Case Report

# Dental Management of Seckel Syndrome in a Child

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**Abstract**

Seckel syndrome is an autosomal recessive disorder characterized by severe intrauterine growth retardation, microcephalic bird-like facies, and a mental retardation. Seckel syndrome is an autosomal recessive disorder characterized by severe intrauterine growth retardation, microcephalic bird-like facies, and a mental retardation. Dental treatment for a 3 year old child diagnosed with Seckel syndrome is discussed

**Keywords:** *Dental, intrauterine growth retardation, Seckel*

## Introduction

Extreme short stature at birth resulting due to intrauterine growth retardation (IUGR) is known as primordial dwarfism. Increased maternal age, syphilis, toxoplasmosis, rubella, cytomegalovirus infections, malnutrition, Vitamin B12, and folic acid deficiency are some of the risk factors of IUGR. Rao PSS, Inbaraj SG in 1977 reported that high mortality rates are seen in children who weigh <1500 g at birth or gestational age <28 weeks. Pinho JR, Filho Fl in 2012 found enamel defects in infants with low birth weight than preterm births.

Primordial dwarfism can be associated with either small (microcephalic) or normal head size (normocephalic). Some syndromes associated with microcephalic primordial dwarfism are Majewski osteodysplastic primordial dwarfism (MOPD) Types I[1] (also called Taybi–Linder syndrome), II, III, Meier-Gorlin, and Seckel. The first symptom of primordial dwarfism is severe IUGR. MOPD I is associated with microcephaly, agenesis of the corpus callosum, seizures, short vertebrae, elongated clavicle, bowed femur, and hip displacement. In MOPD II, microcephaly is associated with squeaky voice, microdontia, widely spaced primary teeth, disturbed sleep patterns, and brain aneurysms. In MOPD III, anomalies of pelvis and clavicles are noted.[2] Meier–Gorlin syndrome is characterized by small ears, variable degrees of deafness and absence of kneecaps, curved clavicles, and microcephaly.[3] Normocephalic head is seen in Russell–Silver syndrome. It is associated with webbed toes, nondescended testicles, hypospadias, weak muscle tone, and delayed bone age.[4] The heights of individuals in Russell–Silver and Meier–Gorlin syndrome exceed when compared to other primordial dwarfism.[4]

Seckel syndrome is marked with IUGR, short stature, and microcephaly; individuals are mentally challenged. The clinical features of Seckel syndrome are shared by other impaired DNA damage responses, such as Nijmegen breakage syndrome and Ligase IV syndrome.[5] Unlike hypopituitary dwarfism, the defect is not caused by deficiency of growth hormone but because of intrauterine growth deficit. Seckel syndrome is distinct from MOPD by less severe growth retardation and proportionate limbs.

## Case Report

A 3-year-old male child was referred from the Department of Pediatrics to the Department of Pediatric Dentistry with a complaint of tooth decay and pain on his upper right back tooth region. He was a prediagnosed case of Seckel syndrome.[6] On physical examination, child was 50 cm in height, weighing 3 kg, and head circumference 34.5 cm. Parental history revealed parents was unaffected, and child was born to consanguineous marriage, born preterm at 32-week gestation. He weighed

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750 g at birth. History revealed that during the gestation period, ultrasonography was not performed to detect fetal defects. Postbirth, all the developmental milestones were delayed. History of child's diet revealed that child was not on regular solid food and primarily dependent on sweetened milk and fed with feeding bottle. The oral hygiene was restricted to cleaning his mouth once a day in the morning. Oral hygiene procedures such as swishing or gargling the mouth after milk consumption were not followed. History of tooth eruption revealed that the first primary tooth to erupt was lower central incisor, which had erupted at 18 months of age.

On examination, microcephalic head, receding forehead, sparse hair, prominent eyes, midface hypoplasia, and a deficient mandible were among the striking features; however, the symmetry was maintained [Figure 1]. Clinodactyly was also evident. The response and behavior of child suggested mild-to-moderate mental retardation. Frankl Behavior Rating Scale was definitely negative with incomprehensible speech. Intraoral examination of primary teeth presented with generalized microdontia and moderately inflamed gingiva with mobility. Lower anterior incisor crowding was evident. Occlusion was in distal step relationship. Dental caries was evident in 54, 64, 74, 75, 84, and 85; all these teeth were symptomatic suggestive of pulp involvement. Fifty-one, 52, and 61 exhibited increased mobility. As the child was of very small stature, uncooperative, and could not sit unsupported, a radiographic examination was decided against. Child was investigated for complete blood picture and reported anemic with 9.2 gm% of hemoglobin. The treatment procedure charted was pulpectomy with obturation of 54, 64, 74, 75, 84, and 85 teeth and extractions of mobile teeth 51, 52, and 61. The treatment was decided to be carried out under general anesthesia and hence performed [Figures 2 and 3].

**Figure 1:** Microcephalic head, short stature, and mid-face hypoplasia

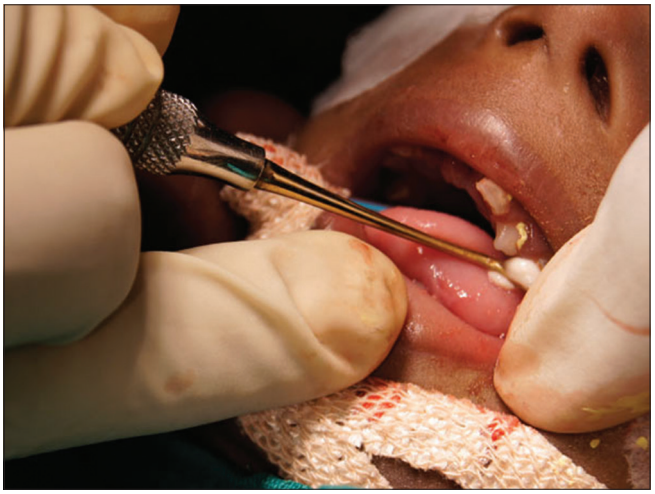

**Figure 2:** Dental treatment under general anesthesia

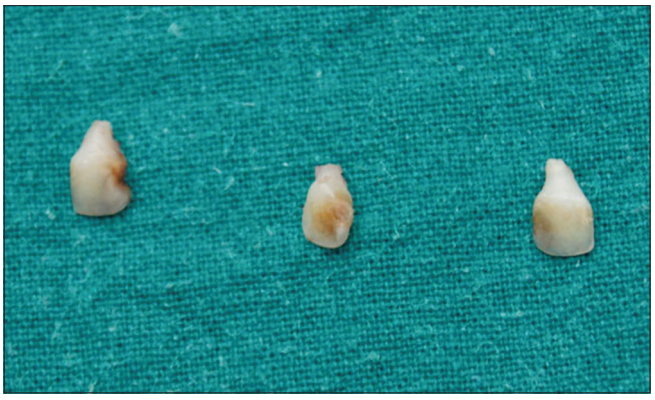

**Figure 3:** Extracted 51, 52, and 61

## Discussion

Seckel syndrome is also called as bird-headed dwarfism and microcephalic dwarfism. It is subclassified into further four types depending on linkage to different chromosomal regions 3q22, 18p11, 14q, and 21q22.3.[6] The pathogenesis seems to be an autosomal recessive condition, and yet the chromosomal study previously done has shown a normal karyotype.[6]

The case is confirmed to be a Seckel syndrome based on the history of birth weight-750 g, phenotypic features, such as proportional body parts, microcephalic head with mental retardation, and characteristic bird-like facies. In the present case, the mental age of the patient was less when compared to chronological age. The child had not been consuming solid foods. He had been given mostly sweetened milk. Cleaning of teeth at regular intervals was not practiced which had increased colonies of cariogenic bacteria and hence tooth decay. Chair-side treatment of the procedure could not be done since the child could not cooperate and could not sit unsupported. Narrow airway and trachea and

fragile veins make administration of general anesthesia a testing task.[7] The gene defect in Seckel syndrome affects bone development and growth but not dental maturation and eruption.[8] There has been a generalized delay in the eruption of teeth in the Indian population probably attributed to nutritional status, socioeconomic status, etc., The eruption of primary teeth in the present case cannot said to be delayed when compared to other Indian studies in control.[9,10] Premature root resorption could be a reason leading to generalized mobility of all primary teeth, whether this is because of IUGR needs to be further investigated. The significance of this case is characterized by profound mental and physical growth deficiency. In a child with carious tooth, intake of food is affected because of the associative symptoms. The inability to consume food and weight loss because of the same becomes a real concern in a severe IUGR child. Thus, the dentist should be well equipped to deal and render the treatment effectively to the patient.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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### Conflicts of interest

There are no conflicts of interest.

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