Hallermann-Streiff syndrome: Case report and literature review

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Hallermann-Streiff syndrome is a rare genetic disorder characterized primarily by head and face abnormalities. Patients show birdlike faces; hypotrichosis; various ophthalmic disorders; and dental abnormalities including absence of teeth, natal and neonatal teeth, enamel hypoplasia, and supernumerary teeth. In addition, delayed eruption of existing teeth and severe agenesis of permanent teeth are frequent findings. Dental and hereditary disorders can be associated with disturbances during tooth development and cause shortened roots. Short roots are a rare developmental anomaly in the permanent dentition, and the etiology is not well established. The generalized form is extremely rare. Generalized diminished root formation can lead to early loss of teeth. This article provides a case report of a 9-year-old boy with Hallermann-Streiff syndrome. Extraoral examination revealed a brachycephalic head, proportionate short stature, sparse hair, and atrophic skin. His face was characterized by a thin beak-shaped nose and retrognathia, resulting in a characteristic birdlike appearance. Radiographically, all teeth of the permanent dentition showed severely underdeveloped roots and partially underdeveloped crowns. The predisposition to severe dental caries and dental malformations makes it imperative to schedule effective prevention measures, especially since root canal treatment to preserve teeth can be hampered due to underdeveloped roots. (Quintessence Int 2011;42:331–338)

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Aubry¹ was the first to publish a report of a patient with Hallermann-Streiff syndrome. Two independent cases of this syndrome were later published, recognizing it as a distinct disease entity. Hallermann² reported two patients with bilateral congenital cataracts and bird face. In 1950, Streiff³ reported a similar case and distinguished the syndrome from progeria and mandibulofacial dysostosis. Although the syndrome had been described previously in the literature, in 1958, Francois⁴ summarized 12 published cases and described an additional one. After analyzing these cases, he delineated the characteristic features of Hallermann-Streiff syndrome and described several essential signs as diagnostic criteria. In 1960, Falls and Schull¹⁵ reported six further cases under the heading of Hallermann-Streiff syndrome. Thereafter, the condition was generally known by Hallermann-Streiff syndrome or Hallermann-Streiff-Francois syndrome. In addition, synonyms including Francois syndrome,⁶⁻⁹ oculomandibulodyscephaly,¹⁰⁻¹³ Francois dyscephalic syndrome,¹¹ and oculomandibulofacial syndrome¹⁴ can be found in the literature.
Hallermann-Streiff syndrome is a rare genetic disorder—only about 180 cases have been published to date. It is a congenital syndrome characterized by distinct craniofacial abnormalities. Dyscephaly with hypoplastic mandible and a small beaked nose leading to a typical birdlike face (89% to 90%); congenital cataracts (81% to 90%); hypotrichosis (80% to 82%); microphthalmia (78% to 83%); skin atrophy (68% to 70%); a short proportional stature (45% to 68%); and dental abnormalities (80% to 85%) (absence of teeth, natal and neonatal teeth, enamel hypoplasia, supernumerary teeth, delayed eruption of existing teeth, and malformed permanent teeth) are the main clinical features of Hallermann-Streiff syndrome. The etiology of this syndrome is unknown. Virtually all cases have been sporadic, and there is no sex predilection. Francois demonstrated marked changes in the structure of the connective tissue (alterations of the elastin) and postulated a primary disturbance in the metabolism of glycoproteins. A dominant succession is presumed. The diagnosis of Hallermann-Streiff syndrome is based on the presence of certain features, including the characteristic facial, eye, hair, skin, and dental findings.

Since the dominant symptom of patients with Hallermann-Streiff syndrome is cataracts, most cases have been reported in the ophthalmology literature. Although over 180 reported cases can be found in the literature, very few considered dental aspects. The present report focuses on the high incidence and variety of dental abnormalities in Hallermann-Streiff syndrome. Most of the patients revealed dental malformations such as supernumerary and neonatal teeth, premature eruption of the primary dentition, partial anodontia, agenesis of permanent teeth, and enamel hypoplasia. In addition, malocclusion, hypoplasia of the mandible, crowding, and high arched palate are typical of this syndrome. Although these dental abnormalities are often observed in Hallermann-Streiff syndrome, they are poorly defined. The particular aspect of the presented case is the occurrence of general underdevelopment of the roots and agenesis of the permanent dentition. The morphogenesis of normal tooth root development is regulated by epithelial and mesenchymal interactions. After the growth of the dental crown is complete, a double layer of cells referred to as Hertwig’s epithelial root sheath is formed by cells of the outer and inner dental epithelium. The formation of root dentin requires proliferation of epithelial cells in the Hertwig’s epithelial root sheath to initiate the differentiation of root odontoblasts and determine the size, shape, and number of the roots. Disturbances can lead to root deformities. Slootweg and Huber conducted histologic examinations of the mandible and a removed erupted tooth of an infant with Hallermann-Streiff syndrome. They reported a root dentin anomaly classified as a combination of irregular dentin and osteodentin, premature disintegration of the dental lamina, and Hertwig’s root sheath.

With regard to dental treatment, malformed crowns and roots confront the practitioner with special problems. Root canal treatment of teeth with severely underdeveloped roots is often impossible. The associated difficulties in dental treatment as well as the necessity of an early diagnosis and multidisciplinary treatment approaches are discussed.

CASE REPORT

A 9-year-old boy with Hallermann-Streiff syndrome and bilateral cataracts was referred to the Department of Restorative Dentistry, Dental School, University of Münster, Münster, Germany, for dental examination. Apart from the syndrome, the boy was in good health. He was the youngest son in a family who emigrated from Kazakhstan in 1994. The two older siblings, a 13-year-old girl and a 10-year-old boy, showed no conspicuousness. The anamnesis did not reveal any other known syndromes in the family of his nonconsanguineous parents. The pregnancy and childbirth took their normal course. His physical growth and development was retarded; however, his intellectual stage of development seemed to correspond to his age. The prime concern was the general mobility and progressing loss of teeth.
The extraoral examination revealed a brachycephalic head, a proportionate short stature, sparse hair, and atrophic skin, particularly in the nose region. His face was characterized by a thin, beak-shaped nose and retrognathia, resulting in a characteristic birdlike appearance, most strikingly in profile.

The clinical intraoral examination revealed reduced mouth opening (24 mm), a small tongue, and good oral hygiene. From an orthodontic point of view, a sagittal and transversal underdeveloped maxillary jaw with a frontal headbite and a crossbite on the right side was diagnosed. In general, the development of the dentition was not in correspondence with the age of the boy. Primary teeth 53, 55, 63, 64, 65, 73, 74, 83, 84, and 85 (FDI) still existed and showed moderate mobility. Permanent teeth 16, 12, 11, 21, 22, 26, 31, 41, and 46 (FDI) were in eruption and showed a higher mobility of grade II to III (Fig 1).

**Fig 1a** Maxillary view.

**Fig 1b** Mandibular view.
A panoramic radiograph taken at 9 years of age (Fig 2) revealed that all teeth of the permanent dentition as well as the primary teeth 55 and 85 showed severely underdeveloped roots and partially underdeveloped crowns. The already erupted teeth 16, 12, 11, 21, 22, 26, 31, 41, and 46 displayed at most one third of the normal root length at this stage of development. The teeth germ 13, 14, 15, 17, 23, 24, 25, 27, 32, 33, 34, 35, 36, 37, 38, 42, 43, 44, 45, and 47 did not show any signs of root development. Comparison of a panoramic radiograph taken at 12 years of age confirmed that no further root development had taken place. During this 4-year period, teeth 53, 64, 85, and 31 exfoliated, but no new tooth eruption was detected (Fig 3).

**DISCUSSION**

Hallermann-Streiff syndrome is a rare congenital anomaly that mainly affects the head and face. The seven characteristic features of this syndrome are congenital cataracts, dyscephaly, proportionate nanism, hypotrichosis, skin atrophy, bilateral microphthalmos, and dental anomalies. In the present case, striking craniofacial findings were noticed, including a small, wide head with a prominent forehead, a characteristic small jaw and mouth with a pinched nose, cataracts, sparse hair, thin skin, short stature, and severe dental abnormalities. Differential diagnosis of Hallermann-Streiff syndrome includes progeria, mandibulofacial dysostosis, and Wiedemann-Rautenstrauch syndrome.
Progeria differs from Hallermann-Streiff syndrome in that the former presents with premature arteriosclerosis, nail dystrophy, acromicria, chronic deforming arthritis, and normal eyes. Wiedemann-Rautenstrauch syndrome patients may have a somewhat similar facial appearance, but do not show the ocular findings of Hallermann-Streiff syndrome. Mandibulofacial dysostosis and Hallermann-Streiff syndrome patients both exhibit micrognathia, high palatal vault, and molar hypoplasia, but the former usually has lower eyelid colobomas and associated ear anomalies.

The radiographic verification of short roots based on the relative root length is based on the ratio between root length and crown length. A ratio of 1:1 or less is defined as a short root anomaly. If the length of the root is shorter than the length of the crown, the term rhizomicry is used,

which is considered extremely rare in generalized form. The patient in the present case revealed a generalized root-to-crown ratio of less than 1:1. Based on the literature, generalized short root anomaly can be divided according to the etiology into familial, syndrome-associated, environmental, and idiopathic. Short roots may also be observed in dental and hereditary disorders such as dentin dysplasia type 1 and dentinogenesis imperfecta. The affected teeth appear clinically normal, but because of their short or almost missing roots, tooth mobility is markedly increased and spontaneous exfoliation may occur. Short roots appear along with rare underlying systemic conditions such as hypoparathyroidism, Stevens-Johnson syndrome, scleroderma, Down syndrome, and Laurence-Moon-Bardet-Biedl syndrome. Some short-stature syndromes (Aarskog syndrome, dwarfism of Seckel, and Rothmund-Thompson syndrome) have also been associated with short roots. In addition, case reports described patients with short stature or short roots or both, but with no recognized syndrome.

Environmental effects during tooth development, such as trauma, periapical infection, and orthodontic or surgical procedures as well as radiation therapy and to a lesser extent chemotherapy for childhood malignancies, may result in diminished root development. An idiopathic generalized short-root anomaly is extremely rare.

The size and morphology of dental roots exert significant implications in several fields of dentistry: periodontology, orthodontics, traumatology and restorative dentistry, and endodontics. Endodontic treatment in teeth with short or nearly missing roots is associated with several risks and special problems. Preparation of access cavities must be carried out with circumspection because the floor of the pulp chamber could at the same time be the end of the root canal system, as in teeth 16, 55, 26, 27, 36, 37, and 46 of the present case (Figs 4a to 4c). Root canal preparation and obturation must be adapted to the situation, especially in teeth with very short, thin roots (teeth 11 and 21) (Fig 4d).

The main problem of the dental treatment in the present case is related to the fact that most teeth of the permanent dentition are retained. The already erupted teeth probably cannot be conserved for a long time due to the disturbed root growth. A toothless jaw will be the consequence. Certainly, an early loss of dentition is associated with anatomical and functional problems. The lack of dental support due to the upgrowth could lead to underdeveloped alveolar processes in addition to problems with the temporomandibular joint because of overload.

In this case, the expected early loss of teeth cannot be treated curatively. The dental therapy approach is to care for and conserve the mixed dentition as long as possible. In patients with short root anomaly, early treatment of caries is essential to prevent endodontic treatment. Preventive measures such as perfecting oral hygiene and fissure sealants are obligatory. A nutrition consultation could be advantageous. Further important aims of dental treatment are reestablishment and normalization of mastication, speech, and swallowing as well as the compensation of the disturbed physiognomy. A complex prosthetic and orthodontic strategy for congenital abnormalities with oligodontia and anodontia is necessary for adequate treatment. In the case of growing patients, fixed partial dentures are not indicated, especially because fixed dentures crossing the center line could lead to transversal growth inhibition. Age of onset, developmental stage, and patient
compliance all may indicate that a removable prosthesis is the treatment of choice.

The three main advantages of a prosthesis are reestablishment of the mastication function,\textsuperscript{52–55} improvement of esthetics\textsuperscript{52–59} through the advancement of a physiologic perioral musculature,\textsuperscript{54,56,58–63} and the advancement of phonetic qualities.\textsuperscript{52,57,61,64,65} The continuous jaw growth and possible dentition change necessitates close monitoring with 3- to 6-month recall intervals for denture adjustments. Potential surgical procedures for the functional and esthetic outcomes are planned for later in life. The final prosthetic restoration will start once the patient reaches adulthood.

In the management of patients with Hallermann-Streiff syndrome, interdisciplinary medical care is important. Periodic ophthalmologic examination should be performed for early diagnosis of eye complications. Although one should pay special attention to ophthalmologic and upper respiratory problems, early consultation with a dentist is important. The predisposition to severe dental caries and dental malformations makes it imperative to perform effective prevention strategies as early as possible. Efforts must be made to prevent root canal treatment.

**REFERENCES**


