



Guidance document for PM JAY package

Truncus Arteriosus Repair

Procedures covered/ Procedure Count: 1

Specialty: CTVS

Package name	Procedure name	HBP code 1.0	HBP code 2.0	Package price (INR)	ALOS
Surgical Correction of Category - III Congenital Heart Disease	Truncus Arteriosus Repair	S1300039	SV003T	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 **Truncus Arteriosus 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:

Persistent truncus arteriosus is characterized by a single arterial vessel that arises from the base of the heart and gives origin to the coronary, pulmonary, and systemic arteries. It accounts for < 1% of all congenital heart defects with a prevalence rate of 0.07/1000 live births. There is associated microdeletion of chromosome 22q11.2 in about 40% of

patients. The aortic arch is right sided in about one-third of patients. Aortic arch interruption or coarctation of the aorta is found in 15%–20% of patients with truncus arteriosus. Physiologic consequences of truncus arteriosus depend on the size of the pulmonary arteries and on the PVR, with truncal valve regurgitation/stenosis adding to the hemodynamic burden on the ventricles. Patients usually present in the first few weeks of life due to congestive heart failure and failure to thrive. Untreated patients have a very high mortality, mainly due to congestive heart failure, with a survival rate of 35% at 6 months and 10% at 1 year.

Classification of truncus arteriosus

- i. Type A1 – Aorta and main pulmonary artery originate from a single large common trunk.
- ii. Type A2 – Both pulmonary arteries arise separately and directly from the truncus.
- iii. Type A3 – One pulmonary artery arises from the truncus and the other is supplied by the PDA or collaterals from the aorta.
- iv. Type A4 – There is an associated obstructive lesion of the aortic arch.

Diagnostic workup

- i. Clinical assessment: Little or no cyanosis with bounding pulses in early infancy.
- ii. Pulse oximetry: For quantifying cyanosis which may give an estimate of PVR.
- iii. X-ray chest: It shows cardiomegaly and increased pulmonary vascular markings. A combination of a right aortic arch and increased pulmonary vascularity is strongly suggestive of truncus arteriosus. A dilated truncal root resembles a dilated ascending aorta. Main pulmonary artery segment may arise at a higher level (Type A1) or may be absent (when pulmonary arteries arise directly from truncus).
- iv. ECG: There is usually a normal QRS axis or minimal right-axis deviation and combined ventricular hypertrophy and left atrial enlargement. When pulmonary blood flow is reduced due to increase in PVR, there is right-axis deviation and predominant right ventricular hypertrophy.
- v. Echocardiography: It is the key tool for the diagnosis and assessment of anatomy, location of VSD, presence and severity of truncal valve regurgitation or stenosis, and for associated lesions such as aortic arch interruption and coronary artery anomalies.
- vi. CTA/cMRI: They are useful in select cases when the anatomy is unclear on echocardiography, especially for the evaluation of aortic arch. These tests are recommended for follow-up imaging after surgical intervention.
- vii. Cardiac catheterization: Indicated in older patients beyond infancy with suspected pulmonary vascular disease, for the assessment of operability.



- viii. Serum calcium levels and genetic testing for microdeletion of chromosome 22q11.2 when clinically indicated.

Ideal age for surgery

Surgery indicated in all, unless the patient is inoperable.

- i. Uncontrolled heart failure: Surgical repair as soon as possible (Class I).
- ii. Controlled heart failure: Surgical repair by 3–6 weeks of age (Class I).
- iii. Bilateral pulmonary artery banding reserved for complex cases and patients with contraindications for cardiopulmonary bypass (Class IIb).

Type of surgery

Total repair using RV-to-pulmonary artery conduit. Non-conduit options (Barbero-Marcial technique) may be possible in select cases. The prospects of repeat surgeries in future for conduit obstruction should be discussed with parents. Truncal valve repair is done if truncal valve is regurgitant.

Contraindication for surgery

Severe pulmonary arterial hypertension with irreversible pulmonary vascular occlusive disease (Class III). Signs of inoperability include age >1 year, resting systemic arterial saturation <85%, and absence of cardiomegaly.

Patients with borderline operability due to pulmonary vascular disease should be referred to a higher center for further evaluation. The decision to operate or not should be made on an individual basis taking into account the history, examination, and results of all the investigations.

Important determinants of long-term prognosis

These include residual or progressive pulmonary hypertension, need for conduit replacement, progressive truncal/neo-aortic valve regurgitation, aortic root dilatation/aneurysm, and recurrent arch obstruction in Type A4.

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	Truncus Arteriosus Repair
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 persistent truncus arteriosus? 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. 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.
3. Mavroudis C, Backer C. Pediatric cardiac surgery. Blackwell Publishing Ltd; 2013 Feb 28.
4. Marino B, Digilio MC, Toscano A. Common arterial trunk, DiGeorge syndrome and microdeletion 22q11. Prog PediatrCardiol2002;15:9-17