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Awareness Of Faces Syndrome Among Dental Students

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Abstract

Introduction
FACES Syndrome, also known as Friedman-Godman syndrome, is a condition which is characterized by unique facial features, anorexia, cachexia, eye and skin lesions. It is a rare, genetic, multiple congenital anomalies/dysmorphic syndrome characterized by facial dysmorphism, severe muscle wasting and cachexia, retinitis pigmentosa, numerous lentigines as well as mild, soft tissue syndactyly.
Aim: The aim of the study was to assess the awareness about FACES syndrome among dental students.
Materials and methods
A cross sectional study involving students of Saveetha Dental College, Chennai, India were taken. A self-structure containing 11 questions was framed and circulated among dental students of 100 people. The questions enquires about the awareness of FACES syndrome among dental students. Google forms were used to circulate the questions and the responses were collected, the data analysis was carried out using SPSS software. Chi square test was used for statistical analysis and p value more than 0.05 was considered as significant.
Results
Out of the participants, 42% were females and the remaining 58% were males. Most of the participants were unaware about the FACES Syndrome. Some questions were wrongly answered and the students opted for ‘not sure’ and ‘none of the above’ as well.
Conclusion
Thus from the study it is clear that most dental students are not aware of FACES syndrome and about the treatment modalities available since it is rare in occurrence.
Keywords
FACES syndrome, facial dysmorphism, cachexia, retinitis pigmentosa, lentigines, syndactyly.

INTRODUCTION

Syndromes are characterised as a group of symptoms which consistently occur together, or a condition characterized by a set of associated symptoms and it's conditions (1). With the advancement of the pharmaceutical industries the side effects of a drug might also be an etiology for a syndrome. Various syndromes that exist around the globe are life threatening. For obtaining a correct pathway for diagnosis is extremely difficult as well as a lengthy process, as physicians or caregivers often lack the appropriate expertise skills in these rarely encountered syndromes (2)

FACES Syndrome also known as Friedman-Goodman syndrome in the society, is an extremely rare condition which is characterized by unique Facial features, Anorexia, Cachexia (body wasting) and Eye and Skin lesions that occur to the patients. With the advancement in the biotechnological field, it has been diagnosed that the underlying prevalence of the syndrome is due to the pattern of genetic inheritance. Other various etiologies for the syndrome are still not established completely(3). FACES syndrome tends to affect various parts of the body. There are particular Facial characteristics in case of this peculiar syndrome. It may include deep-set eyes adding on with mild eyelid drooping known as ptosis, fat vesicles deposition around the eye known as xanthelasma, and a nose with a central groove at the tip(4).

Individuals suffering from FACES syndrome are extremely thin in appearance. This variant feature is termed as cachexia which includes severe wasting of the muscles of limbs in particular. In addition to the clinical features, their skin may be affected with freckle-like spots termed as lentigines along with light brown cafe- au-lait spots which occur in different parts of the patient. 80% of the Individuals suffering with FACES syndrome can have bowed legs known as genu varum, with 20% of occurrences of flat feet termed as pes planus. On the other hand, people might also suffer from mild webbing of fingers and toes known as syndactyly. Other clinically reported features include nasal speech, vision loss due to severe retinitis pigmentosa, thyroid problems, and a sunken chest bone also known as pectus excavatum(5).

The prevalence of this syndrome is extremely rare and estimated to occur in less than 1 in 1 million people. The heterogenous pathophysiology with an addition of dispersed nature of these rare syndrome means research and development efforts along with the patients populations are scattered around the globe. A scarcity of expertise for this Friedman-Godman syndrome poses a huge challenge to patients who seek access to diagnostic testing as well as appropriate treatment(6).

The aim of the study is to assess the awareness of FACES syndrome among the dental students.

MATERIALS AND METHODS

A cross sectional study involving students of Saveetha dental college ,Chennai ,India were taken.Ethical approval was obtained from the international review board prior to the start of the study.This study excludes the age criteria and the year of study of the students.A questionnaire was set up and circulated among dental students of 112 people. The sampling method used in this study was non- probability convenient random survey sampling. To minimize the bias certain measures were taken that include, to avoid leading questions, use of simple language to frame the questions and avoidance of difficult concepts among common people. A self-structured questionnaire containing 15 questions was framed which was checked for validity by three internal experts (from Saveetha Dental College) and also by three external experts (outside Saveetha Dental College). The questions enquired about the awareness of flat face microstomia among dental students. Google forms were used to circulate the questions and the responses were collected, the data analysis was carried out using SPSS software. Chi square test was used for statistical analysis and p value < than 0.05 was considered as significant.

RESULTS

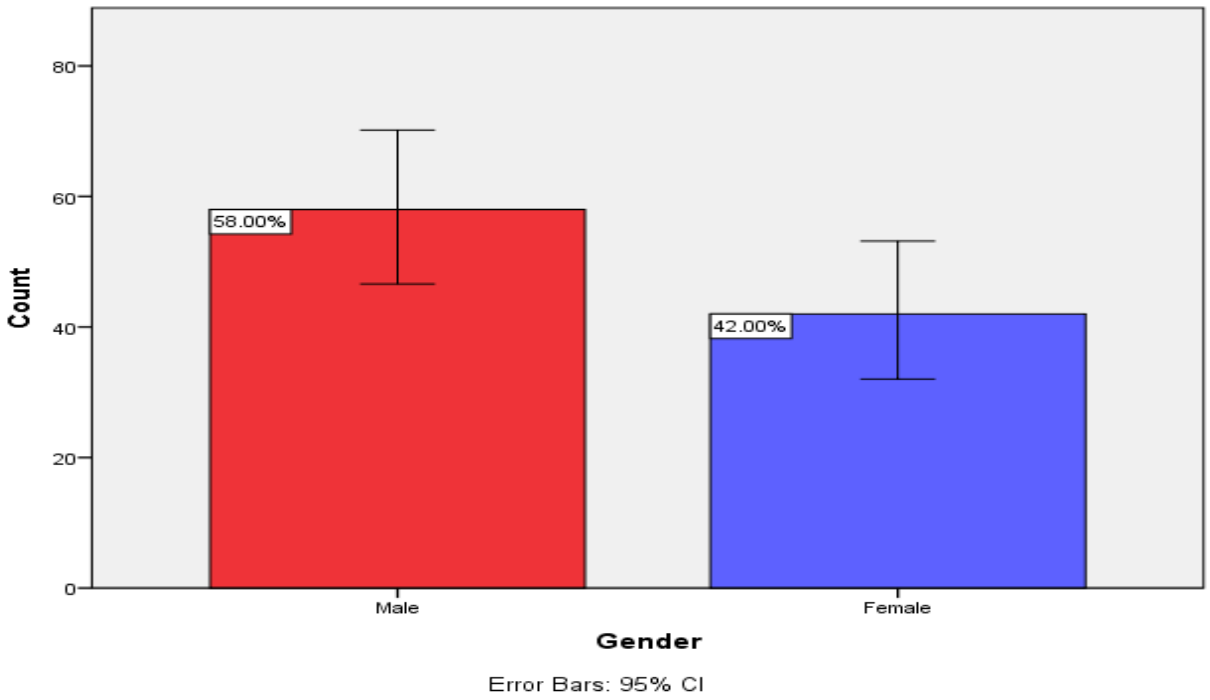

Figure 1 represents the bar chart of the participants who were involved in the survey. Blue colour denotes female and red colour represents male. 42% were female and 58% were male.

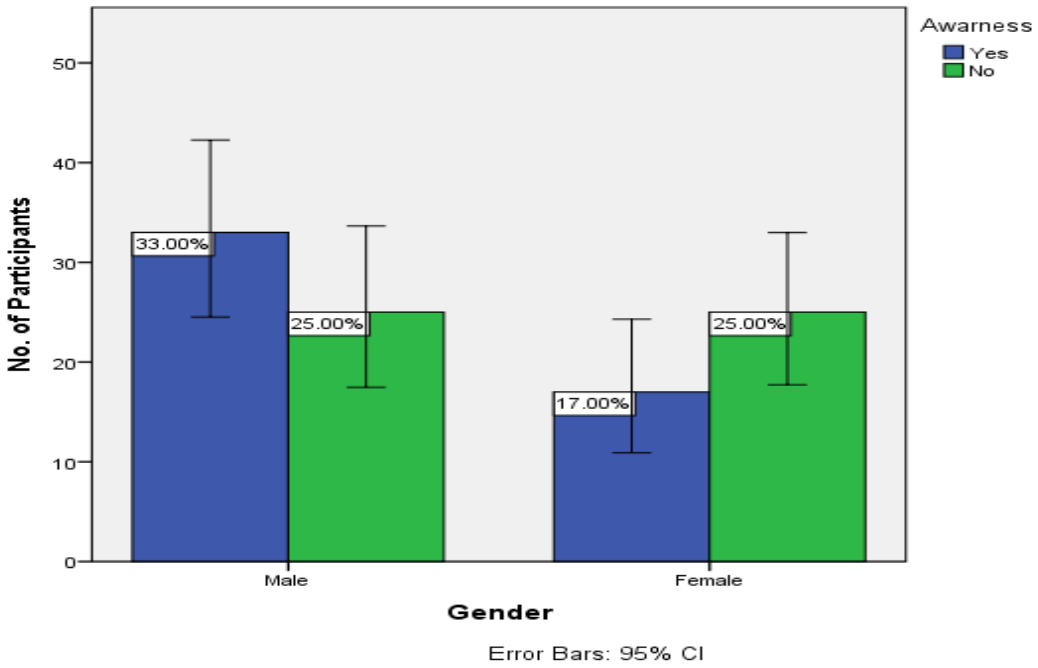

Figure 2. depicts the correlation graph between gender of the participants and whether they are aware of FACES syndrome. Blue colour denotes yes, green colour denotes no. Among females, 25% answered as no, 17% answered yes. Whereas among the males, 25% responded as no,

33% of males answered yes. Chi square test was evaluated for this graph with a p value of p=0.105 (p>0.05). Hence the value is statistically insignificant.

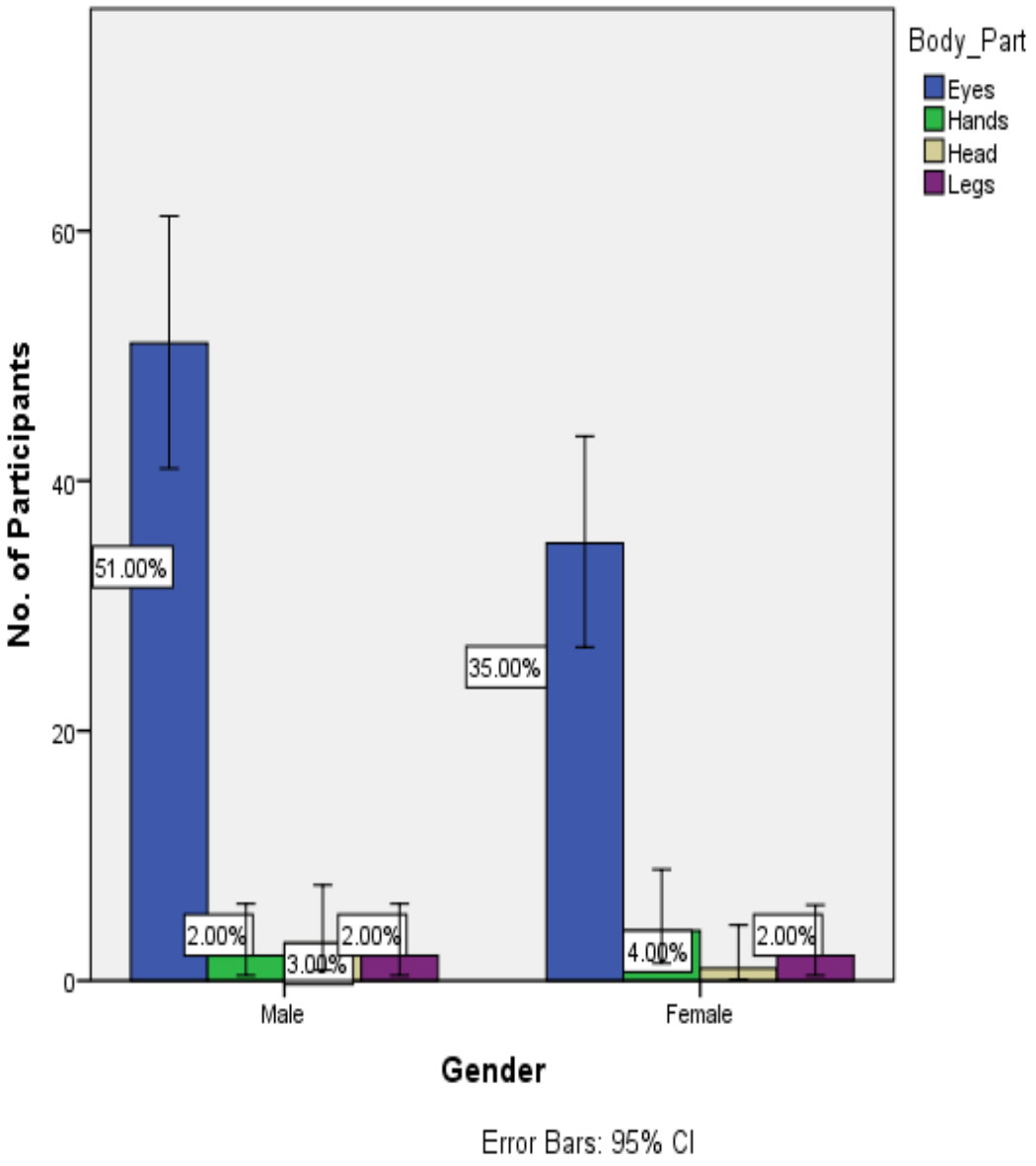

Figure 3. depicts the correlation graph between gender of the participants and their body parts which is being affected in case of FACES syndrome. Blue colour denotes eyes, green colour denotes hands, yellow colour denotes heads and purple colour denotes legs. Among females, 35% answered eyes, 1% answered hands and head respectively, rest of the 2% answered legs. Whereas among the males, 51% responded with eyes, and the rest of the 2% responded as hands, head and legs respectively. Chi square test was evaluated for this graph with a p value of p=0.544 (p>0.05). Hence the value is statistically insignificant.

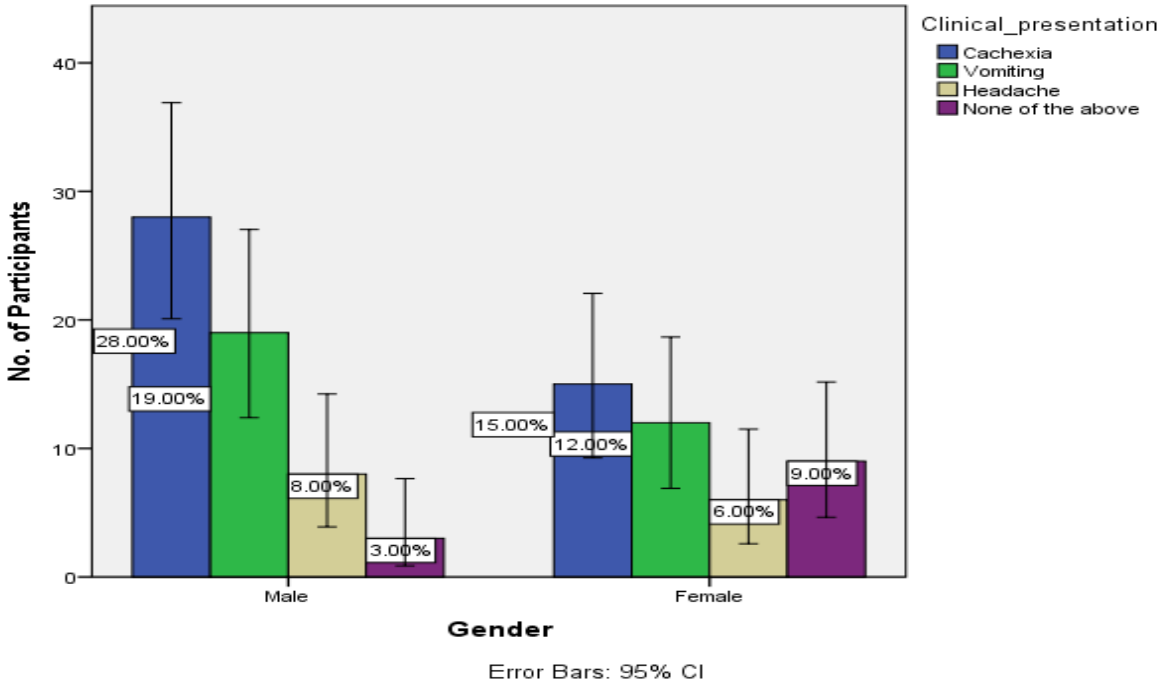

Figure 4. depicts the correlation graph between gender of the participants and the clinical presentation that occurs at the time of FACES syndrome. Blue colour denotes cachexia, green colour denotes vomiting, yellow colour denotes headache and purple colour denotes none of the above. Among females, 15% answered cachexia, 12% answered headache Whereas among the males, 28.28% responded cachexia, 11.11% of males answered as vomiting and 8.08% as headache. Chi square test was evaluated for this graph with a p value of p=0.123 (p>0.05). Hence the value is statistically insignificant.

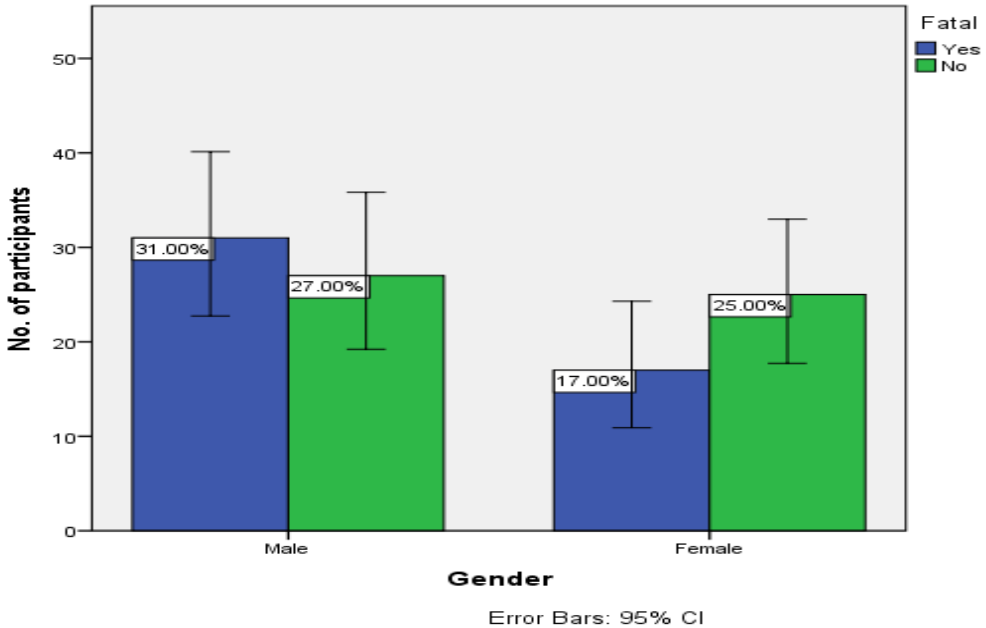

Figure 5. depicts the correlation graph between gender of the participants and awareness about the rate of fatality in case of FACES syndrome. Blue colour denotes no and green colour denotes

yes. Among females,39.39% answered yes and 13.13% answered no. Whereas among the males, 34.34% answered as yes and 13.13% as no Chi square test was evaluated for this graph with a p value of p=0.764 (p>0.05). Hence the value is statistically insignificant.

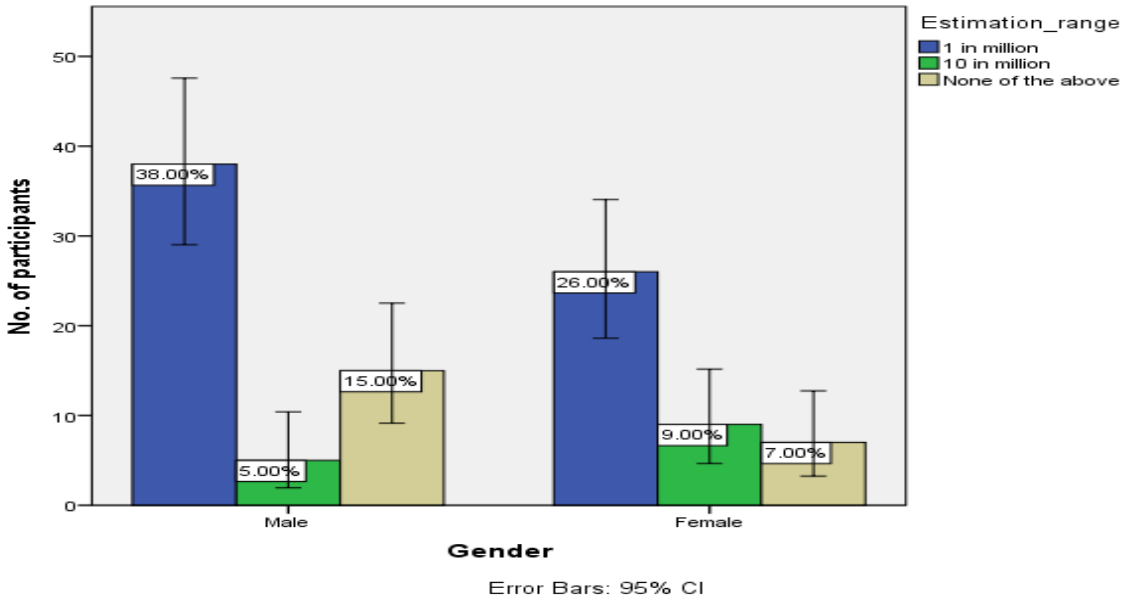

Figure 6. depicts the correlation graph between gender of the participants and estimation range about the FACES syndrome. Blue colour denotes 1 in million, green color denotes 10 in million and yellow colour denotes none of the above. Among females, 38% answered 1 in million, 15% answered none of the above and 5% answered 10 in million. Whereas among the males, 26% answered as 1 in million, 9% answered 10 in million and 7% as none of the above. Chi square test was evaluated for this graph with a p value of p=0.147 (p>0.05). Hence the value is statistically insignificant.

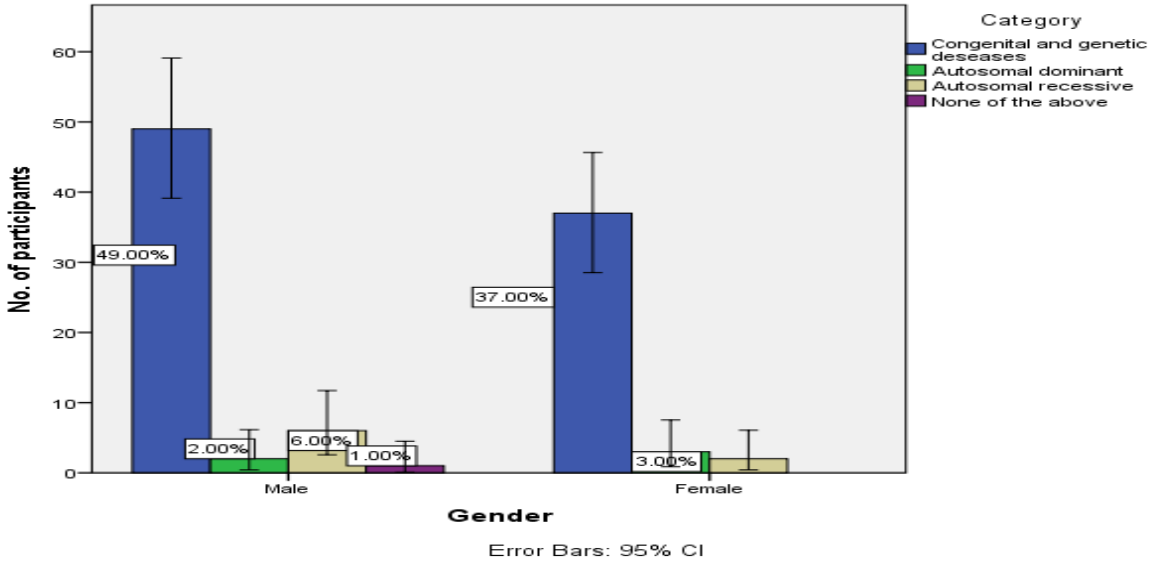

Figure 7. depicts the correlation graph between gender of the participants and category of the syndrome. Blue colour denotes congenital and genetic disease, green color denotes autosomal dominant, yellow colour denotes autosomal recessive and purple for none of the above. Among females, 49% answered congenital and genetic disease, 6% answered autosomal

recessive, 2% answered autosomal dominant and 1% none of the above. Whereas among the males, 37% answered as congenital and genetic diseases, 3% answered as autosomal dominant and the rest for autosomal recessive. Chi square test was evaluated for this graph with a p value of p=0.498 (p>0.05). Hence the value is statistically insignificant.

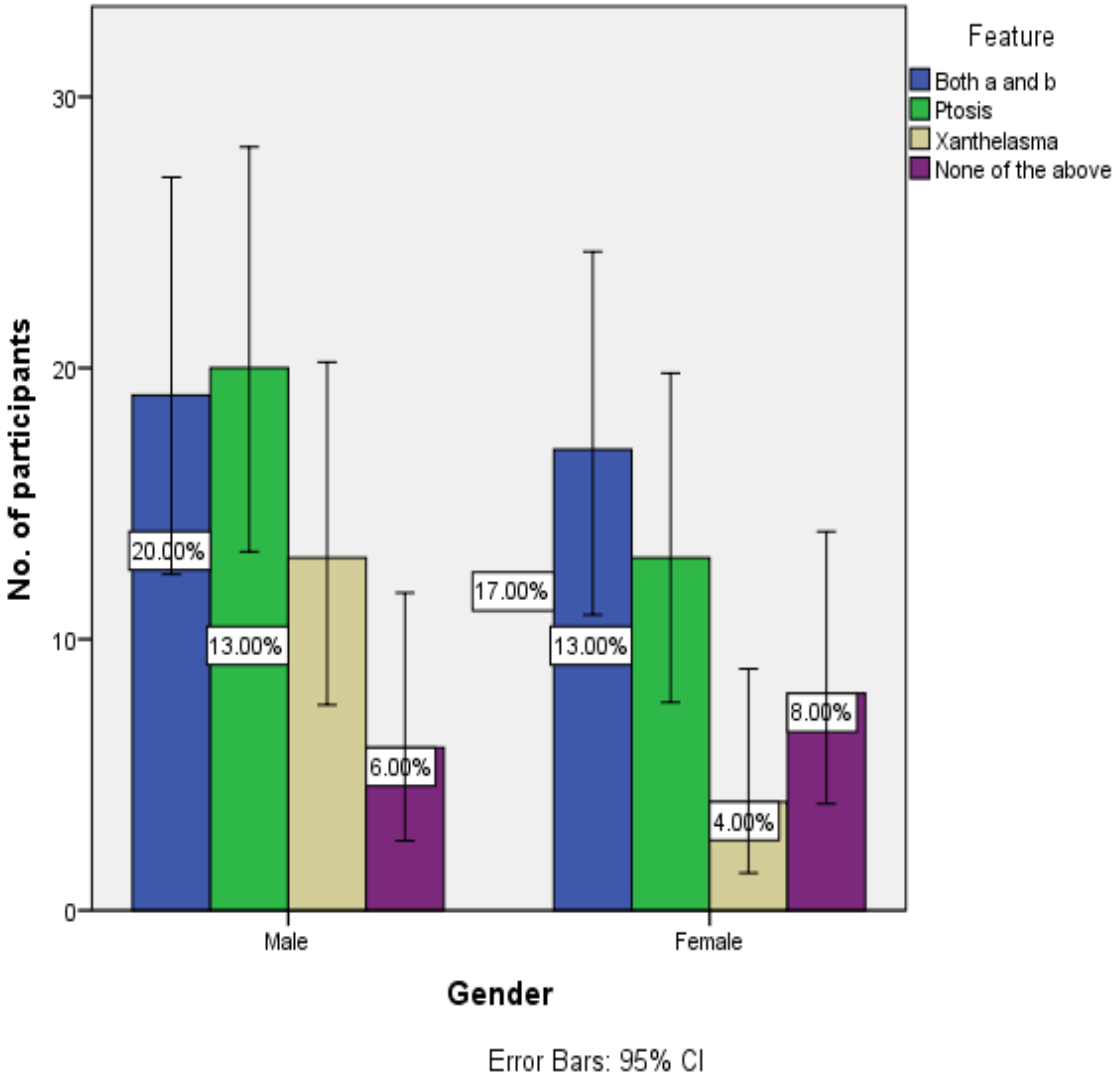

Figure 8. depicts the correlation graph between gender of the participants and features of the FACES syndrome. Blue colour denotes both a and b, green color denotes ptosis, yellow colour denotes xanthelasma and purple for none of the above. Among females, 19% answered both a and b, 20% answered ptosis, 13% answered xanthelasma and 6% none of the above. Whereas among the males, 17% answered as both a and b, 13% answered as ptosis, 8% as none of the above and 4% for xanthelasma. Chi square test was evaluated for this graph with a p value of p=0.241 (p>0.05). Hence the value is statistically insignificant.

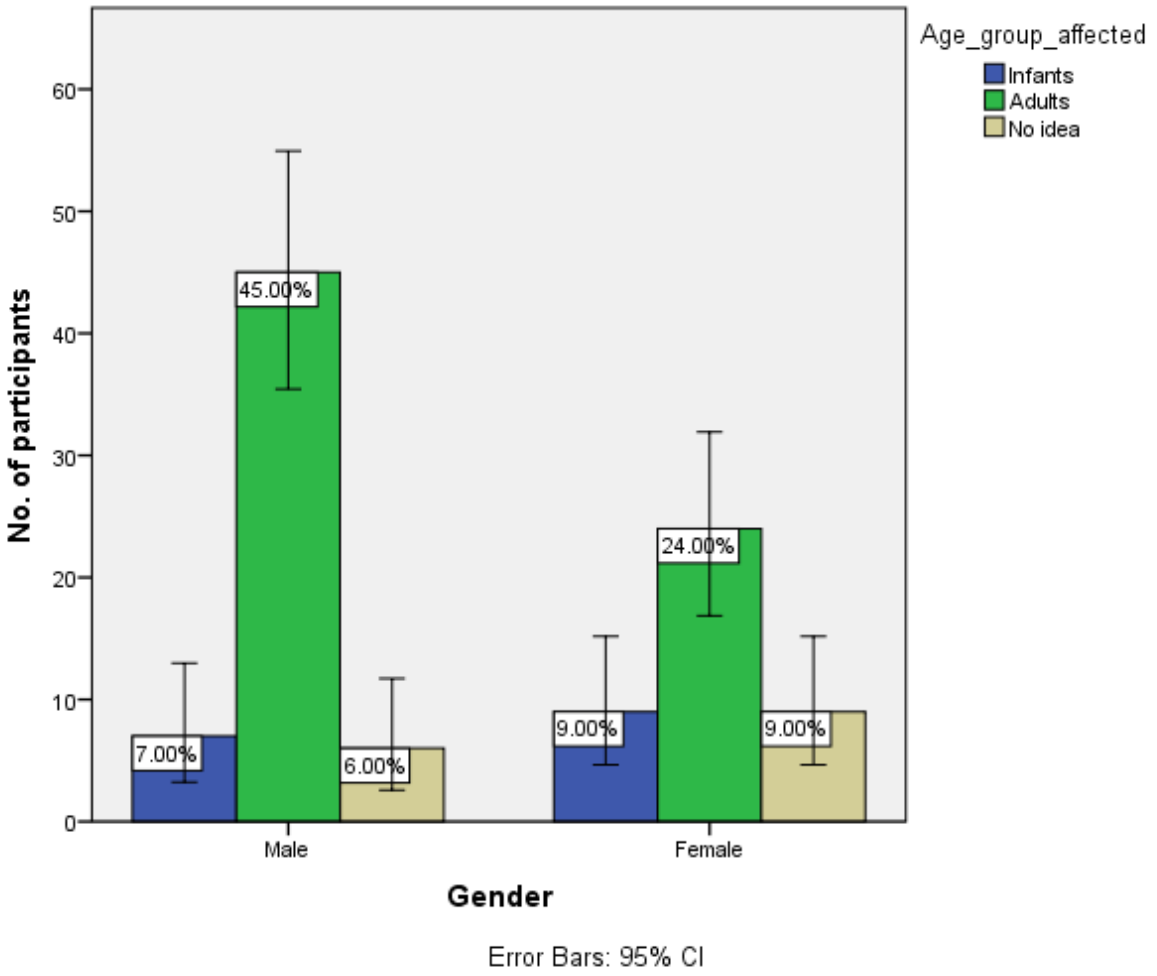

Figure 9. depicts the correlation graph between gender of the participants and age group affected with the FACES syndrome. Blue colour denotes infants, green color denotes adults and yellow colour denotes no idea. Among females, 45% answered adults, 7% answered infants and 6% answered with no idea. Whereas among the males, 24% answered as infants, 9% was the same for infants and no idea. Chi square test was evaluated for this graph with a p value of p=0.091 (p>0.05). Hence the value is statistically insignificant.

DISCUSSION

A total of 100 dental students participated in this study, where 58% were male and 42% were female (figure 1). From the result we can see that most of the people were unaware about the syndrome. Though some people guessed about the syndrome by the name and 33% of the male were aware about it. Most of them were unaware about its manifestations, etiology and treatment provided to prevent its occurrence in the society (figure2). The FACES syndrome often makes the patient compromise on his/ her basic lifestyle therefore resulting in inferior complexity among themselves(7).

According to figure 3 FACES syndrome affects various parts of the body like that of eyes, hands, head and legs. Most of the students opted for eyes as the best option which is affected the most during the course of syndrome. Different clinical features are diagnosed on the later end in the other parts of the body as well (8). That's the reason why students were confused between hands, legs and hands. This chronic illness leads to weakness and wasting of muscles from the body known as Cachexia which was selected by maximum participants. Vomiting also affects certain

individuals on a long term ratio, followed by headache as illustrated in figure 4.

The male ratio was uncertain about its fatality rate and most of them opted for a yes (figure 5). This was due to the lack of awareness of the unknown and uncertain FACES syndrome. On the other hand, females were clear about the picture to a certain extent and selected no, which was the correct option. The Feiderman Godman Syndrome comprises numerous hidden clinical features which ones treated may lead to the cure of the rare syndrome. According to many studies, the syndrome being fatal is still an unclear concept because of the restriction of articles.

Usually such kinds of rare syndromes affect less than 1 in millions of people from society(9). This was predicted to be corrected by both the ratios of male and female dentists(figure 6). The genetic background was due to congenital and genetic disease as correctly chosen by the maximum participants. Autosomal recessive and dominant background led to mislead of response by few (figure 7).

Many of the patients suffer with major eye abnormalities. Ptosis, which refers to the condition of drooping of eyelids due a congenital cause and xanthelasma refers to yellowish plaques that occur most commonly near the inner canthus of the eyelid, more often on the upper lid than the lower lid. Xanthelasma palpebrarum is the most common cutaneous xanthoma. Xanthelesma of four eyelids in patients with hyperlipidemia. These are the two gross main important occurrences seen in the eyes at the course of syndrome (figure 8).

Considering the availability of limited data online regarding the syndrome it has been observed that the more chances of this syndrome being associated with adults followed by infants (figure 9). Pectus excavatum is a condition in which a person's breastbone is sunken into his or her chest. Severe cases of pectus excavatum can eventually interfere with the function of the heart and lungs. Pectus excavatum is a condition in which a person's breastbone is sunken into his or her chest. The condition is related to the syndrome to a greater extent thus causing difficulty in breathing as seen in most clinical scenarios.

Our team has extensive knowledge and research experience that has translate into high quality publications(10–18),(19–24),(25–31)

CONCLUSION

It can be concluded that the majority of the dental students are unaware of the syndrome. The population needs to be knowledgeable about FACES syndrome in order to manage the clinical manifestation and treat the disease. Long- term documentation, review articles, multicentric audit will enhance our understanding and improve our future management of FACES Syndrome and its associated conditions.

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CONFLICT OF INTEREST

The authors declare that there were no conflicts of interest in the present study.

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