

Guidance document for PM JAY package

Arterial switch operation

Procedures covered/ Procedure Count: 1

Specialty: CTVS

Package name	Procedure name	HBP code 1.0	HBP code 2.0	Package price	ALOS
Surgical Correction of Category - III Congenital Heart Disease	Arterial Switch Operation (ASO)	S1300035	SV003W	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 **Arterial switch operation**, 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:

Transposition of great arteries (TGA) is the most common cyanotic CHD at birth, accounting for approximately 5% of all CHDs. In 70% of cases, there is no associated defect apart from ASD, PDA, or insignificant VSD; these cases are labeled as having simple TGA. Association of TGA with other defects such as large VSD and left ventricular outflow tract obstruction or coarctation (CoA) is referred to as complex TGA. It is a serious disease and most patients

with TGA present very early in life, within few days or weeks after birth. The average life expectancy for an untreated newborn is 0.65 years, with mortality rate at 1 year being close to 90%. With the advent of improved surgical techniques and postoperative care, long-term survival is > 90% with very low reintervention rates. The best surgical option for patients with TGA is an arterial switch operation (ASO) where both arteries are switched so that ventriculoarterial concordance is achieved. However, ASO should be performed early, preferably within 3–4 weeks of life, before the left ventricular mass and volume regresses secondary to fall of PVR after birth.

Diagnostic workup

- i. Clinical assessment: In simple TGA, cyanosis is the dominant feature; no heart murmur is audible in most cases. Those with large VSD and/or PDA present with features of heart failure. Weak femoral pulses indicate associated CoA.
- ii. Pulse oximetry and blood gases measurement: Oxygen saturation should be measured in the right arm (preductal), because lower-limb saturation may be higher. In critically ill newborns, arterial (or venous) partial pressure of oxygen should be measured to assess degree of hypoxemia, lactic acidosis, and circulatory failure.
- iii. ECG: It shows right-axis deviation and right ventricular hypertrophy, which, however, may be normal for a newborn. Those with large VSD may show good left ventricular forces in addition.
- iv. X-ray chest: It can be completely normal or may have narrow pedicle with cardiomegaly (egg-on-side appearance) and increased pulmonary blood flow.
- v. Echocardiography: Key diagnostic imaging tool. It shows the following features:
 - a. Atrioventricular concordance with ventriculoarterial discordance.
 - b. Anterior and rightward position of aorta compared to pulmonary artery.
 - c. Site and adequacy of intermixing: size of ASD, VSD, and PDA.
 - d. Associated malformations: VSD, left ventricular outflow tract obstruction, coarctation/interruption of aortic arch, and mitral valve abnormalities.
 - e. Size of aortic and pulmonary valve annulus and coronary artery origin and course, for planning ASO.
- vi. Adequacy of left ventricle to support systemic circulation after an ASO.
- vii. Cardiac catheterization: Generally done for performing balloon atrial septostomy. Occasionally, it may be required for the assessment of adequacy of left ventricle for an ASO or to assess PVR in those presenting late.
- viii. CTA and cMRI: Rarely performed to clarify anatomy of the aortic arch or to evaluate for a surgically relevant coronary anomaly suspected on echocardiography.

Indications and timing of surgery

Surgery is indicated for all patients with TGA except in those with irreversible pulmonary vascular disease.

Timing and type of surgery

- i. TGA with intact ventricular septum presenting soon after birth: ASO is the best option (Class I)
- ii. Timing of surgery: 7 days to 3 weeks. Surgery indicated earlier, if baby is unstable or has associated persistent pulmonary hypertension of the newborn. Exact timing based on institutional preference, but is best done before 4 weeks.
- iii. TGA with intact ventricular septum presenting beyond 3–4 weeks of life with regressed left ventricle:
 - a. Presenting between 1 and 2 months: ASO; extracorporeal membrane oxygenation (ECMO) support may be required in some cases (Class IIa)
 - b. Presenting between 2 and 6 months: ASO with ECMO support or rapid two-stage ASO¹ or an atrial switch (if rapid two-stage or ECMO not feasible) (Class IIa)
 - c. Presenting between 6 months and 2 years: Atrial switch operation (Senning or Mustard operation) (Class IIa). Rapid two-stage ASO¹ to be considered in select cases after detailed evaluation (Class IIb).
 - d. TGA with a large VSD and/or a large PDA: ASO with VSD and/or PDA closure by 6 weeks of age (Class I). These patients develop early pulmonary vascular disease and may become inoperable by 6 months to 1 year of age.
 - e. TGA with VSD and CoA: ASO with VSD closure and arch repair as soon as possible (Class I). It is preferable to repair all lesions in a single stage.
 - f. TGA with VSD and significant left ventricular outflow obstruction (Class I):
 - a. Sub-valvular pulmonary obstruction with normal or near-normal pulmonary valve and pulmonary annulus: ASO with resection of sub-valvular stenosis
 - b. If obstruction involves pulmonary valve or is sub-pulmonary but not amenable to resection:
 - i. Neonates and infants presenting with significant cyanosis: The options depend on patient's age and surgeon's preference
 - ii. Systemic-to-pulmonary shunt (at any age) followed by Rastelli-type repair or root translocation (at 2–3 years of age or when the child weighs >10 kg)
 - iii. Réparation à l'Étage Ventriculaire (REV) procedure (usually done at 4–6 months)
 - iv. Pulmonary root translocation (usually done at 6–12 months)
 - v. Nikaidoh procedure (usually done beyond 6–9 months of age).

- vi. In older, stable patients, presenting beyond 2–3 years of age: One of the following surgeries: Rastelli-type repair, Nikaidoh procedure, or root translocation surgery.
- c. If the VSD is remote and not amenable to one of the biventricular repairs: Multistage palliative cavopulmonary connection (Class IIa).

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	Arterial switch operation
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 Transposition of Great Arteries? Yes

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

References

1. Saxena A, Relan 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. Kouchoyos 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.