SurgiMed PUBLISHERS

Journal of Public Health and Medicine

Volume 1; Issue 1
Case Report

Dental Management of Crisponi/Cold-Induced Sweating Syndrome1 under General Anesthesia: A Case Report

Ahmed I. Ghibban[1,2] and **Jihan M. Turkistani**[1,2*]

[1]Department of Dental Services, King Abdulaziz Medical City, Ministry of National Guard Health Affairs, Jeddah, Kingdom of Saudi Arabia

[2]King Abdullah International Medical Research Center, Riyadh, Kingdom of Saudi Arabia

Received Date: 17 February 2023; **Accepted Date:** 06 March 2023; **Published Date:** 28 March 2023

Correspondence Address: Jihan Turkistani, Department of Dental Services, King Abdulaziz Medical City, Ministry of National Guard Health Affairs, PO Box 9515, Jeddah, 21423, Kingdom of Saudi Arabia.

Abstract

Background: Crisponi/cold-induced sweating syndrome-1 (CS/CISS1) is an infrequent and severe autosomal recessive syndrome. It is initiated by a mutation in the cytokine receptor-like factor1 (*CRLF1*) gene. This report details the dental management under general anesthesia provided to a 6-year-old male patient at King Abdulaziz Medical City in Jeddah who had several dental cavities and poor oral hygiene.

Case presentation: This is the first documented incident of dental management and treatment of typical CS/CISS1 in a Saudi population. To reduce the risk of malignant hyperthermia, treatment for CS/CISS1 requires pre-operative management, including consultation with the patient's physician and anesthesia team. In addition, precautions concerning the positioning of the dental chair and room temperature should be between 22-25˚C.

Conclusions: In order to direct patients to the proper medical professionals, dentists must be aware of this disorder. Collaboration between various specialties is necessary to enhance the quality of life for patients and comprehend the clinical traits and management of CS/CISS1 cases entirely.

Keywords: Crisponi syndrome; Dental management; Cold-induced sweating; Scoliosis.

Abbreviations: ASA: American Sociological Association; CS/CISS1: Crisponi/Cold-induced sweating syndrome 1; *CRLF1*: Cytokine receptor-like factor1; SSC: Stainless steel crowns; GERD:Gastroesophageal reflux disease; KAIMRC: King Abdullah International Medical Research Center; KAMC-J: King Abdulaziz Medical City-Jeddah; MH: Malignant hyperthermia.

Background

A pattern of several anomalies known as a syndrome is those that are assumed to be pathogenetically related and do not appear to result from a single sequence or polytopic field defect [1]. Deviations in the morphology and development of the teeth and other cranial tissue are signs of craniofacial syndrome [1]. Many syndromes have been linked and related to the

oral features, for example, Apert syndrome, Cleft lip palate syndrome, Down syndrome, Crouzon syndrome, and Fragile X syndrome [2]. Craniofacial anomalies challenge many healthcare professionals since they demand ongoing follow-up. Based on the severity of the medical condition and the dental needs, dental management varies from case to case [3].

One of the common syndromes is Down syndrome, an extra chromosome of the twenty-first group called Trisomy 21 [4]. It is frequently seen with other medical problems such as epilepsy, diabetes, leukemia, hypothyroidism, and other conditions [4]. The dental manifestations include underdevelopment or hypoplasia of the midfacial region, open bite, class III malocclusion, spacing, periodontal diseases, delayed eruption of the permanent teeth, and macroglossia [4]. On the other hand, Crisponi/cold-induced sweating syndrome-1 (CS/CISS1), is a rare and severe autosomal recessive syndrome [5]. It is caused by a mutation in the cytokine receptor-like factor1 (*CRLF1*) gene [5].

In 1996, Giangiorgio Crisponi made the initial discovery of this condition in Italy [6]. The clinical features of this syndrome range from anteverted nostrils, plump cheeks, broad nose, hyperthermia, facial muscle contractions, and muscle spasms [7]. In the neonatal period, Crisponi's patients show facial muscle contractions in response to tactile stimulation and crying mimics a tetanic spasm that disappears after the child becomes calm and relaxed [8]. Moreover, they may experience significant sucking difficulties that cause nutritional deficiencies [9]. Small and skinny mouth and micrognathia are anomalies seen in the orofacial region, in addition to abundant saliva stagnation due to defective swallowing [9].

In late infancy and childhood up to adolescence, CS/CISS1 patients develop scoliosis, sometimes mild psychomotor retardation [10]. The massively elevated plasma noradrenaline levels in early childhood are the cause behind the cold-induced sweating, which is related to the (*CRLF1*) gene that is the key role of the center pathway in the function of the autonomic nervous system [10].

However, there isn't much research or reported cases in Saudi Arabian or Arab world literature about the dental rehabilitation of these patients. One incident was described for treating a nine-year-old female child under local anesthesia on the dental chair [10]. Due to the limited mouth opening and the neck muscles contracting, it was necessary to administer the treatment across several visits [10].

The current report aims to describe the dental considerations and management of a patient diagnosed with CS/CISS1 at King Abdulaziz Medical City-Jeddah (KAMC-J) under General Anesthesia.

Case study

A 6-year-old male patient presented with his father to the Pediatric Dentistry clinics at King Abdulaziz Medical City on November 2021 with a chief complaint stated by his father "My child has pain and multiple carious teeth; I want you to treat his teeth."

Medical history revealed that the patient was diagnosed with CS/CISS1 when he was born. Also, the patient was diagnosed with hypotonia, Gastroesophageal reflux disease (GERD), and bronchial asthma. Apart from his older sister, who has the same condition, his parents and siblings were all in fairly good health. Without any issues, the patient was delivered vaginally at full term. He had an adenotonsillectomy in April 2018. No known drug or food allergies were reported, and his vaccinations were up to date.

Additionally, the patient is following up with ophthalmology and orthopedics for scoliosis and cornea scar.

An extraoral clinical examination of the patient showed scoliosis and a cornea scar. However, the patient had a symmetrical face with an average mandibular plane angle and competent lips (Figure 1). The skin, hair, and temporomandibular joint were within normal limits, with non-palpable lymph nodes.

Figure 1: The patient's craniofacial manifestations include a symmetrical face with an average mandibular plane angle and competent lips.

Intraoral examination showed poor oral hygiene, generalized plaque-induced gingival disease, and physiological pigmentation. No pathology was detected in the labial and buccal mucosa, soft and hard palate, tongue, and floor of the mouth. The patient was in the early mixed dentition stage with multiple carious teeth. Occlusion examination showed class I molars classification, Class I primary canines classification, 2 mm overjet, and 20% overbite (Figure 2). Radiographically, the panoramic radiograph showed that the dental age of the patient coincides with the chronological age; two bitewings radiographs showed multiple interproximal and occlusal caries (Figure 3).

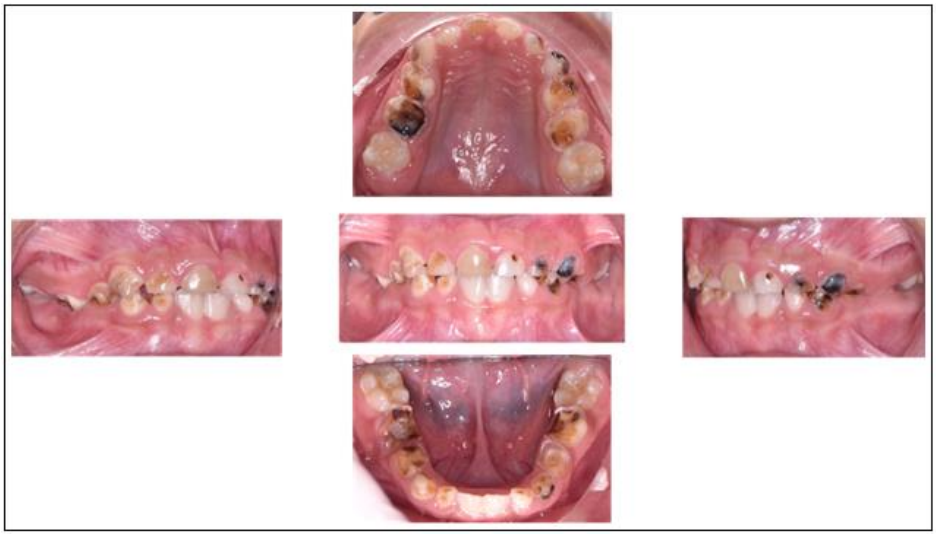

Figure 2: Intraoral examination shows early mixed dentition stage with Multiple carious teeth.

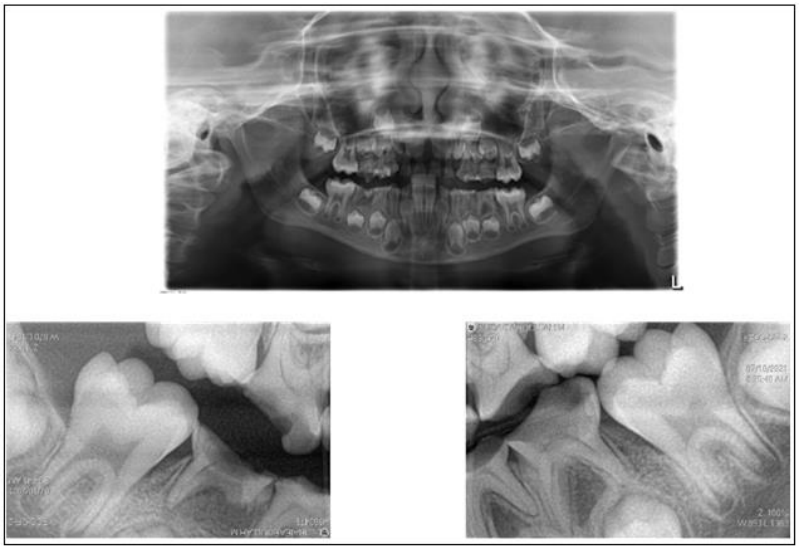

Figure 3: Panoramic and bitewings radiographs show multiple carious teeth; dental age coincides with chronological age.

The patient was seen by a general dentist once when he was 5 years old, but he was uncooperative. However, this was his first time seeing a pediatric dentist, so this experience was unique. Despite having normal amounts of growth hormone, he continuously fell below the 5th percentile on a relevant growth chart when examined. He was hesitant to undergo treatment, and it was clear that he had a negative attitude. He could not tolerate an extended period on the dental chair, starting to sweat after a couple of minutes at 22-25˚C room temperature.

When the patient was three years old, his mother mentioned that he fell to the ground and injured his upper right primary central incisor. At that time, neither a dentist visit, nor a treatment was given. Over time, the only color change was observed without discomfort or pain.

Phases of the treatment plan were developed, beginning with a consultation with the General Pediatric physician to confirm that the proposed dental treatment plan is safe for him and to offer the appropriate measures before doing any necessary dental work. The physician from the pre-anesthesia clinic gave the clearance for dental work to be carried out under general anesthesia, classifying his physical condition as ASA II according to the American Sociological Association (ASA) Physical Status Classification System. Informed consent was acquired from the patient's parents, who approved of the proposed treatment plan and accepted it. The parents received detailed dental hygiene guidelines as well as diet counseling.

Careful preoperative and postoperative monitoring of numerous parameters was performed on the patient. We also considered utilizing lubricant, a small intubation equipment, and gentle patient manipulation. Multiple restorations, pulp therapy, stainless steel crowns (SSC), and extractions were done (Figure 4 and 5).

The patient was discharged with a stable condition on the same day of therapy after being closely observed. The patient was then instructed to have routine follow-ups beginning two weeks after the general anesthesia.

The ethical research committee at King Abdullah International Medical Research Center (KAIMRC) approved to conduct of this case report (#NRJ22J/268/10). Written, informed consent was acquired from the parents.

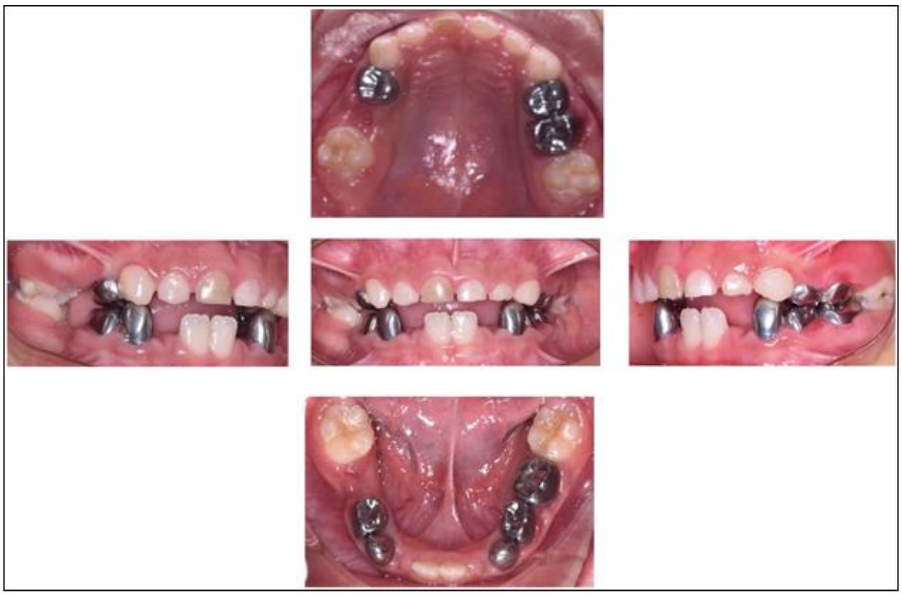

Figure 4: Post-restorative treatment under General Anesthesia.

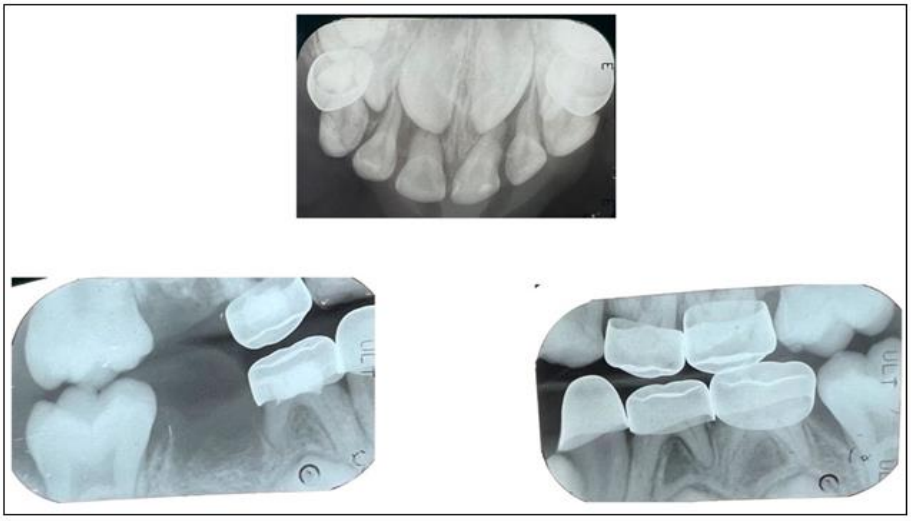

Figure 5: Post-radiographic treatment under General Anesthesia.

Discussion

Studies on children with CS/CISS1 regarding the condition of their oral cavity are lacking. In light of this, the present case report describes the oral presentation and dental treatment of a young patient with CS/CISS1.

Due to the sporadic cases diagnosed with this syndrome worldwide, there is a lack of literature regarding anesthetic management. Hence, specific measures were taken before administering anesthesia to individuals susceptible to malignant hyperthermia (MH). During the treatment of our patient, the

Anesthesia team aimed to maintain the room temperature, avoid the use of triggering or volatile agents, and monitor expired CO_2 levels, in addition to the immediate availability of dantrolene to avoid any complication. As a further precaution against complications, they advised us to reduce the procedure time. According to studies, operating under general anesthesia for longer periods of time considerably raised the risk of complications; at 2 hours or more, the risk of challenges approximately doubled [11]. Practitioners should be aware that the younger the patient, the more serious the complications [9].

Children can respond adversely to unfavorable conditions such as hypoxia, hypothermia, or fluid imbalance. Therefore, in order to safely provide general anesthesia to those children, a thorough understanding of pediatric physiology is required [12].

Early MH symptoms include tachycardia, increased expired CO_2, generalized rigidity after succinylcholine administration, along with skin discoloration, cyanosis, and excessive sweating. These clinical indicators taken together are sufficient to support the diagnosis of MH and the start of MH therapy immediately. The ultimate treatment is dantrolene sodium, a nonspecific muscle relaxant [13,14].

The patient presented in this report was diagnosed with CS/CISS1 since birth. Orally, he had poor oral hygiene and significant teeth decay because of inadequate diet and GERD, which increased acid production in the mouth and accelerated tooth loss. Studies found increased evidence of associations between GERD and tooth erosion [15-17]. Gastric acid will displace saliva easily from tooth surfaces, and proteolytic pepsin will remove protective dental pellicles [18]. Repeated or prolonged exposure of teeth to acid leads to the selective dissolution of specific components of the tooth surface, with eventual loss of tooth substance [19]. Therefore, covering these teeth with SSCs was the treatment of the choice to enhance longevity and maintain oral funcation [19]. A systematic literature review showed SSCs demonstrated excellent clinical performance as restoratives for posterior primary teeth [20]. Moreover, it was revealed that SSCs might increase salivary nickel and chromium concentrations and reduce saliva acidity [21].

According to the dental history of our patient, the upper right primary central incisor was traumatized two years ago. The tooth was left for follow-up because no symptoms or indicators called for immediate treatment [22]. Dark coronal discoloration in primary incisors after traumatic injury is a sign of pathology or pulp necrosis. However, this does not require removal of the pulp unless clear clinical evidence of infection of the pulp is present [23].

The patient was discharged home with oral hygiene instructions, including brushing his teeth twice a day, using dental floss, and maintaining follow-up appointments for future space management after the extraction of some primary teeth. Space maintenance is essential in premature loss of primary dentition to prevent malposition, supra-eruption, impaction, or crowding of the developing permanent teeth [24].

To achieve the best possible treatment outcomes, it is crucial to take a complete medical history, communicate with the patient's physician, guide the child, and reinforce good oral hygiene. Various studies found that special healthcare needs patients with irregular checkup visits were approximately four times more likely to undergo repeated general anesthesia than those with regular checkups and following oral hygiene instructions [25-27]. Additional studies are desirable to determine various treatment options for CS/CISS1 and other dental recommendations.

Conclusion

Pre-operative management is essential for treating CS/CISS1, and it should include consultations with the patient's physician and the anesthesia team to reduce the risk of malignant hyperthermia.

Ethical committee approval

The ethical approval for this case report was obtained from the Institutional Review Board of King Abdullah International Medical Research Center with IRB No. IRB/2581/22.

Availability of data and materials

The datasets used during the current case report are available from the corresponding author on reasonable request.

Competing interests

The authors declare that they have no competing financial, professional, or personal interests that might have influenced the performance or presentation of the work described in this case report.

Funding

The case report did not receive any funding from any sources.

Authors' contributions

JT has a substantial contribution to the Conceptualization, Data Curation, Writing-Review & Editing, Visualization, Supervision and Project administration.

AG has a substantial contribution to the Investigation, Data Curation, Writing-Original Draft.

All authors read and approved the final manuscript.

Acknowledgements

The authors would like to express their deep appreciation and thanks to the dental department and medical records staff for their cooperation and assistance during the case report preparation.

References

1. Kjær I. Dental Approach to Craniofacial Syndromes: How can Developmental Fields Show us a New Way to Understand Pathogenesis? Int J Dent. 2012.
2. Bartzela TN, Carels C, Maltha JC. Update on 13 Syndromes Affecting Craniofacial and Dental Structures. Front Physiol. 2017;8:1038.
3. Buers I, Persico I, Schöning L, Nitschke Y, Di Rocco M, Loi A, et al. Crisponi/Cold-Induced Sweating Syndrome: Differential Diagnosis, Pathogenesis and Treatment Concepts. Clin Genet. 2020;97(1):209-21.
4. Reeves J. Dental Care for the Patient with Down's Syndrome. Dental Nursing. 2006;2(10):490-2.
5. Crisponi G. Autosomal Recessive Disorder with Muscle Contractions Resembling Neonatal Tetanus, Characteristic Face, Camptodactyly, Hyperthermia, and Sudden Death: A New Syndrome? Am J Med Genet. 1996;62(4):365-71.
6. El-Assy OS, Al-Sulaimani AA, Mujahed AA. Crisponi Syndrome: A New Mutation in a Saudi Family.
7. Accorsi P, Giordano L, Faravelli F. Crisponi syndrome: report of a further patient. Am J Med Genet A. 2003;123(2):183-5.
8. Crisponi L, Crisponi G, Meloni A, Toliat MR, Nürnberg G, Usala G, et al. Crisponi Syndrome is caused by Mutations in the CRLF1 Gene and is Allelic to Cold-Induced Sweating Syndrome Type 1. Am J Hum Genet. 2007;80(5):971-81.

9. Allary C, Caruselli M, Fabre A, Audic F, Michel F. General Anesthesia for Crisponi Syndrome: Case Report. Braz J Anesthesiol. 2020;70:299-301.
10. Akleyin E, Özata MY, Crisponi G. Dental Management in a Patient with Crisponi/Cold-Induced Sweating Syndrome Type 1: A Case Report. Clin Dysmorphol. 2022;31(3):136-40.
11. Cheng H, Clymer JW, Chen BP, Sadeghirad B, Ferko NC, Cameron CG, et al. Prolonged Operative Duration is Associated with Complications: A Systematic Review and Meta-Analysis. J Surg Res. 2018;229:134-44.
12. Messieha Z. Risks of General Anesthesia for the Special Needs Dental Patient. Spec Care Dentist. 2009;29(1):21-5.
13. Krause T, Gerbershagen MU, Fiege M, Weisshorn R, Wappler F. Dantrolene-A Review of Its Pharmacology, Therapeutic Use and New Developments. Anaesthesia. 2004;59(4):364-73.
14. Abraham R Ben, Adnet P, Glauber V, Perel A. Classic Diseases Revisited Malignant Hyperthermia. Postgrad Med J.1998; 74(867):11-7.
15. Dundar A, Sengun A. Dental Approach to Erosive Tooth Wear in Gastroesophageal Reflux Disease. Afr Health Sci. 2014;14(2):481-6.
16. Yoshikawa H, Furuta K, Ueno M, Egawa M, Yoshino A, Kondo S, et al. Oral Symptoms Including Dental Erosion in Gastroesophageal Reflux Disease are Associated with Decreased Salivary Flow Volume and Swallowing Function. J Gastroenterol. 2012;47:412-20.
17. Wang GR, Zhang H, Wang ZG, Jiang GS, Guo CH. Relationship Between Dental Erosion and Respiratory Symptoms in Patients with Gastro-Oesophageal Reflux Disease. J Dent. 2010;38(11):892-8.
18. Watanabe M, Nakatani E, Yoshikawa H, Kanno T, Nariai Y, Yoshino A, et al. Oral Soft Tissue Disorders are Associated with Gastroesophageal Reflux Disease: Retrospective Study. BMC Gastroenterol. 2017;17(1):92.
19. Ranjitkar S, Kaidonis JA, Smales RJ. Gastroesophageal Reflux Disease and Tooth Erosion. Int J Dent. 2012.
20. Seale NS, Randall R. The Use of Stainless-Steel Crowns: A Systematic Literature Review. Pediatr Dent. 2015;37(2):145-60.
21. Basir L, Meshki R, Behbudi A, Rakhshan V. Effects of Restoring the Primary Dentition with Stainless-Steel Crowns on Children's Salivary Nickel and Chromium Levels, and the Associations with Saliva Ph: A Preliminary Before-After Clinical Trial. Biol Trace Elem Res. 2019;187:65-73.
22. Malmgren B, Andreasen JO, Flores MT, Robertson A, DiAngelis AJ, Andersson L, et al. International Association of Dental Traumatology Guidelines for the Management of Traumatic Dental Injuries: 3. Injuries in the Primary Dentition. Dent Traumatol. 2012;28(3):174-82.
23. Holan G. Pulp Aspects of Traumatic Dental Injuries in Primary Incisors: Dark Coronal Discoloration. J Endod. 2019;45(12):S49-51.
24. Albati M, Showlag R, Akili A, Hanafiyyah H, AlNashri H, Aladwani W, et al. Space Maintainers Application, Indication and Complications. Int J Community Med Public Health. 2018;5(11):4970-4.
25. Oh TJ, Nam OH, Kim MS, Choi SC, Lee HS. Oral Health of Patients with Special Health Care Needs after General Anesthesia: A 25-Year Retrospective Study. Pediatr Dent. 2018;40(3):215-9.
26. Kim J, Nam O, Kim M, Lee H, Choi S. Comparison of Dental Outcomes After General Anesthesia According to the Follow-Up Pattern in Special Health Care Needs Patients. J Korean Acad Pediatr Dent. 2016;43(2):145-50.
27. Solanki N, Kumar A, Awasthi N, Kundu A, Mathur S, Bidhumadhav S. Assessment of Oral Status In Pediatric Patients with Special Health Care Needs Receiving Dental Rehabilitation Procedures Under General Anesthesia: A Retrospective Analysis. J Contemp Dent Pract. 2016;17(6):476-9.