



Primary Failure of Eruption

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Introduction

Primary Failure of Eruption (PFE) is a rare disorder in which the dentition do not adequately erupt into a typical occlusion. (1) It may be divided into 2 variations: Type I and Type II. Type I features a distally slanting open bite from anterior to posterior. Type II offers a varied eruption pattern, in more than one quadrant, with greater eruption than Type I, but ultimately less second molar height than a normal occlusion.(2) Incisors, canines, and premolars may be involved to a reduced degree.(3) Other variations may include a Class III occlusion and/or poor root malformations, including blunting and dilacerations.(4) Missing dentition may also be a coincident finding.(1, 5) Permanent (more commonly reported) and primary teeth may be affected, ultimately failing to gain vertical function. Primary teeth that may be affected are noted as teeth erupting into an established, initial occlusion and then failing to erupt to the final growth level.(2, 3, 5, 6)

Genotypic and Phenotypic Interpretations

The patient may not recognize the malocclusion due to its longstanding nature. Family members may have similar presentations.(3) Genetically, PFE involves a mutation that disrupts bone homeostasis (osteoclastogenesis, chondrogenesis, bone remodeling, and renal function) and the dental follicle interaction.(1, 2, 5, 7) Inheritance follows an autosomal dominant pattern with complete penetrance and variable expressivity.(5, 8) A multi-factorial involvement, environmental and physiological relational cause is also suspected.(9) Due to this genetic mutation, a cascade of normal eruptive events is hindered.(10) Phenotypically, its presentation includes an infraoccluded supracrestal first molar, which may extend posteriorly, affecting adjacent teeth. Potentially, bilateralism and/or involvement of the opposing arch may be seen.

Differential Diagnosis

PFE appears similar to other pathology involving reduced eruption (cyst- induced, traumatically impaired, tongue thrust, or ankylosis).(2, 4) The cause of ankylosis is the union of bone and cementum due to the loss of the periodontal ligament often noted in traumatized areas, with pathological complications. Generally, only a single tooth is affected. PFE, in contrast, is more generalized. One may note a slanting infraocclusion, anteriorly to posteriorly. PFE may cause a lateral open bite.(4) Confounding the diagnostic picture, there are rare reports of PFE and ankylosis seen in different arches of the same individual.(2) Additional clinical conditions similar to PFE include: Cleidocranial dysplasia, osteopetrosis, Rutherford syndrome, GAPO syndrome, osteoglyphonic dysplasia, and dental non-eruption.(3)

Diagnostic Methods

Salivary studies are ongoing and may involve samples screened for a PTH1R mutation which is thought to cause PFE. The saliva sample kits do not have FDA approval to screen for PFE and therefore results are not considered definitive at this point.(11) However, this information can be paired with clinical factors such as the patient's history, radiographs, and treatment(s) rendered to more fully understand the patient's presentation and develop a potential diagnosis.

Clinical Management

As previously stated, the innate ability of the dentition to erupt and offer subsequent function is impaired. Misinterpretation of the clinical presentation may lead to orthodontic intervention which results in little to no movement.(8) If unchecked, orthodontic ankylosis is a likely outcome. The provider must determine what can be reasonably achieved, be it slight movement or maintenance of the presented state. The orthodontist's role is to hold the present occlusion or to offer slight variations via segmental mechanics. The orthodontist must rely on the abilities of their fellow dental colleagues to render an improved function and esthetic outcome once growth is complete.(6) The oral surgeon may offer a segmental osteotomy/regional distraction osteogenesis after removal of the unerupted dentition. The periodontist may offer supportive implants, with the prosthodontist's role serving to introduce a functional occlusion.(1, 3) Milder cases may be treated with limited coronal build-ups or crowns.(7) All of this is not possible without an accurate screening when PFE is suspected.(12) Furthermore, the recognition of PFE may alleviate years of unfavorable attempted treatment of the patient and family members.(7) It has been said, "When you hear hoofbeats, think of horses not zebras," meaning that rare cases do not generally present themselves. But in some cases, listen for a galloping zebra.

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Lateral cephalogram – note bilateral open occlusion.

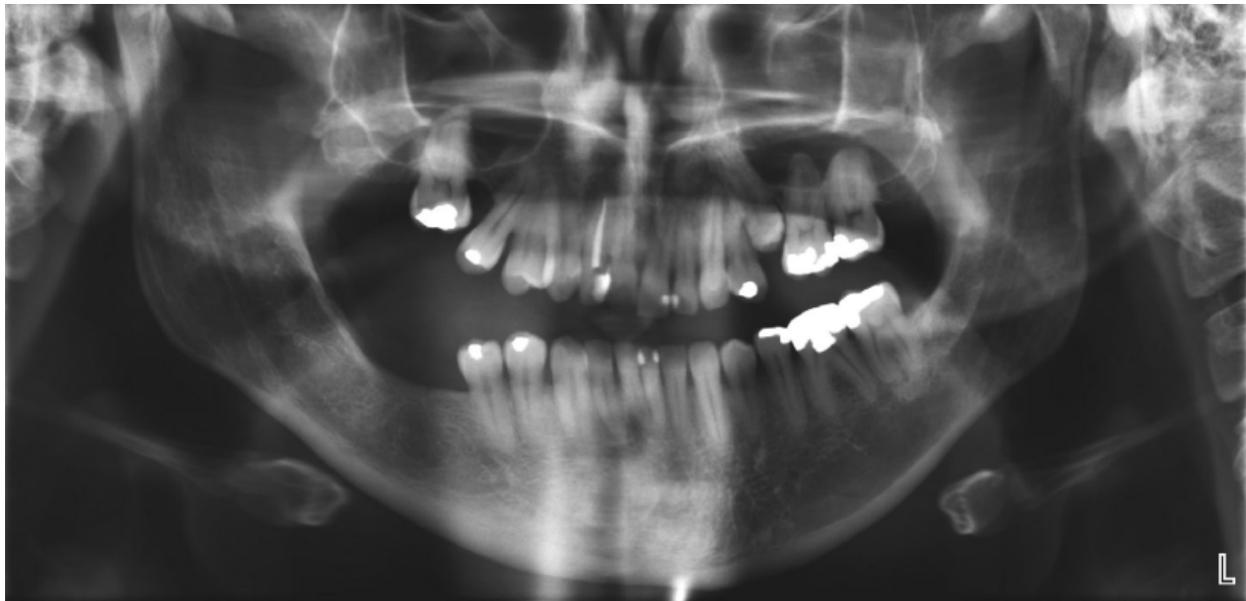


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Lateral right, Central, and Lateral left intraoral presentation, prior to initial coordinated treatment.



Panograph presentation.



Lateral right, Central, and Lateral left intraoral presentation. Limited orthodontics, OMFS, Prosthodontic and Periodontic treatment in progress.