

## Awareness Of Crouzon Syndrome Among Dental Students

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### Abstract

**INTRODUCTION:** Crouzon syndrome is a rare genetic disorder that may be evident at birth or during infancy. The disorder is characterized by distinctive malformations of the skull and facial (craniofacial) region . It is a form of craniosynostosis, a condition in which there is premature fusion of the fibrous joints (sutures) between certain bones of the skull.

**AIM:** To assess the awareness of crouzon syndrome among dental students.

**MATERIALS AND METHODS:** A questionnaire based survey was conducted among dental students to assess the level of knowledge and awareness about crouzon syndrome. The sample size of this study is 100. The questionnaire consisted of 15 questions. Online study setting was used to collect data . Results were analysed using SPSS software.

**RESULTS:** From this study, we come to know that only 24% of the participants are familiar or heard the term crouzon syndrome The participants were asked about the abnormalities caused by crouzon syndrome for which 26% said “ premature closure of fibrous joints between certain bones in the skull”, 14% said “ affected intelligence “, and 58% said they are not sure. Also the participants were asked about the symptoms associated with oral cavity, 22% said underdeveloped jaw, 22% said protruded lower jaw and overcrowded teeth and 52% said all the mentioned symptoms. Participants were asked about the treatment for crouzon syndrome for which 50% said surgery and 50% said that they may not need to be treated.

**CONCLUSION :** From this study, we come to know that only a low population of the dental students involved in the study are aware about crouzon syndrome and majority of them are not aware. Hence more awareness is required as dental clinicians as they may encounter such cases and should be able to diagnose it.

**KEYWORDS:** Crouzon syndrome, awareness, dental students, survey, diagnosis.

## INTRODUCTION:

Crouzon syndrome is a rare genetic disorder that may be evident at birth or during infancy. The disorder is characterized by distinctive malformations of the skull and facial (craniofacial) region (1). It is a form of craniosynostosis, a condition in which there is premature fusion of the fibrous joints (sutures) between certain bones of the skull. The sutures allow an infant's head to grow and expand. Eventually, these bones fuse together to form the skull. In Crouzon syndrome, the sutures fuse prematurely affecting the proper growth of the skull and head and potentially altering the shape and development of the skull. Certain bones in the face may be affected as well. Such abnormalities may vary greatly in range and severity from case to case, including variations among affected family members.

Crouzon syndrome is caused by mutations in the fibroblast growth factor receptor-2 (FGFR2) gene, which is mapped to chromosome locus 10q25-10q26.8. The current research indicates fibroblast growth factor receptors (FGFR) FGFR2 and FGFR3 (2) as the leading factors in causing the autosomal dominant Crouzon syndrome. These two transmembrane proteins are two of four fibroblast growth factor receptors involved in osteoblast differentiation during embryonic development; mutations amongst these receptors are involved in several genetic disorders (3). Fifty percent of incidents of Crouzon syndrome are not inherited and are the result of new mutations (4). The aim of the study is to assess the awareness of crouzon syndrome among dental students.

**MATERIALS AND METHODS:** A questionnaire based survey was conducted among dental students to assess the level of knowledge and awareness about crouzon syndrome. The sample size of this study is 100. The questionnaire consisted of 15 questions.

Online study setting was used to collect data . Results were analysed using SPSS software.

**RESULTS AND DISCUSSION:** In this study , we assessed the level of awareness about crouzon syndrome among dental students. In this study, 100 participants are involved out of which 90% of them are undergraduates and 10% of them are postgraduates. From this study, we come to know that only 24% of the participants are familiar or heard the term crouzon syndrome and the remaining 76% are not familiar with that term (fig.1). On assessing what crouzon syndrome means, 40% said that it is a rare genetic disorder, 14% said it is a rare acquired disorder and 46% have no idea about it (fig.2). Only 20% of the participants are aware of the effects caused by crouzon syndrome and the remaining 80% are not aware.

On assessing the awareness of symptoms of crouzon syndrome, 20% said head appear usually short and broad or may appear long and narrow or triangular, 16% said prominent forehead and some of them said curved nose and a short upper lip and unusual shallowness of orbit and 56% of them said all the symptoms mentioned above are seen in crouzon syndrome (fig.3). The participants were asked about the abnormalities caused by Crouzon syndrome for which 26% said “ premature closure of fibrous joints between certain bones in the skull”, 14% said “ affected intelligence “, and 58% said they are not sure (fig.4). The complications of crouzon syndrome included Hearing loss, vision loss, inflammation in eyes and sleep apnoea and most of them said all the mentioned complications are involved in crouzon syndrome (fig.5). 54% of them said that the life expectancy of someone with crouzon syndrome will not be affected and 46% of them said that the life expectancy is shortened. Out of 100 participants m 52% said that the intelligence

is affected due to this syndrome and the remaining said the intelligence is not affected.

Also the participants were asked about the symptoms associated with oral cavity, 22% said underdeveloped jaw, 22% said protruded lower jaw and overcrowded teeth and 52% said all the mentioned symptoms (fig.6). The probability of affecting population in crouzon syndrome is assessed for which 66% said it affects both males and females equally, 24% said it affects mostly males and 10% said it affects mostly females. Of 100 participants 32% said the crouzon syndrome is diagnosed at birth or infancy, 38% said at the age of 5 years and 30% said above the age of 5 years. The diagnostic method for frozen syndrome is “CT” said by 20%, “MRI” said by 24% and “both” said by 56%. Participants were asked about the treatment for crouzon syndrome for which 50% said surgery and 50 said that they may not need to be treated. From this study we come to know that most of the dental students are not aware of crouzon syndrome and the effects of such syndrome.

Craniofacial abnormalities are often present at birth and may progress with time. Family history may reveal mildly affected individuals. Decreased mental function is present in approximately 12% of the patients. Craniosynostosis commonly begins during the first year and usually completes by the second or third year. Coronal and sagittal sutures are most commonly involved, resulting in a high prominent forehead. Ridging of the skull is usually palpable.

The most common ocular abnormalities reported are shallow orbits, ocular proptosis, orbital

hypertelorism, strabismus, optic atrophy, exposure keratitis, and an unexplained loss of visual acuity (5). Crouzon syndrome with a reported incidence of 1:25000 live births is the most common of over 70 conditions in which premature fusion of the cranial sutures may be a feature (6). A positive family history is reported to occur between 44-67% of cases (7)

A defining characteristic of Crouzon syndrome is craniosynostosis, which results in an abnormal head shape. This is present in combinations of: turricephaly, frontal bossing, trigonocephaly (fusion of the metopic suture), brachycephaly (fusion of the coronal suture), dolichocephaly (fusion of the sagittal suture), plagiocephaly (unilateral premature closure of lambdoid and coronal sutures), oxycephaly (fusion of coronal and lambdoidal sutures), and complex craniosynostosis (premature closure of some or all sutures) (8). Diagnosis of Crouzon syndrome usually can occur at birth by assessing the physical appearance of the infant. Further analysis, including radiographs, magnetic resonance imaging (MRI) scans, genetic testing, X-rays and CT scans can be used to confirm the diagnosis of Crouzon syndrome (9) Treatment by a multidisciplinary team working together with the family provides the best results with any craniofacial disorder. The goal is to stage reconstruction to coincide with facial growth patterns, visceral function, and psychosocial development. Multiple staged surgeries (10) are the general treatment plan for patients with Crouzon syndrome (11). Our team has extensive knowledge and research experience that has translate into high quality publications(12–20),(21–26),(27–33)

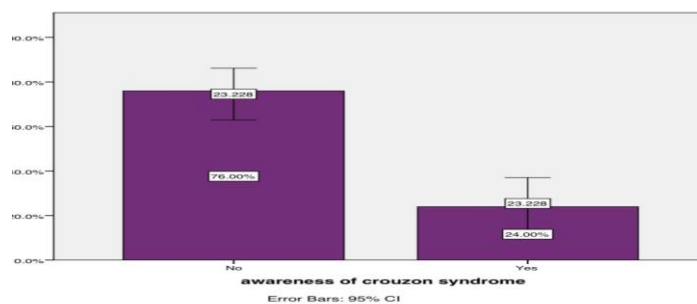


Fig.1 Bar chart representing the percentage of awareness among dental students in which 76% of them are not aware.

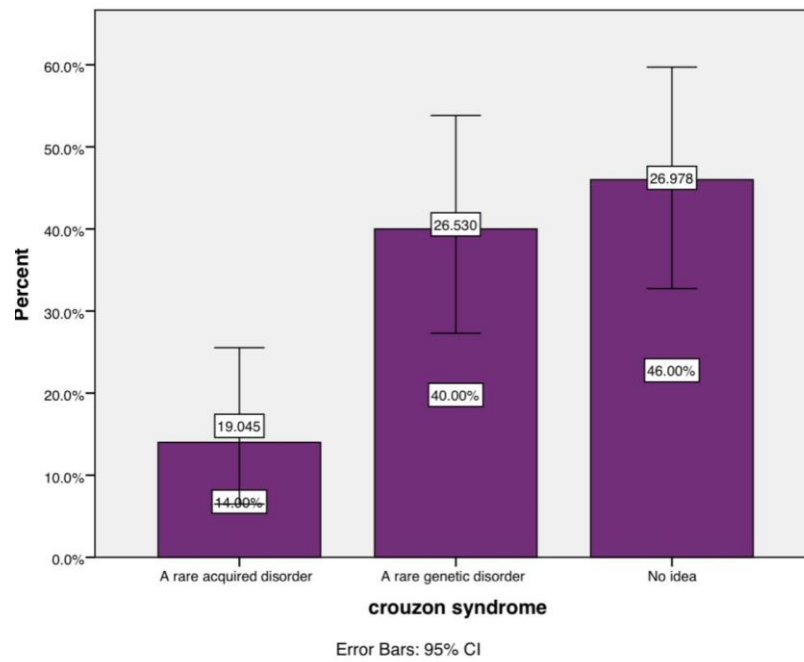


Fig.2 Bar chart representing the percentage of answers given by dental students for “what crouzon syndrome means”. Only 40% of the students are aware that crouzon syndrome is a rare genetic disorder.

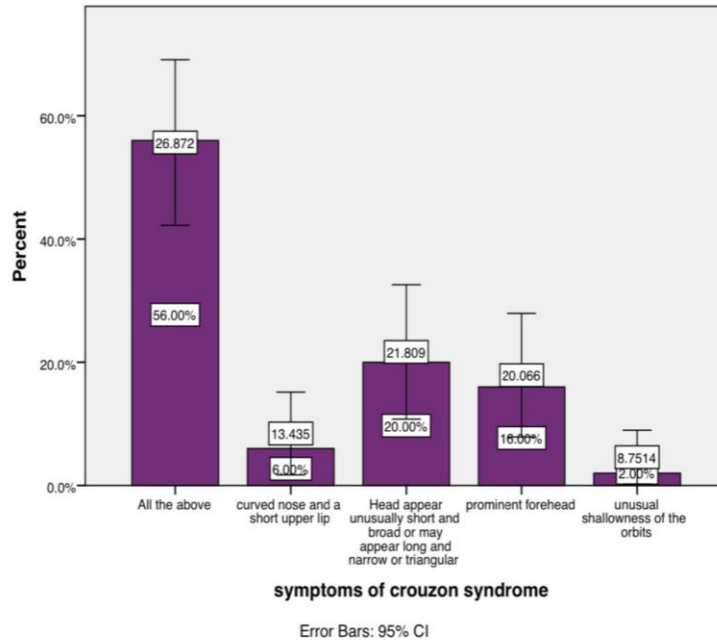


Fig.3 Bar chart representing the percentage of choices given by dental students for the symptoms of crouzon syndrome. Most of them said all the above mentioned symptoms are seen in crouzon syndrome.

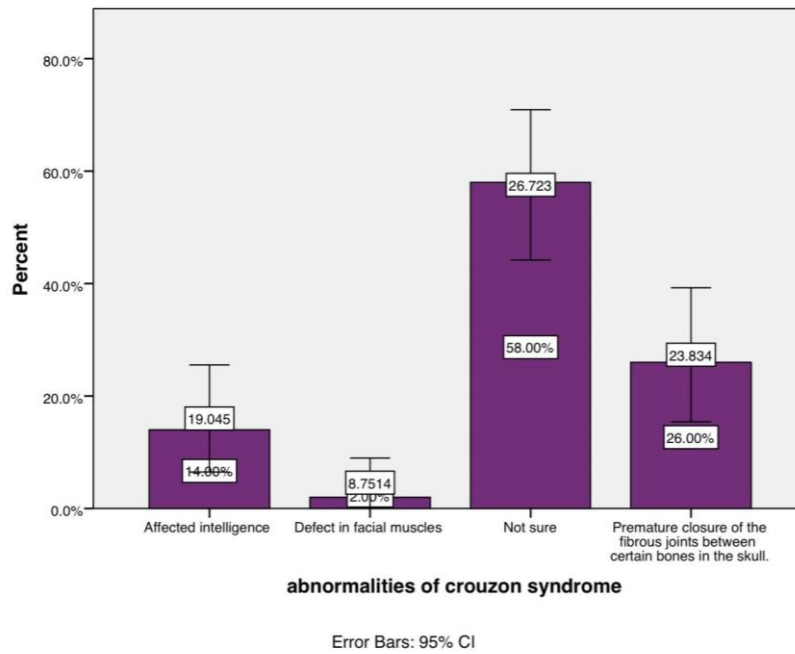


Fig.4 Bar chart representing the percentage of choices given by dental students for the abnormalities caused by Crouzon syndrome. Most of them are not sure about the abnormalities.

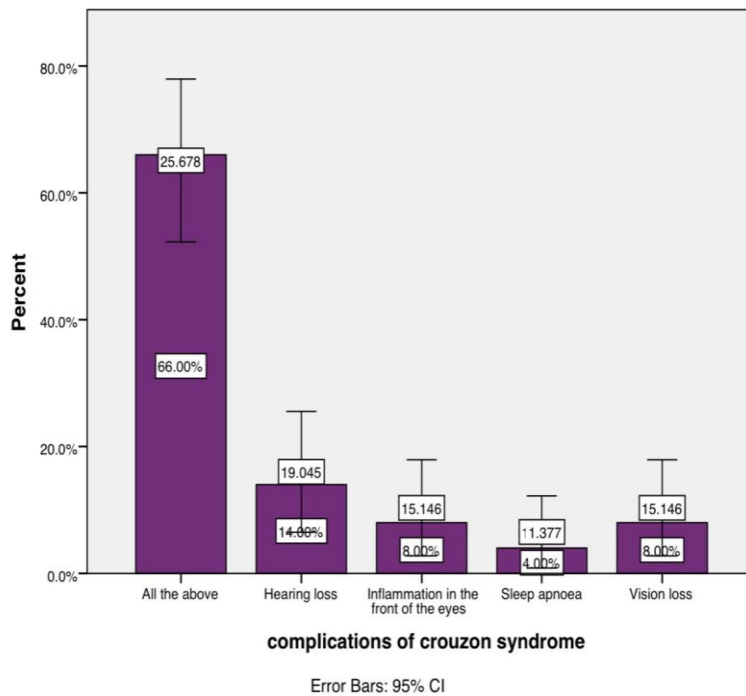


Fig.5 Bar chart representing the percentage of choices for the complications related to Crouzon syndrome. Most of them said all the mentioned complications are seen in Crouzon syndrome.

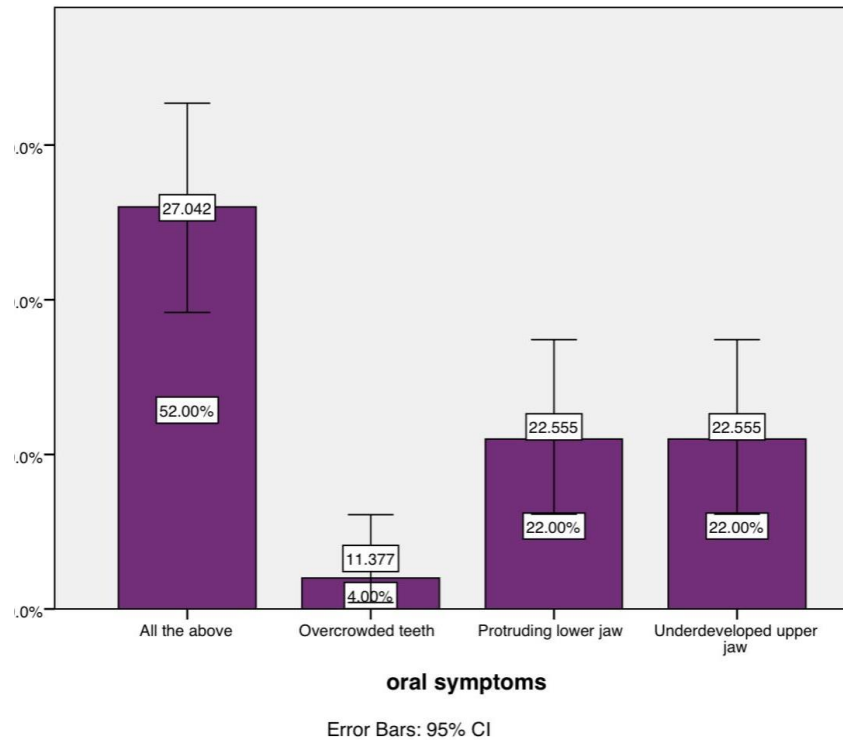


Fig. 6 Bar chart representing the percentage of choices for the symptoms associated with the oral cavity.

**CONCLUSION:** From this study, it is evident that only a very low population of dental students were aware of crouzon syndrome and their effects, symptoms, complications etc.. and most of them are not aware of the syndrome. Much awareness is needed in order to identify or diagnose the syndrome as dental clinicians.

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**CONFLICT OF INTEREST:** The authors have none to declare.

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