

## Guidance document for processing PM-JAY packages

### Neural Tube defects

Procedures covered: 5

Specialty: Neurosurgery

Package name	Procedure name	HBP 1.0 code	HBP 2.0 code	Package price (INR)
Meningocele	Anterior	S800019	SN005A	36,000
Meningocele	Lumbar	S800020	SN005B	36,000
Meningocele	Occipital	S800021	SN005C	50,000
Anterior Encephalocele	Anterior Encephalocele	S800001	SN052A	50,000
Spina Bifida Surgery	Spina Bifida Surgery	S800064	SN053A	36,000

**ALOS:** 10 days

**Minimum qualification of the treating doctor:**

**Essential:** MCh/DNB/Equivalent (in Neurosurgery) involvement of other specialty based on Etiology

**Special empanelment criteria/linkage to empanelment module:** Care at Tertiary Hospital

**Disclaimer:**

For monitoring and administering the claim management process of **Meningocele/Anterior Encephalocele/Spina Bifida Surgery**, 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:**

Neural tube defects are a spectrum of disorders that can affect the brain or the spinal cord. They are caused by interaction of genetic and environmental factors and are prevented with folate supplementation. Optimal care of a patient with spina bifida surgery (major/minor), meningocele and encephalocele in the long term requires a comprehensive coordinated plan of treatment,

usually in a dedicated setting with facilities for neurosurgery, urology, orthopedic surgery, neurology and rehabilitation.

Follow-up should include periodic evaluation of status of any associated anomalies.

1. Hydrocephalus
2. Tethered spinal cord
3. Chiari II malformation
4. Syringomyelia

Any deterioration of function at any time needs to be investigated. Proper CSF diversion with endoscopic third ventriculostomy or ventriculo-peritoneal shunt has to be ensured, whenever necessary.

Multiple surgical procedures which usually include urological and orthopedic operations, Chiari decompression, tethered cord releases, or spinal fusions may be required in follow-up. Revision surgeries for wound infections and CSF leakage from the wound may be required in early postoperative period. Rehabilitation, with training in Clean Intermittent Catheterization (CIC) for bladder may be required in early postoperative period or in follow up.

So, it is advisable to follow up at the treating hospital at the end of one month after discharge. Further follow up can be done at district hospital by general surgeon at 3rd, 6th and 12th month follow up. If necessary, he/she may refer back to the treating surgeon. Other follow up evaluations include

- a. Urological evaluation with Ultrasonogram after one month, 3 months, 6 months and one year.
- b. MRI of spine/brain at 3rd to 6th month, if required, as assessed by treating surgeon.
- c. Additional surgical procedures and revision surgeries may be required, whenever the need arises, in follow up.

Follow-up frequency	<ol style="list-style-type: none"> <li>1. One month after discharge at the treating hospital</li> <li>2. 3 months</li> <li>3. 6 months</li> <li>4. 1 year</li> <li>5. Two visits, as and when required, at treating hospital</li> </ol>
Investigations	<ol style="list-style-type: none"> <li>1. Ultrasonogram to assess Urological status at 1 month, 3 months, 6 months and one year follow-up (when indicated)</li> <li>2. MRI of Brain and/or Spine, if necessary, on recommendation of the treating surgeon</li> </ol>
Medication	<ol style="list-style-type: none"> <li>1. Treatment of urinary or wound infections, with necessary antibiotics, whenever required</li> <li>2. Clean intermittent Catheterisation-Catheters, if CIC is indicated</li> </ol>

## **SPINA BIFIDA**

Spina bifida is a condition in which the neural tube, a layer of cells that ultimately develops into the brain and spinal cord, fails to close completely during the first few weeks of embryonic development. Mostly Spina bifida are diagnosed by antenatal ultrasound.

Spina bifida presents clinically with variable paralysis and sensory loss in legs, orthopedic deformities, neurogenic bowel and bladder, hydrocephalus, and Chiari II malformation (caudal displacement of the cerebellar tonsils and vermis, caudal medulla, and occasionally the fourth ventricle, into the cervical spinal canal).

### **Classification**

- Open spina bifida (not covered by skin)
- Closed spina bifida (covered by skin)
- Spina bifida occulta (skin covered with no visible abnormalities of the back)

### **Spina Bifida Surgery Major:**

#### **Clinical presentation**

- muscle weakness of legs
- bowel & bladder problems
- seizures
- orthopedic problems

### **Spina Bifida Surgery Minor:**

#### **Clinical presentation**

- hairy patch, dimple, dark spot, swelling in the back at the site of the gap in spine,
- may associated with poor ability to walk, problems with bladder or bowel control, hydrocephalous

## **MENINGOCELE**

Meningocele is due to failure of closure during embryonic life of bottom end of the neural tube, the structure which gives rise to the central nervous system (the brain and spinal cord). Herniation of the meninges without involvement of spinal elements. Meningoceles are considered to be neural tube defects and are a rare form of spina bifida.

### **Clinical presentation**

- Lump in back
- Paraparesis
- Severe headache
- Severe vomiting
- Stiffness of neck

- Giddiness
- Bladder bowel syndrome

### **Management**

- To close meningoceles at the skull base or top of the spine, may use the minimally invasive Endoscopic Endonasal Approach (EEA).
- For closing meningoceles lower in the spine, meningocele excision

**Cranial meningoceles**, by definition, are congenital herniations of meninges and CSF through the skull and are bereft of any cerebral tissue. Common location for these lesions is directly over the anterior fontanelle (vertex meningocele). They are often diagnosed on cranial ultrasound. The surgical resection involves a circumferential dissection following what is typically a thick capsule to the dural surface.

### **ANTERIOR ENCEPHALOCELE**

Encephalocele is a defect in calvarium with protrusion of brain, most often in occipital region. Patients with frontal encephaloceles are a rare condition but have better developmental outcomes. The anterior encephaloceles are subdivided further into “sincipital” when the herniation is anterior to the cribriform plate, and “basal,” when the herniation is through the sphenoid sinus.

#### **Types of anterior encephalocele**

- Frontoethmoidal
- Nasofrontal
- Nasoethmoidal
- Naso orbital
- Orbital
- Transethmoid nasopharyngeal
- Transsellar transsphenoidal
- Anterior fontanelle
- Interfrontal

#### **Clinical features**

- Swelling over nose
- Hypertelorism



- Nasal obstruction
- Proptosis
- Neurofibromatosis
- Leaking encephalocele
- Enlarged head
- History of meningitis

## Management

The principles of surgical treatment of encephaloceles are reduction of the herniation with preservation of as much viable brain as possible, water-tight dural closure with adequate skin coverage, repair of any cosmetic deformity, and cranial or craniofacial reconstruction. With anterior encephaloceles in particular, the goal is to remove the lesion before it can further distort facial growth. The approach to anterior encephaloceles often involves a multidisciplinary approach including surgeons who specialize in neurosurgery; oral-maxillofacial surgery (OMFS); ears, nose, and throat (ENT) surgery; and plastic surgery.

## Types of surgery

- One-stage surgery
- Two-stage surgery
- Tessier's operation
- Hemiorbital advancement
- Repair of orbital encephalocele
- Transpalatal repair
- Repair of nasopharyngeal encephalocele
- Ventriculo peritoneal shunt
- Thecoperitoneal shunt

## 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	Meningocele	Anterior Encephalocele	Spina Bifida Surgery
<b>i. At the time of Pre-authorization</b>			
Clinical notes with signs, symptoms, indications, planned line of management and advice for admission	Yes	Yes	Yes

Clinical picture	Yes	Yes	Yes
Plain X-ray skull CT/MRI Brain/Spine	Yes	Yes	Yes
<b>Optional</b> Cranial ultrasound / Spinal ultrasound	Yes	Yes	Yes
<b>ii. At the time of claim submission</b>			
Detailed Indoor case papers (ICPs)	Yes	Yes	Yes
Detailed Procedure / operative notes	Yes	Yes	Yes
Post-operative photographs (optional)	Yes	Yes	Yes
Detailed discharge summary	Yes	Yes	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 clinical presentation and imaging indicative of surgery? Yes

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

### **References**

- Clinical protocol guidelines. Mahatma Jyotiba Phule Jan Arogya yojana. Maharashtra <https://www.jeevandayee.gov.in/MJPJAY/RGJAYDocuments/NEUROSURGERY.pdf>
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- Mahapatra AK. Anterior encephalocele - AIIMS experience a series of 133 patients. *J Pediatr Neurosci.* 2011;6(Suppl 1):S27-S30. doi:10.4103/1817-1745.85706
- BMJ Best Practice. Spina bifida and neural tube defects. Last updated – February 2018
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