



Guidance document for PM JAY package

Ebstein 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	Ebstein Repair	S1300031	SV003A	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 **Ebstein 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:

Ebstein's anomaly of the tricuspid valve is defined as failure of delamination of tricuspid leaflets from the myocardium. Mostly, the septal and the posterior leaflets are involved, and the anterior leaflet remains mobile. This results in the apical displacement of the functional tricuspid annulus and a rotation toward the outflow tract. A part of the RV inflow gets

“atrialized.” Tricuspid regurgitation of varying degree is invariably present, and the right atrium is enlarged. Some of the patients may be cyanosed due to right-to-left shunting at the atrial level. Ebstein's anomaly is rare, seen in 1 in 20,000 live births, and forms <1% of all congenital heart defects. Associated lesions include accessory AV conduction pathways (15%–20%), and an interatrial communication is present in 80%–94% of cases. Patients usually present either in neonatal period or during adolescence or adult life. Neonates may present with cyanosis and heart failure and at times with functional pulmonary atresia with duct-dependent pulmonary circulation. Older patients present with murmur, arrhythmias, or cyanosis. The estimated survival is 76% at 10 years and 53% at 15 years. Prognosis is poor in those diagnosed during fetal or neonatal life. A number of classifications have been described for Ebstein's anomaly; the most commonly used classification was described by Carpentier et al. in 1988.

Diagnostic workup

- i. Clinical assessment.
- ii. Pulse oximetry: For quantifying cyanosis.
- iii. ECG: It is always abnormal. Ebstein's anomaly can sometimes be diagnosed by a typical ECG showing tall (Himalayan) P waves, prolonged PR interval, right bundle branch block, and deep q waves in leads V1–V4. ECG may show evidence of preexcitation due to accessory AV pathway. Some patients may present with an episode of supraventricular tachycardia. Holter monitoring is done when suspecting an arrhythmia.
- iv. X-ray chest: The heart size varies from normal to marked cardiomegaly. The pedicle is narrow, and the cardiac borders are sharp. The heart has a box-like configuration, the right atrium is enlarged, and the lung fields may be oligemic.
- v. Echocardiography: It is the key diagnostic tool and shows the following features:
 - a. Apical displacement of the septal tricuspid leaflet in the four-chamber view. For diagnosing Ebstein's anomaly, the displacement should be $> 8 \text{ mm/m}^2$ in adults as some degree of displacement occurs in conditions with right ventricular volume overload.
 - b. Severity of tricuspid regurgitation and the velocity of jet
 - c. Type of Ebstein's anomaly
 - d. Size and function of the functional RV
 - e. Presence or absence of ASD and other lesions

Echocardiography also helps in assessing the feasibility of valve repair. Transesophageal echo is rarely required.



- vi. Cardiac catheterization: It is rarely performed, unless done for evaluating coronary arteries in older patients (>40 years) undergoing surgical repair of Ebstein's anomaly. Pulmonary artery pressure assessment may be required in those planned for bidirectional cavopulmonary anastomosis (Glenn).
- vii. cMRI: Provides quantitative measurement of right ventricular size, volume, and function, which are important for planning surgical repair.
- viii. Other tests in select patients: Exercise testing, electrophysiological studies.

Indications and timing for treatment

Neonates

- i. Presenting with significant cyanosis: Intravenous prostaglandin infusion (Class I)
- ii. Presenting with heart failure: Diuretics (Class I)
- iii. Presenting with arrhythmias: Appropriate antiarrhythmic drug (Class I)
- iv. Surgery for those not stabilized with medical therapy (Class IIa).

Older children and adults

Tricuspid valve repair (Cone repair) is best done at about 2 years of age for stable cases.

- i. Surgery is indicated (Class I) in those with:
 - a. Symptoms or deteriorating exercise capacity
 - b. Cyanosis (oxygen saturation < 90%)
 - c. Paradoxical embolism
 - d. Progressive cardiomegaly on chest X-ray (CT ratio > 0.65)
 - e. Progressive dilation or dysfunction of the RV on echocardiography.
- ii. Symptomatic with arrhythmias: Catheter ablation. Surgery, if not amenable to catheter ablation (Class IIa).

Types of surgery

Depends on the underlying anatomy and size of the functional ventricle.

- i. Tricuspid valve repair; replacement only if repair cannot be achieved.
- ii. Tricuspid valve repair with bidirectional cavopulmonary anastomosis (one and a half ventricle repair).

- iii. Single ventricle repair (aortopulmonary shunt(BTT Shunt)/Glenn followed by Fontan surgery).

Important determinants of long-term prognosis

These include recurrence of tricuspid valve regurgitation with need for reoperation, right ventricular dilatation and dysfunction, supraventricular and ventricular tachyarrhythmias, and need for pacemaker implantation.

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	Ebstein 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 Ebstein Anomaly? Yes

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

References

1. Saxena A, Relan J, Agarwal Ret.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. Danielson GK, Driscoll DJ, Mair DD, Warnes CA, Oliver WC Jr. Operative treatment of Ebstein's anomaly. J Thorac Cardiovasc Surg 1992;104:1195-202
3. Attie F, Rosas M, Rijlaarsdam M, Buendia A, Zabala C, Kuri J, et al. The adult patient with Ebstein anomaly. Outcome in 72 unoperated patients. Medicine (Baltimore) 2000;79:27-36
4. Carpentier A, Chauvaud S, Macé L, Relland J, Mihaileanu S, Marino JP, et al. A new reconstructive operation for Ebstein's anomaly of the tricuspid valve. J Thorac Cardiovasc Surg 1988;96:92-101