



Guidance document for processing PM-JAY packages

Congenital lobar emphysema

Procedure covered: 1

Specialty: Pediatric Surgery

Package name	Procedure name	HBP 1.0 code	HBP 2.0 code	Package price
Surgery for Congenital lobar emphysema	Surgery for Congenital lobar emphysema	S1400032	SS018A	25,000/-

ALOS: 5-7 Days

Minimum qualification of the treating doctor:

Essential: MCh/ equivalent (in pediatric surgery)

Special empanelment criteria/linkage to empanelment module: None

Disclaimer:

For monitoring and administering the claim management process of **Congenital lobar emphysema**, 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:

Congenital lobar emphysema (CLE), also known as congenital alveolar overdistension, congenital lobar over inflation and infantile lobar emphysema is a developmental anomaly of the lower respiratory tract that is characterized by hyperinflation/overdistension of one or more of the pulmonary lobes. The left upper lobe is affected most often. Congenital lobar emphysema is most frequently the result of a check-valve obstruction with gas trapping, not true emphysema. It can also, on rare occasions, be the result of a hyperalveolar lobe with an excessive number of alveoli of normal size.



Proceed with Congenital lobar emphysema only if diagnosis made is backed by clinical manifestation:

- Breathlessness
- Cough
- Wheezing
- Respiratory distress in neonatal and early infancy

Infants (usual presentation)

- Tachypnea and increased work of breathing/retractions
- Wheezing
- Cyanosis
- Recurrent respiratory tract infections
- Poor feeding with failure to thrive
- Chronic cough

Progressive respiratory distress develops rapidly in some infants, while others have a more gradual, insidious onset and some may remain asymptomatic for years. The severity depends upon the size of the affected lobe, the compression of surrounding lung tissue, and the extent of mediastinal shift.

Diagnosis

A strong clinical suspicion is required to diagnose this rare anomaly which may mimic other causes of respiratory distress.

Congenital lobar emphysema is characterized by:

- (1) difficulty in breathing or very rapid respiration (respiratory distress) in infancy
- (2) an enlarged chest due to over inflation of at least one lobe of the lung
- (3) compressed normal lung tissue in the section of the lung nearest to the diseased lobe
- (4) bluish color of the skin due to a lack of oxygen in the blood (cyanosis)
- (5) underdevelopment of the cartilage that supports the bronchial tube (bronchial hypoplasia)

Investigations

- A posteroanterior chest X-ray is the first choice for an examination
- Lung computed tomography (CT) is the gold standard in the diagnosis of CLE

Management:

- Treatment of CLE is essentially surgical, i.e., lobectomy
- CLE is rarely treated with thoracoscopy, while thoracotomy is still the most common surgical technique for resection of CLE in childhood
- Asymptomatic patients may be managed conservatively but should be kept in close follow-up as they may require surgical intervention in cases of worsening respiratory distress

1.4 Mandatory documents- For healthcare providers

Lung computed tomography (CT) is the gold standard in the diagnosis of CLE. The choice of other investigation depends on the clinical situation for which the investigation is asked for:

Following documents should be uploaded by the concerned hospital staff at the time of pre-authorization and claims submission:

Mandatory document	Congenital lobar emphysema
i. At the time of Pre-authorization	
Clinical notes	Yes
Posteroanterior chest X-ray	Yes
CT Chest	Yes
ii. At the time of claim submission	
Indoor case papers (ICPs)	Yes
Detailed Procedure / operative notes	Yes
Posteroanterior chest X-ray/ CT chest	Yes
Detailed discharge summary	Yes

PART II: GUIDELINES FOR PROCESSING TEAM

PART III: GUIDELINES FOR IT

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:

- Was the CT chest report and post-procedure X-ray stills submitted? Yes
- Does Chest radiograph show the evidence of lobar over aeration, mediastinal shift, and compression of the adjacent lobe for diagnosis? Yes

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



References:

1. [Christopher M Oermann](#). Congenital lobar emphysema – UpToDate. Last updated (July 2019)
2. Demir OF, Hangul M, Kose M. Congenital lobar emphysema: diagnosis and treatment options. *Int J Chron Obstruct Pulmon Dis*. 2019;14:921-928. Published 2019 May 1. doi:10.2147/COPD.S170581 <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6507121/#!po=30.3571>
3. Chinya A, Pandey PR, Sinha SK, Sarin YK. Congenital lobar emphysema: Pitfalls in diagnosis. *Lung India*. 2016;33(3):317-319. doi:10.4103/0970-2113.180883. http://www.lungindia.com/temp/LungIndia333317-3453319_093533.pdf
4. <https://rarediseases.org/rare-diseases/emphysema-congenital-lobar/>