD is a complex congenital heart defect which requires early diagnostic and surgery

treatment. New imaging technologies (MSCT, catheterization) are useful in operative indication.

In type I and type II patients, one-stage repair is preferred, while type IV patients should be indicated two-stage repair (MAPCA unifocalization followed by complete repair). In type III patients, the choice of operative method should be based on the anatomy of pulmonary arteries.

## REFERENCES


1. Kirklin JW; Boyes B; Kouchoukos NT, (2003), Ventricular Septal Defect and Pulmonary Stenosis or Atresia, in *Cardiac Surgery*, 1, Churchill Livingstone, Philadelphia, pp. 946-1074.
2. Balaguru D, Dilawar M. Pulmonary atresia with ventricular septal defect: systematic review. Heart Views 2007,8: 52-61
3. Gupta A, Odim J, Levi D, Chang RK, Laks H. Staged repair of pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries: experience with 104 patients. J Thorac Cardiovasc Surg. 2003 Dec. 126(6): 1746-52.
4. Ben Davies et al. Unifocalization of major aortopulmonary collateral arteries in pulmonary atresia with ventricular septal defect is essential to achive excellent outcomes irrespective of native pulmonary artery morphology. The Journal of Thoracic and Cardiovascular Surgery. Volume 138, Number 6.
5. Iyer KS, Mee RB. Staged repair of pulmonary atresia with ventricular septal defect and major systemic to pulmonary artery collaterals. Ann Thorac Surg. 1991 Jan. 51(1): 65-72.
6. Carrillo SA, Mainwaring RD, Patrick WL, Bauser-Heaton HD, Peng L, Reddy VM, Hanley FL. Surgical Repair of Pulmonary Atresia With Ventricular Septal Defect and Major Aortopulmonary Collaterals With Absent Intrapericardial Pulmonary Arteries. Ann Thorac Surg. 2015 Aug; 100(2): 606-14.
7. Jéôme Soquet, Yves d'Udekem, Christian Brizard. Atrésie pulmonaire avec communication interventriculaire et collatérales systémicopulmonaires: unifocalisation versus reshabilitation. Journal de Chirurgie Thoracique et Cardio-Vasculaire. 2016. vol 20-Septembre.