



Guidance document for PM JAY package

Norwood Procedure/Glenn Procedure/ Fontan Procedure

Procedures covered/ Procedure Count: 3

Specialty: CTVS

Package name	Package name	HBP code 1.0	HBP code 2.0	Package price (INR)	ALOS
Surgical Correction of Category - III Congenital Heart Disease	Norwood Procedure	New Package	SV003K	150,000 + cost of implant	12 days
Surgical Correction of Category - I Congenital Heart Disease	Glenn procedure	S1300050	SV001C	100,000	10 days
Surgical Correction of Category - III Congenital Heart Disease	Fontan Procedure	S1300032	SV003D	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 **Norwood Procedure/Glenn Procedure/ Fontan Procedure**, 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:

Univentricular hearts are defined as the presence of one ventricle instead of two, or the second ventricle is rudimentary without an inlet portion. The main ventricle may be of left (more common) or right ventricular morphology or, in rare cases, indeterminate. The classical examples include double-inlet left ventricle, tricuspid atresia, and hypoplastic left heart syndrome. This term also includes a number of congenital heart defects where two-ventricle (or biventricular) repair is not possible. The examples include unbalanced AVSD, double-outlet RV with a non-routable VSD, straddling of an AV valve, or a very large/multiple VSDs, not amenable to closure. These defects are often grouped together as “functional univentricular” heart, because the management is on the lines of single ventricle. The clinical presentation depends on the ratio of pulmonary-to-systemic blood flow. Infants with unrestricted pulmonary blood flow present with predominant heart failure and mild cyanosis and those with obstruction to pulmonary blood flow have dominant cyanosis and no heart failure. Rarely, there could be a balanced circulation.

Timing and type of intervention

Preamble

Surgery for univentricular heart is a palliative procedure. The life expectancy is less than normal (exact age cannot be predicted) and is interposed by interventions over these years. Treating physicians must inform and discuss the details with the parent/guardian before surgery.

Norwood Procedure: Stage I palliation is the Norwood procedure. Its aims are to 1) allow for an unobstructed blood flow from the right ventricle to the systemic circulation, 2) establish a connection between the systemic and the restricted pulmonary circulation, 3) provide an unobstructed flow from the pulmonary veins through the atrial septum to the right atrium.

Glenn Procedure: The second stage is a transition from a high pressure arterial supply to the pulmonary blood flow to a venous one, through creating an anastomosis between the superior vena cava and the pulmonary artery. The original parallel conduit of pulmonary and systemic circulations is rearranged into series. During the Glenn operation the superior vena cava is separated from the right atrium and connected to the pulmonary artery, cardiac end of the superior vena cava is closed. In cases where, BT shunt was performed earlier, it has to be taken down at the time of Glenn procedure. Thus, the pulmonary circulation is perfused with the fully desaturated, rather than mixed, blood from the systemic venous flow, while the pulmonary venous flow is conveyed to the systemic circulation alone. Antegrade pulmonary blood flow can be interrupted or left depending on the on-table surgical evaluation or pre-operative clinical condition of the child.

Fontan Procedure: In the third stage a total cavo-pulmonary anastomosis or full Fontan circulation is formed. The Fontan operation, in which the systemic arterial and venous blood flow is completely separated, is the final stage, and the full Fontan circulation is the end goal

of the palliative surgical therapy for Single Ventricle physiology. During the Fontan operation the inferior vena cava is connected to the pulmonary artery bypassing the heart, thereby the entire systemic venous flow enters the pulmonary circulation rather than the right atrium. Thus, the systemic circulation is perfused exclusively with fully saturated blood, enhancing oxygen delivery.

The timing and type of intervention depends on age at presentation and presence or absence of obstruction to pulmonary blood flow.

- i. Those presenting in neonatal period or within 2–3 months of life (Class I):
 - a. With increased pulmonary blood flow:
 - Type of surgery: Pulmonary artery banding (usually combined with ligation of PDA).
 - Timing of surgery: At 4–6 weeks of age, preferably before 3 months.
 - Additional procedures may be required if systemic outflow obstruction is present.
 - b. With decreased pulmonary blood flow (PS group):
 - Type of surgery: Systemic-to-pulmonary artery shunt or stenting of ductus arteriosus (depends on institutional policy and clinical scenario).
 - Indications of surgery:
 - When systemic arterial saturation is consistently below 70%–75%.
 - In case of pulmonary atresia with duct-dependent pulmonary circulation (baby is usually on prostaglandin infusion).
 - c. With balanced pulmonary circulation: The baby usually maintains saturations above 80% and is not in failure. Such infants should be followed up closely. If saturation falls below 70% and the age is:
 - Below 4–5 months: A systemic-to-pulmonary artery shunt (Class I)
 - More than 4–5 months: Bidirectional Glenn procedure if branch pulmonary arteries are of good size (Class I).

Those presenting later in life or have undergone first surgery earlier:

- With pulmonary hypertension and no PS: Most patients who present beyond 3–4 months would become unsuitable for pulmonary artery banding or any definitive repair in the future due to irreversible increase in PVR. A minority of patients may continue to have low resistance and should be offered

pulmonary artery banding after full investigations, which includes cardiac catheterization to test for operability.

- a. With normal pulmonary pressure and resistance due to PS/previous pulmonary artery banding/previous aortopulmonary shunt:
 - Bidirectional Glenn procedure between 6 and 12 months of age (Class I) (can be considered between 4 and 6 months in certain circumstances).
 - Additional source of pulmonary blood flow (not completely interrupting the antegrade flow across pulmonary outflow) can be kept in the presence of adequate ventricular function, no AV valve regurgitation, small-sized pulmonary artery branches, and heterotaxy syndrome.
 - Total cavopulmonary connection or completion of Fontan procedure (preferably extracardiac): Between 4 and 7 years of age when the child weighs 15–20 kg. If Fontan required earlier (due to increasing cyanosis or due to pulmonary arteriovenous malformations), a lateral tunnel Fontan can be performed at the age of 3–4 years. Fenestration of Fontan circuit is indicated in high-risk cases.
 - Additional procedures such as repair of AV valve regurgitation, atrial septectomy, branch pulmonary artery stenosis repair (plasty), and TAPVC repair may have to be done at the time of bidirectional Glenn procedure or total cavopulmonary connection.

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	Norwood Procedure	Glenn Procedure	Fontan Procedure
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. Indoor case papers	Yes	Yes	Yes
b. Procedure / Operative notes	Yes	Yes	Yes
c. Post procedure stills of ECHO with report	Yes	Yes	Yes
d. 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 single ventricle morphology or hypoplastic heart syndrome? Yes

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

References

1. 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.
2. Mavroudis C, Backer C. Pediatric cardiac surgery. Blackwell Publishing Ltd; 2013 Feb 28.
3. 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