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## Systematic Review

# A systematic review on dental perspectives on pycnodysostosis

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### ABSTRACT

Pycnodysostosis (PYND) is an inherited bone disorder known as autosomal recessive. Patients with this disorder can be distinguished by the presence of a height smaller than expected for adults as well as smaller limbs with other dental deformities. As this disorder is rarely diagnosed, this review aimed to highlight the various dental findings from case reports and determine the treatment for the same. To date, no definitive therapies have been developed for this disorder. New treatment modalities are needed for the disease, and proper written protocol guidelines must be adopted while treating patients from dental perspectives. Continuous medical education should be provided to undergraduate students so that they would become familiar with the disease. More large-scale clinical trials are needed to determine better outcomes in general.

**Keywords:** Pycnodysostosis, Short stature, High arched palate, Dental

### INTRODUCTION

In 1962, Lamy and Maroteaux classified an inherited bone disorder called autosomal recessive, PYND, or Maroteaux-Lamy syndrome in both males and females.<sup>1,2</sup> The main features are diffuse thickening of the skeleton with cortical thickening, and constriction of the medullary canal. The patient had a short stature, short limbs, and malformed nails. According to Elmore, the first characteristic case of PYND was reported by Montanari in 1923 in a 1.5-year-old male patient with an aberrant variant of achondroplasia. Approximately 200 people with this disorder have been described in the medical literature.<sup>1</sup>

The general consensus is that it affects approximately 1 in 1.7 million people. Some people with PYND have a growth hormone deficiency, and another hormone deficiency, insulin-like growth factor 1 (IGF-1). Affected people reach a height smaller than expected for adults (short stature); their arms and legs are usually abnormally small. Other symptoms include loose (flabby) joints, underdeveloped sinuses (hypoplasia), enlarged liver and spleen (hepatosplenomegaly), and underdeveloped pituitary gland.<sup>2</sup> The other findings were noted as slow closure of the suture was commonly seen in infants with disorders. The usual facial attributes involve an abnormally protruberent forehead (frontal bossing), a

spearlike nose, a highly arched palate, prominent eyes, and bluish sclerae, and due to the underdevelopment of the bones of the jaw, may result in a small jaw (hypoplasia of the maxilla and mandible).<sup>1</sup> The significant oral and dental attributes are midfacial hypoplasia, grooved palate, dual row of teeth, late eruption of permanent teeth, numerous caries, retained deciduous dentition, hypodontia, enamel hypoplasia, unusual tooth configuration, and poor oral hygiene. There are chances of fractures of the bones pathologically are consequence of sclerosis, and radiologically, skull bones shows condensed with unclosed fontanels resembled as 'bone's lake,' osteosclerosis, suture of frontal and cranial are unlocked, non pneumatization of paranasal sinuses, hypoplasia of facial bones and fractures are seen in lower extremities.<sup>1-3</sup> As this disorder is genetically inherited, the detection and diagnosis of syndrome could be demonstrated in a proband with extensive clinical and radiographic factors, and molecular genetic testing can be conducted for biallelic pathogenic variants in the cathepsin K gene CTSK.<sup>4,5</sup>

This review was conducted with the aim of creating awareness of PCND among dental professionals regarding its common dental findings and management.

## METHODS

The search was conducted in accordance with the preferred reporting items for systematic reviews and meta-analyses and adhered to IJBCP formatting and manuscript drafting guidelines (Figure 2).<sup>6</sup>

A strategic search was carried out by using Medical Subject Heading (MeSH) term such as "Pcynodysostosis" AND "Case report", AND "dental perspective" were searched within the databases such as PubMed, CINAHL, Cochrane library and APA PsycInfo by using BOOLEAN operator "AND" in between keywords (Table 1).

## Inclusion criteria

The inclusion criteria were applied for thorough scrutiny of the case reports, such as the years 2017 to 2022, free full text, case reports, associated data, and English language. The studies primarily focused on the case studies with the title on the PYND and its impact on dental aspects.

## Exclusion criteria

Studies that did not assess PYND for informational purposes and relevant clinical outcomes excluded from this review. Also excluded randomized controlled trials, observational studies not involving PYND, paid articles, and materials misaligned with research objectives. Articles published in languages other than English were omitted.

## Data extraction

The authors meticulously carried out data extraction to ensure the accuracy of pertinent information from the literature. Each author used a predetermined technique to gather specific data, including study characteristics, intervention details, and outcomes. Data then transferred to a MS excel spreadsheet from CSV files and cross-checked by authors to resolve inconsistencies, enhancing accuracy of extraction. Data analysis conducted manually based on review's inclusion and exclusion criteria.

## Quality assessment

The study's assessment involved two independent reviewers utilizing predefined inclusion criteria. Conventional quality assessment tools were deemed unsuitable due to inclusion of case studies. Methodological quality was evaluated using the CARE checklist, which measures clarity, diagnostic reasoning, intervention detail, clinical data completeness, and outcome description.

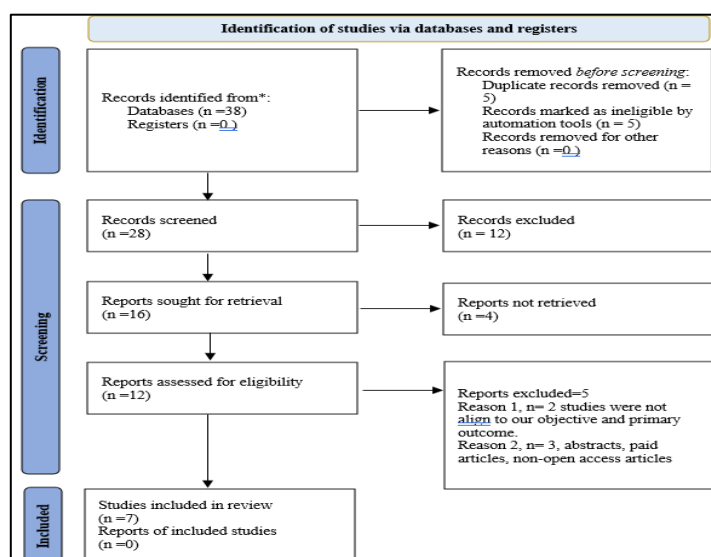


Figure 1: Flow diagram provides overview of selection process, search terms, eligibility criteria, and data extraction by using PRISMA-2 tool.

**Table 1: Comparative keywords and synonyms used in the search strategy for dental perspectives on PYND.**

Concepts	MeSH / Controlled vocabulary terms	Free-text keywords and synonyms (title/abstract)	Boolean within concept
<b>PYND (disease)</b>	"Pycnodysostosis" [Mesh]	Pycnodysostosis OR pyknodysostosis OR "Maroteaux-Lamy syndrome"	OR
<b>Dental perspectives</b>	"Dentistry" [Mesh] OR "Oral Health" [Mesh] OR "Tooth Diseases" [Mesh] OR "Mandible" [Mesh] OR "Maxilla" [Mesh]	dental OR dentistry OR oral OR teeth OR tooth OR mandible OR maxilla OR orthodontic* OR palate OR "high arched palate" OR hypoplasia OR eruption OR crowding OR "open bite" OR osteomyelitis OR "dental finding*" OR "oral manifestation*" OR "dental perspective*"	OR
<b>Case reports/ study type</b>	"Case Reports" [Publication type]	"Case report*" OR "case series" OR "case study*"	OR

**RESULTS**

After scrutinizing total 7 relevant articles were included in the study with the associated history, demographic details

and its dental findings are included in the Table 1. For the better diagnostic perspective on the condition, intraoral and radiographical findings are illustrated in the below Table 2.

**Table 2: Demographic details along with associated medical history.**

Author name	Year and place of the study	Patient details	Associated history
<b>Subramaniam and Muthukrishnan<sup>7</sup></b>	2017, India	20 year/male	No associated history was mentioned in the report
<b>Bhoyar et al<sup>8</sup></b>	2020, India	58 year/ male	Non-consanguineous marriage of parents, history of (H/o) treated osteomyelitis of left posterior body of mandible, frequent bone fracture
<b>Jawa et al<sup>9</sup></b>	2020, India	16 year/female	H/o leg fracture thrice H/o consanguinity of parents was positive
<b>Khoja et al<sup>10</sup></b>	2015, Pakistan	13 year and 11 month/ Female	H/o marble bone disease, multiple and recurrent bone fracture
<b>Rodrigues et al<sup>11</sup></b>	2017, Brazil	13 year/female	H/o recurrent accidents due to lower limb fracture
<b>Costa et al<sup>12</sup></b>	2020, Uberlandia	30 year/ male	Family h/o two cousins have same syndrome
<b>Aghili et al<sup>13</sup></b>	2017, Iran	5 year and 8 months/ female	No associated history was mentioned in the report

**Table 3: Intra-oral and radiographical findings.**

Authors name	Intra-oral and radiographical findings
<b>Subramaniam and Muthukrishnan<sup>7</sup></b>	Intraoral assessment disclosed elevated arched palate, tooth crowding anterior open bite, posterior crossbite, retention 53, 63, 73, 83, 84, 85, 74, 75, 65. Defect 13, 23, 33, 43, 34, 35, 44, 45, not fully erupted 47 pericoronal flap overlies distal portion, periodontal pocket 47 to 6 mm distal, partial exudation of pus confirmed the presence of pericoronal abscess, compared with eruption 47. Panoramic radiographs showed crowded teeth, retained of teeth numbers 53, 63, 65, 73, 74, 75, 83, 84, 85 and impacted teeth 18, 13, 23, 25, 28, 35, 34, 43, 44, 45. Postero-anterior lateral skull radiographs showed marked hyperostosis, open fontanels, loose sutures, and airless sinuses
<b>Bhoyar et al<sup>8</sup></b>	The radiographic findings showed hypoplasia of the maxilla, frontal sinuses were non-pneumatized, and recessed nasal bridge and undeveloped mandible with blunt jaw angle. There was presence V-shaped arches of the upper and lower jaws, class III incisor relationship with a reverse overjet of 2 mm, bilateral open bite, narrow and grooved palate, ununiform and misplaced teeth, and V-shaped arches of the upper and lower jaws. The overall oral health were good, with slight calculus and plaque deposits on the upper anterior surface of the lingual side and on the lower anterior surface of the lingual and labial area, no periodontitis. Even so, slight swelling and color change were noticed. A mandibular first permanent molar had a class I carious lesion that was restored with a composite filling.

Continued.

Authors name	Intra-oral and radiographical findings
Jawa et al <sup>9</sup>	The oral observations included entire absence teeth on maxillary and mandibular arches and the presence of an intraoral drain sinus in the upper maxillary region of the third molar on right side. The patient had a small palate with furrow and a macrotongue. Under radiographic examination, orthopantomogram [OPG], skeletal probe, and non-contrast computed tomography (NCCT). OPG showed vague osteolytic lesions including maxillary right posterior alveolar, alveolar ridge atrophy, and a general elevate in bone thickness. NCCT showed marked atrophy of the maxillary and mandibular alveolar arches with increased bone density. A destructive lesion of sclerotic bone surrounded by a radiolucent rim suggestive of dead bone was existed on the maxilla of right periphery (PYND with maxillary osteomyelitis)
Khoja et al <sup>10</sup>	Intraoral examination showed in the both jaw with the appearance of whole permanent dentition, narrowed maxilla, narrowed furrow palate, and narrowed mandible. Arches represents the existence of non-uniform placement of teeth. On oral and radiographic intervention, on 55 confinement of teeth were observed, 37 with pulp-involving decay, 46 with pulp-involving caries, and entire loss of crowns clinically were not present
Rodrigues et al <sup>11</sup>	Ogival palate, and generalized dental crowding, and typical enamel hypoplasia, with the presence of caries, prognathic mandible, Micro-teeth, impacted teeth, missing teeth, and an obtuse mandibular angle almost horizontal in relation to the mandibular body were identified on the panoramic radiograph. Cephalometric radiography showed frontal bossing, an obtuse mandibular angle, and a horizontally positioned mandibular condyle, corresponding to Angle class III malocclusion
Costa et al <sup>12</sup>	Mandibular hypoplasia with loss of mandible angle abnormal dentition and craniofacial disproportion. In the intraoral clinical examination, it was detected an absence of occlusal stability increase of the free functional space, inadequate dental positioning, atypical and arched palate with fibrous tissue development typical of the syndrome.
Aghili et al <sup>13</sup>	The radiographs revealed a very obtuse mandibular angle, general increase in bone density, and open fontanels and sutures. The radiologist's report included general increase in bone density, craniofacial disproportion, open fontanels, wormian bones, hypoplasia of clavicle, hypoplastic distal phalanges with absence of distal tufts, mild coxa valga deformity

## DISCUSSION

Combining the Latin words "con" for together or with and "sanguineous" sanguineous' for blood, we obtain the English word consanguinity. Consanguineous marriage is defined as the union of two people who are second cousins or closer in a clinical genetic setting.<sup>13,14</sup> In some fields, close ties that are not second cousins are considered to fall under the definition of consanguineous marriage. According to estimates, one billion people in the world now favour consanguineous unions for a variety of socioeconomic factors.<sup>15</sup> The Middle East, Northern India, and North Africa have the highest rates of consanguineous unions. It is also known that genetic aberrations are more frequently seen in areas where consanguineous marriage is a common occurrence.<sup>16</sup> These marriages have several harmful effects on the children born from them unless the parents contain gene mutations, as it facilitates the passage of the congenital malformations, reported problems such as birth defects, genetic diseases, heart and blood diseases, mental disability, hearing problems, asthma, congenital head and neck malformation, and cleft lip or palate, which affect the quality of life of the individual. This review reported that most patients were born through consanguineous marriages.<sup>17,18</sup>

Patients with this disorder are mainly managed with a growth hormone regime; moderation in the environment and occupation are required; for patients with fracture and scoliosis, orthopedic treatment may be needed for other anomalies such as cleft palate, craniosynostosis can be treated by craniofacial and neurosurgical management, for obstructive sleep apnea, pulmonology, and sleep medicine

specialist are required, useful advice from specialists and anesthetists before any surgery, dental care for dental abnormalities, quality care for fractures and scoliosis related to the eye regions, and vision concerns need guidance from the ophthalmologist.<sup>2,19</sup>

Hence, the above are the common findings and treatment for the patients with this disorder, in this review our aim was to highlight the general dental findings and their management in the patient with the pcyndysostosis.<sup>4</sup> Dental practitioners must be aware of the definitive characteristics of this disorder in order to make accurate and prompt detection and examination, especially when meeting these patients for the first time, and have in-depth knowledge of differential diagnosis. This aids in proper dental care of these patients.<sup>10</sup> The major complication reported was osteomyelitis (OSA), which usually affects patients with dysostosis, osteoarthritis due to decreased angiogenesis, and elevated bone density, posing challenges in its medical therapy and care. Treatment of osteomyelitis is considered as per the proportion, position of the bone deformity, and emergence; hence, antibiotics are generally adopted, whether related to surgical resection or not. Instead, buccal fat pad (BFP) grafts, both pedicle and free, were utilized to ensure complete curing of the mucosa and consistent coverage of the altered area following isolated or incomplete excision, preserving the bone from infection.<sup>10,21</sup> Due to presence of Brittle bone is prone to fracture during dental treatment, especially tooth extraction. Many oral appliances are currently being used to treat OSA. They work by opening the vertical dimension, pushing the lower jaw forward, sticking out the tongue, or elevating the soft palate to increase the space in

the upper airway. Although there have been no reports of success with such devices in patients with PYND, anterior positioning of the mandible may prevent obstruction of the oropharyngeal airway. The use of intraoral appliances should be considered in patients with PYND and respiratory failure. Surgical correction of maxillofacial abnormalities by osteotomy has been successful in young adults; however, there are no reports of this approach.<sup>2,10,21</sup>

Given the medical and oral assessment of osteomyelitis in patients with osteonecrosis, the preferred therapies were sequestrum resection and primary cessation by buccal fat pad (BFP) used as a graft due to the expanded loss of bone. In addition, 300 mg clindamycin was administered twice a day for 14 days. Surgery was conducted under local anesthesia succeeding antibiotics use for 1 week, followed by eradication of decomposed bone tissue, surgical removal of dead bone, and creation of a flap to accommodate the BFP. The graft was harvested from the ipsilateral upper second molar area by straight and careful dissection. BFP grafts are conditioned within the operative area to modify healing and conservation and allow adequate wound edge contact and primary closure.<sup>10</sup> Another appliance used as treatment regime were upper bonded expander constructed with the centerline screw expander which was triggered 0.25 mm on every second day using a passive expansion approach.<sup>22</sup>

The device was kept passive to allow tissue remodeling for the next two months. After the passive expansion approach, the latter approach leveled and equalized the upper teeth with massive fixation using Roth 0.022 slots therapy. After alignment of the upper dentition, the lower teeth were glued. Further treatment planning included extraction of the 3<sup>rd</sup> molar and right lower lateral incisor under general anesthesia and aggressive distalization of the upper right 1<sup>st</sup> permanent molar and mandibular 1<sup>st</sup> molar using a mini-implant fixture, including the correction of permanent molars. The overall goal of orthodontic treatment is to achieve normal overjet, overbite, and class I buccal occlusion to improve optimal function and esthetics.<sup>9</sup> For the patient complained of pain, and treated with 5-day regimen of antibiotics: C. amoxicillin + clavulanic acid, T. metronidazole 400 mg twice a day, anti-inflammatory T. aceclofenac + T. paracetamol thrice a day, and astringent gums. Topical dye (tannic acid + potassium iodide + glycerin + menthol) was also added.<sup>9</sup>

Radiographic findings are also beneficial for the good prognosis of patients. Radiographic findings were corroborated using axial imaging. Axial imaging defines the precise relationships between non-erupted teeth that are not possible with Panorex alone. Coronal and sagittal reformatting facilitate the interpretation of craniocaudal anatomical relationships. Surface rendering provides an overview of underlying bony abnormalities, whereas volume rendering helps determine the 3D relationships between abnormal teeth by obscuring alveolar processes. Showing the exact relationship between abnormal teeth

helps surgeons plan extractions and/or reconstructions.<sup>24</sup> Furthermore, improved visualization of the cortical bone helps distinguish infected follicles and tooth abscesses from normal, well-cortical follicles in unerupted teeth. This helps the surgeon more fully understand the abnormal anatomy and plan extractions and/or prosthetic implants; hence, it is considered as the modality of orthodontic treatment.<sup>10,25</sup>

To date, no definitive therapies have been developed for this disorder. The utmost risk to the patient's PYND is fractures; therefore, it is crucial to take the necessary measures to avert or decrease the occurrence of fractures. These precautions include caution when handling an affected child and precautions to avoid accidental injury during daily activities, especially for patients with abnormalities, which would be beneficial.<sup>10</sup>

PYND is a rare disease that is primarily detected on the basis of its attributes based on examination. Recognizing these features is important for making the correct diagnosis, which enables the treatment and precautions for other upcoming ailments, improving the patient's quality of life.<sup>26</sup> While handling and treating patients, careful and vigilant awareness must be given because brittle and delicate bone may be the outcome of defects in osteoclast function. Sole focus must be given to maintaining oral hygiene as good as possible, and precautionary actions must be taken by the dentist while the patient is on a dental chair. Needful expansion of the affected jaw and arches can be performed using passive expansion guidelines, and orthodontic equalization of teeth can be conducted; however, the prognosis of orthodontic therapy may differ at the individual level.<sup>19,27</sup> For treating short stature, growth hormone therapy may prove very effective if used at a young age.<sup>28</sup>

Hence, this review highlights the general examination and oral findings of the disease that require strict vigilance by practitioners. The positive outcome of this review was every aspect of dental and oral findings, along with radiographic detection.

A drawback of this review was the small number of cases considered, and fewer treatment modalities were examined. To understand the intervention in more detail, a randomized controlled trial is needed for review and analysis.

## CONCLUSION

The review concluded that treating PKND at an early age after finding dental anomalies would be effective and beneficial for performing common tooth functions in children. Orthodontic and intraoral appliances were found to be effective in these patients. Complications such as osteomyelitis were treated surgically with BFP. Hence, the treatment and new intervention methods required for studies, further case reports, and randomized controlled trials directing medical management are new calls.

### Future perspective

New treatment modalities are needed for the disease, and proper written protocol guidelines must be adopted while treating patients from dental perspectives. Continuous medical education should be provided to undergraduate students so that they would become familiar with the disease. More large-scale clinical trials are needed to determine better outcomes in general.

### Forensic medicine perspective

From a forensic medicine perspective, the systematic review on PYND offers crucial insights into the identification, recognition, and differential diagnosis of this rare genetic disorder. Forensic pathologists and medical examiners may encounter individuals with PYND in cases involving sudden or unexpected deaths, necessitating familiarity with the distinctive skeletal and dental features described in the review, such as short stature, cortical bone thickening, high arched palate, and dental anomalies. Clear documentation and comprehensive reporting of PYND-related findings are essential for accurate record-keeping during postmortem examinations, aiding in subsequent medico-legal proceedings. Additionally, forensic experts must consider injury patterns and fracture risks associated with PYND, given the increased susceptibility to bone fractures due to characteristic skeletal abnormalities and fragility. Analysis of medical interventions and complications, including osteomyelitis and respiratory concerns, may be pertinent in forensic evaluations, requiring examination of medical records and treatment outcomes to determine contributory factors relevant to the cause of death. Furthermore, forensic investigations involving PYND may prompt considerations of familial inheritance patterns and genetic testing for affected family members, highlighting the importance of genetic counseling services for surviving relatives. By integrating this knowledge into forensic practice, practitioners can enhance the accuracy of medico-legal assessments involving individuals with PYND, contributing to comprehensive forensic evaluations and informed decision-making processes.

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