

## Williams syndrome — oral presentation of 45 cases

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### Abstract

Forty-five patients with Williams syndrome (WS) were evaluated for oral abnormalities. The mean age of the patients was 9.25 years, the median age was 6.7 years, and the majority (62.2%) were male. Hypodontia was present in 11.1% of the patients. Abnormal tooth morphology was noted in 12.5% of the primary dentitions and 40.7% of the permanent dentitions. With the exception of the primary mandibular central incisors of males, all mesiodistal incisor crown dimensions were statistically significantly smaller when compared with norms ( $P < 0.05$ ). At least one hypoplastic enamel defect was present in 9.4% of patients with primary teeth and in 18.5% with permanent teeth. No patients exhibited generalized enamel hypoplasia. More than half of the patients (59.1%) were both caries and restoration free, while only 13.6% presented with clinically active caries. Tongue thrusting was present in 67.7% of the sample, while more than 50% of the patients presented with excessive interdental spacing. Patients exhibited a higher than normal prevalence of Class II and III occlusions, open and deep bites and anterior crossbites. No single dental finding was pathognomonic of WS, however two constellations of findings, each occurring in approximately one-third of the sample, were observed: 1) microdontia, anterior crossbite, tongue thrusting, and excessive interdental spacing, and 2) microdontia, deep or open bite, and excessive interdental spacing. (*Pediatr Dent* 16:262–67, 1994)

### Introduction

Williams syndrome (WS) is a multisystem congenital disorder estimated to affect 1/20,000–1/50,000 individuals.<sup>1</sup> Williams syndrome was initially described three decades ago as a characteristic triad of anomalies: distinctive facial appearance, supravalvular aortic stenosis (SVAS), and mental retardation.<sup>2,3</sup> We now realize that WS produces a broader array of problems including vascular stenosis other than SVAS, hypertension, infantile hypercalcemia, poor growth and/or failure to thrive during infancy, musculoskeletal problems, renal anomalies, learning disabilities/attentional disorders, and/or mental retardation.<sup>1,4–13</sup>

Although mentioned in some previous reports, oral findings in WS have received scant attention in the literature. Williams et al.<sup>2</sup> noted that some patients demonstrated malocclusion and mandibular prognathism, however they did not consider these findings part of the syndrome. Beuren et al.<sup>4</sup> were the first to suggest that dental abnormalities were a consistent component of WS. Two of the WS males in their sample presented with hypodontia, microdontia, abnormal bud-shaped molars, and a broad upper dental arch causing bilateral posterior buccal crossbite. Dental changes in their eight female patients were less pronounced. They also noted that the average mesiodistal measurements of the upper four incisors for three patients equaled 26 mm, which was well below the average norm of 35 mm.

Other studies have reported that certain dental abnormalities appear to be more common in WS. Most of these studies report hypodontia in the majority of WS patients. Other abnormalities reported with less consistency include microdontia, a high caries rate, and a

myriad of other findings.<sup>4,6,8,12,14–24</sup> These findings generally were reported as part of a multisystem review of WS patients, and few involved a systematic and comprehensive oral examination.

The purpose of this study is to report on the oral findings from comprehensive oral examinations of 45 subjects diagnosed with WS. It is hoped that these results will provide more accurate prevalence rates of dental findings in this syndrome, and thus will help families, physicians, and dentists monitor dental development and provide anticipatory guidance. Furthermore, our results suggest that selected dental findings can aid in the diagnosis of WS.

### Methods

Forty-five patients with WS, previously evaluated and diagnosed by one of the authors (BP) in a multidisciplinary WS clinic, were referred to the department of dentistry, Children's Hospital, Boston, for oral evaluation. Dental histories were obtained from the patient's parents to assess general dental care, dental development, orthodontic treatment, and use of a nursing bottle. Radiographs were not exposed as part of this study.

Oral examinations by two of the authors (JH and LN) and photographs were used to evaluate patients for:

1. Hypodontia
2. Anomalous tooth morphology
3. Mesiodistal dimensions of the maxillary and mandibular incisors
4. Enamel hypoplasia
5. Presence of clinically active caries and/or restored teeth

6. Occlusion including excessive spacing or crowding, and swallowing pattern.

Excessive spacing was considered as generalized lack of interproximal tooth contacts in the permanent dentition and generalized spacing greater than 2 mm interproximally in the primary dentition. The mesiodistal dimensions of the maxillary and mandibular incisors were obtained by authors JH and LN measuring the greatest mesiodistal width of each tooth using a Boley gauge. The measurements obtained were compared with Moorrees' norms<sup>25</sup> using an unpaired *t*-test. Interexaminer reliability evaluations were performed by two examiners on 25 randomly selected patients.

## Results

Forty-five WS patients, with a mean age of 9.25 years (median age of 6.7 years; range of 13 months to 28 years) were examined. Sixty-two percent (28/45) were male and 38% (17/45) were female. At the time of presentation 40.0% (18/45) of the WS patients were in the



Fig 1. Typical facies of patient with Williams syndrome.

primary dentition, 31.1% (14/45) in the mixed dentition, and 28.9% (13/45) in the permanent dentition. Seventy-one point one percent (32/45) presented with at least one primary tooth, and 60.0% (27/45) presented with at least one permanent tooth. General observation and parental history indicated that all patients were grossly within normal limits for dental eruption timing and sequence. Fig 1 demonstrates the characteristic facial appearance of a child with WS;



Fig 2. Primary dentition exhibiting microdontia, excessive interdental spacing, anterior crossbite, posterior crossbite, and deep overbite.

Fig 2 and 3 demonstrate selected dental findings in the primary and mixed dentitions, respectively.

## Tooth abnormalities

Hypodontia was present in 11.1% (5/45) of the patients — 3.1% (1/32) of the patients with primary teeth (absent lower left central incisor) and 14.8% (4/27) of the patients with permanent teeth. The missing permanent teeth included maxillary right lateral incisors ( $N = 2$ ), a mandibular left central incisor ( $N = 1$ ), and a mandibular right second premolar ( $N = 1$ ).

Abnormal primary incisor morphology was evident in 12.5% (4/32) of the patients, with three of these presenting as peg-shaped incisors and the other as a bilateral fusion of the mandibular primary lateral incisor and canine. Abnormal primary molar morphology was noted in 9.4% (3/32) of the patients, which included a bud-shaped primary maxillary molar ( $N = 1$ ), a triangular maxillary second molar missing a distolingual cusp ( $N = 1$ ), and a maxillary second molar with an exaggerated trapezoidal shape ( $N = 1$ ).

Abnormal permanent incisor morphology was noted in 40.7% (11/27) of the patients with the majority (8/11) presenting as peg-shaped lateral incisors. Abnormal permanent molar morphology presented in 7.4% (2/27) of the patients as either absence of distolingual cusps on the first or second maxillary molar ( $N = 1$ ) or as an exaggerated trapezoidal shape of the maxillary first molar ( $N = 1$ ).

Table 1 demonstrates the analysis of the mesiodistal crown dimensions of the primary and permanent maxillary and mandibular incisors for both male and female patients. With the exception of the primary mandibular central incisors, the mesiodistal crown dimensions of all primary and permanent maxillary and mandibular incisors for males were statistically significantly smaller when compared with Moorrees' norms. For females, all of the primary maxillary and mandibular incisors were also statistically significantly

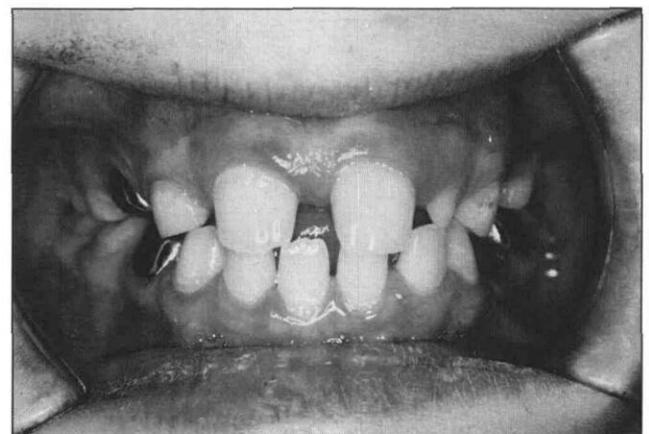


Fig 3. Mixed dentition exhibiting microdontia, hypodontia (maxillary right lateral incisor), peg-shaped maxillary left lateral incisor, enamel hypoplasia, and open bite tendency.

**Table 1. Mesiodistal crown dimensions**

	Primary				Permanent			
	N	WS x̄	Moorrees x̄	P	N	WS x̄	Moorrees x̄	P
<b>Female</b>								
Max. central incisor	6	5.80	6.44	≈ 0.001	8	8.09	8.40	> 0.10
Max. lateral incisor	7	4.81	5.23	< 0.01	7	6.14	6.47	> 0.10
Mand. central incisor	6	3.56	3.98	< 0.01	8	4.87	5.25	< 0.01
Mand. lateral incisor	5	4.17	4.63	< 0.02	7	5.50	5.78	< 0.09
<b>Male</b>								
Max. central incisor	13	5.77	6.55	< 0.001	10	7.90	8.78	< 0.001
Max. lateral incisor	13	4.82	5.32	< 0.001	9	5.88	6.64	< 0.001
Mand. central incisor	12	3.74	4.08	> 0.05 < 0.1	12	4.89	5.42	< 0.01
Mand. lateral incisor	12	4.35	4.74	< 0.05	11	5.39	5.95	< 0.001

smaller when compared with Moorrees' norms. In the permanent dentition of females, only the mandibular central incisors were significantly smaller. There was a tendency for the permanent mandibular lateral incisors to be smaller, however the difference did not reach statistical significance. Investigator inter-reliability of these measurements yielded a weighted Kappa score of 0.92.

At least one hypoplastic enamel defect was present in 9.4% (3/32) of the patients with primary teeth and 18.5% (5/27) of the patients with permanent teeth. Examples of localized hypoplastic enamel defects noted in the primary dentition include notching of the central incisors (N = 1), pitting of the facial surface of the primary canine (N = 1), and enamel opacities on the facial surfaces of all primary second molars (N = 1). None of the patients who presented in the primary dentition exhibited generalized enamel hypoplasia. Enamel hypoplasia of permanent teeth included incomplete cuspal formation of the permanent mandibular first molars (N = 2), incomplete formation of incisal edges of mandibular central incisors (N = 1) and enamel opacities on the facial surface of a permanent incisor (N = 1). The patient with the enamel opacity had a history of trauma to the preceding primary teeth, which may have caused the hypoplastic defect.

**Caries**

More than half the patients, 59.1% (26/44), were free from both caries and restorations, while an additional 27.3% (12/44) had no clinically active caries but at least one dental restoration. Clinically active caries was evident in 13.6% (6/44) of the patients. One patient was excluded from the dental caries analysis because of incomplete data.

**Occlusion**

Of the patients with a primary molar occlusion, 81.8% (9/11) presented with a mesial step terminal plane and 18.2% (2/11) with a flush terminal plane. Seven patients did not have sufficient primary molar eruption to allow for molar classification. Of the patients with a permanent molar occlusion, 59.1% (13/22) presented

**Table 2. Vertical, horizontal, and transverse occlusal findings**

	Primary (N = 15)		Permanent/Mixed (N = 22)		Total* (N = 37)	
	No.	%	No.	%	No.	%
Overbite						
Normal	1	6.7	6	27.3	8	21.6
Deep	9	60.0	8	36.4	17	45.9
Open	4	26.7	8	36.4	12	32.4
Overjet						
Normal	6	40.0	6	27.3	12	32.4
Excessive	3	20.0	8	36.4	11	29.7
Zero/negative	6	40.0	8	36.4	14	37.8
Anterior crossbite	6	40.0	9	40.9	15	40.5
Posterior crossbite	4	26.7	4	18.2	8	21.6

\* Eight patients were excluded from these occlusal observations due to either a history of orthodontic therapy (N = 5) or lack of documentation (N = 3).

with an Angle's Class I occlusion, 27.3% (6/22) with a Class II, 4.5% (1/22) with a Class II subdivision, and 9.1% (2/22) with a Class III. Five patients had orthodontic therapy and were not included in permanent molar classification.

Table 2 demonstrates the vertical, horizontal, and transverse occlusal findings. Eight patients were excluded from these occlusal observations due to either a history of orthodontic therapy ( $N = 5$ ) or lack of documentation ( $N = 3$ ).

The distribution of the anterior bite depth of the primary incisors was: normal overbite — 6.7% (1/15), deep bite — 60.0% (9/15), and open bite — 26.7% (4/15). The distribution of anterior bite depth of the permanent incisors was: normal overbite — 27.3% (6/22), deep bite — 36.4% (8/22), and open bite — 36.4% (8/22). When both the primary and permanent incisor vertical dimensions were combined, the most prevalent anterior vertical relationship was a deep bite (45.9%, 17/37), with the remaining presenting with an open bite (32.4%, 12/37), or a normal overbite (21.6%, 8/37).

The distribution of the anterior horizontal relationship in the primary dentition was: normal overjet — 40.0% (6/15), zero or negative overjet — 40.0% (6/15), or excessive overjet — 20.0% (3/15). The distribution of the anterior horizontal relationship in the mixed/permanent dentition was: normal overjet — 27.3% (6/22), excessive overjet — 36.4% (8/22), or zero or negative overjet — 36.4% (8/22). When both the primary and permanent dentitions were combined the most prevalent relationship was a zero or negative overjet — 37.8% (14/37), with the remainder presenting with either a normal overjet — 32.4% (12/37), or an excessive overjet — 29.7% (11/37).

Anterior crossbites were present in 40.5% (15/37) of the WS patients examined. The prevalences for the primary dentition and mixed/permanent dentitions were similar, 40.0% (6/15) and 40.9% (9/22), respectively. Posterior crossbites were present in 21.6% (8/37) of the patients — 26.7% (4/15) in the primary dentitions and 18.2% (4/22) in the mixed/permanent dentitions. Both anterior and posterior crossbites were present in 8.1% (3/37) of the patients.

The majority of WS patients, 51.4% (19/37), presented with excessive interdental spacing, with similar proportions in the primary dentition, 46.7% (7/15), and the mixed/permanent dentition, 54.5% (12/22). Normal spacing was present in 40.0% (6/15) of the patients in primary dentition, while crowding was evident in 13.3% (2/15) of the patients. None of the patients in the mixed/permanent dentition presented with crowding, while 45.5% (10/22) had normal spacing.

A tongue thrust pattern of swallowing was present in 67.7% (28/42) of the patients. The prevalences of tongue thrusting for WS patients in various stages of development were: primary dentition — 66.7% (10/

15), mixed dentition — 92.9% (13/14), and permanent dentition — 38.5% (5/13). Three patients were excluded from this analysis because of lack of cooperation during the examination.

## Discussion

This is the first study to examine systematically and comprehensively the oral findings of a large sample of patients with a confirmed diagnosis of Williams syndrome. Although most of our patients were male (62.2%), this is likely due to small sample size, as WS affects males and females equally.

Our most common oral findings included microdontia, tongue thrusting, excessive interdental spacing, anterior crossbite, anterior vertical dental discrepancies (deep or open bite), abnormal incisor morphology, and hypodontia.

The single most common abnormality noted in this study was microdontia. Our measurements confirmed that microdontia is an almost universal finding in WS. Primary and permanent incisors are approximately 10% smaller than the dimensions presented by Moorrees.<sup>25</sup> Since some of our patients were uncooperative and some had incomplete dentitions we only measured incisors, but our clinical impression was that all primary and permanent teeth were also smaller. Since both primary and permanent dentitions are affected comparably, the etiology is probably genetic rather than environmental in origin.

A tongue thrust pattern of swallowing was present in 67.7% of our sample. With the transition to the permanent dentition, 38.5% of our sample still swallowed with a tongue thrust, having never made the transition to an adult swallowing pattern. In the general population, the maturation of the swallowing pattern normally occurs between the ages of 3 to 6 years with only 10–15% of patients retaining this infantile swallowing pattern.<sup>26</sup>

More than 50% of the patients in our population have excessive interdental spacing in either the primary or permanent dentitions. This characteristic may be the result of both microdontia and tongue thrusting forces often present in WS patients.

The prevalence of Class I normal occlusions plus Class I malocclusions of WS patients (59.1%) was less than that found in the general population (80–85%).<sup>26–28</sup> In contrast, the prevalence of Class II malocclusions (27.3%) plus Class II subdivision malocclusions (9.1%) is higher than the Class II malocclusions (15–20%) found in the general population.<sup>26–28</sup> Class III malocclusion prevalence (9.1%) in WS patients was much greater than the 1% noted in the general population.<sup>26–28</sup>

More than 40% of our patients presented with anterior crossbites, which is extremely high compared with the United States Health Examination Survey finding of 0.8%.<sup>26–28</sup> Since no cephalometric radiographs were

exposed, it is difficult to ascertain whether these anterior crossbites are related to a skeletal discrepancy or are merely dental in nature. However, only two of our WS patients presented with Class III molar relationships, therefore the anterior crossbites are most likely related to incisor position rather than skeletal disharmonies. It is possible that the microdontia might have an adverse effect on tooth bud position, producing abnormal inclinations of the maxillary and/or mandibular incisors. Tongue thrusting can cause labial positioning of the lower incisors and thus contribute to the prevalence of anterior crossbites. In addition to those findings, other studies have reported that the majority of WS patients in their samples had Class II malocclusions.<sup>16, 29-31</sup> Cephalometric analyses of the horizontal skeletal relationships of WS patients are needed to clarify tooth and jaw relationships.

Anterior vertical bite analysis also demonstrated a characteristic pattern. WS patients had a much higher prevalence of both anterior open (32.4%) and deep (45.9%) bite tendencies compared with the general population (1.2-1.5% and 7.6-11.7%, respectively).<sup>26-28</sup>

In contrast to Beuren,<sup>4</sup> abnormal molar morphology was not a common characteristic among our WS patients. Bud-shaped molars were present in only one patient and minor abnormalities in molar shape were present in an additional four patients. Beuren reported the occurrence of bud-shaped molars in two of eight males, and stated that this unusual finding was possibly "specific" for WS. We did find that abnormal incisor morphology, such as peg-shaped permanent incisors (29.6%), was more common in our patients than in the general population, a finding not previously emphasized.<sup>33</sup>

The 14.8% prevalence of hypodontia in the permanent dentition was higher than the 6.4% prevalence reported by Lai and Seow<sup>32</sup> in the general population. We also noted a higher prevalence of hypodontia in the primary dentition (3.1%) than reported by Gorlin et al.<sup>33</sup> in the general population. These findings probably underestimate the true frequency of hypodontia in WS patients since radiographs might have revealed agenesis of permanent teeth that were not yet due to erupt.

Contrary to previous studies, it is our impression (although no specific data were collected) that none of our WS patients has abnormally delayed eruption patterns or abnormal eruption sequences.<sup>18, 19, 23</sup> Future study is needed to accurately document this clinical impression.

Despite previous studies that report that generalized enamel hypoplasia is common in WS patients and leads to extensive dental caries, our results provide no evidence for this finding.<sup>6, 12, 16, 18, 20-22, 24</sup> In fact, our patients presented with a lower prevalence of dental caries than reported in the 1986-87 National Survey of Dental Caries in US School Children.<sup>34</sup>

No single dental finding was pathognomonic of WS, however patterns of oral findings may be characteristic of the syndrome. The most common constellation of findings — microdontia, anterior crossbite, tongue thrusting, and excessive interdental spacing — occurred in approximately one-third of all patients. Another constellation consisted of microdontia, excessive interdental spacing, and either a deep or open anterior vertical overbite, which also affected about one-third of WS patients. These patterns of common dental findings in WS patients — especially when noted in the primary dentition — can help physicians diagnose WS. Most of these dental findings appear to be intrinsic in nature and at the present time cannot be attributed to any pre-, peri-, or postnatal insults.

Routine dental examinations and care are a necessary part of the overall medical supervision for patients with WS. Our findings can assist dentists when counseling parents and caring for patients with WS. Parents should know that these dental findings are not unusual, are also found in the general population, and are amenable to treatment.

Given the variety of occlusal abnormalities in WS, orthodontic treatment may benefit a large number of patients. Orthodontic evaluation and early guidance should begin as soon as patient behavior allows for treatment. Early intervention may minimize or prevent developing malocclusions such as anterior crossbites. Mucogingival problems caused by anterior crossbites and pseudo Class III malocclusions should be treated in either the primary or mixed dentition.

## Conclusions

1. Hypodontia was present in 11.1% of the patients.
2. Abnormal tooth morphology was noted in 12.5% of the primary dentitions and 40.7% of the permanent dentitions.
3. With the exception of the primary mandibular incisors of males, all mesiodistal incisor crown dimensions were statistically significantly smaller compared with norms.
4. At least one hypoplastic enamel defect was present in 9.4% of patients with primary teeth and 18.5% with permanent teeth. No patients exhibited generalized enamel hypoplasia.
5. More than half of the patients (59.1%) were free from both caries and restorations, while only 13.6% presented with clinically active caries.
6. Tongue thrusting was present in 67.7% of the sample, while more than 50% of the patients presented with excessive interdental spacing.
7. The WS patients exhibited a higher than normal prevalence of Class II and Class III malocclusions, open and deep bites and anterior crossbites.
8. No single dental finding was pathognomonic of

WS, however two constellations of findings, each occurring in approximately one-third of the sample, were observed: 1) microdontia, anterior crossbite, tongue thrusting, and excessive interdental spacing, and 2) microdontia, deep or open bite, and excessive interdental spacing.

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