

Oral and dental rehabilitation under general anesthesia in a pediatric patient with Smith-Magenis syndrome: first case from Turkiye

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ABSTRACT

Smith-Magenis syndrome (SMS) is a rare, genetically-based syndrome affecting multiple systems. It is characterized by developmental delay, behavioral disorders, and characteristic facial features. While various descriptions of the craniofacial and oral findings of SMS exist in the literature, studies involving dental treatment requirements are quite limited. This study aims to present the oral and dental rehabilitation performed for the second time under general anesthesia in a 9-year-old pediatric patient diagnosed with SMS during the mixed dentition period, who presented to our clinic with pain complaints, and to discuss the challenges of dental management of this rare syndrome in light of the literature.

Keywords: Smith-Magenis syndrome, chromosome deletion (17p11.2), general anesthesia, oral and dental rehabilitation

INTRODUCTION

Smith-Magenis syndrome (SMS) is a complex neurodevelopmental disorder affecting multiple systems, occurring sporadically and with a frequency of approximately 1/15,000-25,000. It was first described in 1986 by Ann C. M. Smith and Ellen Magenis. This genetic disorder is mostly associated with a deletion on the short arm of chromosome 17 (17p11.2). The most common cause of SMS is the deletion of the RAI1 (Retinoic Acid Induced 1) gene.¹

Behavioral problems such as self-harming behaviors, aggression, temper tantrums, and sleep disorders are common in individuals with this syndrome. Sleep disorders are prevalent and can exacerbate other clinical symptoms. Hearing loss may be detected in some individuals. Skeletal anomalies, particularly scoliosis and other bone development disorders, are also reported. Intellectual disability is generally mild to moderate. Delays in communication skills significantly affect social interaction and learning processes.²⁻⁵

The facial appearance of patients with SMS is often unique. The main craniofacial findings are brachycephaly, frontal bossing, hypertelorism, fissure midline hypoplasia, tent lip (upward retraction of the midline of the upper lip), and micrognathia, which can develop into relative prognathism in later years.^{6,7}

Oral findings of the syndrome have been reported as weakness of the tongue muscles, inadequate lip closure, prominent jaw structure, and abnormal palate morphology. Cleft lip and palate are rarely seen. Dental findings reported in SMS include micrognathia, hypodontia, microdontia, taurodontism, delayed eruption, persistent primary teeth, ectopic eruption, enamel hypoplasia/hypomineralization, bruxism,

malocclusion, open bite, crowding, posterior crossbite, maxillary hypoplasia, relative prognathism, macroglossia, mouth breathing, and drooling. Dental anomalies, particularly agenesis of the lower second premolar teeth, taurodontism, and root anomalies, are frequently reported. The incidence of caries and gingivitis increases with age.^{4,5,8} Due to behavioral and cognitive difficulties, non-pharmacological behavioral guidance techniques are not feasible for dental treatment in this patient group. Alternative treatment approaches such as conscious sedation or general anesthesia are needed.⁹⁻¹¹

This case report presents the comprehensive dental treatment process and clinical management of a 9-year-old pediatric patient diagnosed with SMS, performed under general anesthesia. While the dental management of a limited number of pediatric cases with this syndrome has been reported worldwide,^{3,12} this presentation is the first case reported from Turkiye.

CASE

A 9-year-old child with special needs presented to the Department of Pediatric Dentistry at Dicle University Faculty of Dentistry in Diyarbakir in 2025 due to aesthetic and pain problems caused by decayed teeth. The patient's medical history revealed a diagnosis of SMS. Necessary informed consent was obtained from the patient's parents.

Medical Anamnesis

The patient was born via cesarean section in 2016, with a recorded birth weight of 3.5 kg. Family history revealed consanguinity between the parents, the patient having one



sibling, and no other family members diagnosed with SMS. The mother’s medical history did not reveal any known teratogenic factors such as smoking during pregnancy or X-Ray exposure. Clinical examination revealed postural abnormalities, overweight, facial asymmetry, and prognathism. The patient’s height (128 cm) and weight (35 kg) were determined to be developmentally advanced compared to peers. It was learned that the patient regularly used neurological medications. Based on the anamnesis obtained from the parents, it was reported that the characteristic findings of SMS included prominent self-harming behaviors, severe temper tantrums, and sleep disturbances. It was stated that the patient was able to self-harm to the point of causing bleeding by banging his head against the wall, and that these behaviors did not stop without family intervention. It was also noted that he exhibited repetitive self-harming behaviors such as throwing objects and biting his hand. It was learned that the patient regularly used Arislow ER oral tablets containing guanfacine hydrochloride and Neurodol oral drops containing haloperidol to control his temper tantrums. It was learned that 1 mg/ml Risperidone syrup (0.3 ml before bedtime) and 3 mg/5 ml Melatonin syrup (5 ml before bedtime) were prescribed to regulate sleep patterns, but due to insufficient clinical response after a certain period despite long-term and regular use, the treatment was discontinued by the parents 6 months ago. Regarding the feeding history, it was reported that the patient’s eating behavior was uncontrolled, limited to the amount given by the family, and that they could not spontaneously terminate the feeling of fullness. No hearing loss was detected, and it was reported that intellectual development was behind that of their peers. The patient’s history revealed that renal and cardiac anomalies and scoliosis, which are frequently reported in SMS, were not present.

Dental Anamnesis

The patient’s history revealed that dental treatments under general anesthesia were performed at an external center 3 years prior. However, due to continued inadequate oral hygiene, secondary caries developed in the existing restorations, along with the formation of new carious lesions. Extraoral examination revealed brachycephaly, short stature with central obesity, midfacial hypoplasia, flat nasal bone structure, and mandibular prognathism. Occlusion assessment showed class III malocclusion according to Angle classification, along with antero-posterior crossbite. Macroglossia was also observed. Facial symmetry was preserved, and the facial profile was assessed as convex. The upper lip appeared full, and no cervical lymphadenopathy was found (Figure 1a, b). The clinical findings to be considered before dental treatment in this very rare patient are presented in the table (Table).

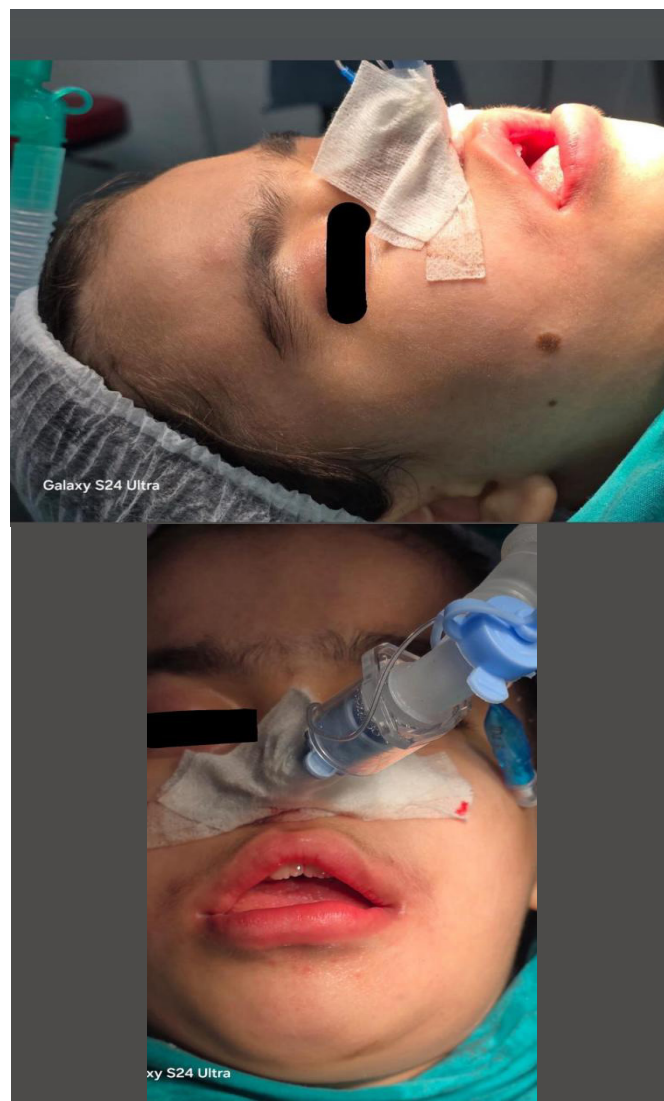


Figure 1. a) Extraoral view of the patient, b) Extraoral view of the patient

Teeth were identified using the FDI two-digit tooth numbering system (ISO 3950).¹³ Intraoral examination revealed caries and mobility in teeth 52 and 62. Mobility was also detected in tooth 82. An enamel fracture was observed in tooth 31, and excessive substance loss due to old restorations and secondary caries was detected in teeth 74 and 75. Deep caries were observed in teeth 16, 26, and 73, while secondary caries beneath existing restorations were detected in teeth 36, 46, and 85 (Figure 2a-d).

In addition to the clinical diagnosis of caries, panoramic and periapical radiographic evaluation revealed the absence of the permanent tooth germ of tooth 45 (Figure 3, 4).

Table. Specific findings observed in our patient’s clinic		
	Clinical findings	Specific features
Craniofacial (face)	Prominent facial phenotype	Square facial structure, broad forehead, flat nasal bone structure, “tent mouth” (upward curved upper lip), brachycephaly, midfacial hypoplasia
Behavioral	Severe maladaptive behaviors	Self-harm, sudden temper tantrums, impulsivity, throwing objects and biting one’s hand, head banging against walls, uncontrolled eating, inability to spontaneously terminate satiety
Neurological/cognitive	Developmental delay	Mild to moderate intellectual disability, delayed speech and language development
Sleep pathology	Reverse melatonin cycle	Excessive daytime sleepiness, frequent nighttime awakenings and hyperactivity, irregular sleep patterns
Characteristic movements	Stereotypical behaviors	The “self-hug” gestures
Dental/oral	Orodental anomalies	Class III malocclusion, anterior-posterior crossbite, mandibular prognathism, macroglossia, hypodontia (missing teeth), bruxism (teeth grinding), and a tendency towards poor oral hygiene

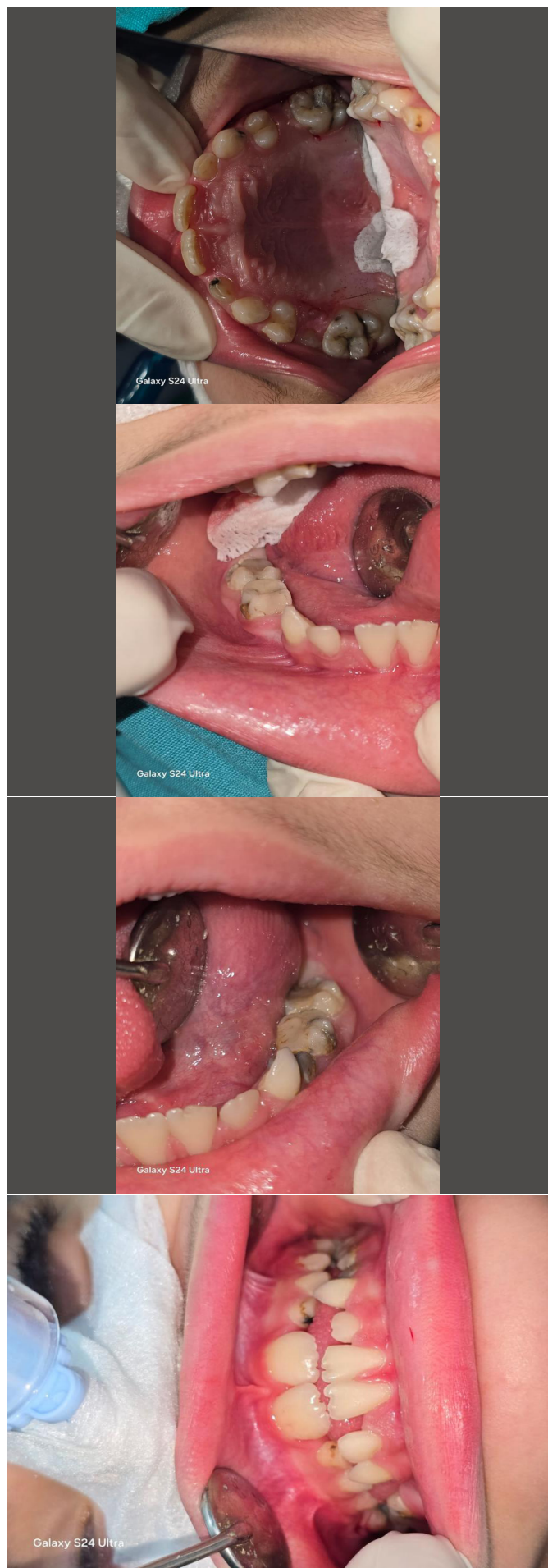


Figure 2. a) Intraoral view of the patient before treatment (maxilla), b) Pre-treatment intraoral view of the patient (right lower mandibular quadrant), c) Pre-treatment intraoral view of the patient (left lower mandibular quadrant), d) Intraoral view of the patient before treatment



Figure 3. Panoramic X-Ray of the patient before treatment



Figure 4. Pre-treatment periapical X-Ray of tooth number 85

Treatments were planned under general anesthesia due to the patient's lack of communication skills. Necessary consent forms were obtained from the patient's parents. Preoperative examinations were performed. Anesthesia approval was obtained as a result of consultation with a general anesthesia specialist at the General Anesthesia and Reanimation Department of Dicle University Faculty of Medicine. It was determined as American Society of Anesthesiologists' (ASA) class II.¹⁴ The anesthesia team evaluated the patient's anesthetic approach and determined monitoring methods. The pediatric patient, who had been fasting for 8 hours, was given intravenous midazolam (Dormicum, La Roche Ltd., Switzerland) at 0.5 mg/kg as premedication 15 minutes before the procedure. Non-invasive blood pressure, electrocardiogram, and peripheral pulse oximetry were used for standard anesthesia monitoring. Intravenous 0.09% NaCl infusion was administered. After 5 minutes of stabilization, the patient's pulse, systolic/diastolic arterial pressure, and mean arterial pressure were recorded as baseline vital signs. The patient's vital signs were closely monitored intraoperatively, and the anesthesia process was managed safely. Oral and extraoral hygiene was performed with 0.12% chlorhexidine, and a pharyngeal tampon was placed. A lip retractor was placed to provide better vision. In our patient, scaling was performed to control plaque.

Composite resin restorations (Palfique Estelite Paste®, Tokuyama Dental Corp., Tokyo, Japan) were placed under rubber dam isolation on teeth 16, 26, 73, and 31. Fissure sealants were applied to teeth 14 and 24 as a preventive measure (**Figure 5a, b**).

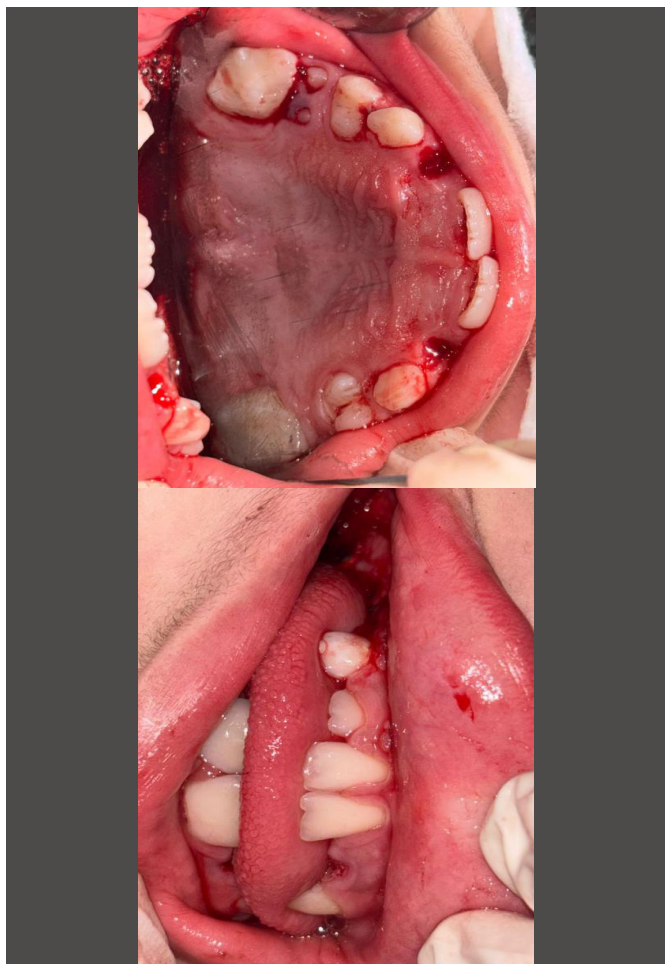


Figure 5. a) Post-operative immediate view (upper jaw), b) Post-operative immediate view (anterior)

Pulpotomy procedures were performed on teeth 36, 46, and 85 using mineral trioxide aggregate (MTA) (Angelus MTA®, Angelus Indústria de Produtos Odontológicos S/A, Londrina, PR, Brazil). A 2.5% sodium hypochlorite solution was used to control bleeding during the procedure. After completion of the composite restorations, stainless steel crowns (SSCs) were placed on the treated molars to ensure long-term durability.

Teeth 52, 62, 82, 74, and 75 were extracted under local anesthesia (Figures 6a-d). At the end of the procedure, the pharyngeal tampon was removed. Before extubation, intravenous paracetamol (10 mg/kg) (Parol Flakon®, MEFAR İlaç San., Türkiye) was administered for postoperative pain control. Following extubation, the patient was monitored in the recovery room. A liquid diet was initiated four hours after transfer to the ward.

Antibiotics and analgesics were prescribed to prevent post-operative infection risk following dental treatments. In the postoperative period, the patient received dietary recommendations to reduce caries-causing eating habits, and parents received oral hygiene education. The patient was discharged the same day. No new tooth decay was observed during the 6-month follow-up examination.

At the follow-up visit, a topical remineralization agent containing casein phosphopeptide-amorphous calcium phosphate with fluoride (CPP-ACPF) was recommended to promote enamel remineralization.



Figure 6. a) Stainless steel crown application to tooth number 36, b) Stainless steel crown application to teeth 85 and 46, c) Periapical radiographs of teeth 85 and 46, d) Periapical radiograph of tooth number 36



DISCUSSION

SMS is a rare genetic disorder characterized by cognitive impairment, resulting from deletions in the chromosome 17p11.2 region or mutations in the RAI1 gene. It is noteworthy that clinical and medical case reports are quite limited in the literature.^{4,15-17} The characteristic craniofacial findings in our case, such as mandibular prognathism, frontal prominence, midfacial hypoplasia (depression of the nose and cheekbone), and macroglossia, are in complete agreement with the SMS phenotype described in the literature.^{6,7}

In the screenings conducted, it is estimated that the prevalence of SMS in Türkiye is between approximately 1/15.000 and 1/25.000, consistent with global data. When the Turkish literature is examined, it is seen that the studies on SMS mostly focus on genetic diagnosis, clinical phenotype and psychiatric findings.¹⁸⁻²⁰

Studies addressing the dental management of patients with SMS are quite limited in the literature.^{3,4,12} No case reports have yet been found regarding the dental management and oral health rehabilitation of individuals diagnosed with SMS in Türkiye. This situation is important because the case we present is the first study in Türkiye to address the dental approach in patients with SMS.

Tomona et al.⁴ report that oral hygiene deteriorates with the transition from childhood to adolescence in individuals with SMS and that the prevalence of caries increases proportionally with age. The fact that our case presented with deep secondary lesions despite being treated under general anesthesia three years ago confirms this progressive picture in the literature.

In the dental management of patients with SMS, pharmacological agents used to treat sleep disorders are a critical risk factor. Soares and Kanungo²¹ reported that the high sugar content of pediatric medications and side effects such as xerostomia (dry mouth) increase caries activity by reducing the protective effect of saliva. The patient's regular use of Risperidone and Melatonin, along with uncontrolled eating behavior, supports this pharmacological and dietary caries risk in the literature. In addition, behavioral problems such as temper tantrums, impulsivity, and attention deficit,²² which are characteristic of the syndrome, accelerate destruction by making it difficult to maintain oral hygiene at home. Therefore, a multidisciplinary approach including the preference for sugar-free drug formulations, personalized hygiene education, and strict dietary control is essential for the success of restorative treatments in children with SMS.

The case of a 5-year-old child reported in Brazil by Ferreira et al.³ shows a complete similarity to our case in terms of frontal prominence, prognathism, and high caries risk due to neurological drug use (Risperidone, Topiramate). While the case reported in Brazil only involved the primary dentition and followed a path focused on extraction and composite restoration; in our case, more comprehensive procedures such as Stainless-Steel Crown (SSC) and fissure sealant were applied to preserve the permanent teeth. Similarly, in the 3-year-old case in India, severe destruction of the primary dentition (13 extractions) necessitated a radical approach, and SSCs were used in the restored teeth. General anesthesia (GA) becomes a necessary option in such cases with physical or mental limitations where classical behavioral guidance methods are insufficient.²³⁻²⁶ In both global examples, GA was

preferred due to the difficulty of cooperation. However, the history of general anesthesia in our case proves the necessity of an individualized long-term follow-up strategy in patients with SMS, instead of focusing only on immediate treatment.

Dental rehabilitation in SMS patients requires advanced medical preparation encompassing the systemic, anatomical, and behavioral manifestations of the syndrome. The case of 3-year-old India,¹² while paralleling our case in terms of severe early childhood caries (ECC), highlights the broad phenotypic spectrum of the syndrome with its complex cardiac anomalies (PFO, tricuspid regurgitation) and hand-foot anomalies (brachydactyly, polydactyly). Furthermore, in terms of anesthesia management, the case of India highlighted the need to be prepared for anatomical anomalies (macroglossia, lymphoid hyperplasia, etc.) by reporting intubation difficulties due to airway obstruction caused by enlarged lingual tonsils. Although there was no cardiac pathology in our case, such complex cases prove the vital importance of preoperative multidisciplinary evaluation. In our case and in Ferreira et al.³ Although nasotracheal intubation, preferred in study, provides surgical field comfort, airway anomalies should always be considered a risk factor in patients with SMS. In addition, the post-operative fever and need for intensive care observation observed in our case are consistent with the literature; confirming the necessity of close monitoring (PICU follow-up) after GA in these children.

Tomona et al.,⁴ in their study examining 15 SMS cases aged between 4 and 19 years, found at least one dental anomaly in more than 90% of the patients and reported that this high prevalence could be a helpful clinical indicator in the diagnosis of SMS. In this presented case, congenital absence of the lower right second premolar tooth was detected. The prognathic appearance frequently observed in individuals with SMS is thought to develop as a result of a combination of factors such as maxillary hypoplasia, protrusion of the lower anterior teeth, increased chin size, and macroglossia, rather than excessive mandibular growth.² In this case, maxillary hypoplasia, class III malocclusion, anterior-posterior cross-bite, and macroglossia were observed, and it was observed that these morphological features contributed to the pronounced mandibular prognathic appearance in the patient.

ETHICAL CONSIDERATIONS

- Ethical approval was not required for this case report according to institutional policies.
- Written informed consent for publication was obtained from the patient's legal guardian.
- Patient confidentiality was strictly maintained.

CONCLUSION

This first reported case from Türkiye highlights the clinical necessity of comprehensive dental treatment under general anesthesia in patients with SMS and aims to be a unique reference from treatment planning to postoperative follow-up. Additionally, it emphasizes the importance of individualized treatment planning and multidisciplinary management to ensure successful dental rehabilitation in patients with complex syndromic conditions.



ETHICAL DECLARATIONS

Informed Consent

Written informed consent was obtained from the patient's legal guardian included in this report. Signed consent forms are retained by the authors and are available upon request.

Peer Review Process

This report underwent external peer review.

Conflict of Interest

The authors declare no conflicts of interest.

Financial Disclosure

This case report did not receive any financial support.

Author Contributions

Concept: HZT, EA,BS; Design: HZT, EA,BS; Control: HZT, EA,BS; Resources: HZT, EA,BS; Materials: HZT, EA,BS; Data Collection and/or Processing: HZT, EA,BS; Analysis and/or Interpretation: HZT, EA,BS; Literature Review: HZT, EA,BS; Writing the Article: HZT, EA,BS; Critical Review: HZT, EA,BS.

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