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內文：

1. INTRODUCTION:
   - Cleidocranial dysplasia (CCD) is an uncommon but well-known genetic skeletal condition, hyperdontia and other developmental abnormalities of the teeth are a major feature and may require special dental management.
   - The purpose of this article was to review the dental manifestations and management of cleidocranial dysplasia(CCD)：The history, genetic background, and general manifestations of CCD are also outlined and an overview is presented.
   - Over the past 40 years, the authors have encountered more than 100 affected persons in Cape Town.
2. CLEIDOCRANIAL DYSPLASIA:
   - 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
   - Cleidocranial dysplasia is inherited as an autosomal dominant trait, with generation-to-generation transmission.
   - Owing to the founder effect, the condition is comparatively common in the mixed ancestry community of Cape Town, South Africa
   - Whereas the worldwide prevalence of CCD is generally regarded as being about 1 per million, in this Cape Town community, the minimum prevalence is 100 per million.
3. CLINICAL MANIFESTATIONS
   - General features
     i. clavicular hypoplasia

     ii. delayed fusion of cranial sutures
     iii. dental abnormalities, The number of teeth may be excessive (hyperdontia)
     iv. Patency of the anterior fontanelle can produce a bulky configuration or a depression in the mid-upper forehead

     v. Skeletal abnormalities, including slight stature, short terminal phalanges, spinal malalignment, genu valgus (knock knees), and pes planus (flat feet)
     vi. Recurrent infections of the upper respiratory tract owing to maldevelopment of the sinuses

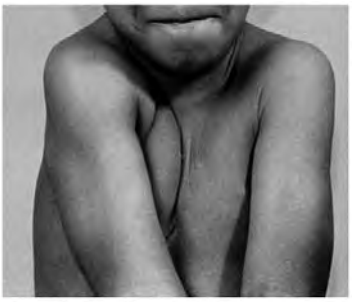
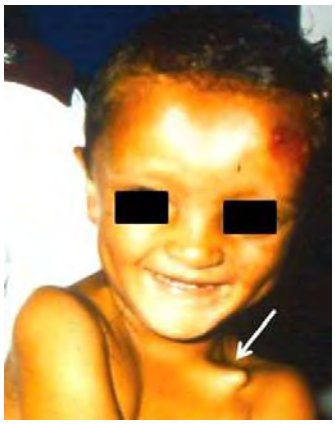


Fig. 3. Anterior apposition of shoulders in facilitated by bilateral clavicular hypoplasia.



vii. Other skeletal abnormalities include a wide pubic symphysis, dysplastic scapulae, coxa vara, and a variety of vertebral anomalies

- Dental features in CCD
  - i. leads to dental impaction, overcrowding, and malocclusion, while midfacial hypoplasia can exacerbate these problems
  - ii. Articulation and mastication may be compromised
  - iii. delayed eruption and retention of the primary and secondary dentition

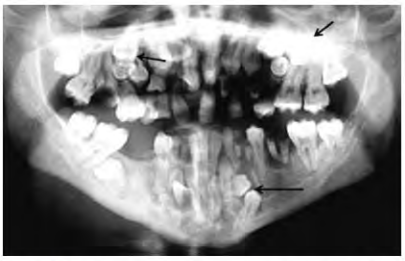


Fig. 6. Hyperdontia: pantamogram of an affected male showing multiple supernumerary teeth (*arrows*).

  - iv. The crowns of the teeth sometimes appear abnormal, the enamel may be hypoplastic, and dentigerous cysts and taurodontia
  - v. Rounded gonion angles, kyphotic sphenoid bones, and Wormian bones in the cranial sutures

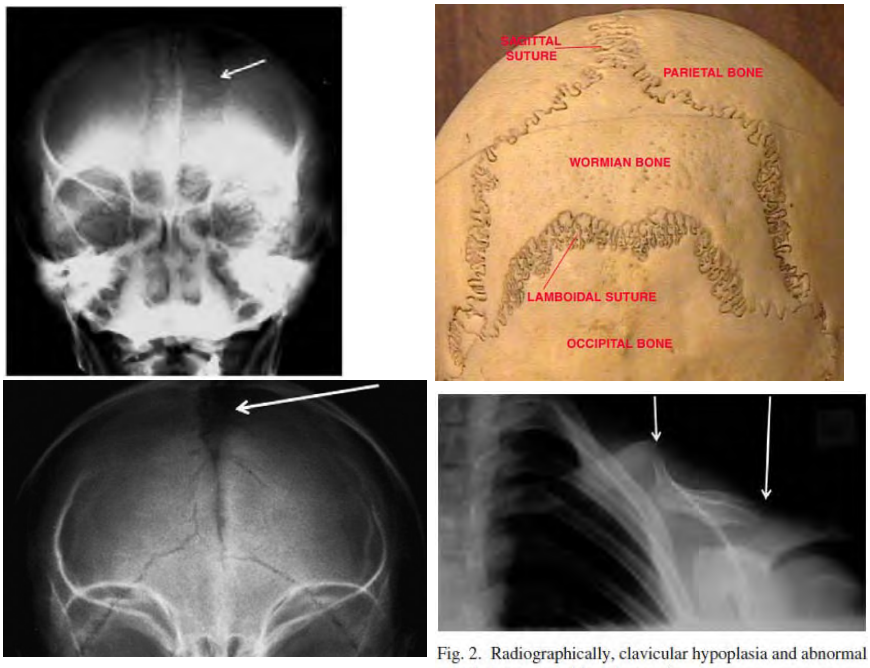


Fig. 2. Radiographically, clavicular hypoplasia and abnormal morphology are evident (*arrows*).

4. DIFFERENTIAL DIAGNOSIS OF CLEIDOCRANIAL DYSPLASIA AND HYPERDONTIA
   - The presence of clavicular hypoplasia is strongly suggestive of CCD
     - i. as an isolated nonsyndromicentity, which is usually unilateral
     - ii. Complete absence of both clavicles is a manifestation of the Yunis-Varon syndrome
       1. rare genetic disorder, intellectual dysfunction and anomalies of the hands and feet are associated with malformations in other systems
   - Defective cranial ossification leading to patency of the anterior fontanelle and Wormian bones in the sutures is an important feature of CCD
     - i. osteogenesis imperfecta (frequent fractures)
     - ii. pycnodysostosis (skeletal density)
     - iii. congenital hypothyroidism (disturbed thyroid metabolism)
   - Hyperdontia may also be a component of specific genetic syndromes, including the

i. Gardner syndrome (familial polyposis of the colon and osteomata),
ii. Hallerman-Streiff syndrome (narrow face, hypotrichosis, microphthalmia)
iii. orofaciodigital syndrome type I

5. DENTAL MANAGEMENT IN CCD:

Table I. Cleidocranial dysplasia: orodental anomalies and management options

Anomaly	*Management option*	*Rationale*	*Reference*
Retained deciduous teeth	Removal	Assist eruption of permanent teeth	49,50,51,64
Supernumerary teeth	Removal	Assist eruption of permanent teeth	49,50,51,56,64
Permanent teeth abnormalities	Removal	Construction of removable full/partial dentures (not indicated in childhood)	54,58,60,61,64,66
	Retention	Abutments for fixed appliances (not indicated in childhood)	63,65
Unerupted teeth	Surgical exposure	Support for overdenture	50,51,55
	Orthodontic eruption	Function and esthetics and alignment	47,48, 51,52
	Implants	Support overdenture	53,64
		Guide impacted teeth into occlusion	51,59,62
	Surgical translocation and/or autotransplantation	Function and esthetics	47,48,52,57,62
Malocclusion	Fixed or removable orthodontic appliances	Function and esthetics	47,49,58
Palatal vault narrow-high arched	Expansion with removable orthopedic appliance	Reduce crowding	64

- The aim of dental management in CCD is to achieve an optimal functional and cosmetic result by early adulthood. Surgical procedures are usually uneventful in CCD but atlanto-axial subluxation with consequent damage to the spinal cord has been documented. This causes potential hazard during anesthesia

Table II. Cleidocranial dysplasia: management approaches

Approach	*Procedure*	*Reference*
Toronto-Melbourne		69,70
Several procedures		
Age: 5-6	Anterior primary teeth are extracted	
Age: 6-7	Primary incisors are exposed and healing is allowed	
	Orthodontic brackets are placed on permanent incisors	
	Posterior primary teeth are extracted	
Age: 9-10	Permanent bicuspids are exposed	
	Surgical removal of supernumerary teeth and healing allowed	
Age: 9-12	Placement of orthodontic brackets on permanent canines and bicuspid teeth	
Jerusalem		47,48
Age: 10-12	Phase 1:	
	Anterior primary teeth are extracted	
	All supernumerary teeth are extracted	
	Permanent incisors are exposed	
	Orthodontic attachments are placed on permanent incisors	
	Surgical flaps are closed completely	
Age: 13 and older	Phase 2:	
	Posterior primary teeth are extracted	
	Unerupted permanent canines and premolars are exposed	
	Orthodontic attachments are bonded	
	Surgical flaps are closed completely	

Belfast-Hamburg Single procedure Age: not specified	All primary and supernumerary teeth are removed All impacted teeth are surgically exposed Surgical packs are placed to prevent healing of bone and soft tissue over teeth Healing by secondary intention Orthodontic attachments are placed Orthodontic appliances placed on fully erupted teeth Elastic thread is placed between brackets on unerupted teeth and the arch wires	73,74
Bronx Two at most 3 Procedures Age: not specified	Phase 1: All primary and supernumerary teeth are removed Surgical flaps are closed Phase 2: Unerupted permanent teeth are exposed Orthodontic brackets are placed Surgical flaps are closed and overdenture is placed Conventional orthodontic appliances are placed Phase 3: Leforte osteotomy-orthognathic surgery Dental implants are placed	74

- These procedures are all undertaken over a long period. It is relevant that patient compliance is essential to a favorable outcome for any of these modalities
- In South Africa, the dental and orthodontic approach to CCD has several constraints. Extensive medical expertise is available, but access is limited and costly

題號	題目
1	鎖骨顱骨發育不全可能影響? (A) 鎖骨和恥骨聯合 (B) 頭顱骨和顏面骨 (C) 脊椎骨和附肢骨 (D) 以上皆是
答案(D)	出處：oral and maxillofacial pathology
題號	題目
2	下列關於牙齒發育異常的敘述，何者錯誤？ (A) Fusion 之牙常有個別的牙髓腔 (B) Germination 之牙有共同的牙髓腔 (C) Cleidocranial dysplasia 常先天多生牙 (D) Macrognathia 可見於軟骨發育不全(anchondroplasia)或鎖骨顱骨(Cleidocranial)發育不全
答案(D)	出處：oral and maxillofacial pathology 101 年第二次專門職業及技術人員高等考試