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Awareness On Silver Russell Syndrome Among the Dental Students

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Abstract

BACKGROUND : Silver Russell Syndrome was first described by silver 1n 1953 and Russell in 1954. Silver Russell Syndrome is a rare disorder characterised by intrauterine growth restriction (IUGR), poor growth after birth, a relatively large head size, a triangular facial appearance, a prominent forehead, body asymmetry and difficulty in significant feeding. Abnormalities involving chromosome 7 or 11 have been found in 60% of the patients.

AIM : The aim of this study is to investigate the awareness of silver russell syndrome among the dental students.

MATERIALS AND METHODS : A questionnaire containing some 20 questions was created using google forms and it is distributed to the dental students with the help of whatsapp. The results obtained are statistically analysed using SPSS Software. The Chi square test was done and Pearson value is noted. Graphs were plotted accordingly.

RESULTS : The results were analysed and it showed that the awareness of silver russell syndrome was very very less as it is a rare disorder.

CONCLUSION : From the obtained results, it has been concluded that dental students are not that much more aware about silver russell syndrome and more knowledge should be required. And it was very helpful to diagnose this syndrome in future without clarification.

INTRODUCTION :

Silver–Russell syndrome is a rare genetic disorder that is characterised by intrauterine or postnatal growth restriction and typical facies. The clinical picture is extremely diverse due to some numerous diagnostic features reflecting a heterogeneous genetic disorder (1). It is characterised by a preserved head circumference, that leads to a lean body habitus, short stature, Facial dysmorphism and asymmetry are considered as a typical features of the silver russell syndrome, although the range of phenotypic variance is unknown (2) intrauterine growth retardation, postnatal growth failure, relative macrocephaly, prominent forehead, body asymmetry and significant feeding difficulties (3). Silver Russell Syndrome is clinically and genetically heterogeneous, and several chromosomal abnormalities that involve chromosomes 7, 8, 15, 17 and 18 have been associated with this syndrome and SRS-like cases. It has been found that only chromosomes 7 and 17 have been consistently implicated in individuals with a strict diagnosis of Silver Russell Syndrome. Maternal uniparental disomy with respect to chromosome 7 occurs in approximately 10% of SRS cases and these individuals generally have a milder phenotype (4)

Previous studies suggest that the normative data in relation to spontaneous growth of children with Silver-Russell syndrome are identified, allowing a better counselling of patients as well as the judgement of the effects of growth promoting therapies (5). It is also considered a clinically and genetically heterogeneous disorder. The SRS was independently described by Silver in 1953 who explained in detail about the short stature and a condition called congenital hemihypertrophy of the affected children and by Russell in 1954 who focused his report on the condition called intrauterine dwarfism and also other condition called the cranio-facial dysostosis associated with this syndrome (6). All the scoring systems for Silver Russell Syndrome have been developed and validated in paediatric cohorts. Recent research explained that a clinical diagnosis is frequently challenged by lack of early growth data. An attempt should be made to obtain photographs of the children, especially of the facial profile, as well as measurements at birth and in the first 2 years and frequently. No current evidence exists to support an alternative approach to diagnosis of Silver Russell Syndrome in adults (7). Silver Russell Syndrome usually leads to a wide spectrum of abnormal physical characteristics and functional abnormalities. Multidisciplinary follow up and early, specific, intervention are necessary for optimum management of this group of patients both adults and children (8).

The short stature of children, which is one of the characteristics of the Silver Russell syndrome is unlikely to make any significant independent contribution to the presence of cognitive impairments. Where short stature syndromes are associated with specific cognitive deficits, as in some cases of hypopituitarism or the Turner's syndrome this is usually due to the underlying pathology rather than the growth disorder itself (9). The influence of genetic factors in the etiology of Silver Russell Syndrome is highly documented by the classical genetic findings such as familial cases of Silver Russell Syndrome and cytogenetic aberrations. Certain genetic and epigenetic disturbances can meanwhile be detected in approximately 50% of patients with typical features of silver russell syndrome (10). In previous study, it has been concluded that although some of the sporadic occurrences and genetic heterogeneity that are involved in the silver russell syndrome, dominant inheritance may be considered as a major causal factor and is explained that dominant transmission cannot be determined until male-to-male transmission is reported or until a specific diagnostic marker for the Silver Russell Syndrome is discovered (11). Some infants or children with Silver Russell Syndrome experience significantly greater problems with feeding than children without growth problems. Children with SRS had significantly more problems in almost every domain. Children with SRS appear to experience several feeding problems that are indicative of OMD, including problems with chewing unfamiliar textures and being slow to eat (12). It is said that the growth pattern in Silver-Russell syndrome is quite homogeneous, and rather accurate predictions are possible, secondly Intersexual genitalia do not seem to be related to

endocrine factors, and thirdly hypergonadotropic hypogonadism appears to be frequent in males (13).Our team has extensive knowledge and research experience that has translate into high quality publications (14–22),(23–28),(29–35)

MATERIALS AND METHODS :

The cross sectional questionnaire study was conducted among the dental students from March 2021 to May 2021. The survey focused on dental students' awareness on the facts about Silver Russel syndrome . A Self structured questionnaire was prepared and has been typed in google forms and distributed to 100 dental students. The 20 questions are based on facts of SR syndrome .The Collected data were verified and imported to SPSS (Statistical Package for the Social Sciences, Chicago,USA) .In SPSS, the individual frequency distributions of each category were assessed and bar charts were obtained. Descriptive statistics and Chi-square-test were used to compare the results.

RESULTS :

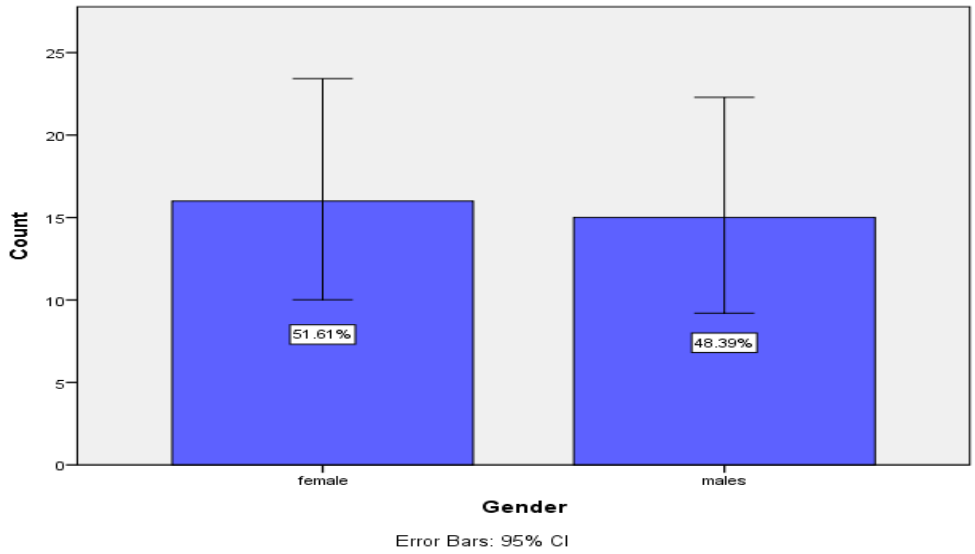

Fig 1 : The pie chart depicts the frequency distribution of gender of the participants. According to the chart 51.16% of the participants were female (Green) and 48.39% of the participants were male (Blue).

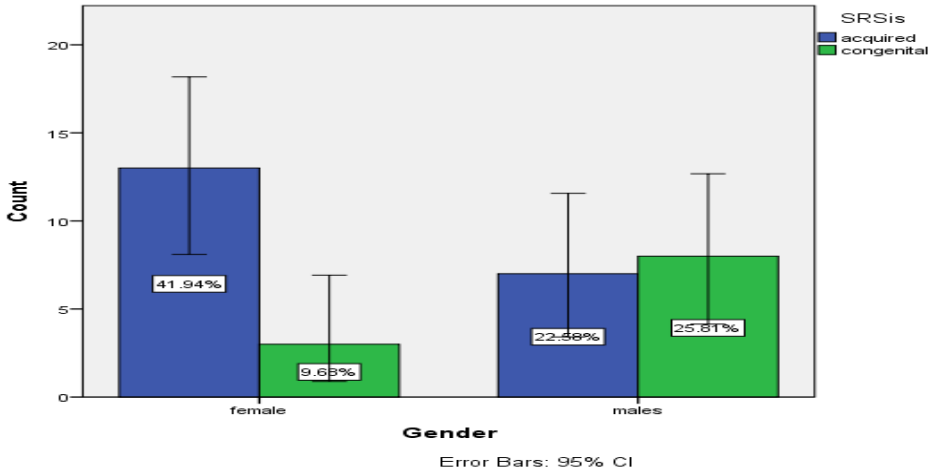

Fig 2 depicts the bar chart comparison between the gender of the participants and the characteristic of silver russell syndrome. It is seen that most of the male participants (25.81%) and some female participants (9.68%) chose that this syndrome is congenital (Green) and most of the females (41.94%) and males (22.58%) chose it as acquired (Blue).

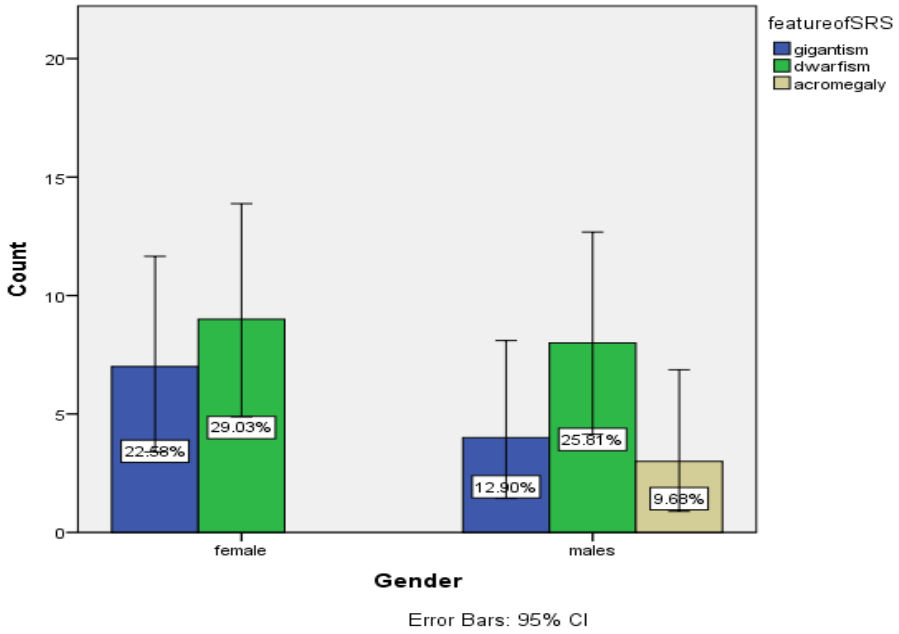

Fig 3 depicts the relationship between gender of the participants and the features of SRS. In that, the majority of the males (25.81%) and females (29.03%) chose that dwarfism (Green) is the common feature of Silver Russell syndrome. Some of the males (12.90%) and females (22.58%) chose gigantism (Blue) as a feature of SRS. And some of the males (9.68%) chose acromegaly (Grey) as a feature of SRS.

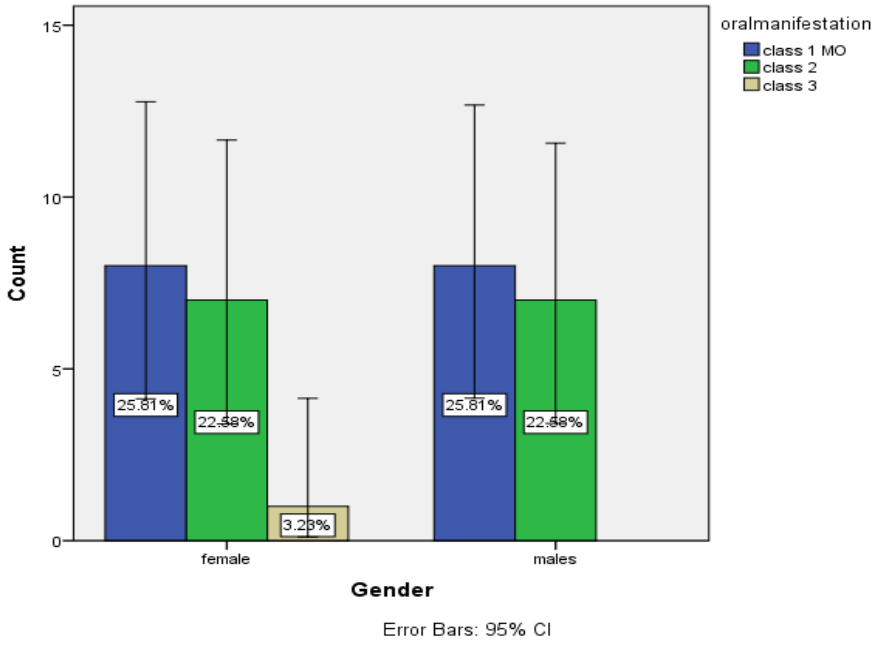

Fig 4 depicts the relationship between the gender of the participants and the oral manifestation of Silver Russell Syndrome. It is seen that the majority of the males (22.58%) and females (22.58%) have chosen class 2 (Green) as the oral manifestation of SRS. Some of the males (25.81%) and females (25.81%) have chosen class 1 MO (Blue) as an oral manifestation of SRS. And about 3.23% of the females have chosen class 3 as an oral manifestation of SRS.

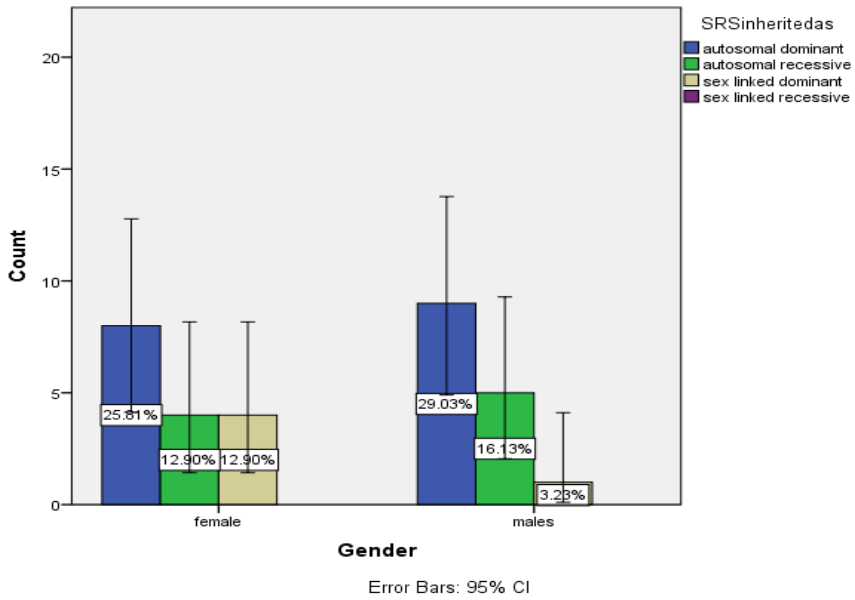

Fig 5 depicts the relationship between the gender of the participants and the inheritance of SRS. It is seen that majority of the males (29.03%) and females (25.81%) have chosen it as autosomal dominant (Blue), About 16.13% of males and 12.90% of the females have chosen it as autosomal recessive (Green), About 12.90% of males and 3.23% of males have chosen it as sex linked dominant (Grey).

DISCUSSION :

A total of 100 dental students participated in the survey, 56 were males and 44 were females (figure 1). From the results, we observe that most of the people were unaware of the syndrome called Silver Russell syndrome. The genetic changes of the syndrome is characterised by the chromosome 7 and 11 and majority of the participants judged correctly and some of the assumptions are wrong. This somewhat shows that there is very less awareness about silver russell syndrome among dental students.

According to figure 2, SRS is an imprinting disorder with congenital growth retardation, relative macrocephaly, body asymmetry, prominent forehead, low BMI and severe postnatal growth failure resembling part of the features of most of the syndrome. Common epigenetic causes of SRS are hypomethylation of the IC1 on 11p15 and maternal uniparental disomy of chromosome 7. In some individuals with SRS, different alterations of the 14q32 imprinted region were detected. Some of these changes were previously associated with Temple syndrome (36) . The main differentiating feature of silver russell syndrome has been body asymmetry, most commonly observed in the Silver dwarf, but occasionally present in the Russell variant.

Previous article concluded that over 70 cases have been reported in that study, very little is known about the genetics and the dermatoglyphics in this disease entity, and the structural abnormalities accounting for the craniofacial dysostosis commonly seen in this syndrome have not been described (37). One of the other studies proved and stated that small forehead, small mandible, skeletal class 2 and a dental phenotype, leading to a special orthopedic and maxillofacial management. Some of the craniofacial features include frontal bossing, reduced vertical facial proportions, triangular face with a narrow and small mandible with short ramus and down-turned corners of the mouth (sharks mouth), low-set posteriorly rotated ears. The dental manifestations include microdontia, high-arched palate and severe crowding due to small-sized jaws, mandible, in particular (38).

Management of SRS requires a multidisciplinary approach with paediatric subspecialists such as an orthodontists, endocrinologist, gastroenterologist, dietician, clinical geneticist,

craniofacial team, orthopaedic surgeon, neurologist, speech and language therapist and psychologist. Early feeding and nutritional support is very important to address low birth weight, low muscle mass and poor appetite. Monitoring of levels of urinary ketones is usually effective in pre-empting hypoglycemia related to fasting, activity or illness (39).

CONCLUSION :

From the obtained results, it has been concluded that dental students are not that much more aware about silver russell syndrome and more knowledge should be required. And it was very helpful to diagnose this syndrome in future without clarification. Furthermore studies are welcomed as this syndrome has wider characteristics and studies can relate with some other disorders..

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CONFLICT OF INTEREST :

The authors declare that there is no potential conflict of interest.

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