Orodental manifestations of facial port-wine stains

Megan B. Dowling, MD,[a,c] Yueqin Zhao, PhD,[b] and David H. Darrow, MD, DDS[a]
Norfolk, Virginia, and Milwaukee, Wisconsin

Background: Patients with facial port-wine stains (PWS) often demonstrate oral manifestations of their disorder; however, the spectrum and prevalence of such findings among a cohort of patients with PWS has not been established. As a result, dermatologists and oral health specialists may be uncertain how to counsel their patients with PWS regarding oral hypervascularity, bony oral changes, and oral hygiene.

Objectives: We sought to identify physical findings and complications involving the teeth, oral cavity, and perioral structures in individuals with facial PWS.

Methods: This was a cross-sectional study of 30 patients with facial PWS. Descriptive data were collected through anonymous paired surveys completed by patients and their dentists, and analyzed (Fisher exact test) for trends based on physical findings and stage of the PWS.

Results: The most common orodental manifestations according to patients were enlargement of the lip (53.3%), stained gums (46.7%), abnormal bite (30%), and spontaneous bleeding of the gums (26.7%). Staining of the gingiva correlated significantly with gingival hyperplasia (P = .006), maxillary hyperplasia (P = .014), and widened interdental spaces (P = .002), and in all cases gingival staining predated these findings. Lip hyperplasia was reported more frequently by patients than by their dentists (50% vs 18.2%, P = .008). Orodental manifestations were more common among patients with darker and thicker PWS. Hemorrhage after dental procedures was rare (4.5%).

Limitations: Modest sample size and difficulty recruiting control subjects are limitations.

Conclusions: Facial PWS commonly affect the orodental structures, and intraoral staining may predict future complications. (J Am Acad Dermatol 2012;67:687-93.)

Key words: bleeding; dental; face; gingiva; gums; malocclusion; mouth; oral; port-wine stain; teeth.

Port-wine stains (PWS) are congenital vascular birthmarks, occurring in an estimated 0.3% of newborns. Described in the literature as either capillary or venular malformations, these lesions are characterized histologically by ecstatic vessels and a deficiency of nerves in the papillary plexus of the skin in the affected area.[1,2] It is hypothesized that the deficient nerves are of sympathetic origin, and that unchecked parasympathetic influence on blood flow through the postcapillary venules results in progressive vascular ectasia.[1,2] The dilated venules produce a discoloration or "stain" of the skin within the affected region that will be with the patient for life.

Abbreviation used:
PWS: port wine stain

PWS lesions are observed more commonly among Caucasians than African Americans and Asians. There is no gender predilection. They usually begin

From the Department of Otolaryngology—Head and Neck Surgery,[a] and Epidemiology and Biostatistics Core,[b] Eastern Virginia Medical School; and Department of Dermatology, Medical College of Wisconsin.[c]
Funding sources: None.
Disclosure: Dr Darrow is a member of the advisory board for www.birthmarks.com. The authors have no commercial associations, current or within the past 5 years, that might pose a potential, perceived, or real conflict of interest.

Accepted for publication November 6, 2011.
Reprints not available from the authors.
Correspondence to: David H. Darrow, MD, DDS, Department of Otolaryngology, Eastern Virginia Medical School, 600 Gresham Dr, Norfolk, VA 23507. E-mail: David.Darrow@chkd.org.
Published online January 10, 2012.
0190-9622/$36.00

doi:10.1016/j.jaad.2011.11.929

as flat and pink, becoming thicker and darker over time. In advanced lesions, nodules or "cobblestones" may be present.[3] Adjacent bone and soft tissue often becomes hyperplastic; this process is not well understood but is thought to result from the release of growth factors from skin affected by the PWS.

Head and neck PWS usually follow the distribution of the branches of the trigeminal nerve, but more than one branch may be simultaneously involved. When the maxillary and/or mandibular divisions are involved, the bone and soft tissue of the oral cavity are often affected.[4-12] As a result, dentists and dental specialists often anticipate complications such as bleeding caused by the hypervascularity of the gingival and oral soft tissues, along with functional and cosmetic deformities from bony overgrowth involving the jaws and teeth. Although there are a number of case reports describing orodental manifestations of facial PWS,[4-12] to our knowledge, no previous study has established the spectrum and prevalence of such findings among a larger population. The goal of this study was to identify and quantify the common orodental complications linked to PWS of the face.

CAPSULE SUMMARY

- Patients with facial port-wine stains (PWS) often demonstrate oral manifestations of their disorder.
- This article identifies physical findings involving the oral and perioral structures in individuals with facial PWS and identifies patients at risk for future complications.
- Familiarity with the orodental manifestations of PWS may result in improved dental care for patients with PWS.

METHODS

Approval for the study was obtained from the Institutional Review Board of the Eastern Virginia Medical School. Study design was a cross-sectional survey of patients with PWS recruited from the practices of collaborating physicians who treat PWS, and from volunteers who frequent www.birthmarks.com, a World Wide Web site devoted to individuals with PWS. A medical database format was used, collecting experimental (rather than observational) data for variables established prospectively. Patients were eligible for enrollment if they were between 1 and 89 years of age and had a PWS involving the face. Patients were specifically excluded if their PWS did not involve the face, or if the PWS was an infantile "salmon patch" (capillary vascular malformation) involving the forehead ("angel kiss") or nape of the neck ("stork bite").

Enrolled participants were provided a questionnaire to be completed by the participant with accompanying instructions, and a paired questionnaire to be completed by the participant's dentist based on a clinical examination. Data were collected anonymously, although surveys were numbered for purposes of pairing participant questionnaires with those of their dentists.

Collected data were analyzed using Fisher exact test for concordance between patient and dentist, and for trends based on physical findings, facial distribution,[13] and stage of progression[2,3] of the PWS as determined by color, thickness, and/or cobblestone formation. Patients with PWS involving the forehead and scalp were intended to serve as a control group because only those with mid-facial and lower-facial lesions were deemed likely to have oral cavity involvement.

RESULTS

In all, 31 patients with facial PWS were enrolled in the study. Only one patient with solitary involvement of the forehead (V1 [first division of trigeminal nerve] dermatomal distribution) responded, effectively eliminating the control group; this patient was not included in the final study group of 30. Mean age of the participants was 28.8 years (SD ± 19.6; range 1-62). Eleven patients had their PWS in the labio-infraorbito-angular or V2 (second division of trigeminal nerve) dermatomal distribution, 4 had a jaw-neck or V3 (third division of trigeminal nerve) dermatomal distribution, and 9 had a labio-infraorbito-angulo-(forehead)-temporal or combined V1/V2 distribution. Six patients had some combination of these distributions (1 bilateral perioral [V2 and V3], 1 forehead and jaw [V1 and V3], and 4 covering the entire half of their face [V1, V2, and V3]. Four patients reported having Sturge-Weber syndrome (facial PWS, glaucoma, seizures, mental retardation, and ipsilateral leptomeningeal angioma). Fifteen participants described their PWS as confluent (consistent discoloration of skin throughout the affected area), whereas 9 patients reported geographic distribution (patches of normal-colored skin within the PWS); 6 patients did not comment on the consistency of their PWS.

Patients' attention to dental hygiene was assessed to illuminate any confounding impact on orodental complications. Of the 30 participants, one patient reported seeing a dentist less than once a year, 4 went to the dentist yearly, and 25 made two or more