

Guidance document for PM JAY package

Double outlet Right ventricle (DORV) Repair

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 outlet Right ventricle (DORV) Repair	New Package	SV003H	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 **DORV 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:

Double-outlet Right Ventricle (DORV) is a condition in which both great arteries are connected completely or predominantly to the RV. By definition, at least more than half of both arteries should arise from the RV. The VSD is the only outlet for left ventricle. The VSD could be sub-aortic (60%–65%), sub-pulmonary (20%–25%), doubly committed (3%–5%), or

remote (noncommitted, 5%). DORV may be associated with a number of cardiac anomalies including PS and CoA. The clinical presentation is variable and depends on the location of VSD, presence or absence of obstruction to pulmonary blood flow, and associated cardiac anomalies. Clinical presentation of DORV can be divided into three types:

- i. TOF-like presentation: When the VSD is subaortic and there is obstruction to pulmonary outflow, the presentation is with progressive cyanosis and is indistinguishable from classic TOF.
- ii. VSD-like presentation: DORV with a large VSD in subaortic location presents with features of heart failure in infancy and mild cyanosis may be present.
- iii. TGA-like presentation: The VSD is subpulmonary in location and the great arteries are malposed (Taussig–Bing anomaly). There may be associated coarctation in some cases. Such patients present very early in life with cyanosis and heart failure.

Patients with subaortic or subpulmonary VSD can have total biventricular repair. Those with noncommitted or remote VSD have complex anatomy and may not be suitable for biventricular repair.

Diagnostic workup

- i. Clinical presentation: Variable as discussed above.
- ii. ECG: Nonspecific and depends on the type of DORV. Those with a large subaortic VSD may show biventricular hypertrophy and left ventricular overload pattern. Patients with VSD and PS may have extreme right-axis deviation which differentiates them from classical TOF.
- iii. X-ray chest: Cardiomegaly and plethoric lung fields in those with large pulmonary blood flow. Normal-sized heart with oligemic lung fields in TOF type of DORV.
- iv. Pulse oximetry: For documenting degree of cyanosis.
- v. Echocardiography: It is the primary imaging technique to confirm the diagnosis and to decide the type of surgical repair in a given case. The following features can be defined by echocardiography:
 - a. Situs, systemic venous, and pulmonary venous drainage (especially in heterotaxy syndrome).
 - b. Location, size, number of VSDs, and their relationship to aortic and pulmonary valve.
 - c. Relationship of great arteries to the ventricles and to each other.
 - d. Location and severity of PS, status of pulmonary annulus, and valve.

- e. Location and severity of subaortic stenosis, status of aortic annulus, and valve.
 - f. Size and competence of AV valves.
 - g. Size and function of both ventricles.
 - h. Associated defects such as CoA.
- vi. Cardiac catheterization: Required in late presenters with pulmonary hypertension suspected of having high PVR. It may also be done in other patients to define the anatomy better.
- vii. CTA and cMRI: Associated arch hypoplasia or coarctation may necessitate CTA or cMRI. 3D reconstruction of CT images helps assess routability of VSD to aorta.

Indication and timing of surgery

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

Timing and type of surgery depends on double-outlet of right ventricle variant (Class I).

- i. DORV with subaortic VSD and PS (TOF-type DORV):
 - a. Presenting with significant cyanosis at <3–4 months: Aortopulmonary shunt.
 - b. Presenting with significant cyanosis at >3–4 months: Total repair with closure of VSD and infundibular resection.
 - c. Stable patients with no or minimal cyanosis: Total repair with closure of VSD and infundibular resection by 6–12 months.
- ii. DORV with large subaortic VSD and pulmonary hypertension (VSD-type DORV):
 - a. VSD closure by 6 months of age.
 - b. Presenting beyond 6 months of age: Assess for operability and close VSD if operable.
- iii. DORV with subpulmonary VSD and pulmonary hypertension (TGA-type DORV):
 - a. ASO with VSD closure by 6 weeks of age.
 - b. If presenting beyond 3 months, should be evaluated for operability. ASO with VSD closure if operable.
 - c. If associated with aortic arch abnormality, arch repair should be done in the same sitting.

- iv. DORV with subpulmonary VSD and PS:
 - a. If pulmonary obstruction is localized, e.g., subvalvular fibrous membrane or ridge: ASO with resection of subvalvular stenosis.
 - b. If pulmonary obstruction is tubular or valvular: One of the following complex surgeries required: Rastelli -type repair, REV procedure, Nikaidoh procedure, or root translocation. A systemic-to-pulmonary artery shunt may be required before these procedures in those presenting early with significant cyanosis. Please refer to the section on “TGA with VSD and left ventricular outflow tract obstruction” for more details.
- v. DORV with remote VSD or associated with other complex anatomy: One should strive to perform biventricular repair by intraventricular baffling of left ventricular connection to aorta. Univentricular palliation (like single stage or multistage palliative cavopulmonary connection) is done in cases where biventricular repair is not possible.

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	DORV 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 Double Outlet Right Ventricle? 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