

## **Cleidocranial dysplasia: a review of the dental, historical, and practical implications with an overview of the South African experience**

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### **Abstract**

Cleidocranial dysplasia (CCD) is an uncommon but well-known genetic skeletal condition. Several hundred affected persons are members of a large extended family in the Cape Town Mixed Ancestry community of South Africa. The clinical manifestations are often innocuous, but hyperdontia and other developmental abnormalities of the teeth are a major feature and may require special dental management.

Over the past 40 years, the authors have encountered more than 100 affected persons in Cape Town. Emphasis has been on dental management, but medical, genetic, and social problems have also been addressed. In this article, we have reviewed the manifestations of the disorder in the light of our own experience, and performed a literature search with emphasis on the various approaches to dental management and treatment options in CCD. Advances in the understanding of the biomolecular pathogenesis of CCD are outlined and the international and local history of the disorder is documented.

The purpose of this article was to review the dental manifestations and management of cleidocranial dysplasia (CCD) [OMIM 119600]. The history, genetic background, and general manifestations of CCD are also outlined and an overview is presented.

The disorder is a genetic skeletal dysplasia in which hypoplasia of the clavicles and deficient ossification of the anterior fontanelle are the major features. Affected persons have a characteristic facial appearance with a bulky forehead, hypertelorism, and midfacial hypoplasia.<sup>1</sup> General health is usually good and the intellect is unimpaired.<sup>2</sup> The adverse general health effects of CCD are usually not very severe or debilitating and there is no associated impairment in cognitive or intellectual functioning in affected persons.<sup>2</sup>

A variety of dental problems may occur in CCD. In particular, supernumerary teeth (hyperdontia) in the primary and secondary dentition may lead to dental crowding and malocclusion. Retention of the deciduous teeth may exacerbate this situation. For these reasons, dental management is a significant aspect of the health care of affected persons.



































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