A REVIEW OF DENTAL FINDINGS AND MANAGEMENT IN SANJAD-SAKATI SYNDROME

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NARRATIVE REVIEW

ABSTRACT

Sanjad-Sakati syndrome (SSS) is an autosomal recessive disorder that is seen commonly in Arab populations and presents with a plethora of orofacial features. While the prognosis of patients with SSS is poor, dental rehabilitation is necessary to improve the quality of life of patients and retain their functional efficiency. Owing to the various dysmorphic features and dental anomalies, oral care and management in patients of SSS can be challenging. This review provides details on the key features of the syndrome along with the dental treatment strategies mentioned in published case reports.

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BACKGROUND

Sanjad-Sakati syndrome (SSS), or hypoparathyroidism-retardation-dysmorphism syndrome is an autosomal recessive disorder. It was first reported in 1988 by Sanjad and Sakati and has since been consistently observed in the Arab population. Case reports from Oman, Palestine, Israel, Qatar, and Saudi Arabia have helped us understand the multifaceted nature of this syndrome which has been linked to mutations in the tubulin folding cofactor E (TBCE) gene on chromosome 1q42-43. The diversified orofacial manifestations of SSS are a major reason in the mortality incidence rate at infancy being 30% for a diagnosed patient.[1,2]

This narrative review highlights the key findings from published case reports that have exclusively reported the dental findings and management of patients with SSS.

METHODS

In order to fulfil our objective, a literature search was carried out across PubMed and MEDLINE databases. The keywords used were ‘Sanjad-Sakati Syndrome’, ‘oral’, ‘dental’, along with necessary truncations. We identified four case reports published in the English language between 1990 to 2021 that met our inclusion criteria.

DISCUSSION

Congenital hypoparathyroidism along with hypocalcaemia and hyperphosphatemia is a common characteristic of SSS. The hormonal imbalance is often associated with dwarfism and mental retardation. Failure of growth in intrauterine and postnatal life makes the patient more susceptible to neurological seizures.[3]

Facial features like depressed nasal bridge are seen in conjunction with other dysmorphic features like microcephaly, prominent forehead, deep sunken eyes, beaked nose and external ear anomalies. Associations between sleep apnoea and SSS have been reported in literature but a definite relation cannot be established.[4,5]

The commonly observed dental features include enamel hypoplasia, which is characterized by reduced enamel thickness, discoloration, and insufficient mineralization of the enamel. Delayed eruption of teeth, microdontia (small sized teeth), micrognathia (reduced jaw size), thin lips, dental agenesis (failure of tooth development), and cessation of root formation directly correspond to retarded growth. Furthermore, a high arched palatal vault is an inconsistent finding.[1-5]

Clinical findings of SSS are similar to Kenny-Caffey syndrome and DiGeorge's syndrome. However, unlike Kenny-Caffey syndrome, SSS does not present with osteosclerosis. SSS can be differentiated from DiGeorge's syndrome as the latter presents with congenital heart disease, cellular immunodeficiency, and absence of growth failure.[1,4]

Out of the four case reports we reviewed, three of the cases for dental care were managed under general anesthesia. In two reported cases, the patient was put under general anesthesia for a second time in a span of two to four years. The decision to put the patient under general anesthesia depends on the amount of treatment needed, and the presence of anatomical complications (microstomia, retrognathism) which might make general anesthesia challenging.

Antibiotic prophylaxis preoperatively, corticosteroid administration during the surgery to prevent postoperative oedema, and post-operative empirical antibiotics are commonly prescribed. Depending on the amount of carious involvement and the health of the surrounding supporting tissues, pulpotomies, stainless steel crowns, resin-bonded restorations, extractions, and dentures need to be planned in different phases. Patients who present with multiple missing teeth or agenesis of permanent teeth are often prescribed antifungal ointments due to the risk of developing denture stomatitis.

Out of the four cases review, only one case reported the death of a patient six months following a second treatment under general anesthesia. However, the reason for death was attributed to pulmonary complications.

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Patient education and preventive measures need to be stressed by dental surgeons. However, patient compliance has often found to be challenging. Rampant caries in patients with SSS may be linked to an etiology of night bottle feeding. Parents of a child with SSS might find it difficult to make their child quit this habit due to poor muscular strength in chewing fibrous foods. Thus, a routine follow-up to the dentist needs to be emphasised.

Table 1 Summary of case reports reviewed

YEAR	AUTHOR	FINDINGS (excluding dysmorphism)	TREATMENT
2004	Al-Malik	Severely decayed teeth	Restorations and extractions under general anesthesia
2010	Wasersprung et al	Sinus tracts from previous pulp therapies, agenesis of permanent teeth	Restorations, extractions, plaque removal, fluoride applications, under general anesthesia
2013	El-Batawi	Recurrent caries, supernumerary teeth	Pulpotomies, stainless steel crowns, extractions under general anesthesia
2018	Hassona et al.	Multiple carious teeth, missing teeth, incomplete root formation	Root canal treatment, resin-bonded restorations, denture fabrication under local anesthesia

CONCLUSION

The prognosis of Sanjad-Sakati syndrome is often poor. Dental treatments are needed to ensure that the quality of life of the patient improves by helping them retain their masticatory efficiency as much as possible.

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