Guidance document for processing PM-JAY packages

Dysmorphic Children

Procedures covered: 1 **Specialty:** Pediatric Medical Management

Package Name	Procedure Name	HBP 1.0 code	HBP 2.0 code	Package price (INR)
Dysmorphic children	Dysmorphic children	M200019	MP034A	General Ward- 1800/- HDU – 2700/- ICU without ventilator– 3600/- ICU with Ventilator– 4500/-

ALOS: 7 days

Minimum qualification of the treating doctor:
Essential: MD/DNB/DCH (Pediatric) and requires a multidisciplinary approach involving geneticist, relevant subspecialties, social workers, physiotherapist, speech therapist

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

Disclaimer:
For monitoring and administering the claim management process for **Dysmorphic Children,** 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 CLINICAL 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 anomalies are a significant cause of neonatal intensive care unit (NICU) admissions. The presence of a particular congenital anomaly may necessitate evaluation for the presence of other specific associated anomalies or genetic syndromes.

An anomaly is a structural defect that deviates from the normal standard and can be categorized as major or minor. A major anomaly has surgical, medical or cosmetic importance and may be a marker for other occult malformations. A minor anomaly has no significant surgical or cosmetic importance; however, many genetic syndromes are recognized based on the pattern of minor anomalies present.

The anomalies and physical features include birth parameters, aplasia cutis congenita, holoprosencephaly, asymmetric crying facies, preauricular ear tags and pits, cleft lip with or without cleft palate, esophageal atresia/tracheoesophageal fistula, congenital heart defects, ventral wall defects, and polydactyly.

Potential causes:

- Chromosomal aneuploidy
- Single gene abnormalities
- Teratogen exposure
- Birth defects
- Environmental

Suspicion

Genetic etiology suspected in any child with:

- Congenital anomalies: at least 1 major/ > 2 minor anomalies.
- Growth deficit (short stature/ failure to thrive)
- Developmental delay, mental deficit or developmental regression
- Failure to develop secondary sexual characteristics
- Abnormal genitalia
- Appears ‘different’/ ‘unusual’

Approach to a dysmorphic child

1. Suspicion
2. Clinical evaluation
    a. history
    b. physical examination

3. Investigations
4. Analysis and diagnosis
5. Confirmation
6. Intervention:
    a. treatment
    b. counseling
    c. prenatal diagnosis
7. Surveillance & follow up

Common presentation:

- Wide spacing between eyes-Hypertelorism
- Narrow spacing between eyes- Hypotelorism
- Palpebral fissure length
- Epicanthal folds
- Common feature: Philtrum Length (upper lip and shape of nose)

Examination:

- Physical examination such as Anthropometry, head, ears eyes, limbs etc.
- Syndromes (FISH: Prader Willi, Angelman, Smith Magenis, Miller Dieker etc.)
- Growing Monitoring (height, weight, head circumference etc.)

Key features on examination and syndromes to consider

Key feature	Syndromes to consider
Short stature	Turner, Noonan, Russell Silver, Williams, skeletal dysplasias
Tall stature	Marfan, Homocystinuria, Klinefelter, Sotos
Obesity	Prader Willi, Bardet Biedl
Pubertal delay	Turner, Klinefelter
Aortic stenosis	Williams
Pulmonary stenosis	Noonan, Williams, Alagille
Conotruncal/ AVSD	22q11 deletion, trisomy 21
Aortic dilatation	Marfan, Ehlers Danlos
Radial ray anomalies	Fanconi anaemia, VACTERL, TAR, Blackfan Diamond
Deafness	Goldenhar, CHARGE, Waardenburg, Treacher Collins, Alport, NF-2
Cleft lip/palate	22q11 deletion (cleft palate), Stickler
Café au lait macules	NF-1, Fanconi Anaemia, McCune-Albright

Kaufman J, White SM. An approach to the child with dysmorphic features. J Paediatr Child Health. 2017 Mar;53(3):221-222.

Investigation:

- General: prenatal & perinatal history
- Skeletal survey
- Chromosomal, DNA, biochemical assay
- ECG/USG can be useful for additional major or minor abnormalities

Management:

- Appropriate medical/ surgical management wherever feasible

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	Dysmorphic Children
i. At the time of Pre-authorization	
Clinical notes including history, evaluation findings, and planned line of management	Yes
Based on Etiology Whole body scanning	Yes

DNA/chromosomal /biochemical assay Skeletal Survey USG Electrocardiogram (ECG) 2D ECHO Thyroid profile Viral Serology markers	
ii. At the time of claim submission	
Detailed Indoor Case Papers (ICPs) mentioning the treatment details	Yes
Investigation reports (if required)	Yes
Detailed Discharge Summary	Yes

PART II: GUIDELINES FOR PROCESSING TEAM

2.1 **Objective:** To provide guidance to the pre-authorization and claims processing team in ascertaining the medical necessity of procedure carried out vis a vis the patient's medical condition as evidenced by supporting documents/investigation reports etc., in deciding the admissibility and quantum of claim and compliance with mandatory documents by the hospital.

2.2 **Following mandatory documents to be diligently reviewed by the pre-auth / claims processing personnel:**

2.2.1 **At the time of pre-authorization processing- For pre-authorization processing doctor (PPD)**:

a. Clinical notes - detailed history especially prenatal, perinatal, maternal medication history and family history, signs & symptoms, vital monitoring, planned treatment line, and advice for admission?
b. Did clinical presentation and imaging/investigation suggestive of the diagnosis?

2.2.2 **At the time of claim processing- For claims processing doctor (CPD):**

a. Are the detailed ICPs with daily vitals and treatment details submitted?
b. Is the Discharge summary with follow-up advice at the time of discharge?

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**:
Dysmorphic Disorder in Children:

I. Did the clinical examination suggest a dysmorphic disorder? Yes
II. Did imaging/investigations support the diagnosis? Yes

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

References

1. Keigman, ST Geme, Shah, Tasker, Blum. Nelson`s Textbook of Pediatric. Elsevier. 21st Edition
2. Ranganath P. Approach to a child with Dysmorphism / Congenital malformation. Indian Academy of Medical Genetics, Genetic Clinics. 2014; 7(3):11-17.
3. Kaufman J, White SM. An approach to the child with dysmorphic features. J Paediatr Child Health. 2017 Mar;53(3):221-222. doi: 10.1111/jpc.13316. PMID: 28251749.
4. Jones KL, Adam MP. Evaluation and diagnosis of the dysmorphic infant. *Clin Perinatol*. 2015;42(2):243-viii. doi:10.1016/j.clp.2015.02.002