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(CASE REPORT)

Full mouth rehabilitation of a pediatric patient with Sturge-weber syndrome under general Anaesthesia: A case report

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Abstract

Sturge-Weber syndrome (SWS) or encephalotrigeminal angiomatosis is a neurocutaneous typified by angiomas that affect the face, choroid, and leptomeninges. It is estimated to affect 1 in 20,000 to 50,000 live newborns. Patients most frequently have ipsilateral parieto-occipital leptomeningeal angiomatosis and calcifications with cortical atrophy, ipsilateral glaucoma, and a unilateral facial hyperpigmented lesion known as the port-wine stain (PWS). About 40% of SWS patients have an oral vascular abnormality. The buccal mucosa, palate, tongue, floor of the mouth, gingiva, and lips may all exhibit purplish-red staining as a result of intraoral angiomatosis. A 9-year-old boy child with a known diagnosis of SWS reported with a chief complaint of decayed teeth, requiring full mouth rehabilitation. As the patient showed minimal response to verbal commands with apprehension and inability to cooperate on a dental chair, it was decided to do full mouth rehabilitation under general anaesthesia. A thorough understanding of SWS is essential due to the significant frequency of oral symptoms of this uncommon congenital condition. This case report added to the knowledge and understanding of full mouth rehabilitation of a child patient with SWS under GA.

Keywords: Sturge Weber Syndrome; Special Child; Full Mouth Rehabilitation; Port Wine Stain

1. Introduction

The neurocutaneous condition known as Sturge-Weber syndrome (SWS), or encephalotrigeminal angiomatosis, is typified by angiomas that affect the face, choroid, and leptomeninges. After neurofibromatosis and tuberous sclerosis, it is the third most prevalent neurocutaneous syndrome.[1] It is estimated to affect 1 in 20,000 to 50,000 live newborns.[2] Multi-systemic symptoms can co-occur including a range of dermatologic, ophthalmologic, neuropsychiatric, and endocrinologic symptoms. The sporadic, somatic mosaic pathogenic R183Q mutation in the GNAQ gene on chromosome 9 is responsible for 80–90% of SWS patients.[3] Patients most frequently have ipsilateral parieto-occipital leptomeningeal angiomatosis and calcifications with cortical atrophy, ipsilateral glaucoma, and a unilateral facial hyperpigmented lesion known as the port-wine stain (PWS).[4]

About 40% of SWS patients have an oral vascular abnormality. The buccal mucosa, palate, tongue, floor of the mouth, gingiva, and lips may all exhibit purplish-red staining as a result of intraoral angiomatosis. Osseous and soft tissue hypertrophy may be seen in the various vascular malformations of the soft tissue. A few cases of SWS have been reported with associated osseous hypertrophy leading to facial asymmetry and malignment of teeth. osteohypertrophy is also seen in cutaneous capillary angioma involving the maxillofacial distribution. [5] Massive gingival growths or mild vascular enlargement are examples of gingival lesions. The acknowledged risks of intraoperative and postoperative haemorrhage makes routine dental and oral surgical treatments appear complex. A thorough understanding of SWS is essential due to the significant frequency of oral symptoms of this uncommon congenital condition.[6]

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2. Case report

Anirudh, a 9-year-old boy with a known medical history of SWS and Cerebral Palsy with 70% permanent disability involving his brain reported to the outpatient wing of Department of Pediatric and Preventive Dentistry, Kottayam with a chief complaint of decayed teeth. The patient was referred to Government Dental College Kottayam from a private dental clinic. Mother reported that the child had pain and swelling in the lower right back tooth region 1 week back and the child had showed extreme aggression, incessant crying and biting in the arm. Mother also reported that the child shows all these behaviors when he is in pain or in extreme discomfort. They had consulted a nearby private clinic and antibiotics were prescribed following which the pain and swelling had subsided.

Only child of a non-consanguineous couple, Anirudh was born at term via caesarean section with a birth weight of 3.4 kg and there was no delay in first cry. Postnatally he was noted to have PWS over right side of the face. There was no history of neonatal seizure or neonatal jaundice or hypoglycemic episode or respiratory distress with no other significant ante natal, natal or post-natal adverse events.

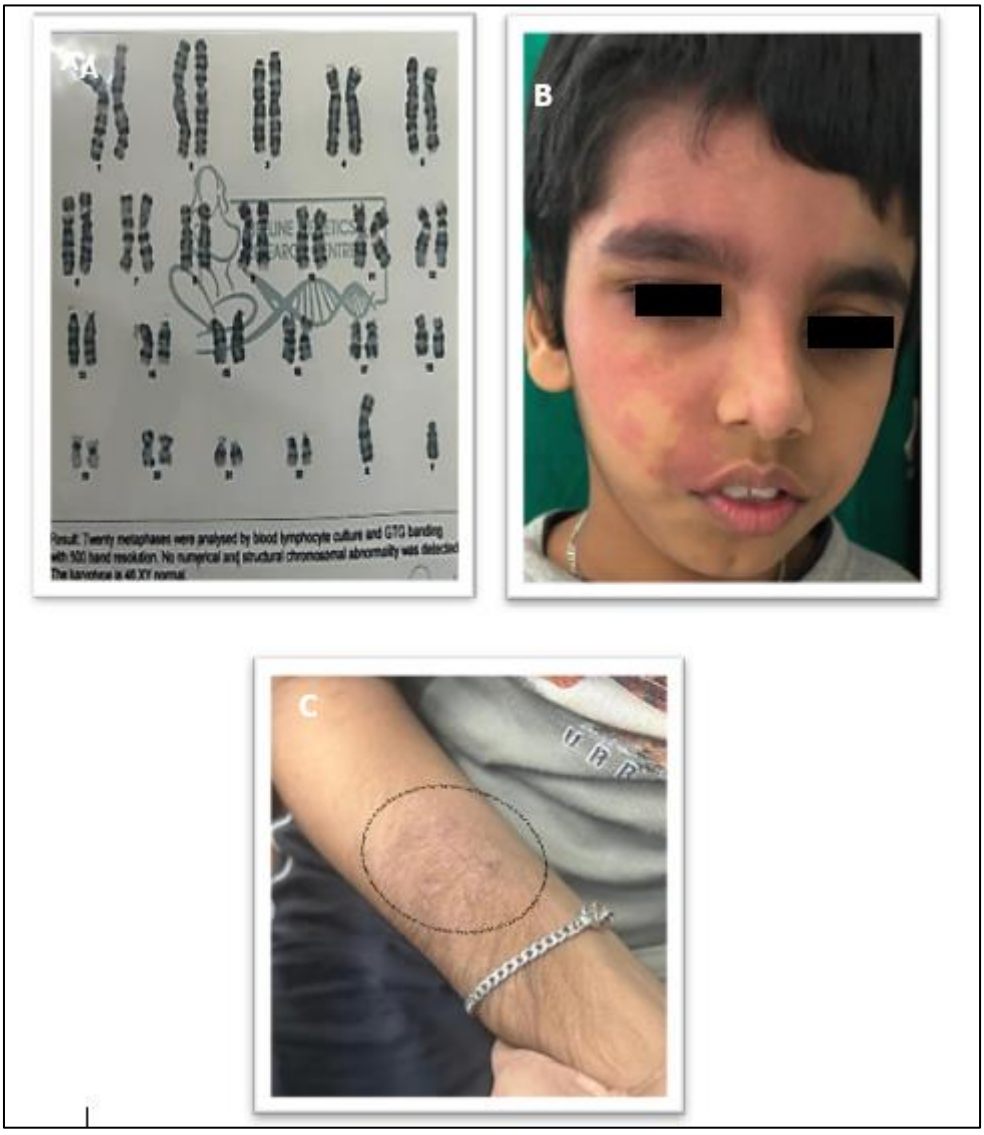

Figure 1 A: Karyotyping showing no numerical & structural chromosomal abnormalities. B: Port wine stain on right side of the face and head. C: Scar of size 3x2 cm on wrist of right hand due to biting, showing self-mutilation behaviour

There was a delay in attaining milestones of development with neck holding attained only after first birthday. Rolling over was started after 1&1/2 years and sitting without support at 2 years. The child had poor eye contact with no verbal communication. Family history revealed that the child's father's sister had died at the age of 4 due to some neurological complication following hydrocephalus.

He had a history of seizure at 56th day of life. It was in the form of rapid jerky movement of left upper limb and flickering of left eyelid with deviation of eye ball to left. Episode had lasted for 5-6 seconds. After this episode he was started on valproate. Since then, he had similar episodes of seizure upto the age of 6 months (7-10 days in a month and 2-4 episodes per day). EEG report at 4 months of age showed asymmetrically reduced background activity over left cerebral hemisphere and frequent spike and wave discharges from left parieto temporal region showed active epileptic discharge arising from same region.

MRI scans were taken at 56 days, 2 years and 8 years under GA. MRI findings were consistent with SWS. Karyotyping was done at the age of 5 years and no numerical & structural chromosomal abnormalities were detected (figure 1A). DNA testing based on the recommendations of American College of Medical Genetics was done at the same time and the genetic test detected a variant interpretation of a hemizygous missense variation in exon 8 of the *TAF1* gene (chrX:g.71378995A>G; Depth: 89x) that results in the amino acid substitution of Valine for Methionine at codon 442 (p.Met442Val; ENST00000423759.6).

On extra oral examination the patient had a unilateral port wine stain centered around right forehead, the right eye, side of the face, nose, right upper lip extending up to the midline (figure 1B). During the anamnesis, his father revealed that the lesion was present since birth with progressive growth. Scar of size 3x2 cm was seen on wrist of right hand due to biting, showing self-mutilation behaviour (figure 1C). Ocular examination revealed no significant findings but the patient has been advised regular ophthalmic check-ups.

In intraoral examination, buccal mucosa, gingiva, tongue, palate, and floor of the mouth showed normal appearance. Patient had a mixed dentition with deep dentinal caries with pulp exposure on 55,65,75,84,85,46, deep dentinal caries on 54,64,74, dentinal caries on 36, preshedding mobility of 73,83. Oral hygiene of the patient was poor with extensive calculus deposits on labial and lingual surfaces of lower anterior teeth (figure 2). As the patient was highly apprehensive and extremely uncooperative intra oral periapical radiograph (IOPAR) and Orthopantomogram (OPG) could not be taken.

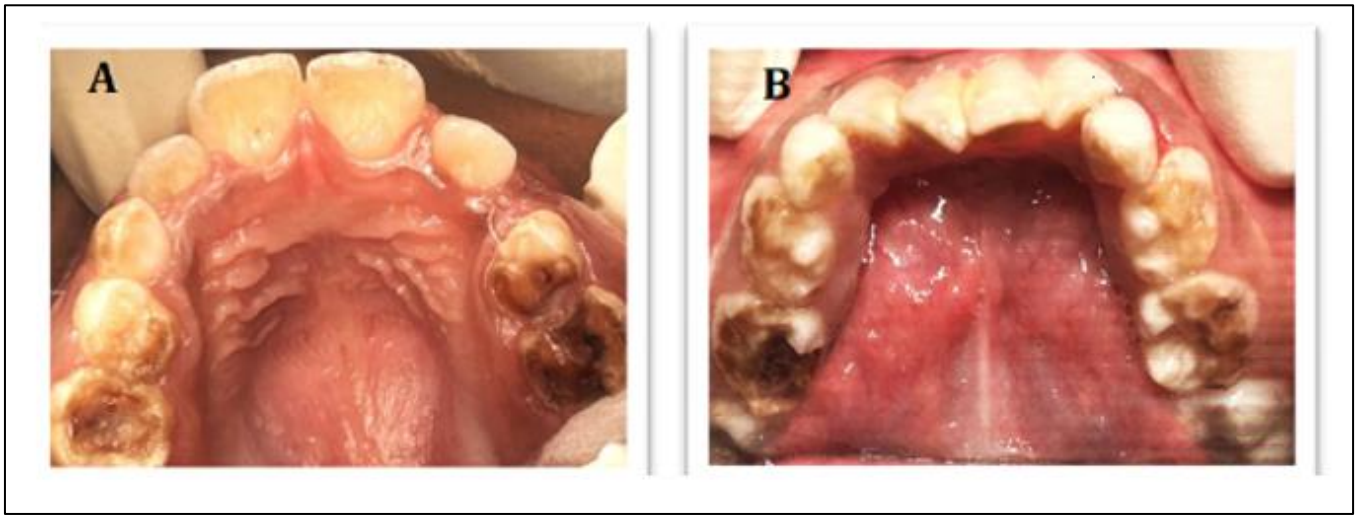

Figure 2 Pre operative occlusal view A: maxillary occlusal view B: Mandibular occlusal view

CT was taken to rule out any intrabony lesions of maxilla and mandible. As the patient was uncooperative, CT was done under midazolam sedation. In the present case both alveolar sockets and teeth of maxilla and mandible showed normal CT morphology for the age. No lytic or sclerotic changes suggestive of osseous hypertrophy were found in the present case. Incidental finding of hemi atrophy of right cerebral hemisphere with gyral calcifications was noted (figure 3).

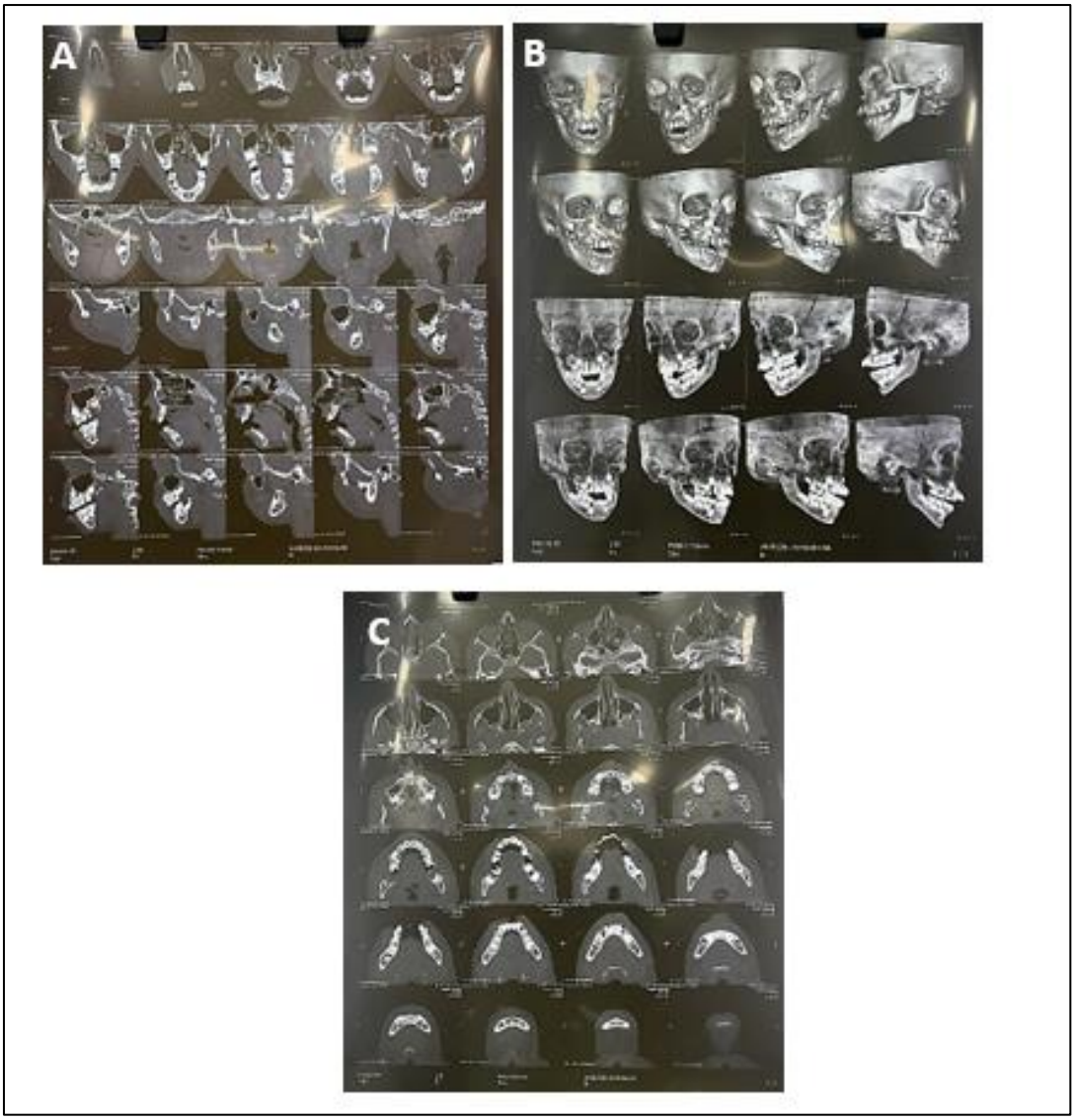

Figure 3 A-C, CT image showing both alveolar sockets and teeth of maxilla and mandible with normal morphology for the age without any osseous hypertrophy

As the patient showed minimal response to verbal commands with apprehension and inability to cooperate on a dental chair, it was decided to do full mouth rehabilitation under GA. Routine blood investigation revealed increased PT/INR values on two occasions (PT=17.6 & INR=1.3 on 12.09.2024 and PT=15.6 & INR=1.15 on 5.10.2024). Pediatric consultation was obtained and was advised to take injection vitamin K, 5mg IM 2 days prior to the procedure. After all the consultations (Neuromedicine, Neurosurgery and Pediatric medicine) patient was provisionally accepted for GA under high risk.

Patient was admitted in Govt Medical College, Kottayam. All the expected complications during the procedure were explained to parents after which high risk informed consent was obtained from them. Monopolar cautery unit was arranged anticipating uncontrolled bleeding following extraction. Patient was naso-tracheally intubated, draped sterile and extra orally painted with 0.5% povidone iodine. Procedure included composite restoration on 36, extraction of 54,55,64,65,73,74,75,83,84,85 and 46 after administering local anaesthetic (2% lignocaine with adrenaline) and suturing with 3-0 vicryl and hand scaling (figure 4). 46 had chronic irreversible pulpitis and it was decided to proceed with extraction of 46 considering the age of the patient (9 yrs) expecting 47 to erupt in the position of 46. The second molars, even if unerupted, start to drift mesially after the loss of the first permanent molar. A greater degree of forward bodily movement will occur with loss of the first permanent molar in 8- to 12-year-old children.[7] Moreover, single sitting root canal treatment in a carious fully formed permanent tooth with chronically infected pulp and without sinus tract is not preferred.[8] Regenerative endodontic procedure requiring long term follow up was also not feasible in this child. Extracted primary teeth had poor crown structure and advanced root resorption, while roots of 46 were incompletely

formed (figure 5). After completion of the procedure patient was extubated and shifted to recovery room. Postoperative period was uneventful. On discharge instructions for home care, including diet counselling and oral hygiene practices, were given to parents.

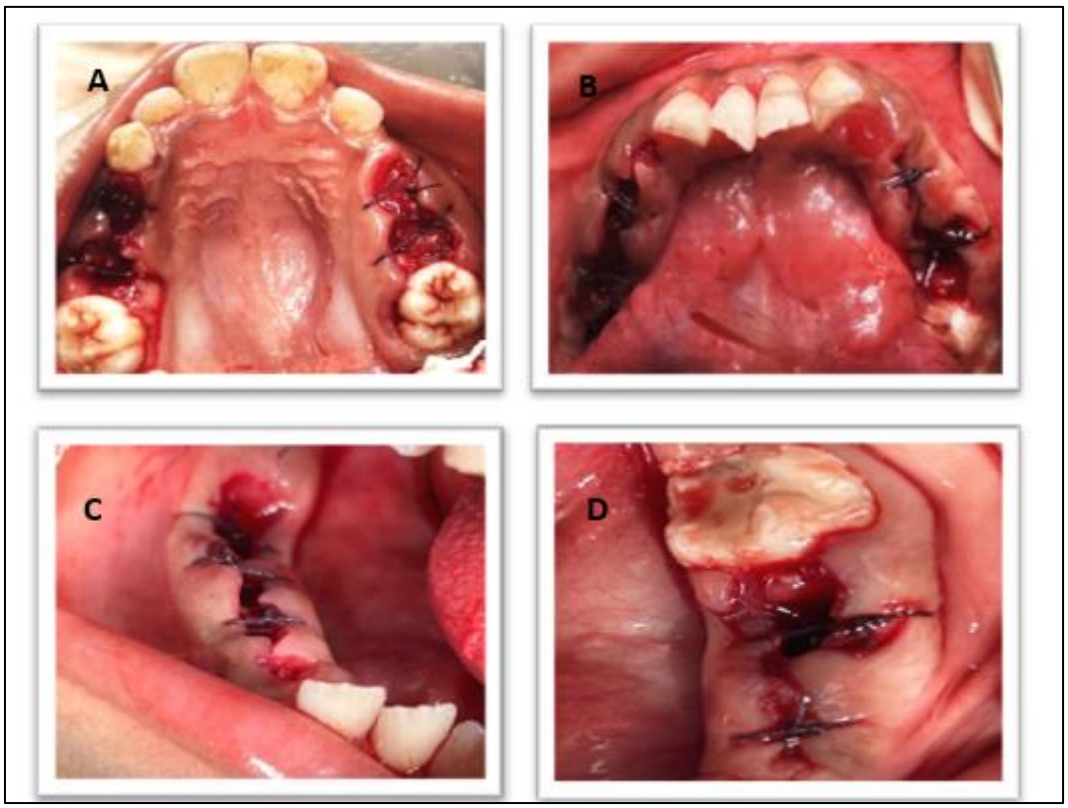

Figure 4 Post operative occlusal view A: Maxillary arch- sutured extracted sockets of 54,55,64 & 65 B & C: Mandibular arch- sutured extracted socket of 74,75,84,85 & 46 D: Composite restoration on 36

Figure 5 Extracted primary teeth with poor crown structure and advanced root resorption & incompletely formed roots of 46

A recall evaluation was planned after 1 week to assess the child's oral cavity and to determine oral hygiene maintenance. Parents reported improvement in child's behaviour as the child became less irritable after full mouth rehabilitation. Periodic evaluations were then arranged every six months.

3. Discussion

Managing a SWS child with Global Developmental Delay and Cerebral Palsy poses a big challenge for the pediatric dentist because of low intelligence, difficulty in communication, uncooperative behaviour, convulsions, poor concentration and difficulty in keeping the mouth open for long time to carry out the dental procedures. SWS is characterized by abnormal blood vessel growth (angiomas), which can lead to significant bleeding during dental procedures, including extractions. Fear and stress can increase intracranial and intraocular blood pressure, and in SWS patients, it is particularly important that these elevations are controlled. General anaesthesia allows for better control of blood pressure and, in some cases, the use of hemostatic agents to minimize bleeding. Hence, in child patients with SWS and intellectual insufficiency, full mouth rehabilitation under GA is justifiable.

According to Inan (1999), 87–90% of the SW cases have PWS localized over the face on the right side. 50% of the patients have lesions extended over the median, and 33% of patients have bilateral involvement.[9] In this case also, the child showed PWS only on the right side of the face without extension over midline.

About 70–80% of patients with SWS experience neurologic complications. Seizures, hemiparesis, stroke-like episodes, intellectual disabilities, and behavioural issues are a few of these.[10] In our case also the child had severe mental retardation, with severe intellectual disability, global developmental delay and self-mutilating behaviour.

According to the Roach scale, SWS is classified as follows:[11]

- Type I: both facial and leptomeningeal angiomas; may have glaucoma
- Type II: facial angiomas only (no CNS involvement); may have glaucoma
- Type III: isolated leptomeningeal angioma; usually no glaucoma.

According to the above criteria, our case was type I SWS as the patient had a PWS on the right side with epileptic convulsions and gyral calcification noted in the right tempero-parietooccipital and left parieto-occipital gyri but glaucoma was absent.

Patients with SWS are more likely to experience oral mucosal bleeding during routine dental procedures because of intra-oral angiomatosis, soft-tissue overgrowth, and gingival hyperplasia (worsened by antiepileptic medication). This can make maintaining proper oral hygiene more difficult and jeopardize physiological hemostasis.[6] Enlargement mainly affects the labial surface of the interdental papilla, though greater extensions can be affected including the gingival margins and lingual and palatal surfaces. The affected mucosa is friable to minimal damage and mild dental procedures due to oral vascular lesions, which can range from localized vascular hyperplasia to severe angiomatous proliferation or pyogenic granulomas.[11] These lesions are located ipsilateral to the face lesion and have a distinct midline demarcation. However oral manifestations were absent in this case.

4. Conclusion

Periodic oral examinations must be performed in children with special health care needs. Dental rehabilitation of such patients is a complicated procedure that calls for multidisciplinary approach and meticulous pre-operative planning before any dental surgery. Application of pharmacological behavior management techniques including GA may be necessary due to their mental impairment.[12] Pharmacological behavior management should be reserved as a secondary measure, used only when all non-pharmacological strategies have failed and should always be delivered in a tailored, safe, well-monitored, and consented framework.

Compliance with ethical standards

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Disclosure of conflict of interest

No conflict of interest to be disclosed.

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Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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