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Surgical Outcomes of Total Anomalous Pulmonary Venous Drainage at Dr. Soetomo General Academic Hospital, Surabaya: A One-Year Case Series

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ABSTRACT

Introduction: Total anomalous pulmonary venous drainage (TAPVD) is a rare anomaly, accounting for 1% to 3% of patients with congenital heart disease. Characterized by the abnormal connection of the pulmonary veins to the systemic venous circulation, rather than to the left atrium. This case series aims to emphasized the significance of early diagnosis, surgical strategy, and outcomes associated with TAPVD.

Methodology: Over a one-year period, we treated 5 cases of TAPVD at our institution, involving patients aged one to nine years, with a male-to-female ratio of 1.5:1. The patients exhibited various clinical symptoms including cyanosis, and respiratory distress. Echocardiography was the primary diagnostic method, supported by cardiac catheterization and computed tomography cardiac imaging for detailed anatomical evaluation.

Surgical correction was performed in all cases, with techniques tailored to the specific anatomical variations of TAPVD, in this cases supra-cardiac, intra-cardiac, and mixed types. The surgeries aimed to redirect the pulmonary veins to the left atrium and close the interatrial defect. Postoperative care involved intensive management of potential complications like pulmonary hypertension.

RESULTS: Three out of five cases experienced postoperative complications due to serious preoperative conditions. Nevertheless, this case series revealed a 100% survival rate, with an average ICU stay of 5.7 days and an overall hospital stay averaging 7.7 days. Short-term follow-up revealed significant symptoms improvement, highlighting the importance of timely surgical intervention for achieving favorable outcomes.

Conclusion: This case series underscores the significance of early diagnosis, meticulous surgical strategy, and effective postoperative care in enhancing outcomes for TAPVD. of tubers (10.20 and 10.30 per plant), were recorded in plots having tuber treatment *Terminila arjuna* bark extract and two foliar sprays of Enviro (botanical virucides)followed by tuber treatment and six sprays with *Tinospora cordifolia* (aerial stem extract) and uber treatment and six foliar spray with *Allium sativum* (garlic clove extract) during 2022-23 and 2023-24, respectively.

1. INTRODUCTION

Total anomalous pulmonary venous drainage (TAPVD) is a rare congenital cardiac anomaly, constituting approximately 1–3% of all congenital heart defects [1]. In this condition, the pulmonary veins fail to connect to the left atrium, instead draining into systemic venous circulation, leading to severe cyanosis, pulmonary hypertension, and congestive heart failure if untreated [2]. Without surgical intervention, 80% of infants die within the first year of life [3].

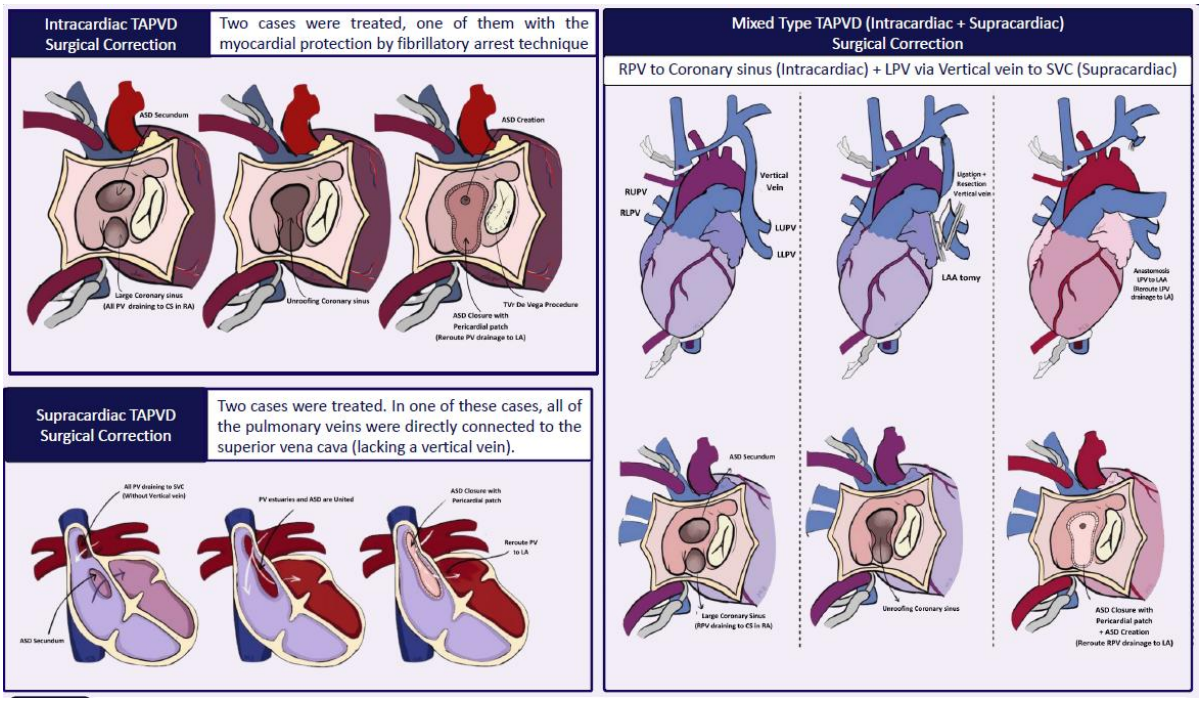

Figure 1. The TAPVD surgical correction technique is applied in a variable manner, according to the specific anatomical variations.

Recent advancements in diagnostic imaging and surgical techniques have improved outcomes, yet challenges persist, particularly in managing postoperative pulmonary vein obstruction (PVO) and pulmonary hypertension [4]. The heterogeneity of TAPVD anatomical subtypes—supracardiac, intracardiac, cardiac, and mixed—demands individualized surgical approaches [5]. TAPVD surgical correction techniques according to the specific anatomical variations are shown in figure 1.

This case series evaluates the one-year surgical outcomes of TAPVD repair at a tertiary care center in Indonesia, emphasizing the roles of early diagnosis, tailored surgical strategies, and intensive postoperative care.

2. METHODOLOGY

Patient Cohort

- Demographics: 5 patients (3 male, 2 female; age 1–9 years).
- Symptoms: Cyanosis (100%), respiratory distress (80%), failure to thrive (60%).

Diagnostic Workup

1. Echocardiography: Initial diagnosis in all cases.
2. Cardiac CT/Catheterization: Detailed anatomy in complex cases (n=3).

Surgical Techniques

- Supracardiac (n=2): Anastomosis to left atrium; one case lacked a vertical vein.
- Intracardiac (n=2): Fibrillatory arrest myocardial protection in one case.
- Mixed (n=1): Combined approach.
- Fenestrated ASD: Created universally for pulmonary hypertension management.

3. RESULTS

Outcome	Value
Survival Rate	100%
Mean ICU Stay	5.7 days (range 4–8)
Hospital Stay	7.7 days (range 6–10)
Complications	60% (3/5)

Complications:

- Pulmonary hypertension crisis (n=2), managed with milrinone and nitric oxide.
- Junctional ectopic tachycardia (n=1), resolved with amiodarone.

Follow-up: All patients showed symptom resolution at 3-month evaluation.

4. DISCUSSION

Our study demonstrated a 100% survival rate among five TAPVD patients, with a mean ICU stay of 5.7 days and hospital stay of 7.7 days. These outcomes align with contemporary studies reporting survival rates exceeding 90% in high-volume centers [6]. However, 60% of our cohort (3/5 patients) developed complications, primarily pulmonary hypertension crises (n=2) and arrhythmias (n=1). This complication rate is comparable to data from Hu et al. (2023), who observed postoperative PVO in 25% of cases despite sutureless repair techniques [7].

Key Findings and Literature Comparison

1. **Fenestrated ASD Creation**
    - We utilized fenestrated ASDs universally to mitigate postoperative pulmonary hypertension, a strategy supported by Mulla & Rahman (2023) [8]. This approach allows right-to-left shunting, reducing acute right ventricular strain.
2. **Surgical Adaptations for Anatomical Variants**
    - In one supracardiac TAPVD case, the absence of a vertical vein necessitated direct anastomosis to the left atrium, a technique with reported success in similar cases [9].
    - Fibrillatory arrest was employed for myocardial protection in an intracardiac repair, minimizing ischemic time, as advocated by Li et al. (2023) [10].
3. **Postoperative Complications**
    - Both patients with pulmonary hypertension responded to milrinone and nitric oxide, consistent with protocols described by Park & Salamat (2020) [3].
    - The single arrhythmia case (junctional ectopic tachycardia) resolved with amiodarone, reflecting standard management [11].

Limitations

- Small sample size (n=5) limits generalizability.
- Short follow-up duration precludes assessment of long-term PVO risks, which affect 10–20% of patients within 5 years [7].

Clinical Implications

- Early diagnosis (via echocardiography ± CT/catheterization) is critical to prevent irreversible pulmonary vascular disease [2].

Institutional protocols for postoperative pulmonary hypertension management (e.g., fenestrated ASD, targeted pharmacotherapy) should be standardized [8].

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