ISSN: 2581-5989
PubMed - National Library of Medicine - ID: 101738774

International Journal of Dental Science and Innovative Research (IJDSIR)

IJDSIR : Dental Publication Service

Available Online at: www.ijdsir.com

Volume – 6, Issue – 6, December - 2023, Page No. : 27 - 32

Dental challenges and triumphs: Managing progeroid syndrome in an 8-year-old

[1]Dr. Divya Lakshmi. G, V.S. Dental College & Hospital, Krishna Rajendra Rd, next to KIMS Hospital, Parvathi Puram, Visvesvarapuram, Basavanagudi, Bengaluru, Karnataka

[2]Dr. Amitha H.A., V.S. Dental College & Hospital, Krishna Rajendra Rd, next to KIMS Hospital, Parvathi Puram, Visvesvarapuram, Basavanagudi, Bengaluru, Karnataka

Corresponding Author: Dr. Amitha H.A., V.S. Dental College & Hospital, Krishna Rajendra Rd, next to KIMS Hospital, Parvathi Puram, Visvesvarapuram, Basavanagudi, Bengaluru, Karnataka

Citation of this Article: Dr. Divya Lakshmi, Dr. Amitha H.A., "Dental challenges and triumphs: managing progeroid syndrome in an 8-year-old", IJDSIR- December - 2023, Volume – 6, Issue - 6, P. No. 27 – 32.

Copyright: © 2023, Dr. Divya Lakshmi, et al. This is an open access journal and article distributed under the terms of the creative common's attribution non-commercial License. Which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given, and the new creations are licensed under the identical terms.

Type of Publication: Original Research Article

Conflicts of Interest: Nil

Abstract

Progeria, or Hutchinson-Gilford Progeria Syndrome (HGPS), is an exceptionally rare genetic disorder causing premature aging, with an incidence of approximately one in eight million live births and a male predominance. The case of an 8-year-old boy with Progeria showcased numerous dental challenges, including mobile teeth, caries, and skeletal abnormalities. The dental aspect of Progeria demands careful attention due to jaw and dental anomalies, requiring individualized treatment plans due to the lack of standardized protocols. The present case stressed the challenges in prosthetic rehabilitation due to skeletal limitations, highlighting the vital importance of oral health maintenance and preservation of natural teeth in managing the distinctive needs of Progeria patients."

Keywords: Pediatric dentistry, progeroid syndrome, Hutchinson-Gilford Progeria Syndrome, oral rehabilitation, genetic disorders, dental anomalies.

Introduction

"Progeria" also known as "Hutchinson-Gilford Progeria Syndrome (HGPS)" is an extremely rare, severe genetic condition characterized by clinical signs that often mimic physiological aging in a premature manner. [1,7]

This disorder has a very low incidence and occurs in one per eight million live births and male predominance with M:F ratio of 1.5:1.[8]

Since its first description in 1886, over 180 cases have been reported in the world with only about 50 being molecularly confirmed. The diagnosis of Progeria is based on the recognition of common clinical features and the detection of p. Gly608Gly mutation in exon 11 of the LMNA gene. [6,8]

The general clinical features include short stature & Stunted growth, Craniofacial disproportion, Prominent veins on scalp, Alopecia, Prominent eyes, Pear shaped thorax, Thin Limbs & tightened joint ligaments. [2-4] (Figure 1 a-c)

The dental clinical features include Anodontia or Hypodontia, Tooth size / Arch Discrepancy, Delayed tooth eruption, Incomplete formation of roots, Agenesis of permanent teeth, High caries incidence Reticular atrophy of the pulp and more. [4,5]

These symptoms usually do not present themselves until the first or second year of the child's life. In the past, diagnoses relied solely on observable physical symptoms. However, contemporary diagnostic procedures involve a genetic test specifically designed to identify an LMNA mutation, thereby corroborating a suspected diagnosis of Hutchinson-Gilford Progeria Syndrome (HGPS).[6]

A dentist plays a crucial role within the medical team for individuals with Hutchinson-Gilford Progeria Syndrome (HGPS). This significance arises from the fact that many characteristics of the syndrome manifest in the dental, head, and neck regions. Given the unique challenges and specific dental issues associated with HGPS, the dentist's expertise becomes pivotal in addressing and managing these aspects of the syndrome as part of comprehensive medical care.

Case presentation

An 8-year-old boy reported to the Out Patient Department of Paediatric Dentistry with chief complaint of moving jaw in the patients front teeth region for 2 months.

Patient is a known case of progeroid syndrome born to a consanguineous married couple. The younger sibling has been diagnosed with thalassemia and is undergoing treatment for the same

Careful history revealed multiple mobile teeth in the maxillary & mandibular arches with dental caries of varying progression.

The extraoral examination recorded patient's height as 100 cm and weight of 11 kgs. Notable features include a convex profile with competent lips. Craniofacial disproportion is observed, accompanied by a triangular head shape. Short stature and stunted growth with short fingers and toes and prominent eyes with a pinched nose noted contributing to the unique facial appearance associated with progeria. The presence of a rachitic rosary is also noted. (Figure 1 a-c)

Intra oral examination revealed the patient was in mixed dentition period. Grade II Mobility noted with respect to multiple teeth and mobility with respect to maxillary & mandibular alveolar bone. Dental caries noted in almost all primary first and second molar of the maxillary and mandibular arch. Root Stump noted with the maxillary primary left second molar. (Figure 1 d-f)

Figure 1a, b

Figure 1c

©2023 IJDSIR, All Rights Reserved

Figure 1d

Figure 1e

Figure 1.f

Investigations

Radiographic investigations, comprising Orthopantomogram (OPG) and Cone Beam Computed Tomography (CBCT), unveiled critical findings. The OPG highlighted hypodontia with incomplete tooth and root formation across multiple teeth. Both the upper and lower jaws exhibited a drastic reduction in both alveolar bone thickness and height. The misshapen teeth displayed radicular anomalies, featuring short and thin roots. In the temporomandibular joint (TMJ) CBCT analysis, views from Panoramic, Axial, and Sagittal perspectives on both right and left sides revealed thinning of the glenoid fossa and condylar head, indicative of Condylar Hypermobility. (Figure 2 a,b)

Figure 2.a

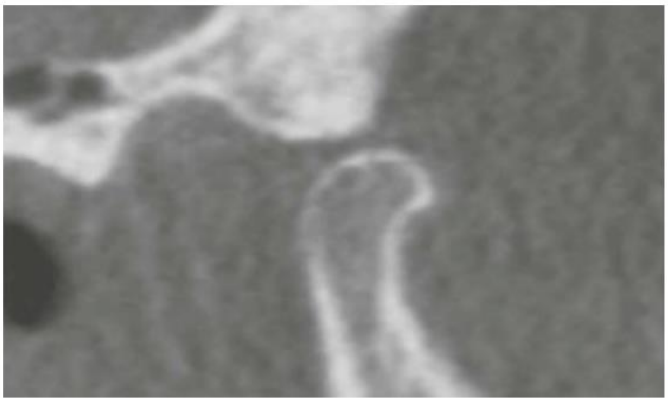

Figure 2.b

Treatment

Currently standardized protocols are not available; each solution is tailored according to the patient's condition. The initial dental management of this case was focused solely on maintaining oral health and preserving the

©2023 IJDSIR, All Rights Reserved

natural teeth in both the arches in order to maintain minimal masticatory function. Prosthetic rehabilitation here was very challenging as the child presented with reduced mouth opening and the radiographic investigations revealed marked reduction in the alveolar bone height and thickness with Condylar Hypermobility which caused the child to show a consistent tendency to protrusive posture that is a disadvantageous feature for the prosthesis use & retention.

The dental interventions for the patient included Glass Ionomer Cement (GIC) restorations for permanent and primary molars. Additionally, Silver Diamine Fluoride (SDF) application was applied for various teeth. Routine oral prophylaxis was performed, emphasizing the importance of maintaining good oral hygiene. (Figure 3a,b) The patient received recommendations to use a fluoridated toothpaste and practice the modified Bass tooth brushing technique at least twice daily. Furthermore, parents were advised to assist the child during home brushing, ensuring consistent and effective oral care practices.

Figure 3.a

Figure 3.b

Discussion

Progeria is listed as a “rare disease” by the Office of Rare Disease (ORD) of National Institute of Health (NIH). It is an extremely rare genetic disorder with a prevalence of 1 in 8 million live births. [1]

Although they are born looking healthy, children with Progeria begin to display many characteristics of accelerated ageing around 18 to 24 months of age. [1,6]

The children have a remarkably similar appearance, despite differing ethnic backgrounds.

The average life expectancy for a child with progeria is about 13, but some with the disease die younger and some live for 20 years or slightly longer.[1]

It is caused by the mutation of the LMNA Gene. These genes produce Lamin A protein which is the structural scaffolding that holds the nucleus of a cell together. This abnormal Lamin A protein called Projerin causes Progeria and it makes the nucleus unstable. That cellular instability leads to the process of premature ageing and disease in progeria. [6,15]

In this syndrome, the oral appearance is highly affected. The maxilla & mandible present variations that are difficult to handle and the frequent dental anomalies, impose an accurate study of each patient. Developmental defects of the enamel, anomalies in tooth number, position, or shape, malocclusion, natal teeth, or delayed

©2023 IJDSIR, All Rights Reserved

eruptions have been reported for these patients in literature. [16,17]

Following the third molars, the second premolars and lateral incisors emerge as the most commonly absent permanent teeth, as indicated by various studies. [10-14]

Our study aligns with previous findings on Hutchinson-Gilford Progeria Syndrome (HGPS), revealing a notably elevated prevalence of congenitally missing permanent second premolars.

Radiographic observations reveal irregularities in both crown and root development, suggesting that, beyond inadequate jaw growth, abnormal tooth development could directly contribute to the clinically observed dental anomalies and this is in agreement with a study done by Domingo DL et al., in 2009 which evaluated 15 confirmed cases for oral findings.[15]

The first dental examination should be planned as soon as the first deciduous teeth erupts in order to promote a tailored preventive plan for the child with progeria.

Standardised protocols for dental management for children with progeria are currently lacking, leading to individualized solutions based on the patient's condition.

In this case, Prosthetic rehabilitation posed challenges due to the child's reduced mouth opening and significant reduction in alveolar bone height and thickness, along with Condylar Hypermobility.[16]

Hence, the primary focus in this case was initially on preserving oral health and maintaining natural teeth to ensure minimal masticatory function.

Conclusion

Individuals with Progeria are very special people on many different levels.

This relatively small group of individuals deserve the support of researchers, healthcare practitioners and government agencies globally.

There is presently no cure for Progeria, but there are new drugs on the horizon which provide a hopeful outlook. These drugs aim to treat the underlying genetic cause of the syndrome while providing an increased life expectancy for these individuals.[9]

Progeria patients typically experience a delayed eruption of teeth and root development. This is most likely due to the retrognathic position of the maxilla and mandible and insufficient jaw growth.

This was certainly a challenging case scenario. The primary goal was preservation of the natural teeth in order to maintain masticatory function.

The importance and implementation of oral health & hygiene maintenance was continuously stressed upon throughout the treatment and for the consequent follow ups.

References

1. Progeria Research Foundation. What is the cause of progeria? Progeria Research Foundation. 2018
2. Wiedemann HR. An unidentified neonatal progeroid syndrome: follow- up report. Eur J Pediatr 1979; 130:65- 70.
3. Rastogi R, Chander Mohan SM. Progeria syndrome: a case report. Indian J Orthop 2008; 42:97-9.
4. Delgado-Luengo WN, Petty EM, Solís- Añez E, et al. Petty-Laxova- Wiedemann progeroid syndrome: fur- ther phenotypical delineation and con- firmation of a rare syndrome of prema- ture aging. Am J Med Genet A 2009;149A:2200-5.
5. Ullrich NJ, Silvera VM, Campbell SE, Gordon LB. Craniofacial abnormalities in Hutchinson-Gilford progeria syn- drome. AJNR Am J Neuroradiol 2012; 33:1512-8.
6. Audrey Gordon & Leslie Gordon (2014) The Progeria Research Foundation: it’s remarkable

©2023 IJDSIR, All Rights Reserved

journey from obscurity to treatment, Expert Opinion on Orphan Drugs, 2:11, 1187-1195

7. Martin GM. Genetic modulation of senescent phenotypes in Homo sapiens. *Cell.* 2005; 120:523–532
8. Pollex RL, Hegele RA. Hutchinson-Gilford progeria syndrome. *Clin Genet.* 2004; 66:375–381.
9. Rehman NA, Rehman AA, Ashraf IN, Ahmed S. Can Hutchinson-Gilford progeria syndrome be cured in the future? Intractable Rare Dis Res.2015; 4:111-112.
10. Nunn JH, Carter NE, Gillgrass TJ, et al. The interdisciplinary management of hypodontia: background and role of paediatric dentistry. *Br Dent J.* 2003; 194:245–251
11. Zhu JF, Marcushamer M, King DL, Henry RJ. Supernumerary and congenitally absent teeth: a literature review. J Clin Pediatr Dent. 1996; 20:87–95.
12. Caldo-Teixeira AS, Puppin-Rontani RM. Management of severe partial hypodontia: case report. *J Clin Pediatr Dent.* 2003; 27:133–136.
13. Caldo-Teixeira AS, Puppin-Rontani RM. Management of severe partial hypodontia: case report. *J Clin Pediatr Dent.* 2003; 27:133–136.
14. Stephen A, Cengiz SB. The use of overdentures in the management of severe hypodontia associated with microdontia: a case report. *J Clin Pediatr Dent.* 2003; 27:219–222.
15. Domingo DL, Trujillo MI, Council SE, Merideth MA, Gordon LB, Wu T, Introne WJ, Gahl WA, Hart TC. Hutchinson-Gilford progeria syndrome: oral and craniofacial phenotypes. Oral diseases. 2009 Apr;15(3):187-95.
16. Cagetti MG, Camoni N, Cetraro F, Scanferla M, Moretti GM. Special-needs patients in pediatric dentistry: Progeroid syndrome. A case of dental management and oral rehabilitation. Pediatric Reports. 2019 May 23;11(2):7951.
17. Maloney WJ. The Dental and Oral Significance of Hutchinson-Gilford Progeria Syndrome. Archives of Dentistry. 2018 Feb 11;1(1):4-6.

©2023 IJDSIR, All Rights Reserved