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Prevalence of dental caries in children with Silver-Russell syndrome

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Abstract

Silver-Russell syndrome (SRS) is a rare hereditary disease that manifests itself in fetal growth retardation during pregnancy, delayed growth after birth, a relatively large head relative to the body, a triangular face shape, a prominent forehead (when viewed in profile), body asymmetry and significant feeding difficulties at a young age. Children with Silver-Russell syndrome have smaller linear facial dimensions and abnormalities in facial proportions, such as a small, retropositioned, and steeply inclined maxilla and mandible, and proportionally greater anterior facial height relative to posterior facial height. The aim of this article is to examine the prevalence of caries lesions in children with Silver-Russell syndrome. The subject of the study were 60 children with Silver-Russell syndrome. The children were divided into three groups, according to age (<6years, 6-12 years, 12-18 years). The prevalence of caries was measured using the dft/dmf (t+T)/ DMFT indices. The prevalence of caries lesions for the children with Silver-Russell syndrome is 4,55±1,86. There is a significant prevalence of D1 and D2 caries lesions in the age group above 12 years old. (p=0.001) For the age group <6 years there is significance in the prevalence of D4 caries lesions. (p=0.007). Children with SRS have a higher prevalence of dental caries as a result from different predisposing factors, which include excessive crowding due to micrognatia, frequent carbohydrate intake due to hypoglycemia. Another reason for increased dental caries prevalence is the deficiency of vitamin D3, which is observed in children with growth hormone deficiency.

Keywords: caries, children, Silver-Russell syndrome

Introduction

Silver-Russell syndrome (SRS) is a rare hereditary disease that manifests itself in fetal growth retardation during pregnancy, delayed growth after birth, a relatively large head relative to the body, a triangular face shape, a prominent forehead (when viewed in profile), body asymmetry and significant feeding difficulties at a young age. (1,2) The cause of the syndrome is still unknown, but is reported to be heterogeneous. (3)

SRS syndrome is a rare disease without a pathognomonic feature. (4,5,6) Worldwide, the incidence of SRS ranges from 1:30,000 to 1:100,000. (7) A variety of dental defects are observed—absence of teeth, small teeth (microdontia), overlapping teeth. Children with Silver-Russell syndrome have smaller linear facial dimensions and abnormalities in facial proportions, such as a small, retropositioned, and steeply inclined maxilla and mandible, and proportionally greater anterior facial height relative to posterior facial height.(8) The development of the teeth is within normal limits, and the periods for the eruption of the teeth are slightly delayed. (9)

Aim

The aim of this article is to examine the prevalence of caries lesions in children with Silver-Russell syndrome

Material And Methods

The subject of the study were 60 children with Silver-Russell syndrome. The children were divided into three groups, according to age (<6years, 6-12 years, 12-18 years). The prevalence of caries was measured using the dft/dmf (t+T)/ DMFT indices. The DMFT index consists of decayed (D), missing (M) and filled (F) teeth. The diagnostic limit used is D1a (non-cavitated enamel lesions). D2, D3 and D4 lesions were also registered. The dental assessment was carried out according to the World Health Organisation (WHO) criteria for epidemiological trials.(10) Observation is done with a dental mirror only, and a probe is not used to determine the depth of the defects and to check the roughness or texture of any of the observed lesions, as irreversible damage may be done to the already demineralized enamel or with the use of a periodontal probe with an atraumatic tip.

D1 – clinically observed non-cavitated enamel lesion
D2 - clinically observed cavitated enamel lesion
D3 - clinically observed dentin lesion
D4 - dentin lesion, with pulp involvement

For every child there was full documentation created to register the data, which allows observation of the caries process and its prevention and treatment. All of the children were examined in a dental clinic with individual dental sets. The registered information was filled in a detailed ambulatory card. Dietary habits and oral hygiene habits were also registered on the card. The parents of the children have signed a declaration of informed consent and a questionnaire, regarding the health status of the children.

Results

The main index dmft/ dmf(T+t)/ DMFT for the children is 4,55±1,86. The prevalence of caries lesions for the children with Silver-Russell syndrome divided into groups is as follows: For the group <6 years for D1 lesions the results are 0,73±0,69, for D2 lesions - 0,44±0,48, for D3 - 2,41±1,47 and for D4 - 0,73±0,6. For the missing teeth we have M- 0,18±0,51 and for the filled F - 0,74±0,88. The caries lesions D3 are most commonly observed in this group, compared to the other two groups.

For the second age group (6-12 years) the results are as follows: D1 - 0,34±0,56; D2 - 0,28±0,44; D3 - 1,78±1,09; D4 - 0,33±0,35; M - 0,23±0,39 and F - 0,93±0,55.

For the third age group we have D1 - 1,19±0,63; D2 - 1,31±1,58; D3 - 1,69±0,89; D4 - 0,32±0,31, M - 0,08±0,17 and F - 1,15±1,01. The results are statistically significant for the prevalence of D1, D2 and D4 lesions with p<0.05. The results are shown in the following table (Table 1)

Table 1. Mean values of DMFT/dmft index by age groups in children with Silver-Russell syndrome

	< 6 years	6-12 years	>12 years	**p<0.05***
D1/d1	0,73±0,69	0,34±0,56	1,19±0,63	**0,001***
D2/d2	0,44±0,48	0,28±0,44	1,31±1,58	**0,001***
D3/d3	2,41±1,47	1,78±1,09	1,69±0,89	**0,172**
D4/d4	0,73±0,6	0,33±0,35	0,32±0,31	**0,007***
M/m	0,18±0,51	0,23±0,39	0,08±0,17	**0,568**
F/f	0,74±0,88	0,93±0,55	1,15±1,01	**0,376**

According to these results, there is a significant prevalence of D1 and D2 caries lesions in the age group above 12 years old. (p=0.001) For the age group <6 years there is significance in the prevalence of D4 caries lesions. (p=0.007)

Discussion

Children with Silver-Russell syndrome often experience feeding difficulties, combined with frequent acid reflux. Reflux of gastric contents may be manifested clinically by arching the child's body backwards and/or vomiting. Reflux can be overcome by more frequent meals with small amounts of food, keeping the child upright after feeding, thus gravity prevents food from flowing back up from the stomach into the esophagus. (11) Because of this more frequent intake of small amounts of food the children tend to have longer intervals of demineralization, which leads to higher caries prevalence. (12) Hypoglycemia is also a common condition for these patients. To avoid episodes of low blood glucose (LBG), children with SRS should never go hungry for a long period of time (even due to performing medical procedures). (13) Children with SRS are prone to develop spontaneous hypoglycaemia especially if they are not fed both frequently and regularly. The most likely explanation is accelerated starvation and/or GH insufficiency. (14) The amount of glucose and the frequency of consumption seem to be a significant contributor to the cariogenicity of the diet and common risk factors for the caries progression for both primary and permanent dentitions. (15) Another reason for increased prevalence of caries is poor oral hygiene. (16) Poor oral hygiene in the presence of dental crowding, which is often observed in these children, can lead to increased risk for dental caries. (17) Overcrowded teeth can make it difficult to brush and floss properly, which leads to easier plaque accumulation and bacteria build up in the mouth. This leads to an increased risk for developing caries lesions. Dental crowding is a result from micrognathia, which is a characteristic for SRS. (18) Orthodontic anomalies are also the reason for severe plaque accumulation, which leads to increased caries frequency. The patient's oral hygiene may be deteriorated due to microphoria. Appropriate dietary control and fluoride application are required. (19) SRS is characterized by craniofacial disproportion, which results in a triangular-shaped face. (20) Delayed dental eruption, microdontia, absence of secondary teeth and blunted

condyles have all been reported in patients with SRS. (4,9) Children with SRS are often treated with growth hormone. (21,22) Growth hormone treatment in children with SRS is recommended for a number of reasons:

- improving body structure, especially to increase muscle mass
- improvement of motor skills and appetite
- reducing the risk of developing hypoglycemia and improving growth.

Vitamin D deficiency is a common risk factor for multifactorial diseases, and it seems to be associated with growth hormone deficiency (GHD). Vitamin D could prevent dental caries. Vitamin D impacts bone and teeth mineralization. With parathormone (PTH) and fibroblast growth factor 23 (FGF 23), vitamin D controls the concentration of calcium and phosphorus in circulating blood and subsequent mineralization of bones. (23) Vitamin D3 is a potentially effective agent, which reduces the number of dental caries, especially among patients with growth hormone deficiency. (24) The increase in vitamin D3 levels reduces the D value by 0.66 per each 10ng/mL of vitamin D3 concentration. Wojcik et al. found a statistically significant correlation between the duration of growth hormone therapy and the DMFT index. Increasing the duration of GH therapy by 10 months resulted in a mean increase in DMFT index of 0.70. (24) A study by Hujoel showed that vitamin D supplementation resulted in a 47% reduction in dental caries. (25) Children with SRS with eating problems often require supplementary dental care, including help with their oral hygiene and fluoride treatment as well as regular check-ups of dental and jaw development. Preventing dental decay is challenging in children with SRS. Promotable oral hygiene is as important as dietary facts which eliminates the biofilm. Moreover, the maxillofacial characteristics of the syndrome such as petite face and small jaw make the oral hygiene unmanageable within the usual methods. There are specific toothbrushes, designed for children with micrognathia – the Collis-Curve toothbrush, which is adapted with several working surfaces, that intend to make brushing easier for these children.(19) The alteration in the development and growth of the face and its jaws implies the presence of malocclusions, crowding and malpositions, plus the type of diet lead to a plaque retention factor and individual risk for the appearance of dental caries in these patients, must be taken into account for the objectives and treatment plan of these patients. Consultation with other specialists in genetics, orthopedics, nutrition, physical therapy, speech therapy and psychology is required.(26) An orthodontist should also be consulted when needed. Feeding and swallowing difficulties are investigated and treated by a specialist team at the hospital or multidisciplinary treatment centre. A speech therapist may provide practical advice regarding feeding, as well as instruction for the stimulation of the mouth muscles. (27,28,29)

Conclusion

Children with SRS have a higher prevalence of dental caries as a result from different predisposing factors, which include excessive crowding due to micrognatia, frequent carbohydrate intake due to hypoglycemia, as well as difficulty in maintaining proper oral hygiene. Another reason for increased dental caries prevalence is the deficiency of vitamin D3, which is observed in children with growth hormone deficiency.

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