



Guidance document for PM JAY package

Aortic Pulmonary (AP) Window Repair

Procedures covered/ Procedure Count: 1

Specialty: CTVS

Package name	Procedure name	HBP 1.0 code	HBP 2.0 code	Package price	ALOS
Surgical Correction of Category - III Congenital Heart Disease	Aortic Pulmonary (AP) Window Repair	S1300029	SV003E	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 **AP Window 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:

Aorto Pulmonary Window (APW) is a rare congenital cardiac malformation, comprising only 0.1% of all congenital heart defects. In most patients with this anomaly, the defect is

moderate to large. Associated anomalies occur in half of the cases and include interrupted aortic arch (most commonly type A interrupted aortic arch), Ventricular septal defect, origin of right pulmonary artery from the aorta, coarctation of aorta (CoA), anomalous right or left coronary artery origin from main pulmonary artery, tetralogy of Fallot [TOF], subaortic stenosis etc. Clinical manifestations depend on the diameter of APW, the relative systemic and pulmonary vascular resistances, and associated lesions. Large APW is associated with very early development of advanced pulmonary vascular disease.

Types of aortopulmonary window

- Type 1 proximal defect (most common): Defect located just above the sinus of Valsalva, with very little inferior aortopulmonary septum above the semilunar valves.
- Type 2 distal defect: Defect located in the uppermost portion of the ascending aorta, with little superior rim of aortopulmonary septum.
- Type 3 total defect: Large defect that spans from semilunar valves to the pulmonary artery bifurcation with little superior and inferior rims.
- Intermediate type: Central defect with adequate superior and inferior rims.

Diagnostic workup

- i. Clinical assessment
- ii. X-ray chest: Cardiomegaly with increased pulmonary vascularity is seen in those with significant left-to-right shunt. There is absence of cardiomegaly with decreased vascularity in outer third of lung fields, prominent pulmonary artery segment and dilated central pulmonary arteries in patients with elevated PVR.
- iii. ECG: Signs of biventricular hypertrophy in response to volume overload of the left ventricle and pressure overload of the RV. Predominant right ventricular hypertrophy is seen if the PVR is elevated.
- iv. Echocardiography: This is the key tool for the diagnosis of APW, assessment of its size and location, relation to semilunar valves and coronary ostia, identification of associated anomalies, assessment of size of cardiac chambers, and estimation of pulmonary artery pressure.
- v. Cardiac catheterization: Performed for diagnostic purposes in those with pulmonary hypertension and suspected pulmonary vascular disease. Rarely, catheterization is performed for interventional purpose, in those with a small defect suitable for device closure.
- vi. Computed tomography angiography (CTA): Rarely performed as a part of preoperative workup in older children, where details of anatomy are not clear on echocardiography.

Ideal age of closure

- i. Uncontrolled heart failure: Surgical repair as soon as possible (Class I)
- ii. Controlled heart failure: Elective surgical repair by 3 months of age (Class I)
- iii. In patients with associated anomalies, single-stage repair of all defects is preferred (Class I).
- iv. Those presenting beyond 6 months of life with severe pulmonary hypertension and suspected elevated PVR should be referred to a higher center for further evaluation to assess operability.

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

Contraindication for closure

Severe pulmonary arterial hypertension with irreversible pulmonary vascular occlusive disease (Class III).

Method of closure

- i. Surgical patch repair is the treatment of choice (Class I).
- ii. Transcatheter device closure in selected cases of intermediate-type APW (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	AP Window 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. Procedure / Operative notes	Yes
b. Post procedure stills of ECHO with report	Yes
c. 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 Aortopulmonary Window? 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. Kutsche LM, Van Mierop LH. Anatomy and pathogenesis of aorticopulmonary septal defect. Am J Cardiol 1987;59:443-7
3. Jacobs JP, Quintessenza JA, Gaynor JW, Burke RP, Mavroudis C. Congenital heart surgery nomenclature and database project: Aortopulmonary window. Ann Thorac Surg 2000;69:S44-9.