

Guidance document for PM JAY package

Atrioventricular (AV) Canal Repair

Procedures covered/ Procedure Count: 3

Specialty: CTVS

| Package name | Procedure name | HBP code 1.0 | HBP code 2.0 | Package price (INR) | ALOS |
|--|------------------------------|--------------|--------------|---------------------------|---------|
| Surgical Correction of Category - II Congenital Heart Disease | Partial AV Canal repair | S1300026 | SV002I | 120,000 + Cost of implant | 10 days |
| Surgical Correction of Category - II Congenital Heart Disease | Intermediate AV Canal repair | S1300026 | SV002J | 120,000 + Cost of implant | 10 days |
| Surgical Correction of Category - III Congenital Heart Disease | Complete AV canal repair | S1300026 | SV003V | 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 **AV Canal Repair (Partial/ Intermediate/ Complete)**, 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:

AV septal defects (AVSDs) account for 4%–5% of all congenital heart defects with estimated incidence of 0.19/1000 live births. These lesions can be divided into partial and complete forms. Clinical manifestations and outcome of patients with AVSD depend on the size of VSD, degree of ventricular hypoplasia (if any), AV valve regurgitation, presence or absence of left ventricular outflow tract obstruction, and presence or absence of associated syndromes. Down syndrome is present in 50% of patients with AVSD. Conversely, about 40%–45% of children with Down syndrome have CHD, and AVSD accounts for almost half of these, mostly in its complete form. Patients with Down syndrome tend to develop early and more severe form of pulmonary vascular disease with irreversible changes appearing as early as 6 months of age. The presence of preoperative left AV valve regurgitation is associated with increased risk of surgery and need for reoperations on follow-up. Complete form of AVSD, if left untreated, has a survival of only 54% at 6 months and 35% at 12 months. Partial form of AVSD has a better survival with 50% alive at 20 years of age.

Types of atrioventricular septal defects

- i. Complete AVSD: Large septal defect with an atrial component (ostium primum defect) and a ventricular component (inlet septal defect), common AV valve ring, and common AV valve. There may be incompetence of the right- and left-sided parts of the common AV valve. Complete form of AVSD is generally associated with large left-to-right shunt, pulmonary artery hypertension, and congestive heart failure.
- ii. Partial AVSD: These patients have separate annuli of right and left AV valve. There is a primum ASD. Cleft of the anterior leaflet of AV valve is common with variable degrees of regurgitation.
- iii. Intermediate AVSD: Two separate AV valves with primum ASD and small restrictive inlet VSD.
- iv. Unbalanced AVSD: One of the ventricular chambers is hypoplastic. This form is usually associated with complex congenital defects such as heterotaxy syndrome (isomerism).

Diagnostic workup

- i. Clinical assessment
- ii. X-ray chest: Cardiomegaly may be present due to dilation of the right or left heart chambers depending on the severity and direction of AV valve regurgitation and the severity and level of left-to-right shunting. Large left-to-right shunts lead to increased pulmonary vascular markings and prominent pulmonary artery conus.

- iii. ECG: PR interval prolongation is present in 50% of cases; occasionally, complete AV block develops. Other findings include moderate-to-extreme left-axis deviation, q waves in leads I and aVL (counterclockwise depolarization), and left atrial and ventricular hypertrophy if significant AV valve regurgitation is present. Right ventricular hypertrophy suggests the presence of pulmonary artery hypertension or right ventricular outflow tract obstruction.
- iv. Echocardiography: It is the key tool for the diagnosis and assessment of size of atrial and ventricular septal defects, size of the ventricles (balanced or unbalanced), estimation of the pulmonary artery pressures, presence and severity of AV valve regurgitation and for associated lesions such as left superior vena cava, left or right ventricular outflow tract obstruction, and heterotaxy syndrome. Transesophageal echocardiography may be rarely required in older patients with suboptimal transthoracic windows.
- v. Cardiac catheterization: Indicated for the assessment of operability in patients with pulmonary hypertension and suspected pulmonary vascular disease.

Ideal age of surgery

- i. Complete AVSD
 - a. Uncontrolled heart failure: Complete surgical repair as soon as possible (Class I)
 - b. Controlled heart failure: Complete surgical repair by 3 months of age (Class I)
 - c. Pulmonary artery banding: May be considered in select patients under 3 months of age (Class IIb).
- ii. Partial or intermediate AVSD, stable, and with normal pulmonary artery pressures: Surgical repair at 2–3 years of age (Class I)
- iii. Associated moderate or severe AV valve regurgitation may necessitate early surgery in partial or intermediate forms.
- iv. Pulmonary artery banding is reserved for complex cases and in patients with contraindications for cardiopulmonary bypass (Class IIb).
- v. Surgery for moderate-to-severe left AV valve regurgitation is recommended as per the guidelines for mitral regurgitation, discussed later (Class I).
- vi. Surgery for left ventricular outflow tract obstruction is reasonable with a peak systolic gradient of ≥ 50 mmHg, or at a lesser gradient if heart failure symptoms are present, or if concomitant moderate-to-severe atrioventricular or aortic regurgitation is present (Class IIa).
- vii. Those presenting beyond 6 months of life with significant pulmonary hypertension and suspected elevated PVR should be referred to a higher center for further evaluation to assess operability.

All patients with AVSD must be advised to maintain good oro-dental hygiene.

Important determinants of long-term prognosis

These include left AV valve stenosis/regurgitation (5%–10%), subaortic stenosis (5%), atrial arrhythmias, late-onset CHB, and issues related to Down syndrome (if present).

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 | Partial AV Canal repair | Intermediate AV Canal repair | Complete AV Canal repair |
|--|-------------------------|------------------------------|--------------------------|
| i. At the time of Pre-authorization | | | |
| a. Clinical notes | Yes | Yes | Yes |
| b. Echo/Doppler report | Yes | Yes | Yes |
| ii. At the time of claim submission | | | |
| a. Procedure / Operative notes | Yes | Yes | Yes |
| b. Post procedure stills of ECHO with report | Yes | Yes | Yes |
| c. Detailed Discharge Summary | Yes | Yes | 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 AV Canal defect? 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. Freeman SB, Taft LF, Dooley KJ, Allran K, Sherman SL, Hassold TJ, *et al.* Population-based study of congenital heart defects in down syndrome. Am J Med Genet 1998;80:213-7
3. Frescura C, Thiene G, Franceschini E, Talenti E, Mazzucco A. Pulmonary vascular disease in infants with complete atrioventricular septal defect. Int J Cardiol 1987;15:91-103
4. Berger TJ, Blackstone EH, Kirklin JW, Barger LM Jr., Hazelrig JB, Turner ME Jr. Survival and probability of cure without and with operation in complete atrioventricular canal. Ann Thorac Surg 1979;27:104-11