



## Guidance document for PM JAY package

### Total Anomalous Pulmonary Venous Connection (TAPVC) Repair

Procedures covered/ Procedure Count: 1

Specialty: CTVS

Package name	Procedure name	HBP 1.0 code	HBP 2.0 code	Package price	ALOS
Surgical Correction of Category - III Congenital Heart Disease	Total Anomalous Pulmonary Venous Connection (TAPVC) Repair	S1300033	SV003S	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 **TAPVC Repair**, 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:**

Total Anomalous Pulmonary Venous Connection (TAPVC) occurs in 0.5–0.9/10,000 live births and accounts for 1% of all patients born with congenital heart defects. It has no specific sex predilection. An ASD is necessary for survival. TAPVC frequently occurs as an isolated lesion,

but may be associated with other more complex CHDs such as heterotaxy syndrome with asplenia. TAPVC is classified into four types depending on the site of drainage. Each type can be obstructive or nonobstructive. The four types and their relative frequencies are: Supracardiac (45%–50%), cardiac (15%–20%), infracardiac (26%–28%), and mixed (where the drainage is at two or more sites, 5%–8%). Obstruction to the drainage of pulmonary veins is most common in the infracardiac variety and least common in the cardiac type. If not treated, TAPVC has a very high mortality with 85% dying in the 1<sup>st</sup> year of life. The median survival was 2 months (range 1 day to 49 years) in the article published by Hazelrig *et al.* However, some of the exceptional survivors present later in life with clinical features suggestive of a large ASD, but have mild desaturation. Pulmonary venous obstruction significantly reduces median survival from 2.5 months in the nonobstructive group to 3 weeks in the obstructive group. Obstructive TAPVC is the only cyanotic CHD where prostaglandin infusion should be avoided.

### Diagnostic workup

- i. Clinical assessment: Depends on whether the obstruction is present or not. Neonates with severe obstruction present with cyanosis and respiratory distress soon after birth, usually within 12 h of birth. Patients with no obstruction present with clinical features of large left-to-right shunts with mild cyanosis.
- ii. Pulse oximetry: For quantifying cyanosis which may give an estimate of PVR.
- iii. ECG: Right-axis deviation with right ventricular hypertrophy.
- iv. X-ray chest: Shows cardiomegaly, prominent pulmonary artery segment, and pulmonary plethora. In older infants and children, one may see a typical figure of 8 or “snowman sign” in cases with supracardiac TAPVC. Neonates with obstructive TAPVC show no or minimal cardiomegaly and severe pulmonary venous hypertension (ground glass appearance).
- v. Echocardiography: It is the investigation of choice and gives complete information in majority of patients. The right atrium, RV, and pulmonary artery are dilated. The most important finding is the inability to show connection of pulmonary veins to left atrium and right-to-left shunting through an ASD. The exact site of drainage and the presence or absence of obstruction can be defined, although may be difficult in mixed variety with > 1 site of drainage. Assessment of pulmonary artery pressure is possible with echo-Doppler, and associated anomalies can also be identified.
- vi. Cardiac catheterization is very rarely performed. It may be required in patients with pulmonary artery hypertension presenting beyond infancy, where operability is in doubt. An arterial saturation of >85% and a PVR (indexed) of <8 WU.m<sup>2</sup> may indicate that the patient is operable.
- vii. CTA/cMRI: They are reserved for patients where echocardiography is inconclusive, as in cases with mixed type of TAPVC.

## Indications and timing of surgery (all are Class I recommendations)

- All patients need surgical repair.
- Patients with obstructive TAPVC should undergo emergency surgery.
- Surgery should be performed as early as possible in nonobstructive TAPVC, even if they are asymptomatic.
- Those presenting late should be evaluated for onset of pulmonary vascular disease and operated if the data suggest operable status.

## Important determinants of long-term prognosis

These include residual pulmonary vein stenosis, residual pulmonary hypertension, progressive stenosis of surgically created anastomosis, and late-onset arrhythmias. Pulmonary venous obstruction occurs in 5%–15% of patients after surgical repair.

### 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	TAPVC repair
<b>i. At the time of Pre-authorization</b>	
a. Clinical notes	Yes
b. Echo/Doppler report	Yes
<b>ii. At the time of claim submission</b>	
a. Procedure / Operative notes	Yes
b. Post procedure ECHO with reports	Yes
c. 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 TAPVC? Yes

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



## References

1. Saxena A, Relan J, Agarwal R, 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
2. Ferencz C, Rubin JD, McCarter RJ, Brenner JI, Neill CA, Perry LW, *et al.* Congenital heart disease: Prevalence at livebirth. The Baltimore-Washington infant study. *Am J Epidemiol* 1985;121:31-
3. Craig JM, Darling RC, Rothney WB. Total pulmonary venous drainage into the right side of the heart; report of 17 autopsied cases not associated with other major cardiovascular anomalies. *Lab Invest* 1957;6:44-64.
4. Seale AN, Uemura H, Webber SA, Partridge J, Roughton M, Ho SY, *et al.* Total anomalous pulmonary venous connection: Morphology and outcome from an international population-based study. *Circulation* 2010;122:2718-26
5. Hazelrig JB, Turner ME, Blackstone EH. Parametric survival analysis combining longitudinal and cross-sectional – Censored and interval-censored data with concomitant information. *Biometrics* 1982;38:1-15.