Prevalence of Dental Anomalies in Patients with Wolf-Hirschhorn Syndrome

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Introduction

- Wolf Hirschhorn syndrome (WHS, OMIM#194190) is a congenital syndrome with characteristic craniofacial features.
- Cooper and Hirschhorn in 1961 first described a case of WHS with defects of midline fusion, low birth weight, and epilepsy with a genetic deletion on chromosome 4p.
- Wolf et al. in 1965 described a case with similar genetic findings and 'Greek warrior helmet facial appearance' (flat forehead, prominent glabella, hypertelorism, the wide nose continuing to the forehead, high arched eyebrows, exophthalmos, short philtrum, and micrognathia).
- WHS is rare with an incidence of 1 in every 20,000 to 96,000 live births.
- WHS is two-four times as common in females as in males.
- There are no studies undertaken in patients with WHS to comprehensively evaluate craniofacial and oral findings.
- The primary objective was to evaluate all the medical findings while the secondary objective was to evaluate developmental dental anomalies and pathoses (DDAP) in patients with WHS.

Methods

- One million electronic medical-dental records of patients (1-18 years) reporting at Children's Hospital Colorado were screened for WHS diagnosis.
- Twenty-six charts identified with WHS diagnosis were systematically screened by a calibrated examiner for medical and dental information including dental anomalies of shape, number, position, structure, and other developmental anomalies or pathoses.
- Twelve patients with WHS had comprehensively reported dental findings which were used for descriptive statistics.

Table 1: Demoraphic Infomration from Patients with Wolf-Hirschhorn Syndrome (N=26)

Sex	n (percent)
F	18 (69.2%)
M	8 (30.8%)
Race	
African American	1 (3.8%)
Asian	1 (3.8%)
Caucasian	24 (92.3%)
Ethnicity	
Hispanic	5 (19.2%)
Non-Hispanic	21 (80.8%)
Status	
Deceased	4 (15.4%)
Live	22 (84.6%)
Percentile Weight	
Over 5th percentile	1 (19.2%)
Under 5th percentile	21 (80.8%)
Percentile Height	
Over 5th percentile	1 (3.8%)
Under 5th percentile	24 (92.3%)
BMI	
Over 5th percentile	5 (19.2%)
Under 5th percentile	21 (80.8%)
Cardiovascular System	14 (53.8%)
Microcephaly	26 (100.0%)
Respiratory System	16 (61.5%)
Muskuloskeletal System	19 (73.1%)
Immunological System	2 (7.7%)
Endocrine System	5 (19.2%)
Genitourinary System	18 (69.2%)
Central Nervous System	21 (80.8%)
Peripheral Nervous System	22 (84.6%)
Gastrointestinal System	22 (84.6%)
Hepatobiliary System	0 (0.0%)
Behavioral	24 (92.3%)
Otorhinolaryngological System	18 (69.2%)
Hematological	6 (23.1%)
Oncological	1 (3.8%)
Opthalmological	13 (50.0%)
Impaired Communcation	25 (96.1%)
Allergies	16 (61.5%)

Table 2: Craniofacial, dental anomalies and habits in patients with Wolf-Hirschhorn Syndrome (N=12)

Extraoral Features	
Category	**n (percent)**
Microcephaly	12 (100%)
Dysmorphic facial features	12 (100.0%)
Maxillary prognathism	8 (66.7%)
Mandibular prognathism	1 (8.3%)
Lip incompetence	4 (33.3%)
Facial Asymmetry	3 (25.0%)
Shape Anomalies	
Taurodontism	6 (50.0%)
Pyramidal molars	4 (33.3%)
Dilacerated roots	4 (33.3%)
Microdontia	2 (16.7%)
Dens Invaginatus	2 (16.7%)
Pulp stones	2 (16.7%)
Localized short root anomaly	1 (8.3%)
Radiculomegaly	1 (8.3%)
Number Anomalies	
Hypodontia	6 (50.0%)
Oligodontia	4 (33.3%)
Hyperdontia	1 (8.3%)
Positional Anomalies	
Rotation	3 (25.0%)
Ectopic eruption	2 (16.7%)
Crowding	2 (16.7%)
Infraocclusion	1 (8.3%)
Distally displaced premolars	1 (8.3%)
Impacted teeth	1 (8.3%)
Structural Anomalies	
Hypoplastic teeth	7 (58.3%)
Other Developmental Dental Anomalies	
Bifid mandibular canal	1 (8.3%)
Delayed eruption	9 (75.0%)
Non-nutritive habits	
Nocturnal or diurnal bruxism	8 (66.6%)
Pica	5 (41.6%)

Results

- All the reviewed WHS patients had otorhinolaryngological, speech or behavioral findings while four-fifths of the patients had gastrointestinal, musculoskeletal, central, or peripheral nervous system findings.
- Two-thirds of the WHS patients had respiratory and genitourinary findings while ophthalmological and cardiovascular findings were documented in about half of the cases.
- The WHS patients had positive craniofacial findings including microcephaly and dysmorphic features while two-thirds had maxillary excess.
- Microdontia (83.3%), pyramidal molars (66.7%), taurodontism (50%), dilacerated roots (33.3%), dens invaginatus (16.7%), pulp stones (16.7%), and root anomalies (16.7%) were the common shape anomalies.
- There was a high prevalence of number anomalies (hypodontia:50.0%) while the most common positional anomalies included rotated (25%), ectopic (16.7%), infra-occluded (8.3%), distally displaced (8.3%), or impacted (8.3%) teeth.

Highlights and Caveats

- This is the first study that comprehensively evaluated ~80 dental anomalies and pathoses in patients with WHS.
- While the small sample size is a caveat, the majority of the reported studies have even smaller sample size and limited number of dental anomalies and pathoses evaluated.

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