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Awareness On Williams Syndrome Among Dental Students

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Abstract

Introduction : Williams syndrome (WS) is a rare genetic and neurodevelopmental disorder.Williams syndrome (WS) is a multisystem disorder caused by the deletion of 26 contiguous genes, including elastin (ELN)on chromosome 7q11.The child with this syndrome also shows distinctive facies (elfin-like features), hypercalcemia, connective tissue abnormalities, growth abnormalities, intellectual disability, behavior deficits, and a gregarious personality.The first cases of Williams syndrome were found in early reports detailing the clinical characteristics of children who had infantile hypercalcemia, short stature, and variable congenital malformations.

Aim: To evaluate awareness level regarding clinical Features, genetic factors, specific dental features associated with Williams syndrome among Dental students .

Materials And Methods: This survey was conducted among 100 dental students of a private dental college by circulating a questionnaire . Google forms and links were shared in various social media. 10 questions consisting related to awareness,clinical features,dental features associated with Williams syndrome were included in this study. Students of the private dental colleges were only included in the study.The data was collected and analysed using SPSS software version 23 and the statistical significance was checked using Chi-Square analysis . ($p < 0.05$ was considered as statistically significant)

Result:.54% students think that it is not possible to cure Williams syndrome 56% of students are aware of clinical features and genetic variations of Williams and Down syndrome.Majority of the dental student population about 66% says that prevalence of tooth abnormalities such as midline spacing , crooked teeth are more common among Williams syndrome

Conclusion : It was noted that there is still a lack of awareness prevailing , hence more updates have to be given among undergraduate students regarding Williams syndrome and it's dental features for better knowledge .
Key words: Williams syndrome, Elfin facies,gregarious personality

INTRODUCTION :

Williams syndrome (WS) is a rare genetic neurodevelopmental disorder. WS often presents at birth itself when the child is discovered to have supra-vascular aortic stenosis. The child also shows peculiar clinical features such as distinctive facies, elfin-like features,hypercalcemia, connective tissue abnormalities, growth abnormalities, intellectual disability, behavior deficits, and a gregarious personality(1). Genetic cause of the William syndrome is due to deletion at chromosome band 7q11.23 that involves the elastin gene (ELN). This gene is present in the Williams-Beuren Syndrome Critical Region (WBSCR). The gene deletion, comprising 26 genes, is detected through dual-color fluorescent in situ hybridization (FISH) or deletion/duplication testing method(2).Microarray analysis is another distinctive diagnostic test that can identify the size of the elastin deletion. Both FISH and microarray analyses utilize a parenteral blood sample to extract DNA for analysis. From 96 to 98% of patients with WS have a deleted elastin gene which is found to be the major etiology(3). The genetic disorder shows autosomal dominant transmission. The defect in elastin leads to generalized arteriopathy and may affect any artery in the body, but it often affects medium to large-sized arteries.(4)

Infants with Williams syndrome often present with failure to thrive, short stature, and supra-vascular aortic stenosis. Due to the defect in elastin, children may also have other elastin arteriopathies, along with peripheral pulmonary stenosis and hypertension.During childhood, patients may have middle ear infections and visual difficulty(5). On a physical exam, all children with Williams syndrome have distinctive facial features with elfin-like features.The death in the WS is largely due to the presence of arteriopathy and congenital heart disease. Eighty percent of patients with Williams syndrome experience cardiovascular abnormalities, such as stenosed large arteries that require cardiothoracic surgery(6). Our team has extensive knowledge and research experience that has translated into high quality publications (7–15),(16–21),(22–27) .

Children with WS commonly have various endocrine abnormalities such as presence of hypercalcemia and hypercalciuria which can result in renal calculi.The patient may also have physical signs of hypothyroidism, delayed growth, or early puberty.They also may develop Connective tissue abnormalities often result in hyperextensible joints or hypotonia, resulting in delayed motor milestones(28). Patients with WS born with blue or green eyes often display a "starburst" or stellate pattern on their iris.This "starburst" pattern characteristically shows a white, lacy appearance.The patient may have comorbid psychiatric disorders, including intellectual disability (ID), attention-deficit hyperactivity disorder (ADHD), obsessive-compulsive disorder (OCD), or generalized anxiety disorder (GAD)(29)

This research is needed to obtain the awareness level prevailing among dental students regarding Williams syndrome and its clinical features ,peculiar dental features and it's genetic factors associated with it.The aim of the study is to evaluate awareness level prevailing among dental students regarding Williams syndrome.

MATERIALS AND METHODS:

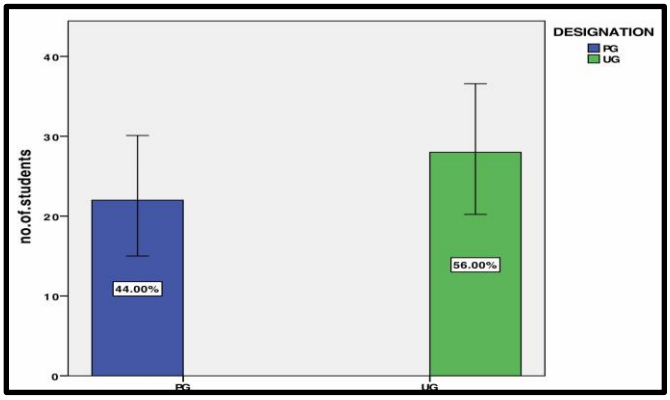
In this study, a survey was conducted among 100 dental students of a private dental college by circulating questionnaire . Google forms and links were shared in various social media. 10 questions related to awareness, clinical Features,and dental features of Williams syndrome were included in the study. Students of the private dental colleges were only included in the study.The data was collected and analysed using SPSS software version 23 and the statistical significance was checked using Chi-Square analysis . ($p < 0.05$ was considered as statistically significant).

RESULTS AND DISCUSSION::

Out of 100 students participating, 44% were postgraduate students and 56% were undergraduate students respectively (fig1) .54% students think that it is not possible to cure Williams syndrome and the remaining 46% are not aware about the cure of disease (fig2). 56% of students are aware of clinical features and genetic variations of Williams and Down syndrome whereas 44% of students are not aware about the difference in clinical features of Williams and Down syndrome (fig 3). 72% of students are aware that diagnosis age of Williams syndrome is below 4 years old of age which is clinically proven and remaining 28% students says that diagnosis above the age of four(fig4).Majority of the dental student population about 66% says that prevalence of tooth abnormalities such as midline spacing , crooked teeth are more common among Williams syndrome and remaining 34% are not aware about its prevalence (fig 5).Majority of the students (82%) of the students are preferred treating patients with Williams syndrome under General anaesthesia (fig6).

On evaluating the association of clinical features of williams syndrome and Down syndrome among the undergraduates and postgraduates (fig7).In the association between prevalence of tooth abnormalities , post graduates are aware 44% (fig8). Fig 9 represents that the majority of the dental students population preferred using performing under GA which is pg (48%) and ug (34%).

Fig 1:



Designation of the students participated

Bar graph represents the percentage distribution of students.Blue colour denotes postgraduate(44%) and green colour denotes undergraduate students (56%)

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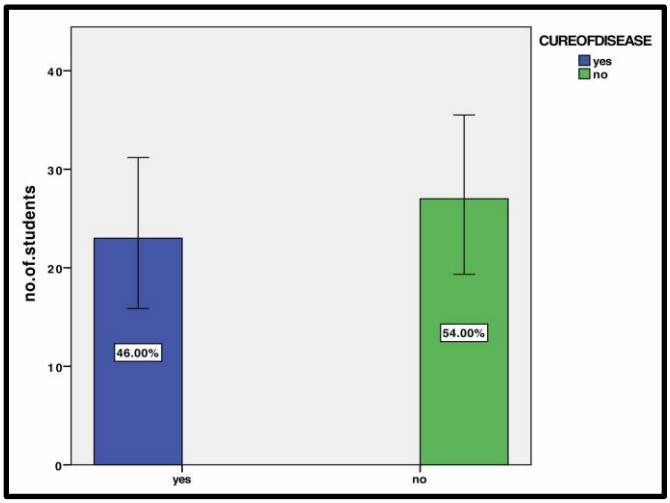
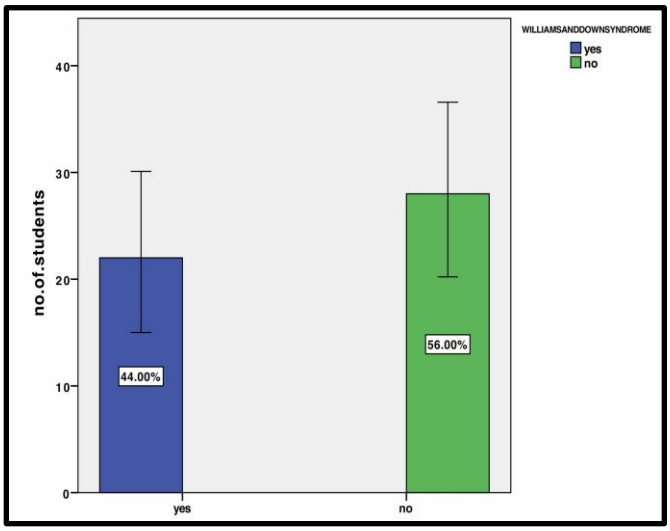
Fig 2:



Awareness level of cure of disease

Bar graph represents the percentage of awareness level of cure of disease of Williams syndrome .Blue colour denotes that students are aware that it is not possible that cure of disease is not possible 54% (green colour) whereas the blue colour denotes that remaining of the students says that the cure of disease is possible (46%).

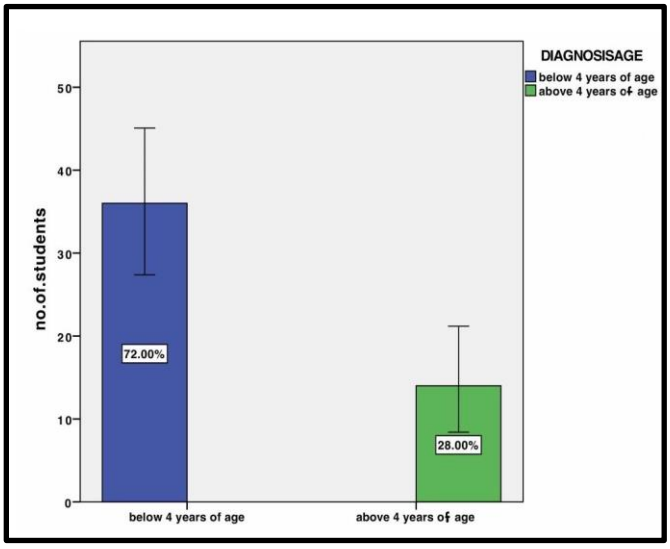
Fig 3:



Awareness level of clinical features of Williams and down syndrome .

Bar graph represents the percentage of awareness level of clinical features of Williams syndrome and Down syndrome .44% of students are aware that clinical features of Williams and Down syndrome are not the same (green colour) whereas the blue colour denotes that remaining of the students says that the clinical features of Williams and Down syndrome are similar (56%).

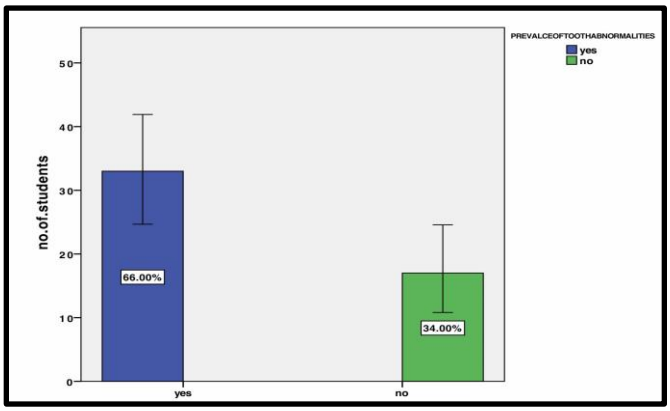
Fig 4:



Awareness level in diagnosing the age

Bar graph represents the percentage of awareness level in diagnosing the age of Williams syndrome .72% of students are aware that diagnosing Williams syndrome is below 4 years of age whereas 28% of the students says that diagnosing age is above 4 years of age,

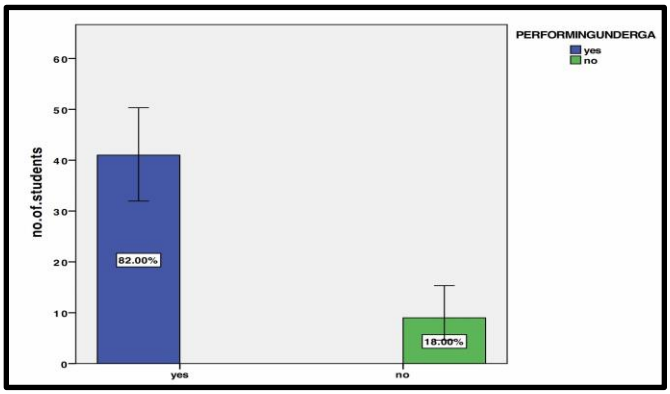
Fig 5:



Awareness level that William syndrome causes tooth abnormalities.

Bar graph represents the percentage of awareness level in diagnosing the age of Williams syndrome .66% of students are aware that prevalence of tooth abnormalities in Williams syndrome is common whereas 34% of the students says that prevalence of tooth abnormalities is not common.

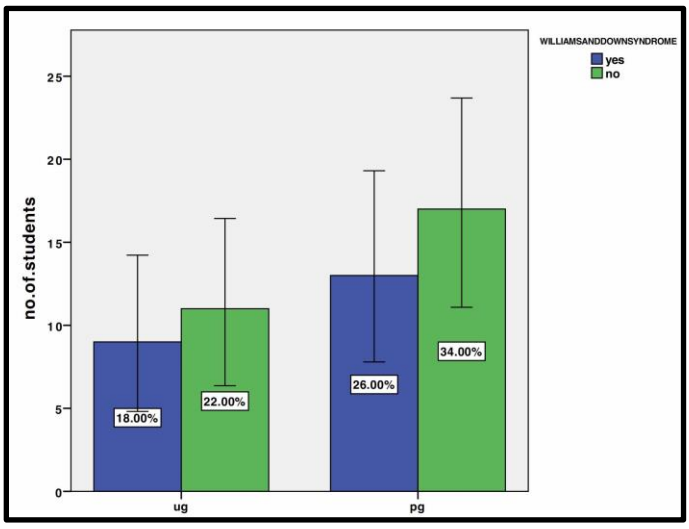
Fig 6:



Performing under GA

Bar graph represents students preferred treating patients with Williams syndrome under General anaesthesia. 82% of students preferred treating patients under GA remaining 18% of students did not prefer treating under GA.

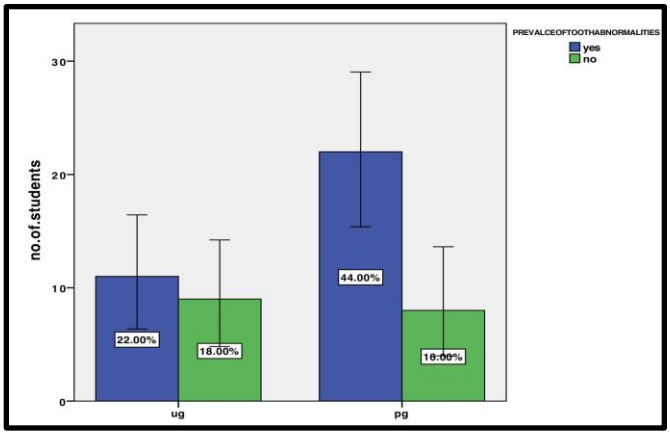
Fig 7:



Correlation between clinical features of William syndrome and Down syndrome and students .

Correlation chart represents the clinical features of Williams and Down syndrome . Among postgraduate students, nearly 34% (green colour)says that clinical features of Williams and Down syndrome are not the same , whereas 26% of students say that clinical features of Williams and Down syndrome are similar . Among undergraduate students 22% of the students are aware of the difference between clinical features of each whereas 18% are not aware .Statistical analysis was carried out with Chi -Square test ; p value = 0.518, which was not statistically significant .

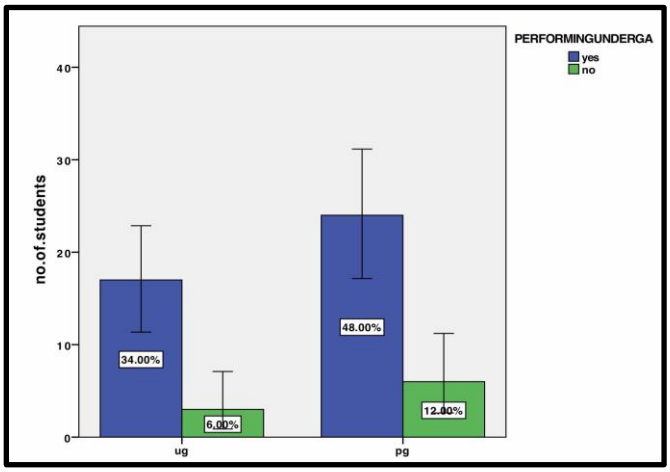
Fig 8:



Awareness level correlation in prevalence of tooth abnormalities.

Correlation chart represents the prevalence of tooth abnormalities among undergraduate students and postgraduate students. Among Ug's 22% students(blue colour) say that the prevalence of tooth abnormalities is common whereas 16% of the students(green colour) are not aware.Among Pg's 44% (blue colour)say that the prevalence of tooth abnormalities is common whereas 18% (green colour) are not aware.Statistical analysis was carried out with Chi -Square test ; p value = 0.385, which was not statistically significant .

Fig 9:



Awareness level correlation in performing under GA

Correlation chart represents the students' preferred treatment under GA.Among postgraduate students 48% of the students(blue colour) preferred that treatment under GA whereas 12% of the students(green colour) are not preferring.Among undergraduates 34% of the students(green colour)are preferred treating patients under GA remaining 6% of the student

(blue colour)not preferred treating under GA.Statistical analysis was carried out with Chi - Square test ; p value =0.364, which was not statistically significant

Many articles suggested that currently there is no specific cure for Williams syndrome but rehabilitation therapy and early targeted educational interventions are extremely important to enhance the development of cognitive and social competencies(30). Some programs have been developed in recent years to optimize the life performances of children or adults with disabilities.Moreover, research has shown that approximately 40% of all people with intellectual disabilities show marked emotional and gregarious behavioural problems that interfere with their daily life(31) .It is therefore crucial, in addition to producing more in-depth scientific knowledge on the Syndrome, to invest on treatment options for an improved quality of life and social functioning in WS patients.

CONCLUSION:

Thus from the preliminary study we can hypothesize that awareness of williams syndrome has been not much prevailing among undergraduate students and much more information updation and awareness program helps in better awareness .We anticipate that this study will serve as a base for further breakthrough and still more awareness among students, which has remained a topic of controversy for months and also prove to be of enormous importance in the dental practice .

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