



## Guidance document for processing PM-JAY packages

### Infantile cholestasis

**Procedures covered:** 1

**Specialty:** Pediatric Medical Management

Package name	Procedure name	HBP code 1.0	HBP code 2.0	Package price (INR)
Infantile cholestasis	Infantile cholestasis	M200037	MP024A	Routine Ward - 1800 HDU - 2700 ICU (without Ventilator) - 3600 ICU (with Ventilator) - 4500

**ALOS:** 2 days (Once diagnosis is established the case can be booked in the relevant package, further stay/admission should be decided based on the level of complications of the disease)

**Minimum qualification of the treating doctor:**

**Desirable:** MD/DNB/DCH (Pediatric Medicine) / Pediatric Surgeon referral (if required)

**Special empanelment criteria/linkage to empanelment module:** None

#### 1.2 Disclaimer:

For monitoring and administering the claim management process of **Infantile cholestasis**, 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:

Cholestasis is defined as reduced bile formation or flow resulting in the retention of biliary substances within the liver normally excreted into bile and destined for elimination into the intestinal lumen. Cholestatic jaundice in infancy is an important clinical condition that results from hepatobiliary dysfunction and can be caused by a number of disorders. Idiopathic neonatal hepatitis (INH) and biliary atresia (BA) are two main causes. It is imperative that the clinician differentiates between intrahepatic and extrahepatic origin of cholestasis.

### Causes of infantile cholestasis:

<i>Obstructive</i>	<i>Infectious</i>
Biliary atresia	Congenital TORCH infection:
Choledochal cyst	Toxoplasmosis
Syndromic and nonsyndromic paucity of interlobular bile ducts	Cytomegalovirus
	Rubella
Inspissated bile syndrome	Herpes virus
Caroli's disease/congenital hepatic fibrosis	HIV
Neonatal sclerosing cholangitis	Bacterial sepsis
	Urinary tract infection
<i>Idiopathic neonatal hepatitis</i>	<i>Metabolic</i>
	Bile acid synthesis defects
	Gestational alloimmune liver disease/neonatal hemochromatosis
	Galactosemia
	Hereditary tyrosinemia
	Hypothyroidism
	Panhypopituitarism
	Storage diseases
<i>Genetic</i>	<i>Toxic</i>
Alpha-1-antitrypsin deficiency	Parenteral nutrition
Alagille syndrome	Drugs
Progressive familial intrahepatic cholestasis	
Cystic fibrosis	
Arthrogryposis–renal dysfunction–cholestasis syndrome (ARC)	

*HIV* human immunodeficiency virus, *TORCH* Toxoplasmosis, Other Agents, Rubella, Cytomegalovirus, Herpes Simplex

Proceed with Infantile cholestasis only if diagnosis made is backed by clinical manifestation

1. Jaundice (yellow discoloration of the skin, sclera, mucous membranes, and body fluids)
  - Conjugated hyperbilirubinemia  $>1.5 - 2$  mg% and/or direct component of more than 20% of total bilirubin in an infant
2. Passage of high colored urine with or without clay stools

### Associated factors:

1. Hepatosplenomegaly
2. Dehydration
3. Failure to thrive (clinically unwell, poor feeding, poor weight gain)

## Investigations

Cholestasis is generally recognized by evaluation of serum studies, with elevation of serum conjugated (or direct) bilirubin and bile acids as central readily identified features of hepatobiliary

dysfunction. Bilirubin is a red flag found in the laboratory evaluation that can help the clinician identify cholestasis.

There is no gold standard investigations for diagnosing infantile cholestasis. The evaluation should be based on etiology.

## Treatment

The therapy for cholestasis is treating the underlying etiology of the disease.

General Management:

- Medium chain Triglycerides (MCT) oil
- Fat soluble Supplements
  - Vitamin A,D,E,K
- Water soluble supplements (Twice the RDA)
- Calcium
- Phosphate
- Zinc
- Ursodeoxycholic acid
- Prophylactic phenobarbitone
- Portal hypertension – propranolol
- Pediatric Surgical consultation if required
- End stage disease – Liver transplantation

### 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	Infantile cholestasis
<b>i. At the time of Pre-authorization</b>	
Clinical notes including vitals, examination findings, planned line of treatment and advice for admission	Yes
Liver function test	Yes
USG Abdomen	Yes
<b>Optional – Based on the etiology and availability</b> Lab investigations: Complete Blood Count, Renal Function Test, Prothrombin Time/Partial thromboplastin time, Sepsis screen, Urinalysis, blood sugar, Sr. Electrolytes, Thyroid profile, Viral Serology, Metabolic screening, Genetic screening	Yes

Diagnostic Imaging: Endoscopic retrograde cholangiopancreatography (ERCP)	
<b>ii. At the time of claim submission</b>	
Detailed Indoor case papers (ICPs) with treatment details	Yes
Total serum bilirubin	Yes
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:**

- I. Is the Direct Bilirubin fraction >20% in Total Serum Bilirubin (evidence of conjugated hyperbilirubinemia)? Yes
- II. Is the patient's age more than 28 days and less than one year? Yes

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

## **References**

1. Standard Treatment Guidelines - Pediatrics & Pediatric Surgery: Ministry of Health & Family Welfare Govt. of India (MoHFW). 2017.
2. Fawaz R, Baumann U, Ekong U, et al. Guideline for the Evaluation of Cholestatic Jaundice in Infants: Joint Recommendations of the North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition and the European Society for Pediatric Gastroenterology, Hepatology, and Nutrition. *J Pediatr Gastroenterol Nutr.* 2017;64(1):154-168. doi:10.1097/MPG.0000000000001334
3. Standard Treatment Protocol for Medical Officers. 2014. Government of Tripura Health & Family Welfare Department
4. Feldman, A., & Suchy, F. (2014). Approach to the infant with cholestasis. In F. Suchy, R. Sokol, & W. Balistreri (Eds.), *Liver Disease in Children* (pp. 101-110). Cambridge: Cambridge University Press. doi:10.1017/CBO9781139012102.009



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6. Dehghani SM, Haghighat M, Imanieh MH, Geramizadeh B. Comparison of different diagnostic methods in infants with Cholestasis. *World J Gastroenterol*. 2006;12(36):5893-5896. doi:10.3748/wjg.v12.i36.5893