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Awareness Of Smith-Magenis Syndrome Among Dental Students

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Abstract

Introduction :
Smith-Magenis syndrome is a developmental disorder that affects many parts of the body. Smith-Magenis syndrome is usually not inherited. Most people with Smith-Magenis syndrome have no history of the condition in their family. People with Smith-Magenis syndrome typically have affectionate, engaging personalities, but most also have behavioral problems. Dental abnormalities like the curving of mouth downwards and the upper lip curves outwards, due to a fleshy philtrum.

Aim:
The main aim of this study is to analyse the knowledge and awareness about Smith - Magenis syndrome among the dental students.

Materials and methods:
To analyze the knowledge and awareness of Smith- Magenis syndrome among dental students, an online survey was conducted. data was collected and then data manipulation was done by clean up in an excel spreadsheet and the results were collected and then analyzed through SPSS software version 22. Chi square test was done to obtain the p value to find the significant differences.

Results and discussion:
The results of this awareness survey shows that most of the students (61.54%) are aware about the smith-magenis syndrome. The study revealed that the students say, males are the highly affected population (46.15%) with smith magenis syndrome when compared to males (15.38%).
Unlike this study, the previous studies show that Smith-Magenis syndrome affects males and females in equal numbers.

Conclusion:
This study observed moderate awareness of the knowledge about the Smith-Magenis syndrome and the need to increase its clinical awareness by presenting the salient features in a proper way to create a better way for treating this syndrome.

Keywords:
Syndrome, Children, mental illness, differently abled

Introduction :

Smith-Magenis syndrome is a developmental disorder that affects many parts of the body(1). The major features of this condition include mild to moderate intellectual disability, delayed speech and language skills, distinctive facial features, sleep disturbances, and behavioral problems(2).

Smith-Magenis syndrome is usually not inherited. This condition typically results from a chromosomal deletion or an RAI1 gene (Chromosome 17) mutation that occurs during the formation of reproductive cells (eggs or sperm) or in early fetal development. Most people with Smith-Magenis syndrome have no history of the condition in their family(3).

People with Smith-Magenis syndrome typically have affectionate, engaging personalities, but most also have behavioral problems(4). These include frequent temper tantrums and outbursts, aggression, anxiety, impulsiveness, and difficulty paying attention(5). Self-injury, including biting, hitting, head banging, and skin picking, is very common. Repetitive self-hugging is a behavioral trait that may be unique to Smith-Magenis syndrome(6). Some people with this condition also compulsively lick their fingers and flip pages of books and magazines(a behavior known as "lick and flip").Most people with Smith-Magenis syndrome have a broad, square-shaped face with deep-set eyes, full cheeks, and a prominent lower jaw. The middle of the face and the bridge of the nose often appear flattened.These facial differences can be subtle in early childhood, but they usually become more distinctive in later childhood and adulthood(7).

Dental abnormalities like the curving of mouth downwards and the upper lip curves outwards, due to a fleshy philtrum. These facial features become more noticeable as the individual ages, as Mandible growth outstrips that of the maxilla leading to a clear midface hypoplasia(8).

Other signs and symptoms of Smith-Magenis syndrome include short stature, abnormal curvature of the spine (scoliosis), reduced sensitivity to pain and temperature, and a hoarse voice. Some people with this disorder have ear abnormalities that lead to hearing loss(9). Affected individuals may have eye abnormalities that cause nearsightedness (myopia) and other vision problems. Although less common, heart and kidney defects also have been reported in people with Smith-Magenis syndrome.Our team has extensive knowledge and research experience that has translate into high quality publications (10–18),(19–24),(25–31) The main aim of this study is to analyse the knowledge and awareness about Smith - Magenis syndrome among the dental students.

Materials and methods:

To analyze the knowledge and awareness of Smith- Magenis syndrome among dental students, an online survey was conducted in a standard manner by simple random sampling method involving all age groups, by distributing 15 questionnaires(Given below) through an online portal called google forms, among 200 dental practitioners including Interns, dental postgraduates and dentists from different locations by acquiring approval from the ethical committee and proper consent from the individuals involved in filling the survey and then the stratified data was collected and then data manipulation was done by clean up in an excel spreadsheet and the results were collected and then analyzed through SPSS software version 22. Chi square test was done to obtain the p value to

find the significant differences.P value was set as 0.05 as a level of significance.

RESULTS AND DISCUSSION::

The results of this awareness survey shows that most of the students (61.54%) are aware about the smith-magenis syndrome (figure 1). The survey shows that prominent forehead is the most common abnormality of smith-magenis syndrome known to students (55.77%) whereas unusual shallowness of orbit and sleep apnoea is the least common abnormality said by the students (figure 2). It is known that most of the students (28.85%) say, all the given symptoms like abnormality of philtrum, hypoplasia and brachycephaly are the prevalent features in smith-magenis syndrome (figure 3). It is also known that most of the students (32.69%) say that inflammation in front of the eyes is the most common complication of smith magenis syndrome whereas vision loss is the least common complication of smith - magenis syndrome told by the students (11.54%) (figure 4). The study revealed that the students say, males are the highly affected population (46.15%) with smith magenis syndrome when compared to males (15.38%) (figure 5). The study graph also shows that most of the students say, Smith-magenis syndrome should be correctly diagnosed at the age of 5 years whereas very less number of students say that it can be diagnosed at infancy (figure 6). This study shows that the students say, fluorescence in situ hybridisation (32.69%) is the most used diagnostic method for diagnosing Smith-magenis syndrome whereas MRI and chromosome analysis (7.69%) are the least used methods for diagnosing Smith-magenis syndrome (figure 7). The study shows that the students say, gene replacement therapy is the treatment method for treating Smith-magenis syndrome whereas few students say that Smith-magenis syndrome need not be treated(figure 8).

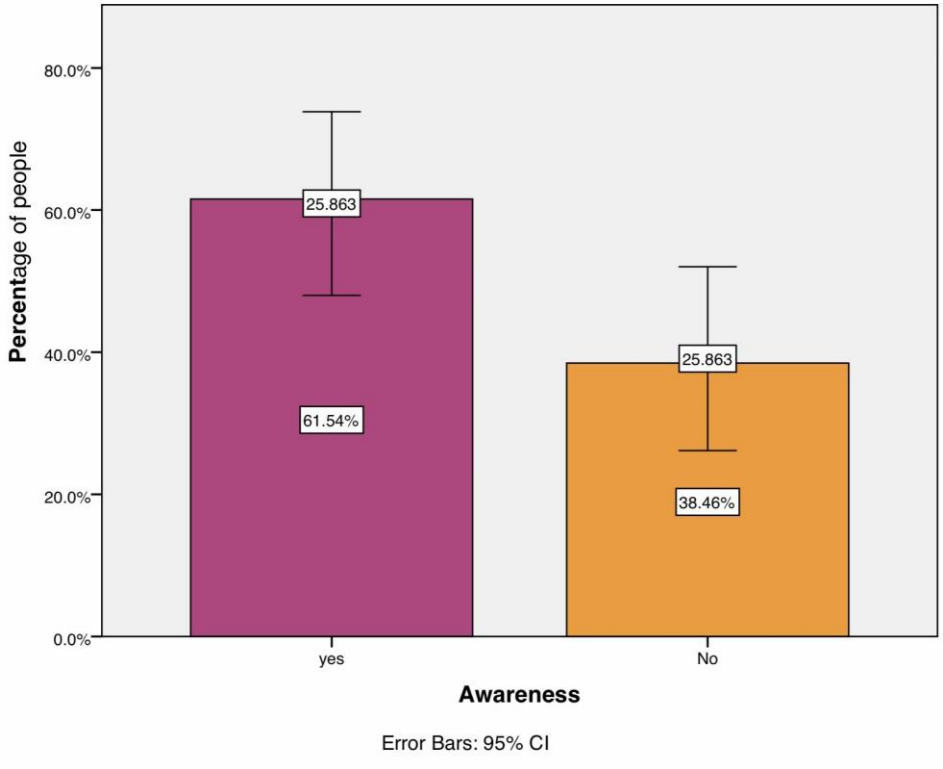

Figure 1: This graph shows the awareness about Smith-magenis syndrome among dental students. The results show that 61.54% of the students are aware about Smith-magenis syndrome and 38.46% of the students are not aware about the syndrome.

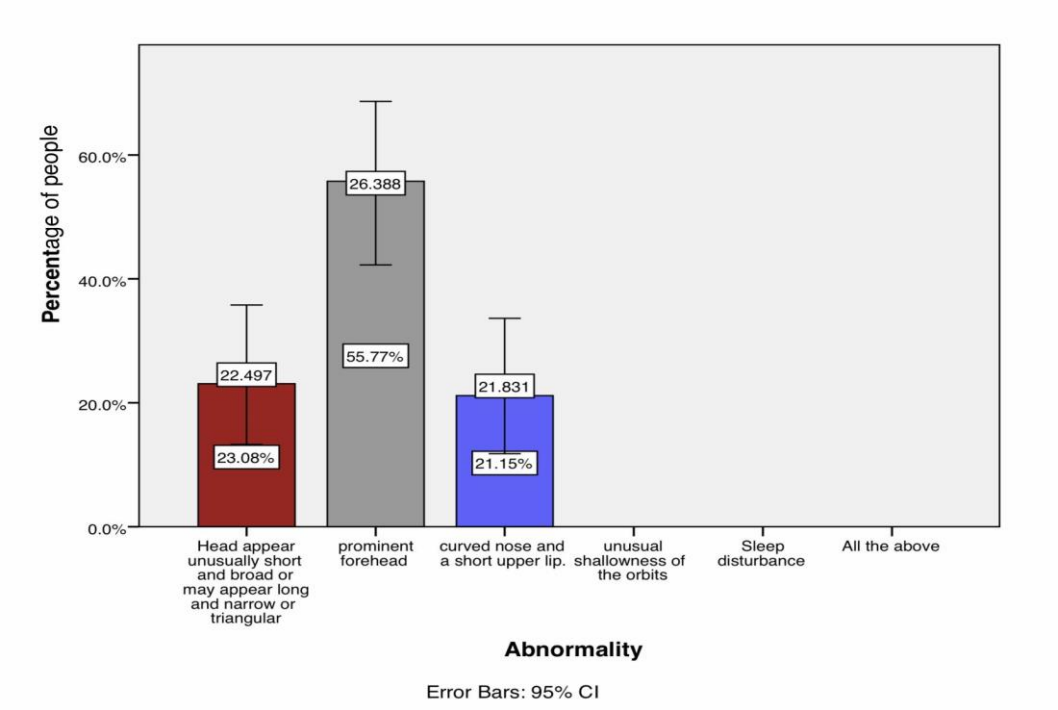

Figure 2: This graph shows the awareness among the dental students about the complications of Smith-magenis syndrome. It is known that 23.08% of the students say that the head appears unusually short and broad , 55.77% of the students say that the head will be prominent and 21.15% say that the people having Smith-magenis syndrome have curved nose and a short upper lip.

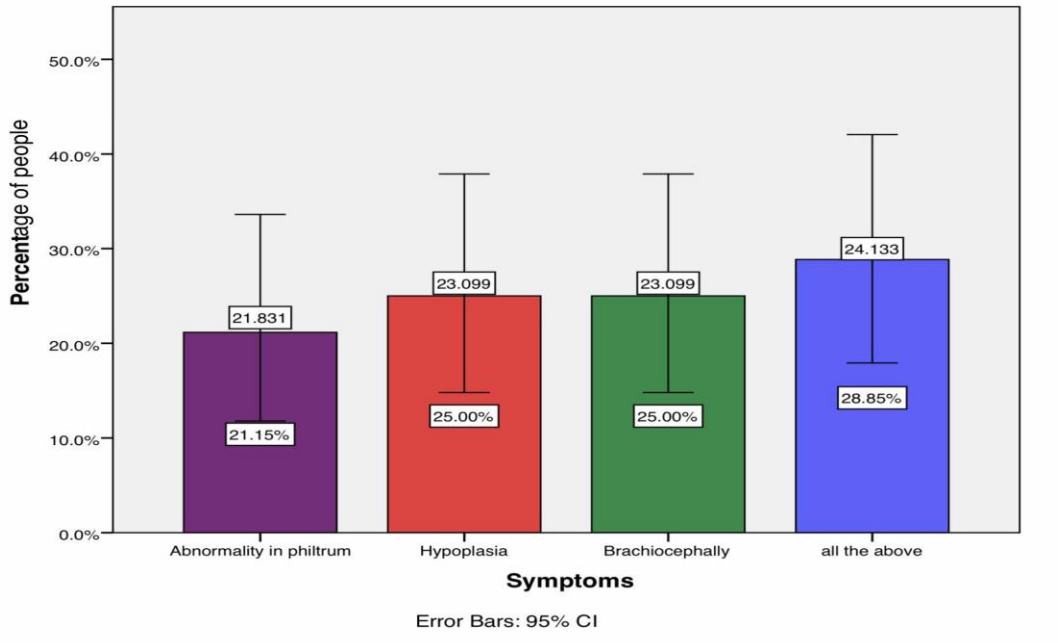

Figure 3: This graph shows the awareness among the dental students about the symptoms of Smith-magenis syndrome. It is known that 21.15% of the students say abnormality of philtrum , 25% of the students say hypoplasia, 25% of the students say Brachiocephally and 28.85% of students say that all the above symptoms are suggestive of Smith-magenis syndrome .

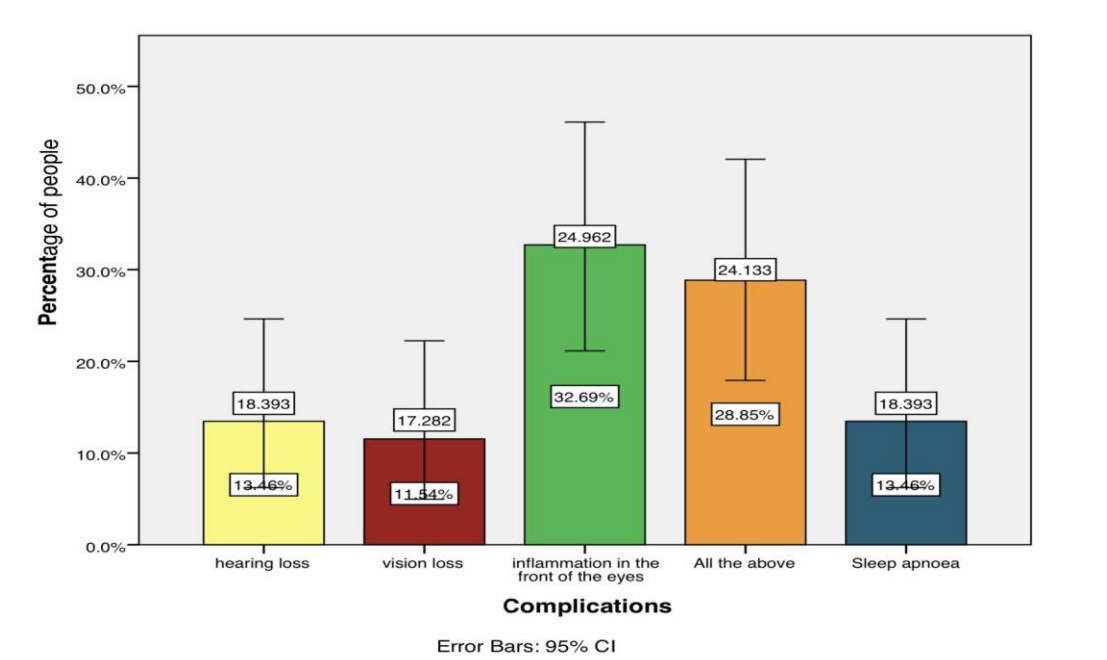

Figure 4: This graph shows the awareness among the dental students about the complications of Smith-magenis syndrome. It is known that 13.46% of the students say hearing loss , 11.54% of the students say hearing loss, 32.69% of the students say inflammation in front of the eyes, 13.46% of the students say sleep apnoea and 28.85% of the students say that all the above complications are suggestive of Smith-magenis syndrome .

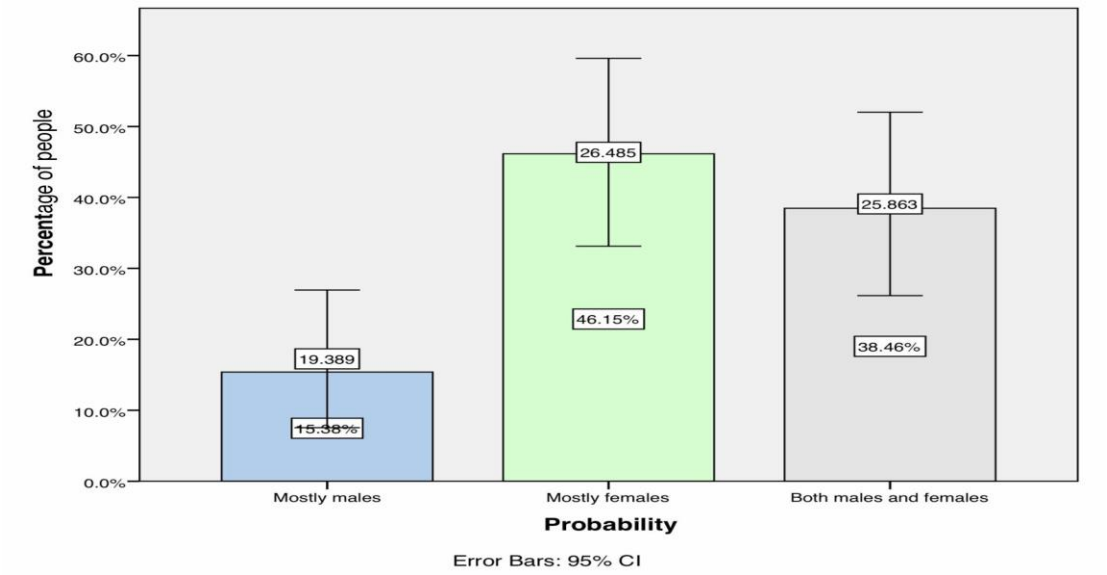

Figure 5: The graph shows the awareness among the dental students about the gender predilection of Smith-magenis syndrome. It is known that 15.38% of the students say that males are the most affected population with smith-magenis syndrome , 46.15% of the students say that females are the most affected population whereas 38.46% of the students say that both males and females are equally affected with Smith-magenis syndrome .

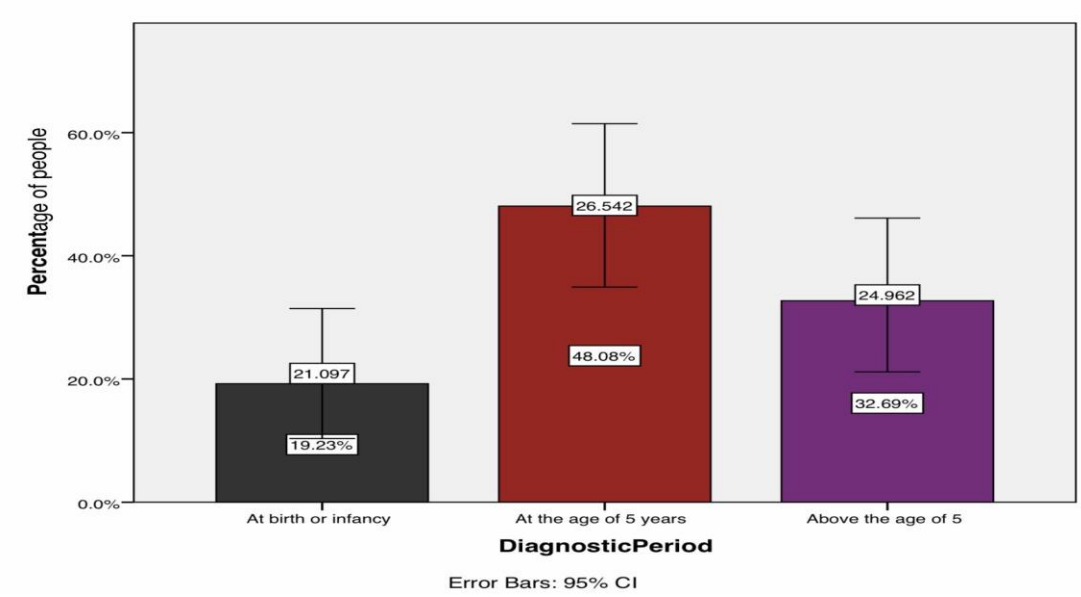

Figure 6: This graph shows the awareness among the dental students about the correct diagnostic period of Smith-magenis syndrome. It is known that 19.23% of the students say at the period of birth and infancy , 48.08% of the students say at the age of 5 years , 32.69% of the students say above the age of 5 years is the correct period for diagnosing Smith-magenis syndrome.

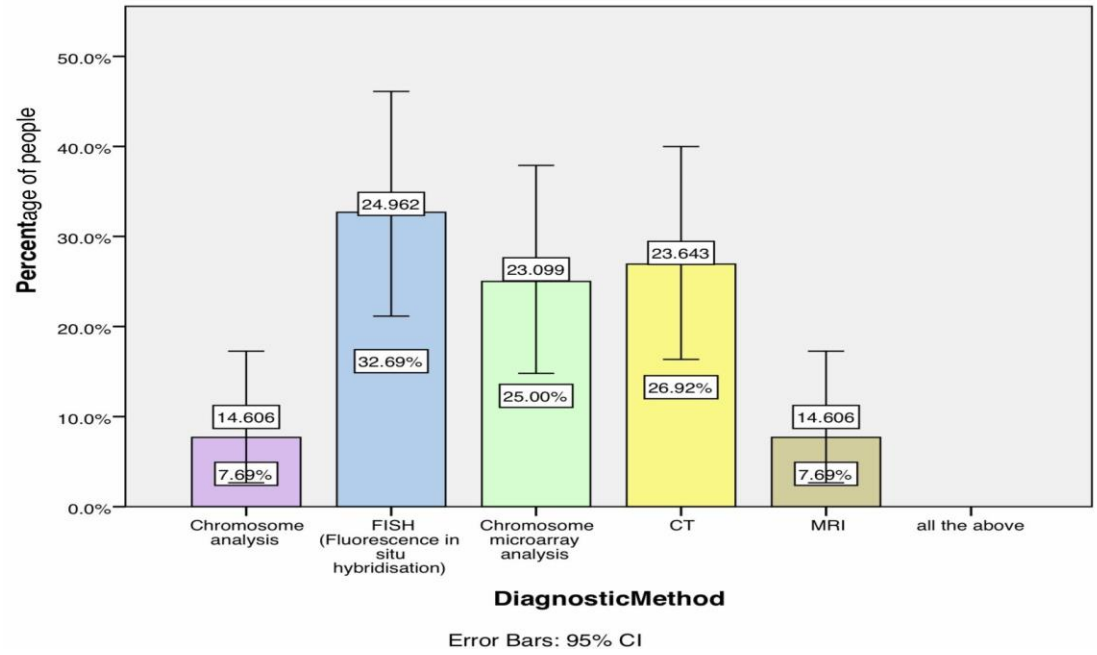

Figure 7: This graph shows the awareness among the dental students about the diagnostic method of Smith-magenis syndrome. It is known that 7.69% of the students say chromosome analysis , 32.69% of the students say fluorescence in situ hybridisation , 25% of the students say chromosome microarray analysis, 26,92% of the students say CT, 7.69% of the students say MRI and none of the students suggest all the above diagnostic methods for the diagnosis of Smith-magenis syndrome

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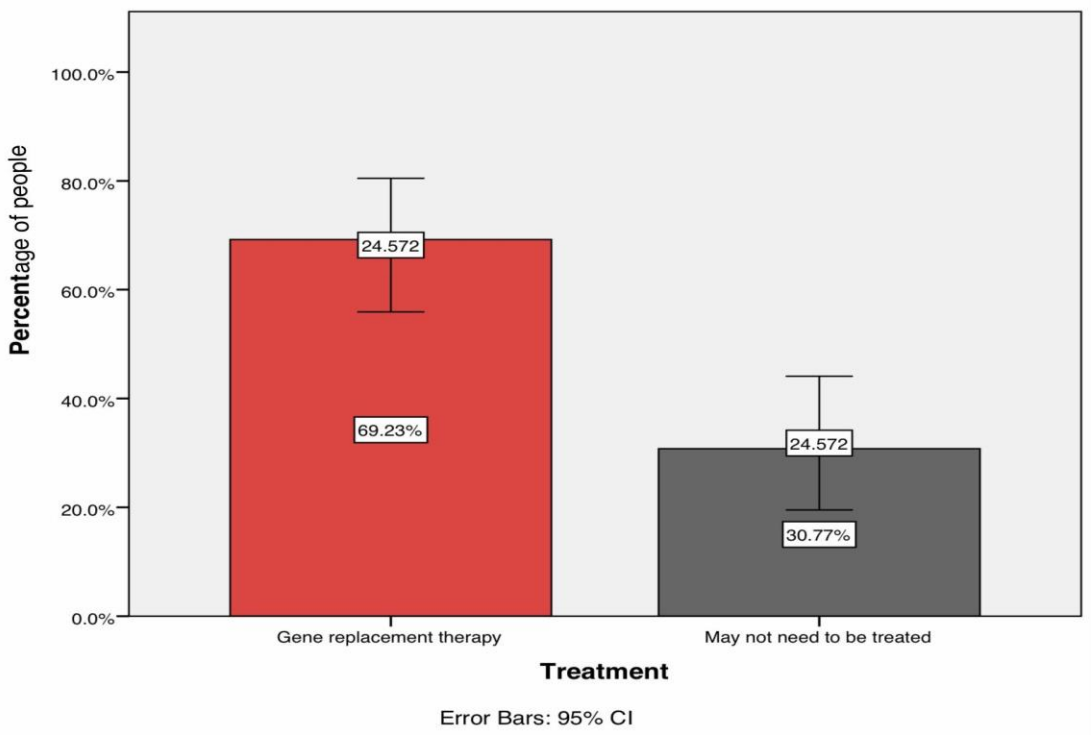

Figure 8: This graph shows the awareness among the dental students about the treatment of Smith-magenis syndrome. It is known that 61.23% of the students say gene replacement therapy is the treatment suggestion for smith-magenis syndrome whereas 30.77% of the students say that smith-magenis syndrome may not be treated.

Smith-Magenis syndrome affects males and females in equal numbers. The incidence is estimated to be 1 in 15,000-25,000 people in the general population in the United States(32). However, cases may go undiagnosed or misdiagnosed, making it difficult to determine the true frequency of SMS in the general population. SMS has been reported throughout the world and in all ethnic groups(33).

Approximately 90% of cases are caused when a portion of chromosome is missing or deleted. This deleted portion within chromosome 17p11.2 includes the RAI1 gene, which is believed to play a major role in the development of the disorder(34). In the remaining cases, there is no deleted material on chromosome 17; these cases are caused by mutations in the RAI1 gene(35). Other genes within the deleted segment may also play a role in variable features in the syndrome, but it is not fully understood how significant a role they play in the development of SMS(36).

Conclusion:

This study observed moderate awareness of the knowledge about the Smith-Magenis syndrome and the need to increase its clinical awareness by presenting the salient features in a proper way to create a better way for treating this syndrome.

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