

# Systemic Diseases and Conditions Affecting Jaws



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## KEYWORDS

- Radiopacity • Radiolucency • Mixed lesion • Ill-defined border • Effaced-border
- Localized lesion • Generalized lesion • Root resorption

## KEY POINTS

- Perform a detailed medical history and physical examination for each patient.
- Understand the potential osseous changes that may be caused by an underlying medical problem.
- Understand normal radiographic anatomy.
- Identify and describe any osseous changes noted on radiographic imagery of the patient.
- Develop a differential diagnosis and treatment plan.

This article is written to highlight and focus on patients with medical conditions seeking dental treatment. History and physical examination findings combined with laboratory and imaging findings lead the clinician to understand the burden of the underlying systemic condition and its effect on the impending dental treatment. It is the responsibility of the dental practitioner to be aware of these systemic conditions, diagnose and appropriately refer to a specialist when needed, and more commonly, manage them as dental patients. Any pertinent data must be used in conjunction with the radiographic investigation or other imaging modality to arrive at a clinical diagnosis. The systemic conditions that are covered range from endocrine abnormalities and developmental conditions to malignancies that have skeletal manifestations. Some of the systemic conditions that may affect the dental treatment or outcomes are described elsewhere in this issue.

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## **SICKLE CELL ANEMIA**

### ***Introduction***

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Sickle cell disease (SCD) comes from a specific form of anemia that is the result of homozygosity for the mutation that causes sickling of hemoglobin (HbS). Sickle cell anemia also has been referred to as HbSS, SS disease, and hemoglobin S. In the heterozygous populations that have only one sickle gene and one normal adult hemoglobin gene, the condition is referred to as sickle cell trait or HBAS. Other forms of sickle disease includes sickle cell hemoglobin c (HbSC), sickle beta plus-thalassemia (HbS/ $\beta^+$ ), and sickle beta zero-thalassemia (HbS/ $\beta^0$ ) According to the Centers for Disease Control and Prevention<sup>1</sup> (CDC data) that looked into the data from 44 states representing 88% of population, the prevalence of SCD was approximately 73 per 1000 births among African American individuals and 7 per 1000 among Hispanic American individuals. The overall rate was approximately 15 per 1000 live births in the United States.

### ***Summary of Clinical Features***

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Anemia, infection, and vaso-occlusion are the most common reasons for the complications related to SCD, episodes of pain in the chest, back, abdomen, or extremities. Multiple areas are often involved simultaneously. The acute chest syndrome leading to respiratory insufficiency will have the cardinal features of fever, pleuritic chest pain, referred abdominal pain, cough, lung infiltrates, and hypoxia. Patients with SCD now survive into their fifth or sixth decades in the industrialized countries.<sup>2,3</sup>

### ***Radiographic Features***

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The vaso-occlusion leads to bone infarcts, osteomyelitis, orbital wall infarction, and subperiosteal hemorrhage. Chronic anemia leads to internal carotid artery stenosis and extramedullary hematopoiesis. The infections lead to osteomyelitis and regional lymphadenopathy. Bone involvement is the most common clinical feature of SCD, mainly the long bones and to a lesser extent the maxillofacial area, due to the small number of marrow spaces within these bones. In the head and neck, the most frequently reported location is the orbital wall, followed by the mandible and skull base. There are both acute and chronic phases of bone involvement; in the acute phase, vaso-occlusive crisis and osteomyelitis are notable, whereas in the chronic phase, bone marrow hyperplasia, osteoporosis, and iron deposition in the marrow due to repeated transfusions may be noted.<sup>4</sup> Avascular necrosis of the bone is a major problem in the long bones, especially the hip, although it is not a common occurrence within the jaws.

### ***Imaging Protocols***

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Bone infarction leading to change in the trabecular pattern may be a radiographic sign noted in patients with SCD. MRI is a much more sensitive imaging technique over computed tomography (CT) for these changes.

### ***Imaging Findings/Pathology***

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Radiographically, cortical defects, adjacent soft tissue fluid collection that communicates with the medullary compartment through cortical defects, and ill-defined bone marrow enhancement are indicative of osteomyelitis.<sup>5</sup> A panoramic radiograph (Fig. 1) of a 34-year-old woman with compensated SCD, who presented to the oral medicine admissions clinic with severe abdominal distension and vasculitis affecting the lower extremities showed inconsistent trabecular changes, enlarged bone marrow spaces, especially in the lower anterior region, and some cortical defects. Hypercementosis of some teeth was noted as an incidental finding.

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