



Guidance document for PM JAY package

Tetralogy of Fallot 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	Tetralogy of Fallot Repair	S1300027	SV003U	150,000 + Cost of implant	12 days
Immediate reoperation (within 5 days)	Tetralogy of Fallot Repair	S1300027	SV031A	75,000 + 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 **Tetralogy of Fallot 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:

TOF is the most common cyanotic congenital heart disease with a prevalence of 0.4/1000 live births constituting about 5% of all congenital heart defects. The clinical signs and symptoms seen in infants generally vary in accordance with the degree of right ventricular outflow tract

obstruction. Almost two-thirds of newborns with TOF are acyanotic at birth, but by 6 months of age, over half of them have cyanosis at rest. This occurs because of worsening infundibular stenosis which increases right-to-left shunting across the VSD. Intermittent hypercyanotic spells are one of the defining features of TOF. Peak incidence of these episodes occurs between the 2nd and 6th months of life, and these become infrequent after 2 years of age. Patients with untreated TOF have an estimated 1-year, 3-year, and 10-year survival of 66%, 49%, and 24%, respectively.

Diagnostic workup

- i. Clinical assessment: Degree of cyanosis is the most important aspect of clinical evaluation.
- ii. Pulse oximetry: Measuring oxygen saturation by pulse oximeter is recommended at each follow-up examination. Anemia may undermine the severity of clinical cyanosis.
- iii. ECG: Typical ECG shows right-axis deviation, right ventricular hypertrophy, and an early QRS transition with abrupt change from an R wave in lead V1 to an rS pattern in lead V2.
- iv. X-ray chest: Normal heart size with an upturned apex (“boot-shaped” heart due to right ventricular hypertrophy), deficiency of the main pulmonary artery segment (“pulmonary bay” seen as a concavity in the upper left cardiac border), and reduced pulmonary vascularity are the cardinal features.
- v. Echocardiography: It is a vital tool for the diagnosis of TOF, and the following features should be noted
 - a. Site and degree of right ventricular outflow tract obstruction
 - b. Pulmonary valve and annulus size
 - c. Size and confluence of branch pulmonary arteries and any evidence of ostial stenosis. McGoon ratio and Nakata index can be calculated to decide suitability for total repair.
 - d. Size and location of malaligned VSD and any additional VSD
 - e. Coronary anomalies which may interfere with surgical repair (especially anomalous left anterior descending coronary artery crossing the right ventricular outflow tract)
 - f. Aortic arch sidedness and branching pattern, aortic dilation, and aortopathy
 - g. Additional anomalies such as ASD, complete AVSD, and persistent left superior vena cava
 - h. Transesophageal echocardiography may not be required for defining details of anatomy, but it is of great help at the time of surgical repair to check for adequacy of repair.

vi. Cardiac catheterization and angiography

Timing of surgery

- i. All patients need surgical repair
- ii. Stable, minimally cyanosed: Total repair at 6–12 months of age or earlier according to the institutional policy (Class I)
- iii. Symptomatic children of <6 months of age with significant cyanosis or history of spells despite therapy: Palliation (by systemic-to-pulmonary artery shunt or stenting of the ductus arteriosus/right ventricular outflow tract or pulmonary valve balloon valvuloplasty) or total repair depending on anatomy and center's experience (Class I)
- iv. Lower threshold for earlier surgery if no requirement of transannular patch is anticipated
- v. Patients having TOF with absent pulmonary valve who are stable: Medical management till 1 year of age followed by total correction with repair of pulmonary artery branch dilation/aneurysm (Class I)
- vi. Patients with anomalous left anterior descending artery from the right coronary artery crossing the right ventricular outflow tract, who are likely to need RV-to-pulmonary artery conduit (Class I):
 - a. <10 kg weight with significant cyanosis: Aortopulmonary shunt
 - b. >10 kg weight: Total repair using conduit, or double-barreled approach after 2 years of age, when the child weighs >10 kg.

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	Tetralogy of Fallot Repair	Immediate reoperation- Tetralogy of Fallot Repair
i. At the time of Pre-authorization		
a. Clinical notes	Yes	Yes
b. Echo/Doppler report	Yes	Yes
ii. At the time of claim submission		
a. Indoor case papers	Yes	Yes
b. Procedure / Operative notes	Yes	Yes
c. Post procedure stills of ECHO with report	Yes	Yes
d. Detailed Discharge Summary	Yes	Yes

e. Detailed Operative notes indicating need for Reoperation	No	Yes
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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 Tetralogy of Fallot Repair? 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. Bonchek LI, Starr A, Sunderland CO, Menashe VD. Natural history of tetralogy of fallot in infancy. Clinical classification and therapeutic implications. Circulation 1973;48:392-7
3. Kouchoukos 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.
4. Mavroudis C, Backer C. Pediatric cardiac surgery. Blackwell Publishing Ltd; 2013 Feb 28.