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The Integral Role of the Dentist in Treating Individuals with Hutchinson-Gilford Progeria Syndrome

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Introduction

Hutchinson-Gilford Progeria Syndrome (HGPS) is a rare genetic disorder in which the individual displays a phenotypic expression similar to that of an aged individual. In addition, these individuals suffer from various afflictions usually seen in the elderly.

HGPS individuals exhibit many dental manifestations as well as a multitude of other craniofacial manifestations of the disorder. Hence, it is a logical conclusion that dentistry should play a very large and vital role in not only the documentation of various traits of a dental and/or head and neck origin but, also, to conduct original research with a goal of unlocking some of the many mysteries of this rare but, scientifically, extremely relevant disorder. It has been suggested in the scientific literature that individuals born with HGPS might hold certain physical characteristics or traits which might enable science to unlock some of the cellular mysteries of natural aging (1).

Hutchinson-Gilford Progeria Syndrome

Sir Jonathan Hutchinson described, in 1886, the case of a 3-year-old boy who had features resembling those of an elderly individual (2). A year later Dr. Hastings Gilford described a case with similar clinical findings (3). This disorder has since become known as Hutchinson-Gilford Progeria Syndrome (HGPS).

Only approximately 100 cases have been documented in the scientific literature sine the time of Hutchinson and Gilford owing greatly to the rarity of HGPS- only 1 in every 8 million live births (4). 97% of all cases are found in Caucasians (5) with a 1.5:1 ratio of males outnumbering females. At birth, infants with HGPS usually appear normal. However, over the first year or two of life typical manifestations gradually develop and become apparent (6). The average lifespan of these individuals is 13 years (7). HGPS individuals often suffer from various ailments usually reserved for the aged (i.e. stiffness of joints, severe and progressive cardiovascular disease, and hip dislocations) (8).

The cause of HGPS is genetic. In most cases of HGPS there is a change from glycine GGC to glycine GGT in codon 608 of the lamin A (LMNA) gene which activates a cryptic splice donor site to produce abnormal lamin A thus disrupting the nuclear membrane and thus causing an alteration in transcription (9). Some hope on the research horizon has been seen in the past few years as Alsina et al. (10) have demonstrated the ability of the farnesytransferase inhibitor tipifarnib (zarnestra) to inhibit protein farnesylation.

DENTAL AND CRANIOFACIAL MANIFESTATIONS

Micrognathia is the primary manifestation of HGPS of which the treating dentist needs to be cognizant. Micrognathia leads to a delayed loss of deciduous teeth and overcrowding thus necessitating, at times, the extraction of overretained primary teeth (7).

A high caries incidence is often seen in HGPS individuals. Accordingly, dentists should see these individuals on a more frequent recall basis as they should be placed in a high-risk category for dental caries. Various fluoride therapies might also prove helpful in fighting dental caries in these individuals. Ora hygiene instructions should be constantly reviewed with HGPS individuals and their caregivers.

Diagnosis of HGPS is made upon the signs and symptoms even though there are presently genetic tests available which look for LMNA mutations (11). Therefore, dentists also play an important role in the diagnostic stage by documenting any clinical and/or radiographic abnormalities.

Due to the many dental manifestations of HGPS, it is absolutely imperative that any dentist who treats an individual with HGPS be fully aware of these many manifestations and various treatment modalities. A thorough list of the dental (Box 1), head and facial (Box 2), and jaw (Box 3) manifestations are provided below (1,12).

Conclusion

The dental profession has a wonderful and proud history of both providing much sought-after relief from physical discomfort and original scientific research

which has provided the scientific community much valuable information. Many times this dental research has led to significant advance in healthcare. Such an opportunity has once agin presented itself to out profession in the form of the individuals afflicted with HGPS.

Unfortunately, through the years HGPS has not garnered the attention it deserves due to short-sighted thinking. Its research has received neither the publicity nor the financial support with which other disorders have been endowed on account of it only affecting a relatively small amount of individuals worldwide at any given time. Ironically, nature might be presenting a gift to the scientific community in the form of HGPS individuals. Current research seems to indicate that HGPS might hold secrets to the mysteries of the process of natural aging which affects every individual. Hence, I urge the dental community to conduct original research in the area of HGPS. This could lead to a new understanding of the process of natural aging and, also, provide some much deserved physical relief to individuals with HGPS.

BOX 1- DENTAL MANIFESTATIONS

-secondary incisors located lingually and palatally
-incomplete formation of roots of primary molars
-delayed tooth eruption of primary and secondary dentition
-calcification along the nerve fibers and the vascular walls
-abnormal tooth formation
-reticular atrophy of pulp
-anodontia
-delay in calcification of the crowns of the permanent teeth
-hypodontia
-irregularity in calcification of the crowns of the permanent teeth
-narrow pulp chambers
-discoloration
-high caries incidence

BOX 2- HEAD AND FACIAL MANIFESTATIONS

-sparse to absent eyebrows and eyelashes
-delayed closure of fontanelles and sutures
-sparse to absent scalp hair
-no subcutaneous fat
-sculpted beaked nose
-relatively large tongue
-peri-oral cyanosis
-large cranium
-prominent forehead and frontal bossing
-small mouth
-prominent scalp veins
-prominent eyes

BOX 3- JAW MANIFESTATIONS

-micrognathia
-comparative paucity of vertical growth
-hypoplastic mandible
-atrophy of alveolar process
-retarded anterior and vertical growth
-narrow and high palatal vault
-small maxillary arch
-short mandibular ramus
-obtuse mandibular angle
-craniofacial disproportion

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