

# Pulmonary atresia with intact ventricular septum: medium term follow-up

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## Introduction and purpose

Pulmonary atresia with intact ventricular septum (PAIVS) is a rare and complex congenital heart disease characterized by imperforate pulmonary valve and intact ventricular septum. Treatments have been improved over time: surgical techniques have become safer during the neonatal period and percutaneous intervention has been developed as an alternative. This resulted in a reduction in mortality over time, but clinical trend, final cardiac physiology (biventricular 2VR, one-and-a-half 1+½VR, univentricular 1VR), long term complications and therapeutic history can be very complex and currently not well known.

Purpose of this study is to clarify this topic by analyzing a surviving population affected by PAIVS in retrospect.

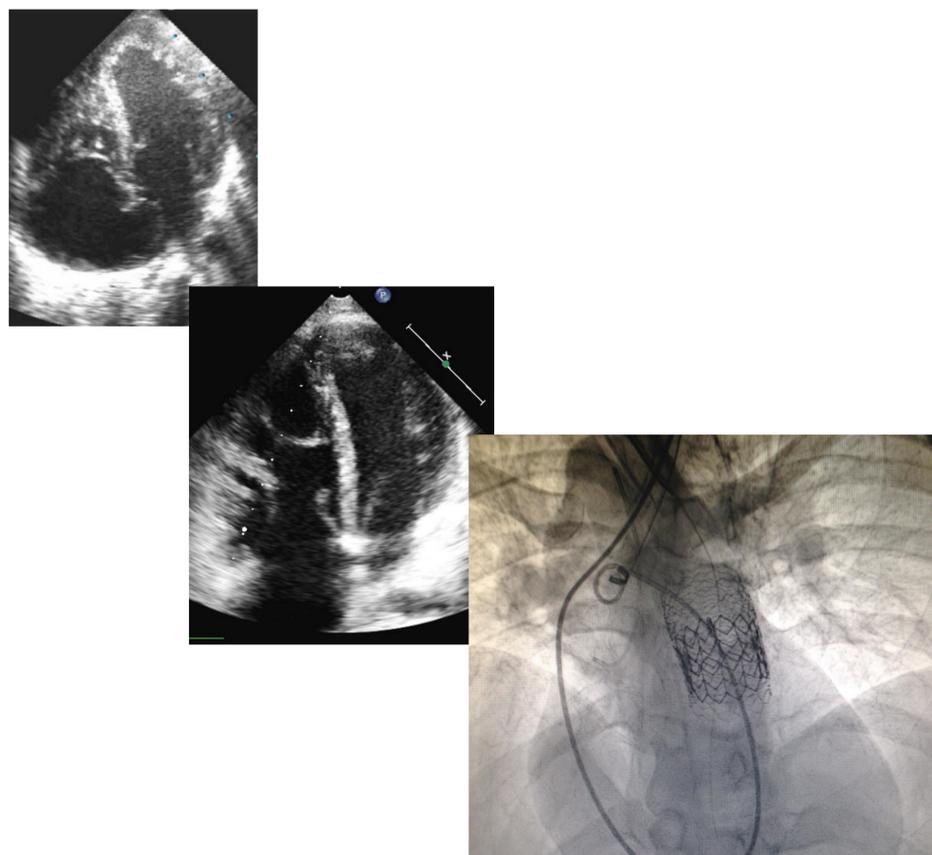
## Methods:

- 59 patients (35 male, 24 female) affected by PAIVS older than 6 years at last evaluation were selected.
- Heart anatomy with echocardiography, angiography and cardiac MRI (when available) was evaluated in the neonatal period (tricuspid valve Detroit z-score, right ventricular morphology, tricuspid valve dysplasia) and during the follow-up.
- Therapeutic history were evaluated (first therapeutic approach, surgical or percutaneous interventions in first year and during follow-up).
- Final cardiac physiology, clinical outcome (NYHA class, O2 sat) and late complications were evaluated in a median follow-up of 16 years.

## Results:

### Therapeutic history

- 9 patients (15%) underwent palliation surgery.
- 16 patients (27%) underwent percutaneous right ventricular decompression;
- Palliated patients had lower tricuspid valve (TV) z-score, smaller RV and less TV regurgitation ( $p < 0,05$ ).
- Patients threatened by percutaneous approach had only higher TV z-score ( $p < 0,05$ ) than patients surgically threatened, no difference in RV morphology or TV regurgitation.



## Conclusion

- Most of PAIVS patients has reached 2VR physiology at follow-up.
- Patients with final 2VR had higher tricuspid neonatal z-score, more neonatal tricuspid valve regurgitation and more represented right ventricle.
- Therapeutic history is characterized by a lot of interventions.
- After several years a significant amount of 2V patients underwent pulmonary valve replacement for severe pulmonary insufficiency and severe right ventricle dilatation.
- Most patients are asymptomatic at last evaluation.
- Arrhythmias is not an issue.

## Statics:

Results are reported as mean±SD or median±SD. T-test for analysis of continuous variables and Chi-squared for categorical variables were used.

## Results::

### Neonatal characteristic

|                              |            |
|------------------------------|------------|
| Nr patients                  | 59         |
| Birth weight (Kg)            | 3,00±0,58  |
| Tricuspid z-score            | -1,55±1,75 |
| Tricuspid dysplasia          | 25%        |
| Significant TR regurgitation | 87%        |
| RV tripartite                | 41%        |
| RV bipartite                 | 44%        |
| RV monopartite               | 15%        |

### Last follow-up

- 39 of 50 eligible patients (78%) at last reached 2VR physiology.
- 14 patients have 1 VR, 6 patients 1½ VR physiology.
- Patients with final 2 VR physiology had higher TV z-score, more TV regurgitation and bigger RV at newborn ( $p < 0,05$ ).
- Final 2 VR physiology is not influenced by the type of RV decompression (surgical or percutaneous).
- Most patients are asymptomatic at last follow-up irrespective of final physiology (80% at NYHA I class).
- Only 10% of patients had arrhythmic problems (all supraventricular tachycardia).

### Pulmonary issue

- 50% of patients with final 2 VR physiology had severe pulmonary valve (PV) regurgitation during follow-up.
- PV regurgitation is not dependent on first procedure/surgical intervention of RV decompression.
- 12 patients underwent pulmonary valve replacement because of severe RV enlargement.
- RV volume decreased after PV replacement (MRI data average 141±19 ml/mq before, 89±31 ml/mq after).
- Neonatal TV z-score was significant higher in patients who had developed significant RV enlargement (0.37 vs 1.29,  $p < 0,05$ ).