Cone-beam computed tomography: An inevitable investigation in cleidocranial dysplasia

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Abstract
Cleidocranial dysplasia is a heritable skeletal dysplasia and one of the most common features of this syndrome is multiple impacted supernumerary teeth. Cone-beam computed tomography, the most recent advancement in maxillofacial imaging, provides the clinician to view the morphology of the skull and the dentition in all three dimensions and help in treatment planning for the patient.

Keywords: Cleidocranial dysplasia, cone-beam computed tomography, impacted teeth

Introduction
Cleidocranial dysplasia (CCD) is a rare hereditary congenital defect which has an autosomal dominant mode of inheritance. The characteristic features of this syndrome include hypoplastic/aplastic clavicles, delayed closure of cranial sutures, and multiple impacted supernumerary teeth.[1,2]

Conventional radiography has been employed until recently to study the morphology of skull and dentition for patients suffering from this syndrome. Panoramic radiology can provide an overview of over-retained deciduous teeth, multiple impacted permanent and supernumerary teeth but it fails to provide an accurate assessment of the proximity of the impacted teeth with vital structures like inferior alveolar nerve canal and floor of maxillary sinus. To overcome these shortcomings, cone-beam computed tomography (CBCT) can be applied to study the location, alignment, proximity of teeth to vital structures as well as the quality and quantity of basal bone available which could help a great deal in orthodontic planning and surgical extraction.

The objective behind performing a CBCT examination for all the patients included in this case series was not only for the diagnosis and study of the peculiar features of this syndrome, but to highlight its importance in treatment planning.

In the present case series, 3 cases of CCD were subjected to orthopantomographs (OPG) examination. On the basis of OPG findings, a three-dimensional (3D) CBCT scan was obtained on Planmeca machine with field of view 16 cm × 16 cm in order to exactly recognize teeth and anatomical anomalies and to plan which teeth are to be extracted. The scans were scrutinized using Planmeca Promax 3D Mid machine, Helsinki, Finland (Planmeca Romexis software) and could be studied in the axial, coronal, and sagittal planes with 0.4 mm sections which helped in imaging the impacted teeth and analyze their orientation to the surrounding teeth and adjacent vital structures

Case Reports
Case 1
An 18-year-old female reported to our institute with a chief complaint of multiple missing teeth since childhood and failure of eruption of permanent teeth. The patient gave no history of any trauma or extractions. On clinical examination, the patient showed features like small built with normal intelligence, brachycephalic skull, frontal bossing, maxillary hypoplasia resulting in a prognathic appearance, ocular hypertelorism, and depressed nasal bridge. She had abnormal mobility of her shoulders toward the midline. On intra oral examination, patient had a narrow high arched palate, multiple missing permanent teeth and over-retained deciduous teeth.

Orthopantomographs were taken which showed multiple impacted teeth and diagnosis of CCD was made.

Cone-beam computed tomography examination was performed and its features were compared with OPG to study findings not clearly identified on OPG.

- At some areas, the position and location of impacted teeth which were difficult to interpret on OPG
could be easily identified through CBCT examination [Figure 1a and b]. The finding of the position and location of the impacted teeth is vital for oral surgeons for any kind of surgical treatment required which makes CBCT examination an important step in such patients.

- Since there are multiple impacted teeth, their relation with the inferior alveolar canal needs to be accurately analyzed before commencing any surgical treatment. OPG being 2D has this disadvantage compared to CBCT that proper nerve positioning and its relation with the tooth cannot be judged perfectly [Figure 2a and b], which makes CBCT examination mandatory.

- In the same patient, OPG showed a radiopaque structure distal to 48 and it was difficult to make out whether it was a developing tooth or an odontome. When CBCT examination was done, the doubt was cleared as root canal within the radiopaque structure was easily visible confirming it as a developing tooth [Figure 3a and b].

Case 2
A 17-year-old male reported with the chief complaint of multiple missing teeth since childhood and failure of eruption of several permanent teeth. The patient had a younger brother who showed similar features. On clinical examination, features similar to the above case, conducive of a diagnosis of CCD were seen which was confirmed on OPG showing multiple impacted teeth. CBCT examination was performed and its features were compared with OPG similar to case 1.

- In cases of CCD, due to multiple impacted teeth and some teeth being outside the focal trough, their exact size and morphology are distorted due to image magnification. This disadvantage can be overcome with CBCT examination [Figure 4a and b]

- It is important to determine the proximity of tooth to vital structures like floor of maxillary sinus prior to any surgical procedure. CBCT can overcome the disadvantage of 2D imaging where in the exact proximity cannot be determined easily [Figure 4a and b].

Case 3
A 16-year-old male reported with complaint of multiple missing teeth similar to above cases. His shoulders were approximating to midline and OPG showed multiple missing teeth. Other features of CCD were also seen so again CBCT examination was performed and features compared with OPG.

- Due to the ability to view the CBCT scans in sections as thin as 0.4 mm, small structures such as over-retained root pieces of deciduous teeth which are missed on OPG can be easily seen on CBCT [Figure 5a and b]

- The exact morphology of deciduous teeth being resorbed due to eruption of permanent teeth can be studied well on CBCT [Figure 6a-c].

Discussion
Cleidocranial dysplasia also known as cleidocranial dysostosis is a genetic condition affecting bone growth. It is an autosomal dominant disorder showing a wide variety of expression.\(^1\)\(^2\) The primary defect lies in the mutation of the gene coding for osteoblast transcription factor Runx2/Core binding factor alpha mapped to short arm of chromosome 6p21. This factor is required for membranous ossification (cranium, clavicles), endochondral ossification (long bones), and skeletal morphogenesis.\(^3\)

Figure 1: (a) In cropped orthopantomographs, the actual position of impacted teeth cannot be determined (b) Sagittal section of cone beam computed tomography clearly demonstrates the position of impacted teeth and its relation to adjacent teeth

Figure 2: (a) In orthopantomographs, the impacted tooth appears closely related to the inferior alveolar nerve canal (b) In coronal section of cone-beam computed tomography, the exact relation of the impacted tooth with the inferior alveolar nerve can be determined
It was first reported in 1760 and until now, more than 1000 cases of CCD have been reported in medical literature. They have a characteristic facial appearance having a short head (brachycephaly) with prominent forehead (frontal bossing). Delayed closure of fontanels is generally seen with some patients showing open fontanels. The eyes may be widely spaced (hypertelorism). The shoulders are narrow, down sloping, and may show abnormal hypermobility. The clavicles may be hypoplastic or completely absent on one or both sides, mostly hypoplasia affects the acromial end. In rare occasions, patients may show unusual positioning of head joints, abnormalities of spine and abnormal formation of bones of fingers and hands, scoliosis, extra ribs, cleft palate, hearing loss, and respiratory problems.

The dentition of these patients is highly affected showing multiple supernumerary and impacted teeth. The teeth may be abnormally formed and positioned. Delayed eruption of permanent teeth is common.

Conventional diagnostic imaging like OPG and cephalometric tracings have been traditionally been studied.

Orthopantomographs show multiple impacted and supernumerary teeth. In PA Caldwell view, defective fusion of frontal and parietal bones leading to open coronal and sagittal sutures can be seen. Lateral skull view shows wormian bones in the occipital region. Lateral cephalogram may show maxillary hypoplasia and a concave facial profile. A PA chest view may show hypoplastic bilateral clavicles and a bell shaped rib cage.
The diagnosis is mainly based upon clinical and radiographic characteristics. Serum alkaline phosphatase activity has been observed to be consistently reduced in patients suffering from CCD.\textsuperscript{[9]}

The limitations in analysis of 2D imaging modalities are magnification, geometric distortion, and superimposition of structures.\textsuperscript{[6]-[7]}

In cases of CCD, multiple impacted teeth cause numerous superimpositions. Moreover, due to many teeth being outside the focal trough causing magnification and geometric distortion interpreting the positions of individual teeth becomes very difficult. Hence, a 3D imaging is important for treatment planning in cases of CCDs.

In 2007, Korbmacher et al.\textsuperscript{[8]} reported the importance of CBCT from orthodontic viewpoint stating that CBCT provides more information than the conventional radiographs for localizing impacted and retained teeth, root resorption, cleft lip and palate, and third molar evaluations. We found similar findings in the cases we studied.

Cone-beam computed tomography scans providing precise location of the tooth help the clinician to decide which tooth to save and which to salvage, the optimal surgical approach to minimize damage to the real tooth.\textsuperscript{[9]}

In 2000, Ericson and Kurol\textsuperscript{[10]} documented that presence and the extension of root resorption of the teeth adjacent to impacted teeth is difficult to interpret on traditional radiology. However, CBCT can provide enhanced visualization of roots from all directions making it easier to confirm presence or absence of root resorption.\textsuperscript{[9]} Even root morphology such as dilacerations, number of canals, and canal morphology can be assessed which may help in endodontic treatment planning.

In cases of CCD, extraction of multiple teeth results in edentulous areas, so CBCT will further act as a guide for implant planning and rehabilitation of the patient.

Cone-beam computed tomography provides accurate assessment of alveolar bone height and helps in analysis of the size, shape, and volumetric differences in bilateral structures as well as growth changes in 3D.\textsuperscript{[11]}

A recent study by Haney et al. in 2010 states that diagnostic information from CBCT scans resulted in substantially different perception of tooth localization and root damage when compared to routine radiographs. Furthermore, it resulted in significantly higher confidence in diagnosis and treatment planning by orthodontists.\textsuperscript{[12]}

The various treatment procedures may include prosthetic replacements, removal of the supernumerary teeth followed by surgical repositioning of the permanent teeth, and a combination of surgical and orthodontic measures for actively erupting and aligning the impacted permanent teeth. Speech therapy may be required during periods of dental treatment. Aggressive treatment of sinus and middle ear infections; tympanostomy tubes are considered for recurrent middle ear infections. If the cranial vault defect is significant, the head needs protection from blunt trauma; helmets may be used for high-risk activities. If bone density is below normal, treatment with calcium and Vitamin D supplementation is considered. Preventive treatment for osteoporosis should be initiated at a young age.\textsuperscript{[9]}

**Conclusion**

We present this case series to conclude and support the use of CBCT technology in CCD patients as an imminent guide for orthodontic and surgical treatment planning. CBCT examination offered the following advantages over conventional radiographic examination [Table 1].

| Table 1: Advantages of CBCT over conventional radiography in imaging impacted teeth |
| --- |
| Number and position of impacted permanent and supernumerary teeth |
| Exact number and position of the impacted teeth and supernumerary teeth could be calculated |
| Crown morphology |
| Study of the crown anatomy and designation of the impacted teeth as permanent or supernumerary and subsequent extraction of the supernumerary teeth |
| Root formation |
| On the basis of completion of root formation, decisions regarding orthodontic extrusion could be made |
| Root morphology |
| The root orientation of the impacted teeth could be traced. Any root morphological changes like dilaceration, supernumerary roots which could affect extraction or extrusion of teeth could be imaged |
| Root canal morphology |
| Presence of additional root canals, accessory root canals could be traced for the teeth which could be saved endodontically |
| Status of alveolar basal bone |
| Proximity to vital structures |
| Relationship to vital structures like the inferior alveolar nerve canal, mental foramen, floor of maxillary sinus, nasal floor could be traced |

Determine any minute periapical radiolucency

Which could be overlooked in conventional radiography due to severe superimposition. This is important as the impacted teeth could be accompanied by follicular and eruptive pseudocysts and thus CBCT examination can aid in the early recognition and treatment of such lesions
Acknowledgment

We would like to acknowledge Dr. H. R. Umarji, Professor and HOD, Department of Oral Medicine and Radiology, Government Dental College and Hospital, Mumbai.

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How to cite this article: Gupta NS, Gogri AA, Kajale MM, Kadam SG. Cone-beam computed tomography: An inevitable investigation in cleidocranial dysplasia. Contemp Clin Dent 2015;6:257-61.

Source of Support: Nil. Conflict of Interest: None declared.