



Guidance document for PM JAY package

Double switch operation

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	Double switch operation	S1300036	SV003B	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 **Double 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:

Congenitally corrected TGA (ccTGA) is a congenital heart defect characterized by atrioventricular and ventriculoarterial discordance. This double discordance results in physiologically corrected circulation, as the great arteries receive appropriate blood. ccTGA occurs due to abnormal looping of the primitive cardiac tube to the left instead of to the

right. Although the physiology is corrected, anatomy is not, and morphologic RV supports systemic arterial circulation. In over 90% of cases, ccTGA is accompanied by other cardiac lesions: VSDs in 80%, PS (outflow obstruction to morphologic left ventricle) in 30%–50%, abnormalities of the conduction system in 15%–50%, ASD in 12%, and tricuspid valve abnormalities (as detected at autopsy) in 90%. Mismatch between visceral situs and cardiac position is common, with dextrocardia/mesocardia in one-fourth of patients. The age of presentation is variable, and the presentation is determined by the associated anomalies. Patients present with heart failure if a large VSD is present and as TOF physiology if large VSD is associated with significant PS. The associated anomalies affect the natural history of ccTGA. Approximately 10% are born with complete heart block (CHB). In the rest, the risk of developing CHB is 2% per year and about 30% develop CHB by adulthood. Function of the RV deteriorates after the second decade even in those without any associated anomaly.

Diagnostic workup

- i. Clinical assessment: Features of heart failure, cyanosis, bradycardia, and systolic murmur of tricuspid regurgitation.
- ii. Echocardiography: Main imaging tool for determining the situs and cardiac position, atrioventricular and ventriculoarterial discordance, relationship of great arteries (aorta anterior and to left of pulmonary artery in situs solitus cases), and associated anomalies. Echocardiography is also very useful for the diagnosis of Ebstein's malformation of left-sided morphologic tricuspid valve, quantification of tricuspid regurgitation and for the assessment of right ventricular function. Older patients may require a transesophageal echo for complete evaluation.
- iii. Cardiac catheterization: Generally not required for making the diagnosis. It may have to be performed for demonstrating coronary anatomy or for the measurement of pulmonary artery pressure and PVR in those with large left-to-right shunts.
- iv. cMRI: In adults, diagnostic imaging alone with echo could be inconclusive; cMRI can provide the required information.

Indications and timing of surgery

The double switch procedure (atrial switch plus arterial switch or Rastelli) is a surgical technique used to repair congenitally corrected transposition of the great arteries (cc-TGA)

The procedure consists of any of the following surgical combinations:

Atrial inversion surgery (either the Senning or the Mustard repair) and Jatene arterial switch

Atrial inversion surgery (either the Senning or the Mustard repair) and Rastelli-type repair (also known as the Ilbawi or Senelli procedure)

The indications and timing of surgery in ccTGA depend on the presence and type of associated anomalies.

General recommendations:

- i. Tricuspid valve (systemic AV valve) surgery for severe regurgitation should be considered before systemic ventricular failure (ejection fraction < 45%) sets in (Class IIa).
- ii. Anatomic repair (double-switch operation-atrial switch plus arterial switch or Rastelli) may be considered when left ventricle is functioning at systemic pressure and when such surgery is feasible (Class IIa).
- iii. Indications and timing for specific groups of congenitally corrected transposition of great arteries
 - a. No associated anomalies: Medical follow-up to look for any development of tricuspid regurgitation or right ventricular dysfunction (Class I). Neonatal double-switch operation may be considered (Class IIb).
 - b. Associated with large VSD:
 - i. <3 months: Pulmonary artery banding followed later by double-switch operation (atrial plus arterial switch) (Class I).
 - ii. >6 months: Double switch (atrial plus arterial switch), provided that the patient has not developed irreversible pulmonary vascular disease (Class I).
 - iii. 3–6 months: Pulmonary artery banding followed by double-switch operation or direct double-switch operation depending on institutional policy (Class IIa).
 - c. Associated with large VSD and left ventricular outflow obstruction (PS):
 - i. VSD routable: Double switch (atrial switch plus Rastelli) (Class I); univentricular repair pathway if the surgeon is not comfortable doing double-switch operation and saturation is low (Class IIa).
 - ii. VSD not routable:
 - i. Saturation good – Medical follow-up after informed discussion with family (Class IIa).
 - ii. Saturation low – Univentricular repair pathway (Class I).
 - iii. Associated with complex cardiac malformations: Physiological biventricular repair, root transfer (for nonroutable VSD), or univentricular repair (Class IIa).
 - iv. Associated with CHB: Permanent, dual-chamber pacemaker implantation (Class I).
 - v. Associated with progressive development of isolated, severe tricuspid regurgitation or right ventricular dysfunction in a child or adolescent: Cardiac catheterization to measure left ventricular pressure (Class IIa).

- vi. If left ventricular pressure is $\geq 80\%$ of right ventricular pressure: Double-switch operation.
- vii. If left ventricular pressure is $< 80\%$ of right ventricular pressure: pulmonary artery banding to prepare left ventricle.

It is associated with severe tricuspid regurgitation in an adults.

Good right ventricular function, prepared left ventricle: Double-switch operation and tricuspid valve repair (Class IIa).

Good right ventricular function, low left ventricular pressure: Tricuspid valve repair/replacement (Class IIb).

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	Double 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
e. Barcode of implant, if used	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 congenitally corrected 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. Kouchochos 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.