Fibro-osseous lesions

Fibro-osseous lesions are a diverse group of processes that are characterized by replacement of normal bone by fibrous tissue containing a newly formed mineralized product.

Fibro-osseous lesions of the jaws include

1. Developmental (hamartomatous) lesions: - fibrous dysplasia
2. Reactive or dysplastic processes: - cemento-osseous dysplasia
3. Neoplasms: - ossifying fibroma

Although these lesions share many histopathological features but they have different etiology, clinical behavior, and prognosis.

Fibrous dysplasia:

Fibrous dysplasia is a developmental tumorlike condition that is characterized by replacement of normal bone by an excessive proliferation of cellular fibrous connective tissue intermixed with irregular bony trabeculae. Fibrous dysplasia result from postzygotic mutation in (GNAS I gene) (**present in 86% of the cases of fibrous dysplasia**). This genetic mutation affects the proliferation and differentiation of fibroblasts/osteoblasts that make up these lesions. The clinical severity of the condition depends on the point in time during fetal or postnatal life that the mutation of GNAS1 occurs.

- If mutation occur during early embryologic life: multiple bone lesions, cutaneous pigmentation, and endocrine disturbances will present.
- at later stages of embryonic development: multiple bone lesions of fibrous dysplasia will develop.
- Postnatal mutation: one bone is affected.

Clinical and radiographical Features:

Fibrous dysplasia may involve a single bone or several bones. When the disease is limited to a single bone, it is termed **monostotic fibrous dysplasia** (accounts for about 80% to 85% of all cases). most examples of monostotic fibrous dysplasia are diagnosed during the second decade of life. Males and females are affected equally. A painless swelling of the affected area is the most common feature. Growth is generally slow (Occasionally rapid). The maxilla is involved more often than the mandible. Maxillary lesions may extend to involve the maxillary sinus, zygoma, sphenoid bone, and floor of the orbit (**craniofacial fibrous dysplasia**).

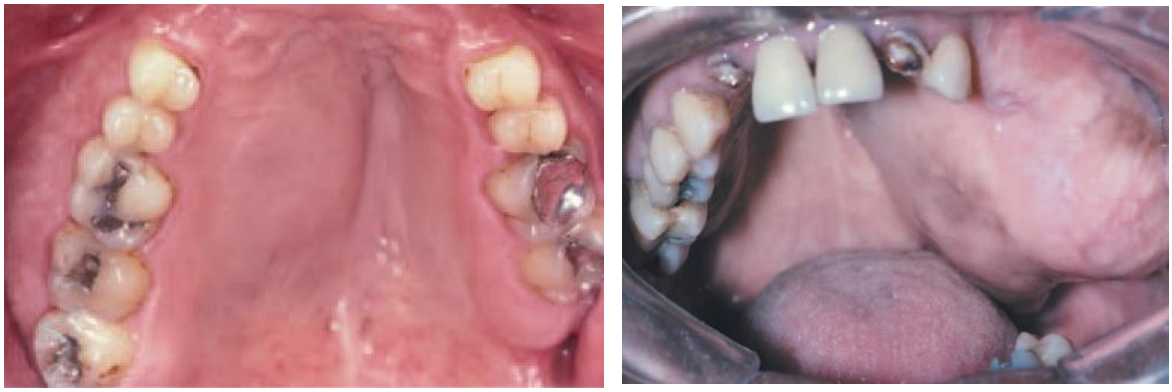
The chief radiographic feature is a fine "ground glass" opacification that results from superimposition of a numerous poorly calcified bone trabeculae arranged in a disorganized pattern. the lesions of fibrous dysplasia are not well demarcated. The margins tend to blend into the adjacent normal bone so that the limits of the lesion may be difficult to define. In the earlier stages, the lesion may be largely radiolucent or mottled. Involvement of the mandible often results in expansion of the lingual and buccal plates and bulging of the lower border. Superior displacement of the inferior alveolar canal is not uncommon. Periapical radiographs of the involved dentition often demonstrate narrowing of the periodontal ligament space with an ill-defined lamina dura that blends with the abnormal bone pattern. When the maxilla is involved, the lesional tissue displaces the sinus floor superiorly and commonly obliterates the maxillary sinus.

Involvement of two or more bones is termed **polyostotic fibrous dysplasia** (No. of involved bones varies from a few to entire skeleton). When polyostotic fibrous dysplasia seen with café au lait (coffee with milk) pigmentation, the process is termed **Jaffe-Lichtenstein syndrome.**

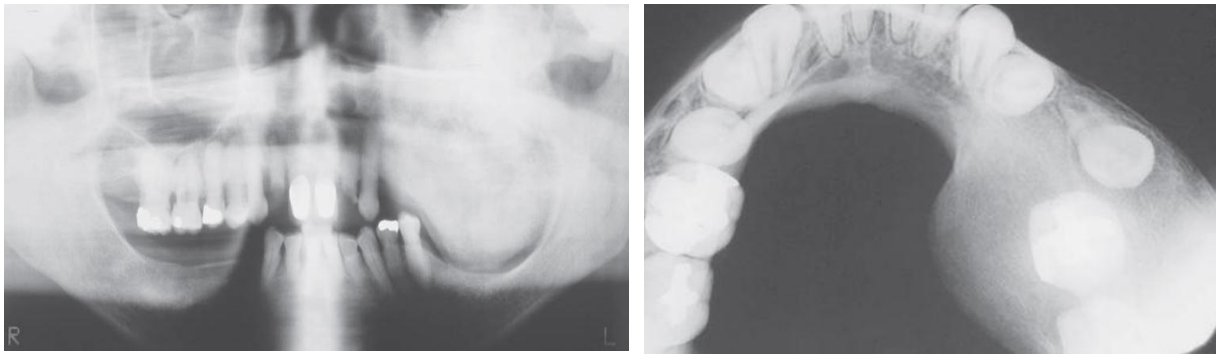
Polyostotic fibrous dysplasia also may be combined with café au lait pigmentation and multiple endocrinopathies (sexual precocity, hyperthyroidism, acromegaly, hyperparathyroidism). This pattern is known as the **McCune Albright syndrome.**

Another rare disorder associated with fibrous dysplasia is **Mazabraud syndrome**, characterized by fibrous dysplasia in combination with intramuscular myxomas.

the café au lait pigmentation consists of well-defined, generally unilateral tan macules on the trunk and thighs. The margins of the café au lait spots are typically very irregular, resembling a map of the coastline of Maine (in contrast to the café au lait spots of neurofibromatosis which have smooth borders (like the coast of California)).



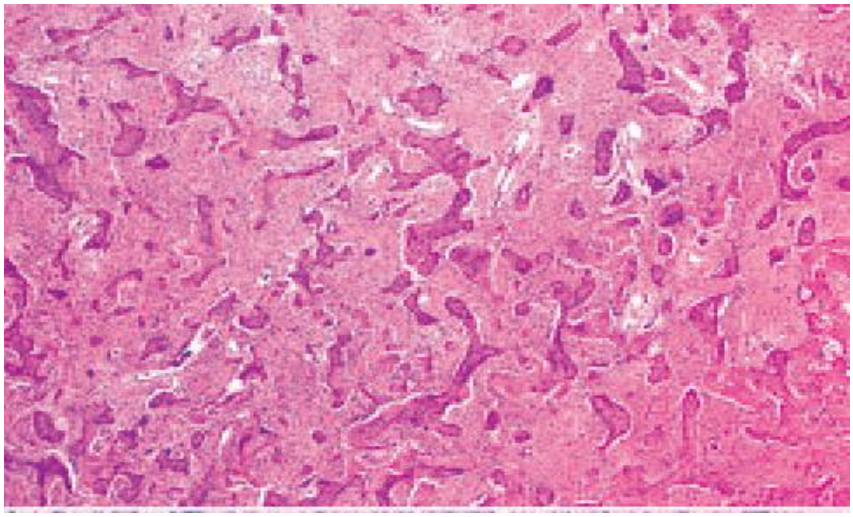
Fibrous dysplasia involving the maxillary bone



Ground glass opacity of fibrous dysplasia

Histopathological features:

Fibrous dysplasia consists of cellular fibrous connective tissue stroma (fibroblasts exhibit uniform spindle-shaped nuclei (no mitotic figures)) that contains foci of irregularly shaped trabeculae of immature woven bone. The bony trabeculae assume irregular shapes (likened to Chinese characters) and haphazardly arranged. The bone is predominantly woven in type and appears to arise directly from the collagenous stroma without osteoblastic rimming. In a mature fibrous dysplasia lesion, lamellar bone may be found. Capillaries typically are prominent and uniformly distributed.



Fibrous dysplasia: Irregularly shaped and disconnected trabeculae of woven bone in a fibrous stroma.

Treatment and Prognosis

- After a variable period of prepubertal growth, fibrous dysplasia characteristically stabilizes.
- Small lesions may require no treatment only periodic follow-up.
- Large lesions that have caused cosmetic or functional deformity may be treated by surgical recontouring (delayed until stabilization of the lesion).

- En bloc resections for complete removal are impractical and unnecessary because the lesions are relatively large and poorly delineated.
- Malignant transformation is a rare complication of fibrous dysplasia (usually in patients with the polyostotic type previously treated with radiotherapy)

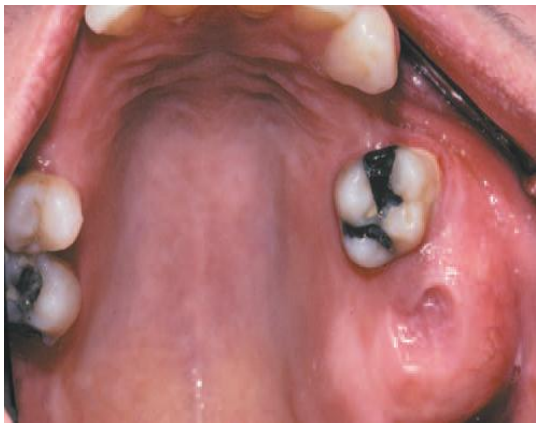
Ossifying fibroma (cementifying fibroma, cemento ossifying fibroma):

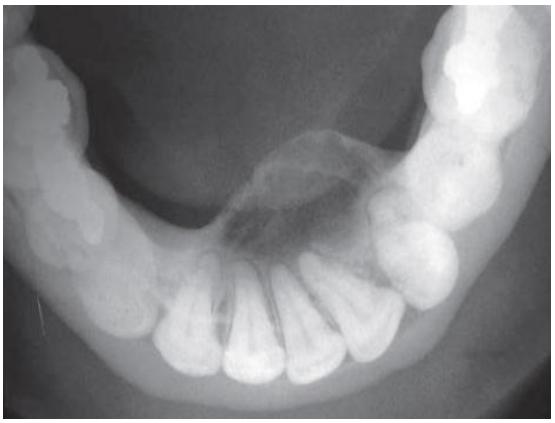
Ossifying fibroma is a benign neoplasm of unknown origin that can occur in any facial bone and has the potential for excessive growth, bone destruction, and recurrence.

Clinical and radiographical Features

- Ossifying fibroma is an uncommon lesion that tends to occur during the third and fourth decades of life.
- women more commonly affected than men.
- It is a generally slow growing, asymptomatic, and expansile lesion.
- In the head and neck, ossifying fibroma may be seen in the jaws, craniofacial bones, and anterior cranial fossa.
- Uncommonly, rapid growth may be seen in children (juvenile ossifying fibroma).
- Lesions of the jaws characteristically arise in the tooth bearing regions, most often in the mandibular premolar molar area
- The slow but persistent growth of the tumor within the jaws may ultimately produce expansion and thinning of the buccal and lingual cortical plates, although perforation and mucosal ulceration are rare.
- Most of these lesions are solitary
- The most important radiographic feature of this lesion is the well-circumscribed, sharply defined border present a variable appearance, depending on the density of calcifications present (unilocular or multilocular radiolucency to mixed radiolucent and radiopaque lesion to mostly radiopaque)

- The roots of teeth may be displaced; less commonly, tooth resorption is seen.

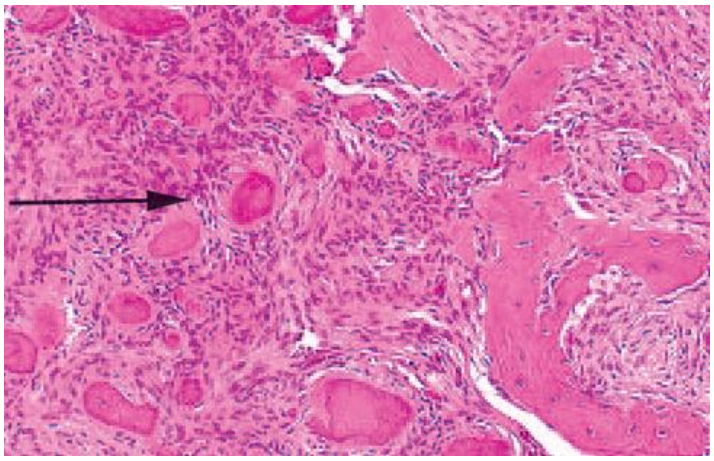




Histopathological features:

At surgical exploration, the lesion is well demarcated from the surrounding bone, thus permitting relatively easy separation of the tumor from its bony bed. A few ossifying fibromas will show, grossly and microscopically, a fibrous capsule surrounding the tumor. Most are not encapsulated but are well demarcated grossly and microscopically from the surrounding bone. On gross examination, the tumor is usually submitted in one mass or as a few large pieces. Ossifying fibromas consist of fibrous tissue that exhibits varying degrees of cellularity and contains mineralized material. The hard tissue portion may be in the form of trabeculae of osteoid and bone or basophilic and poorly cellular spherules that resemble cementum. Admixtures of the two types are typical. The bony trabeculae vary in size and frequently demonstrate a mixture of woven and lamellar patterns. Peripheral osteoid and osteoblastic rimming are usually present. The spherules of cementum-like material often demonstrate peripheral brush borders that blend into the adjacent connective tissue. Significant intralesional hemorrhage is unusual. Variation in the types of mineralized material (bone and cementum) produced may be helpful in distinguishing

ossifying fibroma from fibrous dysplasia, which has a more uniform pattern of osseous differentiation.



Ossifying fibroma shows a mixture of woven bone and cementum-like material. Note the spherules demonstrating peripheral brush borders (arrow).

Treatment and prognosis:

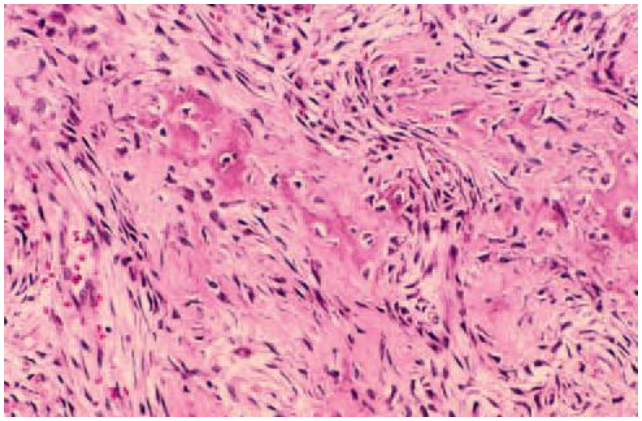
The circumscribed nature of the ossifying fibroma generally permits enucleation of the tumor with relative ease. Some examples, however, which have grown large and destroyed considerable bone, may necessitate surgical resection and bone grafting. The prognosis, is very good, and recurrence after removal of the tumor is rarely encountered. There is no evidence that ossifying fibromas ever undergo malignant change.

Juvenile ossifying fibroma

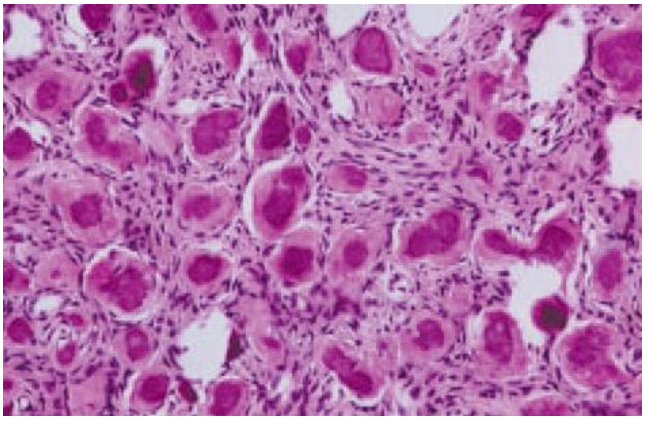
The term juvenile (aggressive or active) ossifying fibroma was used to describe two variants of ossifying fibroma (juvenile trabecular ossifying fibroma (JTOF) and juvenile psammomatoid ossifying fibroma (JPOF)) that occur in younger patients. Juvenile trabecular ossifying fibroma (JTOF) typically occurs in children and adolescents (younger than 15 years). The lesion occurs almost exclusively in the maxilla and mandible. JTOF is characterized by progressive and sometimes rapid growth but rarely pain. Radiographically, the tumor has a defined border and can range from radiodense to radiolucent. Following complete excision, recurrences of JTOF are infrequent. By contrast, the juvenile psammomatoid ossifying fibroma (JPOF) occurs principally in the extra-

gnathic craniofacial bones, particularly the paranasal sinuses and periorbital bones, where it may cause exophthalmos, sinusitis, and nasal symptoms. JPOF occurs in a slightly older population compared with JTOF (average 22 years). Treatment consists of surgical excision, but up to 30% of cases will show recurrences, sometimes multiple and over a span of many years.

Histopathologically, the trabecular variant shows irregular strands of highly cellular osteoid encasing plump and irregular osteocytes. These strands often are lined by plump osteoblasts and in other areas by multinucleated osteoclasts. In contrast, the psammomatoid pattern forms concentric lamellated and spherical ossicles that vary in shape and typically have basophilic centers with peripheral eosinophilic osteoid rims. A peripheral brush border blending into the surrounding stroma is noted in many of the ossicles. Occasionally, individual ossicles undergo remodeling and form crescentic shapes.



Juvenile trabecular ossifying fibroma



juvenile psammomatoid ossifying fibroma

Cemento-osseous dysplasia

Cemento-osseous dysplasia occurs in the tooth-bearing areas of the jaws and is probably the most common fi bro-osseous lesion encountered in clinical practice. Based on the clinical and radiographic features, cemento-osseous dysplasias are separated into three groups:

1. Periapical cemento-osseous dysplasia
2. Focal cemento-osseous dysplasia
3. Florid cemento-osseous dysplasia

Periapical Cemento-osseous Dysplasia

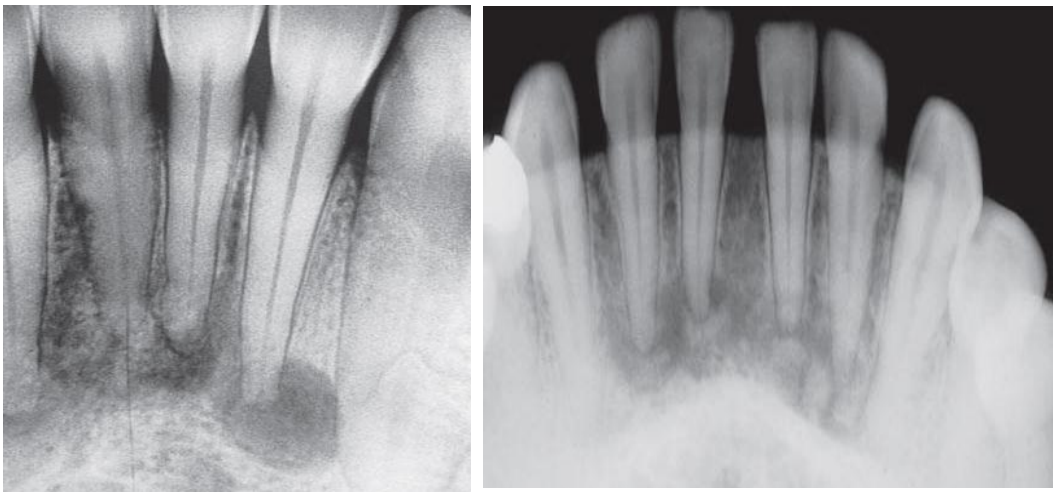
This lesion appears to be an unusual response of periapical bone and cementum to some undetermined local factor. Populations most at risk include East Asians and those of African origin. When not associated with a tooth apex, the term focal cemento-osseous dysplasia is used.

Clinical and radiographical Features

Relatively common phenomenon occurs at the apex of vital teeth. Women, especially black women, are affected more than men. Periapical cemento-osseous dysplasia appears mostly in middle age adult and rarely before the age of 20. The mandible, especially the anterior periapical region, is more commonly affected than other areas. Often, the apices of two or more teeth are affected. This condition typically is discovered on routine radiographic examination because patients are asymptomatic. It appears first as a periapical lucency that is continuous with the periodontal ligament space (similar to periapical granuloma or cyst) **the teeth are always vital**

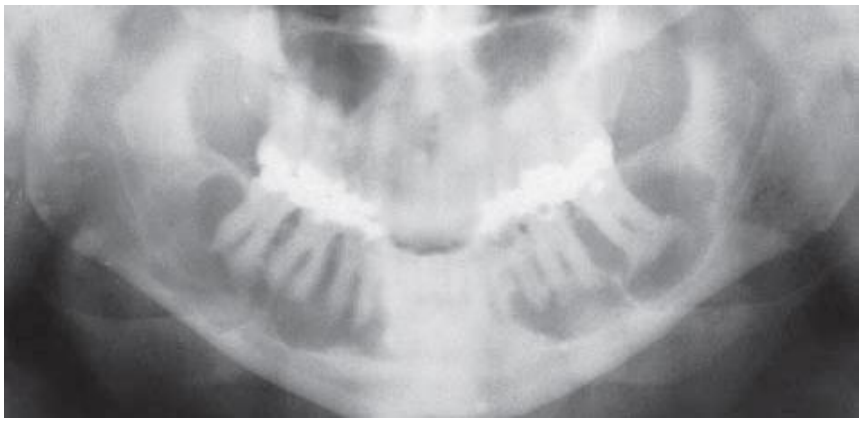
As the condition progresses or matures, the lucent lesion develops into a mixed or mottled pattern because of bone repair. In its final stage, the tumor appears as a solid, opaque mass

that is often surrounded by a thin, lucent ring. This process takes months to years to reach the final stages of development and, obviously, may be discovered at any stage.



Periapical cemento-osseous dysplasia

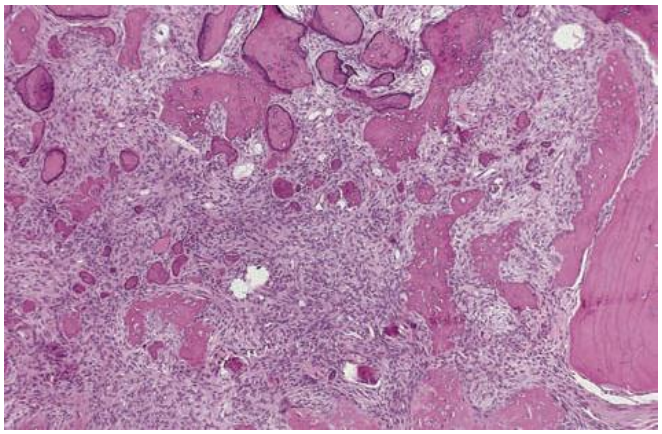
A related but less common condition is known as **florid cemento-osseous dysplasia (FCOD).** No cause is apparent and patients are asymptomatic, except when the complication of osteomyelitis occurs. Women, especially black women, are predominantly affected, usually between 25 and 60 years of age. The condition typically is bilateral and may affect all four quadrants. A curious finding has been the concomitant appearance of traumatic (simple) bone cysts in affected tissue. Radiographically, FCOD appears as diffuse radiopaque masses throughout the alveolar segment of the jaws. A ground-glass or cyst like appearance may be seen.



Florid COD

Histopathology

Periapical cemento-osseous dysplasia represents a mixture of benign fibrous tissue, bone, and cementum. Calcified tissue is arranged in trabeculae, spicules, or larger irregular masses. Reversal lines eventually are seen, and osteoblasts, cementoblasts, or both, line the islands of hard tissue. Chronic inflammatory cells may also be seen. Microscopically, periapical cemento-osseous dysplasia may appear very similar to chronic osteomyelitis and ossifying fibroma. Microscopically, FCOD is a heterogeneous lesion consisting of a benign fibrous stroma that contains irregular trabeculae of mature and immature bone and cementum like material. Because FCOD is an asymptomatic, self-limited process, no treatment is required. In cases in which secondary infection occurs, antibiotics and sequestrectomy may be necessary.



Florid cemento-osseous dysplasia

Treatment

No treatment is required for periapical cemento-osseous dysplasia or FCOD. Once the opaque stage is reached, the lesion usually stabilizes and causes no complications. Because teeth remain vital throughout the entire process, they should not be extracted, and endodontic procedures should not be performed.