Journal of Positive School Psychology
2022, Vol. 6, No. 3, 718–727 http://journalppw.com

Awareness Of Sturge Weber Syndrome Among Dental Students

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Abstract

Introduction
Sturge-Weber syndrome is a rare, congenital, neuro-oculo-cutaneous disorder which is characterised extra-orally by unilateral port wine stains on the face, glaucoma, seizures and mental retardation, and intra-orally by ipsilateral gingival haemangioma which frequently affects the maxilla or mandible.The aim of the study is to create awareness about Sturge Weber syndrome among dental students.

Materials and methods
An online study questionnaire of 15 questions was circulated among 112 dental students of Saveetha Dental College .The data was compiled in Excel and the results were statistically analysed using spss software.

Results and discussion
The results shows that 81.82% of respondents heard of Sturge-Weber Syndrome, 57.58% knew it was a birth defect, 54.55% knew SWS is caused by mutation in GNAQ gene(, 81.82% think hemangiomas in the region of oral cavity are always of clinical importance to the dental profession,36.36% are only aware that it is a lifelong condition,24.24% among the PG respondents(34.34%) are aware that port wine stains on the face and 34.34% of PGs knew Intra-Orally SWS is characterized by ipsilateral gingival haemangioma.

Conclusion
Within the limits of study,PG respondents have greater awareness than UG respondents. Among all, results of this survey shows that half of them are aware about the Sturge-Weber Syndrome while others are not.

Keywords: Sturge-Weber syndrome,port wine stains,innovative technology.

INTRODUCTION

Sturge-Weber syndrome is a rare, congenital, neuro-oculo-cutaneous disorder which is characterised extra-orally by unilateral port wine stains on the face, glaucoma, seizures and mental retardation, and intra-orally by ipsilateral gingival haemangioma which frequently affects the maxilla or mandible.Sturge-Weber syndrome is also called encephalotrigeminal angiomatosis.It is the third most common neurocutaneous syndrome after neurofibromatosis and tuberous sclerosis(1).The neurologic manifestations of SWS include atonic, tonic, or myoclonic seizures.Sturge-Weber syndrome is a sporadic developmental disorder caused by somatic mosaic mutations in the GNAQ gene which is located on the long arm of chromosome 9 (2).The incidence of Sturge-Weber syndrome is not well known and estimated to be 1 in 20,000-50,000 live births. SWS affects males and females equally and there is no race predilection.The facial nevus is composed of multiple thin-walled vessels that resemble capillaries. The neuropathological finding is an angioma that consists of multiple capillaries and small venous channels and is usually confined to the pia mater. There is a relative lack of superficial cortical veins, and the blood is shunted to the deep venous system by the enlarged medullary veins which result in stasis and ischemic changes.

A seizure is usually the first neurological manifestation of Sturge-Weber syndrome. Infantile spasms are seen in approximately 90% of affected patients in the first year of life followed by atonic, tonic, or myoclonic seizures(3). Facial nevus (port-wine stain) is another common finding and is typically seen along the ophthalmic or maxillary segment of the trigeminal nerve (forehead, cheeks)(4). A child with a facial port-wine stain has a 10% to 35% risk of brain involvement. If there is involvement of both upper and lower eyelids, then the risk of glaucoma increases up to 50%. Glaucoma is almost always ipsilateral to the facial port-wine stain. Not all patients with port-wine stains have Sturge-Weber syndrome. Patients with Sturge-Weber syndrome may present with cerebral symptoms without facial findings. Ocular involvement in infancy may present with increased vascularity of the conjunctiva, eye enlargement, strabismus, and increased tearing. Other symptoms are intellectual disability, early handedness, and gaze preferences. Diffuse choroidal hemangioma is seen in about 20% of patients with Sturge-Weber syndrome and is usually on the same side as a facial port-wine stain(5)

Diagnosis of Sturge-Weber syndrome is based on typical clinical symptoms, facial appearance, and brain magnetic resonance imaging (MRI) findings(5,6).An ophthalmic examination is required to rule out glaucoma. Ocular ultrasound can demonstrate diffuse choroidal thickening which suggests choroidal hemangioma. Gyriform calcifications can be seen on the skull radiographs and are classically described as a "tram-track sign." Computed tomogram (CT) is the best modality to detect calcifications and also show the other changes such as cortical atrophy and leptomeningeal enhancement on the post-contrast studies. Fluorodeoxyglucose-positron emission tomography (FDG-PET) may be a useful modality to study cerebral metabolism in patients with Sturge-Weber syndrome. The affected area is usually hypermetabolic in the early stages with hypometabolism in the late stage(7).PET may be useful in surgical planning when cortical resection is required for the treatment of intractable seizures.

There is no specific treatment for Sturge-Weber syndrome. The primary aim is to minimize seizure activity with anticonvulsant medications. Surgery may be considered in patients who fail medical management and continue to have refractory seizures(8).Low-dose aspirin has also been shown to be effective in decreasing the frequency of seizures and stroke-like episodes(9).An annual ophthalmologic examination is recommended even if the early evaluation does not detect glaucoma. Topical medication is considered first for late-onset glaucoma. The surgery is considered for patients with early-onset glaucoma and associated angle abnormalities and includes goniotomy or trabeculotomy(9,10).The port-wine stain can be treated with laser photocoagulation which results in irreversible damage to the blood vessels without damage to the other skin components. Our team has extensive knowledge and research experience that has translated into high quality publications(11–20),(21–26),(27–33). The aim of the study is to

assess the awareness of Sturge Weber syndrome among dental students.

MATERIALS AND METHOD:

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.Inclusion criteria-Students of saveetha dental college.Exclusive criteria-Students studying other than saveetha dental college.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 Sturge Weber 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 < than 0.05 was considered as significant.

RESULTS :

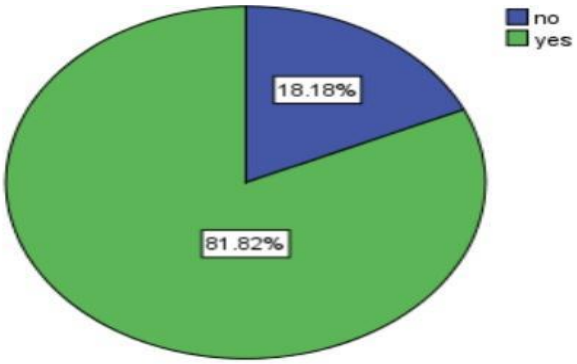

FIG1:Pie chart showing percentage distribution in response to the question (Have you ever heard of Sturge-Weber Syndrome),81.82%(green) represents 'Yes' that they are aware about the syndrome while 18.18%(blue) represents 'No'.

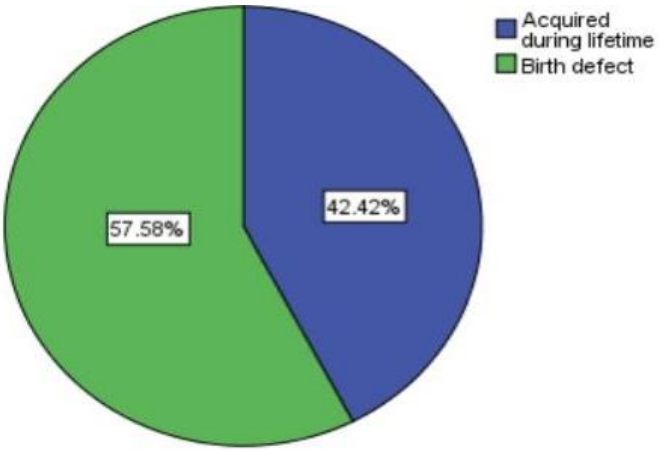

FIG2:Pie chart showing percentage distribution in response to the question (What type of disorder it is), 57.58% (green) represents that they are aware it is a birth defect while 42.2%(blue) it is acquired during lifetime.

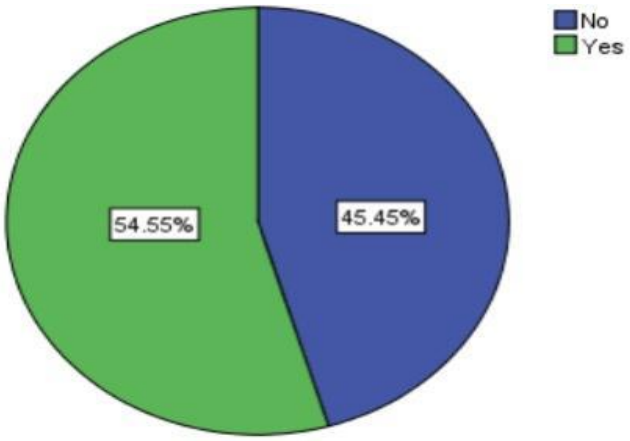

FIG3:Pie chart showing percentage distribution in response to the question (Do you think SWS is caused by mutation in GNAQ gene?),54.55%(green) represents 'Yes' that they agree to it,while 45.45% (blue) represents 'No'.

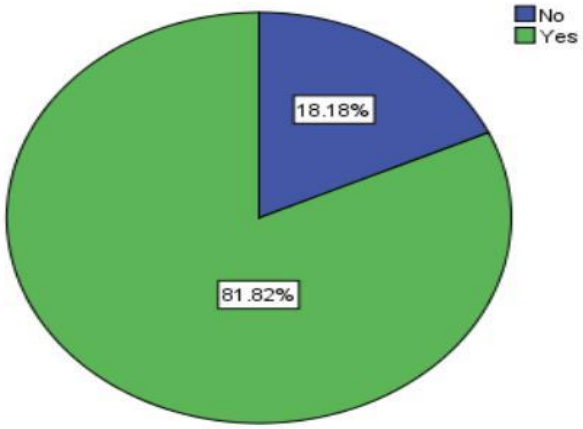

FIG4:Pie chart showing percentage distribution in response to the question(Do you think hemangiomas in the region of oral cavity are always of clinical importance to the dental profession?),81.82%(green) responded 'Yes' that they agree to it, while 18.18%(blue) responded No'.

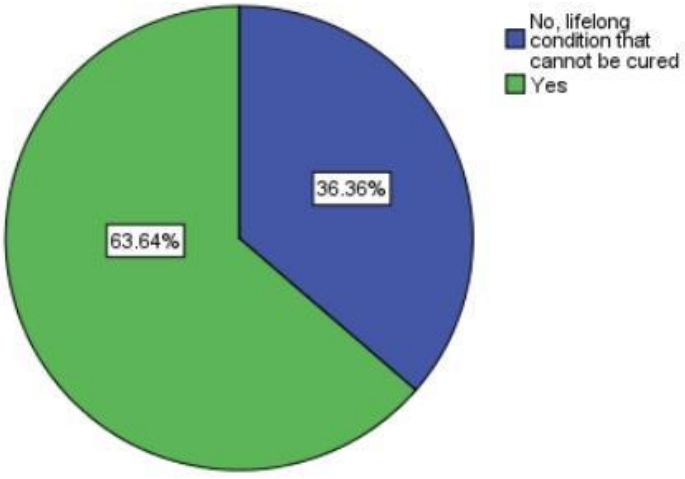

FIG5:Pie chart showing percentage distribution in response to the question(Is there a cure for Sturge Weber?),36.36% (blue) are aware that it is a lifelong condition that cannot be cured while 63.64% are not aware.

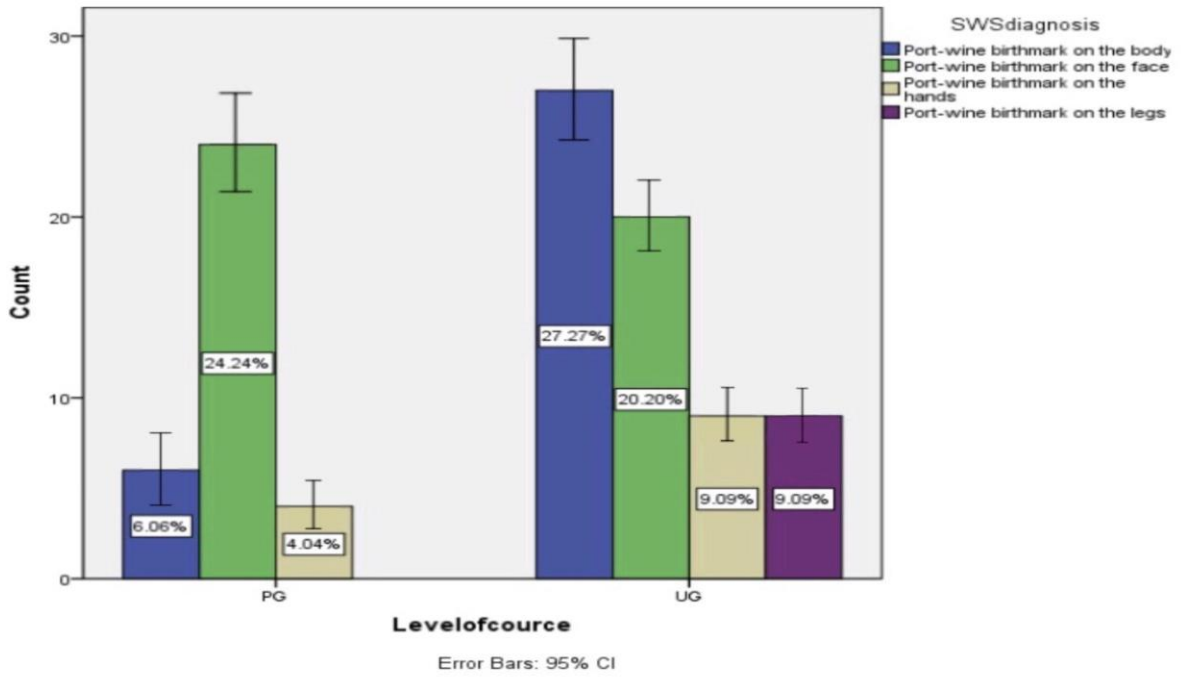

FIG6:This Bar chart represents the correlation between level and where the port wine stains occurs.X-axis represents the level of study ; and Y-axis represents the number of participants. Chi square test was done and the association was found to be statistically insignificant. Pearson's chi-value is 16.568, df:3, p value: 0.345(>0.05)hence statistically insignificant.Blue colour represents port wine stains on the body, green represents port wine stains on the face, gold represents port wine stains on hands, violet represents port wine stains on the legs. Providing PGs have better awareness than UGs.

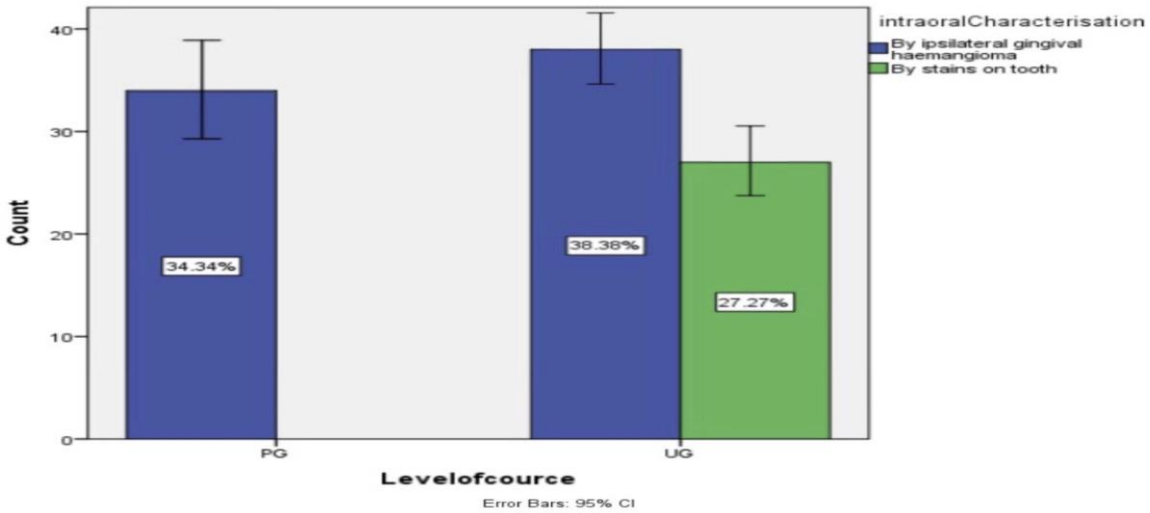

FIG7:This Bar chart represents the correlation between level and How Intra-Orally SWS is characterized.X-axis represents the level of study ; and Y-axis represents the number of participants. Chi square test was done and the association was found to be statistically insignificant. Pearson's chi-value is 2.190, df:1, p value: 0.345(>0.05)hence statistically insignificant.Blue colour represents ipsilateral gingival haemangioma,green represents stains on the tooth . pGs have better awareness than UGs.

The results shows that 81.82% of respondents heard of Sturge-Weber Syndrome(fig-1), 57.58% knew it was a birth defect(fig-2), 54.55% knew SWS is caused by mutation in GNAQ gene(fig-3), 81.82% think

hemangiomas in the region of oral cavity are always of clinical importance to the dental profession(fig-4),36.36% are only aware that it is a lifelong condition(fig-5),24.24% among the PG respondents(34.34%) are aware that port wine stains on the face(fig-6) and 34.34% of PGs knew Intra-Orally SWS is characterized by ipsilateral gingival haemangioma(fig-7).

DISCUSSION:

A total of 112 dental students who participated in the survey, 35 were males and 77 were females.
From the results, we observe that most of the people (81.82%)were aware of the Sturge-Weber syndrome (fig-1) . From the results we observe that though some people guessed the syndrome, most of them were unaware what type of disorder it is, 57.58% are aware it is a birth defect while 42.2% think it is acquired during lifetime(fig-2).This stain is a birthmark caused by an overabundance of capillaries near the surface of the skin. Blood vessels on the same side of the brain as the stain may also be affected(34).

According to the question, Do you think SWS is caused by mutation in the GNAQ gene? 54.55% are aware of it while 45.45%are not aware .This graph thus shows the unawareness of half of the students towards this disease (fig-3). The blood vessel formations associated with SWS start when a baby is in the womb. Around the sixth week of development, a network of nerves develops around the area that will become a baby's head.Normally, this network goes away in the ninth week of development. In babies with SWS, however, this network of nerves doesn't go away(35). This reduces the amount of oxygen and blood flowing to the brain, which can affect brain tissue development(36).

According to the study, The oral manifestation of the sturge Weber syndrome was discussed among the dental students. 81.82% of the respondents are aware that hemangiomas in the region of the oral cavity are always of clinical importance to the dental profession (fig-4). Hemangiomas are common tumors characterized microscopically by proliferation of blood vessels. The congenital hemangioma is often present at birth and may become more apparent throughout life(37). They are probably developmental rather than neoplastic in origin. Despite their benign origin and behavior, hemangiomas in the oral cavity are always of clinical importance to the dental profession and require appropriate clinical management(38). Surgery and other treatment modalities are not always satisfactory and have a higher morbidity, recurrence, and complication rate (39,40).

According to the question "is there a cure for Sturge-Weber syndrome", Most of the dental students answered that there is cure for the syndrome(63.64%) while 36.36% are aware that it is a lifelong condition that cannot be cured.This graph thus shows the unawareness of the students towards this treatment (fig-5).Sturge-Weber is a lifelong condition that can't be cured. However, treatment of symptoms can help prevent complications and improve your child's quality of life (41).

A correlation graph was obtained between the level of study(UG and PG)and where the port wine stains occur.Among full strength (34%) of the PG's and remaining of the UG's, 24% and 20% of the PG and UG respectively are aware that it is a birthmark on the face.While most of them misled where it occurs.Providing PGs have better awareness than UGs(fig-6).SWS is a neurological disorder marked by a distinctive port-wine stain on the forehead, scalp, or around the eye (42)**.**

A correlation graph was obtained like the above figure to the question How Intra-Orally SWS is characterised,full strength of PG(34%) and 38%of the UGs are aware that it is characterised by ipsilateral gingival haemangioma, while 27%of the UGs misled that it is stains on the tooth .Providing PGs have better awareness than UGs (fig-7).The most common oral manifestations in SWS are mainly seen in the unilateral side of the gingiva and finish abruptly at the midline. Angiomatous gingival lesions range from slight vascular hyperplasia to severe hemangiomatous proliferation due to abnormal increases in the vascular component. As a result of these changes, gingiva bleed on slight provocation or minor trauma (43)

CONCLUSION:
Within the limits of study,PG respondents have greater awareness than UG respondents. Among all, results of this survey shows that half

of them are aware about the Sturge-Weber Syndrome while others are not.In SWS the challenge for the oral physician is to be aware of its other manifestations and institute treatment and referral accordingly. The key in the management of SWS is to prevent or reduce the intensity of complications as the underlying pathology cannot be treated. Early identification and early institution of treatment is imperative for the patient to have a better quality of life.

ACKNOWLEDGEMENTS:
Saveetha Dental College and Hospital,Saveetha Institute of Medical and Technical Science,Saveetha University.

CONFLICT OF INTEREST:
None declared.

SOURCE OF FUNDING:
The present study was supported by the following agencies

- Saveetha Dental College,
- Saveetha Institute of Medical and Technical Science,
- Saveetha University
- G.A.S electricals private LTD.

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