



Guidance document for PM JAY package

VSD closure + RV - PA conduit

Procedures covered/ Procedure Count: 1

Specialty: CTVS

Package name	Procedure name	HBP 1.0 code	HBP 2.0 code	Package price (INR)	ALOS
Surgical Correction of Category - III Congenital Heart Disease	VSD closure + RV - PA conduit	New Package	SV003L	150,000 + Cost of implant	12 days

Minimum qualification of the treating doctor:

Essential: M.Ch./DNB/equivalent (Cardiothoracic Surgery)

Special empanelment criteria/linkage to empanelment module: Cardiothoracic Surgery OT

Disclaimer:

For monitoring and administering the claim management process of **VSD closure + RV - PA conduit**, NHA shall be following these guidelines. This document has been prepared for guidance of PROCESSING TEAM and TRANSACTION MANAGEMENT SYSTEM of AB PM-JAY for the claims of procedures mentioned above. The hospitals can also refer to this document so that they have the insight on how the claims will be processed. However, this document doesn't provide any guidance on clinical and therapeutic management of patient. In that respect the hospitals and physicians may refer to any other relevant material as per the extant professional norms.

PART I: GUIDELINES FOR CLINICIANS AND HEALTHCARE PROVIDERS

1.1 Objective:

The purpose of this section is to act as a guidance & a clinical decision support tool for the clinicians in deciding the line of treatment, plan clinical management of patient and decide referral of cases to the appropriate level of care (as required) for treatment of patients under PMJAY and selection of corresponding Health Benefit Package.

It will also serve as a tool for hospitals to determine and submit the mandatory documents required for claiming reimbursement of health benefit package under PMJAY.

1.2 Clinical key pointers:

VSD with pulmonary atresia (VSD-PAt) is the most severe form of Tetralogy of Fallot (TOF). In the Bohemia survival study, a prevalence of 0.07 per 1000 live births was observed for VSD-PAt, accounting for 20% of all forms of TOF. About 20% of cases are associated with a

syndrome/genetic defect. This CHD usually presents in the neonatal period with cyanosis due to right-to-left shunt at ventricular level. The degree of cyanosis depends on the magnitude of pulmonary blood flow, which in turn, depends on the size of PDA and/or the number and size of aortopulmonary collaterals. Closure of the ductus in the early neonatal period can be lethal due to acute severe cyanosis. After the neonatal period, cyanosis gradually increases. Rarely, however, patients may present in congestive heart failure due to multiple aortopulmonary collaterals (MAPCAs) or a large PDA.

Anatomical types

VSD-PAt is a complex disease with varying anatomy, especially related to pulmonary artery branches and sources of pulmonary arterial blood supply. The detailed anatomy must be defined in each case to plan management. For the sake of simplicity, VSD-PAt is classified into four types.

- Type A – Short-segment valvular atresia, pulmonary arteries confluent, and good sized, supplied by a PDA
- Type B – Long-segment pulmonary atresia with absent main pulmonary artery. Branch pulmonary arteries confluent and good sized, supplied by a PDA
- Type C – Long-segment pulmonary atresia with absent main pulmonary artery. Branch pulmonary arteries confluent, but pulmonary blood flow dependent predominantly on MAPCAs
- Type D – Long-segment pulmonary atresia with absent main pulmonary artery. Nonconfluent branch pulmonary arteries with MAPCA-dependent pulmonary blood flow.

Diagnosis:

Echocardiography is a vital tool for the diagnosis; however, it may not delineate the distal pulmonary arterial tree or the sources of pulmonary arterial supply. Hence, additional imaging in the form of cardiac catheterization, CTA/cMRI, or a combination of these is essential for planning definitive repair.

Indications and timing of intervention:

Management depends on the type of VSD-PAt, the institutional experience, and the clinical presentation. In general, this lesion requires a multistage management. Patients with Type C and D have a more complex anatomy and are best referred to a specialized center for further treatment.

- Type A (short-segment VSD-PAt with PDA)

- i. Presentation with significant cyanosis at < 1 year of age: Aortopulmonary shunt (Class I) or PDA stenting (Class IIa)
 - ii. After 1st intervention or those presenting at ≥1 year of age: Total correction at about 1 year of age, since a RV-to-pulmonary artery (PA) conduit is not required (Class I).
- Type B (long-segment pulmonary atresia with PDA):
 - i. Presentation with significant cyanosis at <1 year of age: Aortopulmonary shunt (Class I) or PDA stenting (Class IIa) depending on the institutional preference and feasibility.
 - ii. After 1st intervention or in those presenting at ≥1 year of age (Class I):
 - a. Optimal pulmonary blood flow with good-sized PAs – Total repair with RV-PA conduit at 3–4 years
 - b. Suboptimal pulmonary blood flow with small PAs – additional shunt followed by total repair with RV-PA conduit at 3–4 years
 - c. Increased pulmonary blood flow with large PAs – total repair with RV-PA conduit by 1 year
- Type C (long-segment pulmonary atresia with confluent branch pulmonary arteries supplied by MAPCAs) (Class I):
 - i. Neonatal presentation – aortopulmonary shunt + unifocalization of MAPCAs or RV-PA conduit keeping VSD open
 - ii. After 1st intervention or late presentation
 - a. Optimal pulmonary blood flow with good-sized PAs – total repair with RV-PA conduit and VSD closure at 3–4 years
 - b. Borderline PAs with large MAPCAs
 - i. Unifocalization + RV-PA conduit at 6–12 months
 - ii. Total repair with RV-PA conduit and VSD closure at 3–4 years.
 - iii. Increased pulmonary blood flow and large PAs – single-stage repair (unifocalization of MAPCAs + RV-PA conduit + VSD closure) at about 1 year of age with a preferable weight of >10 kg.
- Type D (long-segment pulmonary atresia with nonconfluent branch pulmonary arteries supplied by MAPCAs) (Class IIa):
 - i. Neonatal presentation – Aortopulmonary shunt + unifocalization of MAPCAs.
 - ii. After 1st intervention or late presentation

- Unifocalization + RV-PA conduit at 6–12 months
- Total repair with RV-PA conduit and VSD closure at 3–4 years.

1.3 Mandatory documents- For healthcare providers

Following documents should be uploaded by the concerned hospital staff at the time of pre-authorization and claims submission

Mandatory document	VSD closure + RV - PA conduit
i. At the time of Pre-authorization	
a. Clinical notes	Yes
b. Echo/Doppler report	Yes
ii. At the time of claim submission	
a. Indoor case papers	Yes
b. Procedure / Operative notes	Yes
c. Post procedure stills of ECHO with report	Yes
d. Detailed Discharge Summary	Yes

PART II: GUIDELINES FOR PROCESSING TEAM

PART III: GUIDELINES FOR TRANSACTION MANAGEMENT SYSTEM (TMS)

3.1 Objective: To enable setting up of cross check mechanisms/rule engines within the IT platform (TMS) to ensure compliance with STGs and to prevent fraud / abuse of the Health Benefit Package.

3.2 Below mentioned are the scenarios where a provision would be built in TMS for pop-ups:

1. Was the Echo/ Doppler report suggestive of Ventricular Septal defect with pulmonary atresia? Yes

Till the time the functionality is being developed, the processing doctors shall check the above manually.

References

- Saxena A, Relan J et al Indian guidelines for indications and timing of intervention for common congenital heart diseases: Revised and updated consensus statement of the Working group on management of congenital heart diseases. Ann Pediatr Card 2019;12:254-86
- Kouchoukos NT, Blackstone EH, Hanley FL, Kirklin JK. Kirklin/Barratt-Boyes Cardiac Surgery: Expert Consult-Online and Print (2-Volume Set). Elsevier Health Sciences; 2012 Oct 26.
- Mavroudis C, Backer C. Pediatric cardiac surgery. Blackwell Publishing Ltd; 2013 Feb 28.