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An unusual pattern of dental damage with salivary gland aplasia

Louis Mandel, DDS

Aplasia, or agenesis of the parotid and submandibular salivary glands, is an uncommon finding that is not always subjectively symptomatic.^{1,2} Any one or a group of these salivary glands may be absent, and their absence can be noted in a variety of unilateral and bilateral combinations.^{1,3-6} Salivary gland aplasia may occur alone^{3,5-10} or in association with other anomalies, particularly defects in the lacrimal apparatus.³⁻⁶ The absence of major salivary glands has been observed in lacrimo-auriculo-dento-digital (LADD) syndrome,^{4,11,12} hemifacial microsomia,^{13,14} mandibulofacial dysostosis (Treacher Collins syndrome),^{6,15,16} multiple facial anomalies¹⁷ and ectodermal dysplasia,^{6,18} and it can be a feature in first and second branchial arch anomalies.^{19,20}

Although heredity is a significant factor,^{3,4,9,10,20,21} there are patients in whom no familial background can be determined.^{2,7,22-24} Males appear to be more prone than females to the development of salivary gland aplasia.^{5,7,10,16,21,25} Facial defects are not obvious because the void caused by the missing gland is filled adequately by fat and connective tissue.^{20,21}

Intraorally, there is an absence of the involved gland's duct orifice and papilla.^{3,16,20,23} Salivary production varies and is dependent on the number of absent salivary glands. Even with the absence of the major salivary glands, mucosal moisture may be apparent, reflecting the continued presence of the minor salivary glands.^{4,20} The subjective symptomatology of xerostomia does not become apparent until unstimulated salivary volume falls below 50 percent.⁶ With the decrease in salivary volume, patients will develop difficulty in swallowing, taste

ABSTRACT

Background. Dental destruction can develop from numerous causes. Major salivary gland aplasia is an uncommon causative factor. The resulting xerostomia can lead to extensive dental demineralization.

Case Description. The author examined a 19-year-old man because of the patient's concern regarding decreased salivary volume and his dental condition. There was extensive loss of tooth structure and an astonishing pattern of dental destruction most notable on the palatal portions of the maxillary molars and premolars that is best described as "chipping." It was only after taking the patient's history, clinically examining the patient and conducting a radioisotope study that the author was able to make a confident diagnosis of the absence of four major salivary glands.

Clinical Implications. Dentists should be aware that salivary gland aplasia is an uncommon cause of dental deterioration. It may manifest itself not by extensive caries but by a dental chipping effect. Early recognition and a therapeutic strategy can prevent progressive dental damage.

Key Words. Salivary gland aplasia; xerostomia; radioisotope.

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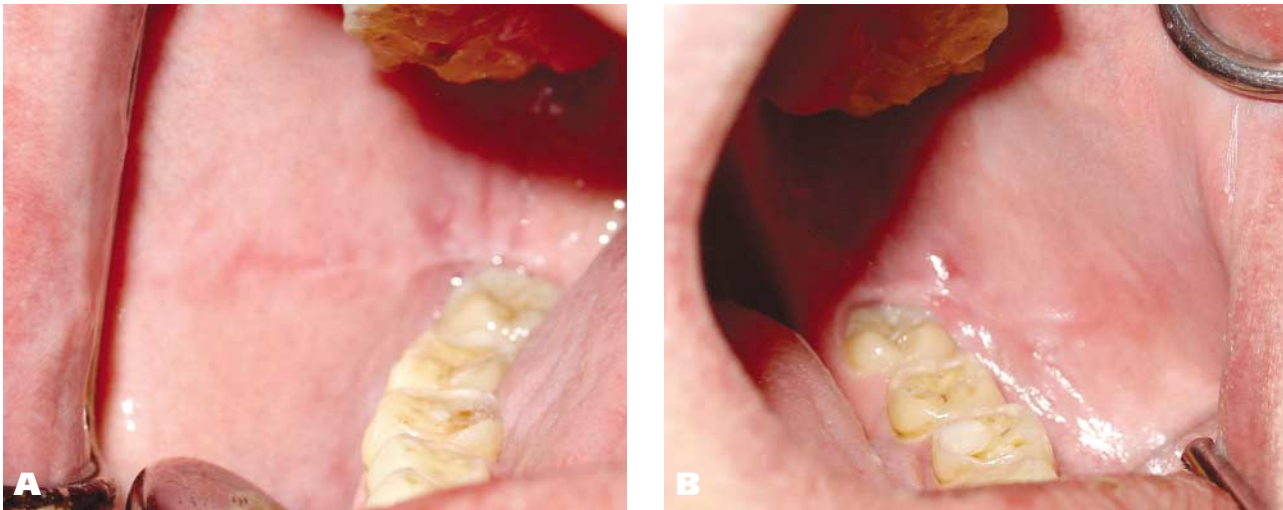


Figure 1. A. Right buccal mucosa with absence of a Stensen's papilla. **B.** Left buccal mucosa with absence of a Stensen's papilla.

alterations, oral burning, lip dryness and, most importantly, dental deterioration. Extensive caries can be anticipated with the loss of saliva's buffering, cleansing and antibacterial capacities.^{8,20,23-25}

The following case report of agenesis of all the major salivary glands in a 19-year-old man derives its significance from the unusual appearance of the dental breakdown. I did not observe rampant caries, but instead noted a pattern best described as "chipping," along with multiple areas of smooth surface enamel loss and dentin exposure. Furthermore, the case is unusual in that I could not determine any familial background, and no associated anomalies were present.

CASE REPORT

A 19-year-old man was referred to the Salivary Gland Center at Columbia University College of Dental Medicine in New York City by his general dentist because of a long history of oral dryness and concern about his dental condition.

A medical history indicated that the patient had no systemic medical problems other than asthma, for which he occasionally used an inhaler. Clinically, I found no extraoral swelling. Although the patient was thin, his facial features were within normal limits. When I palpated the soft tissues in the parotid and submandibular areas, they appeared to be normal, though I could not distinguish a parotid gland bulk or discrete submandibular gland outline. There was no cervical lymphadenopathy.

Intraorally, the mucosa was not inflamed, though it did appear to be somewhat, but not com-

pletely dry. No saliva could be delivered intraorally from either parotid gland or either submandibular gland despite my aggressive massaging of the glands. The patient was missing both his right and left Stensen's papillae (Figure 1). Also, I was not able to identify the orifices of the right and left submandibular ducts or the sublingual carunculae on which these ducts exit. I measured stimulated whole saliva from the patient. I obtained only .2 milliliters per minute (normal mean stimulated whole saliva volume, as determined in the Salivary Gland Center, is 2.1 mL/minute).

The most eye-catching intraoral abnormality was the obvious disintegration of tooth structure, particularly the occlusal-palatal aspects of the maxillary dentition (Figure 2). I noted what could best be described as "chipping" of the palatal cusps and palatal surfaces of the maxillary molars and premolars, which exposed large segments of the underlying dentin. Enamel loss was extensive on all surfaces of the maxillary and mandibular dentition. It was most marked on those surfaces subject to masticatory stress.

A panoramic radiograph revealed almost complete loss of occlusal enamel on all of the teeth except the recently erupted third molars (Figure 3, page 987). Proximal enamel was present. No widespread caries was evident.

To determine the real-time function of all of the patient's major salivary glands, I ordered a radioisotope study with 99m technetium pertechnetate (TPT). The scintiscan revealed the absence of both parotid and both submandibular salivary glands (Figure 4, page 988).

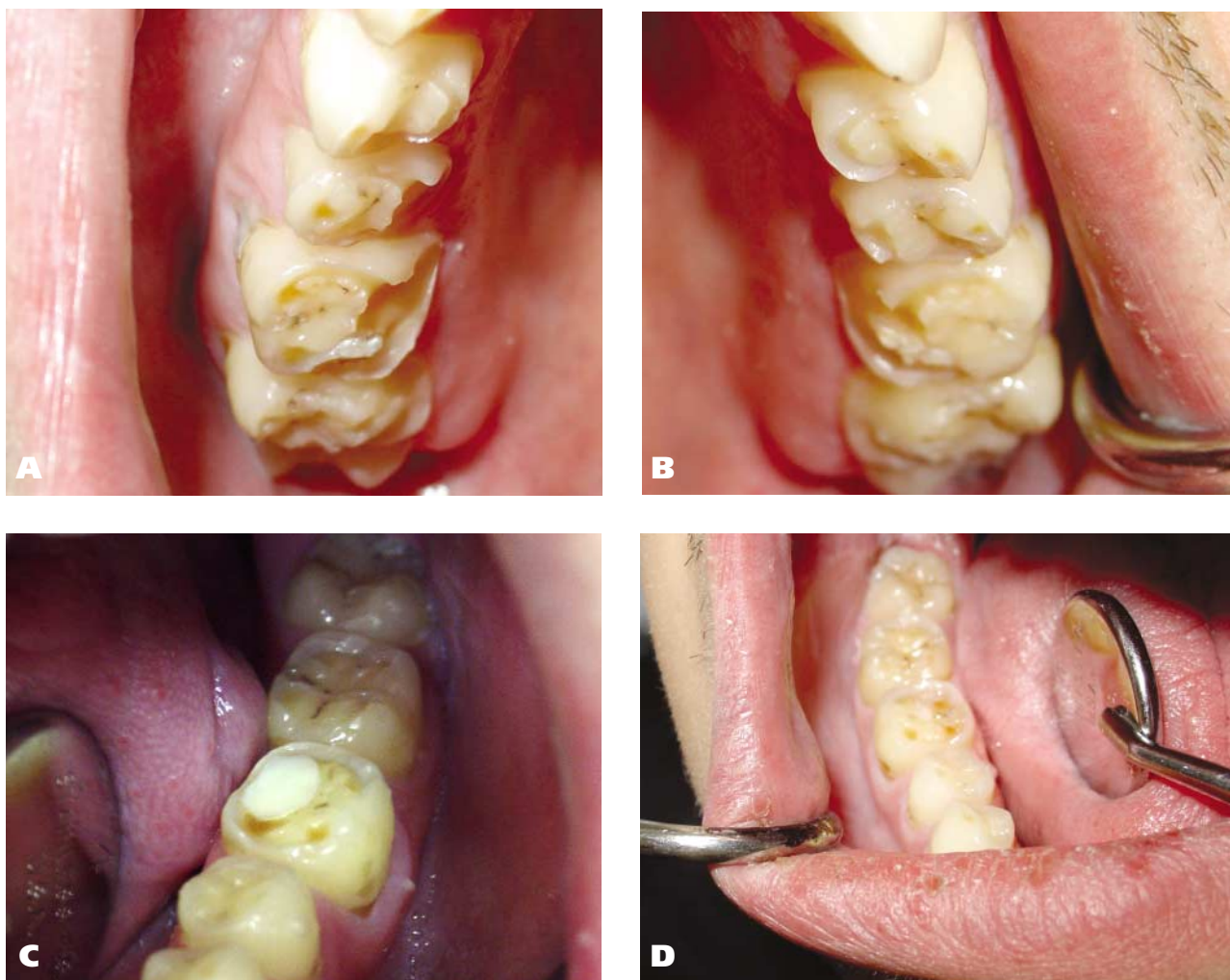


Figure 2. **A.** Maxillary right side. Chipping of palatal cusps and palatal cervical areas exposing large areas of dentin on teeth nos. 2, 3, 4 and 5. Note the peephole effect, or “cupping,” on the buccal cusps of teeth nos. 3, 4 and 5. Buccogingival enamel loss can be seen on teeth nos. 2 and 3. **B.** Maxillary left side. Chipping is most evident on teeth nos. 13 and 14. Peripheral enamel rim is evident on tooth no. 12. Note the peephole effect on the buccal cusps of teeth nos. 12 and 13. **C.** Mandibular left side. Loss of occlusal enamel with exposed dentin encircled by a peripheral white enamel rim on teeth nos. 18 and 19. A resin-based composite restoration is present in tooth no. 19. Buccogingival enamel loss is evident on tooth no. 19. **D.** Mandibular right side. Peripheral white enamel rim surrounding occlusally exposed dentin on tooth no. 30. Buccogingival enamel loss also is visible on tooth no. 30. Note the dry appearance of the lip.

DISCUSSION

Patients with a wide variety of salivary gland disorders, secretory dysfunction or both often are seen in the dental office. Attaining a definitive diagnosis requires an organized investigation into the patient’s complaint. Such an approach involves a complete review of the patient’s history followed by a clinical examination using multiple modalities. The oral examination must be thorough and include a close scrutiny of salivary flow, mucosal state and dental status. A variety of laboratory and imaging procedures are available, and their use is mandatory in

achieving a diagnosis.

Aplasia of the salivary glands is observed most frequently with LADD syndrome, but it often is partnered only with abnormalities of the lacrimal apparatus, which some consider to be an incomplete form of LADD syndrome.^{2,16,23,26} Aplasia only of both parotid and both submandibular salivary glands is an unusual finding in patients with no other anomalies. In a review of the literature, Matsuda and colleagues²¹ found 44 cases of salivary gland aplasia, all of which were associated with a variety of congenital anomalies. Only 13 of these cases had aplasia of all four major salivary glands. Their report also stated that it was diffi-

cult to determine the status of the sublingual salivary gland, which was my experience with my patient in the case I report.

Despite repeated questioning of the patient and his mother, his family history did not include salivary gland aplasia. In cases in which heredity is a factor, salivary gland aplasia is considered autosomal dominant^{2,21,26,27} and may be the result of the effect of a single pleiotropic gene.²⁶ Regardless, salivary gland aplasia can develop in the absence of a familial history,^{2,7,22-24} and it may exist with no associated anomalies.^{1,4-6,8-10,19}

A radioisotope study with TPT is an accurate method of evaluating simultaneously the real-time functioning of all the salivary glands. The radioisotope is introduced intravenously, and then it is picked up and secreted largely by the salivary glands. A gamma camera reads the glandular concentration of the gamma-radiating TPT and digitally illustrates it. In the case I report here, the resulting scintiscan confirmed the absence of both parotid glands and both submandibular salivary glands. Correlation of the radioisotope imaging study with the clinical findings pointed to a definitive diagnosis of agenesis of four major salivary glands.

Despite the patient's long-standing history of oral dryness and the absence of four major salivary glands, his mucosa did not appear to be totally dry. Compensatory salivary flow from the minor salivary glands^{4,19-21,24} probably served to produce enough saliva to prevent the rampant caries that can be anticipated with prolonged salivary diminution. However, there was insufficient saliva to actively buffer the acids in the patient's diet. Therefore, the enamel and to a lesser extent the exposed dentin experienced the demineralizing effects of these dietary acids.

With its buffering capacity and its ability to form a protective enamel pellicle,²⁸⁻³⁰ saliva can control dental decalcification. This physiological protection fails when there is inadequate saliva to prevent demineralization. The time frame in which the enamel is lost depends on the extent of salivary loss, the duration of exposure to the decreased saliva, masticatory stresses, dietary acids and oral hygiene practices.

With demineralization, the softened enamel becomes susceptible to episodes of chipping.⁶ The

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Figure 3. Panoramic radiograph. Occlusal enamel loss is extensive except on recently erupted teeth nos. 1, 16, 17 and 32. Interproximal enamel can be seen on all teeth.

maxillary molars and premolars of the patient in the case I report demonstrated this unusual dental phenomenon (Figures 2A and 2B). During mastication, the palatal occlusal aspects of these teeth are subject to the significant functional stress exerted by the mandibular teeth.

Inevitably, chipping can occur, and large segments of dentin becomes exposed. In my patient, loss of occlusal enamel was apparent in varying degrees on all maxillary and mandibular dental surfaces subjected to physical forces. Exposure of the yellow-hued dentin surrounded by a rim of frosty-white enamel was most pronounced occlusally, where the stresses of mastication resulted in the removal of the acid-softened enamel (Figures 2B, 2C and 2D).

“Cupping,”^{29,31-34} the development of a hollowed-out area that occurs when softer exposed dentin dissolves faster than the surrounding enamel, was present on many teeth (Figure 2). Such a peephole configuration results from point contact during mastication with the cuspal height of an opposing tooth.

The buccal and cervical aspects of all of the patient's teeth, particularly the mandibular posterior teeth, had an increased yellow hue. The softened demineralized enamel, which normally is thin along the gingival tooth surfaces, is thinned further by abfraction and toothbrush abrasion. With enamel loss, the yellow dentin becomes closer to the surface where it becomes more visible or exposed.

Two areas of normal enamel still were evident and could be seen on the panoramic radiograph (Figure 3). Interproximal enamel was present. Although it can be assumed that some demineralization with softening had occurred, I noted that the interproximal location protected most of

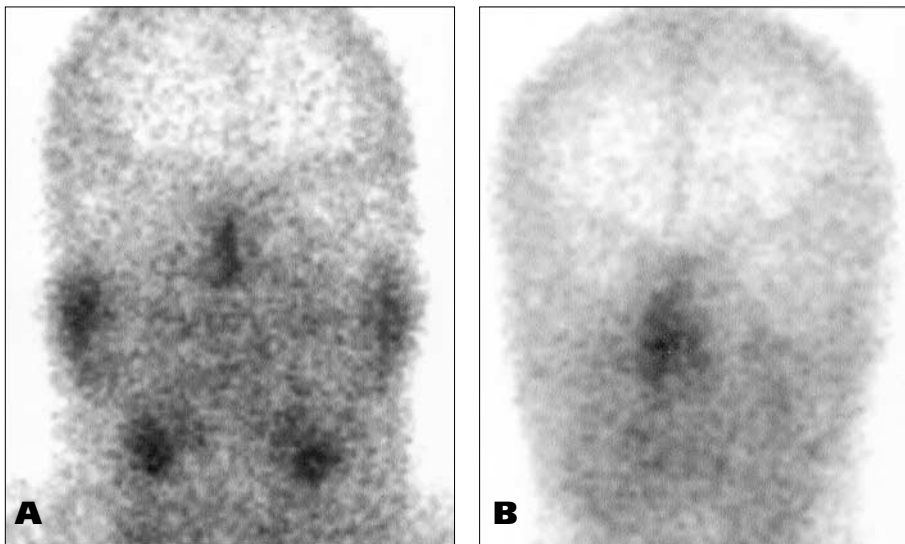


Figure 4. A. A scintiscan of the head of a healthy patient displays bilateral pickup of the radioisotope by the two parotid glands and two submandibular glands. **B.** A scintiscan of the head of the patient in the reported case reveals the absence of the two parotid glands and two submandibular salivary glands. There are no glands present to concentrate the radioisotope.

this enamel from the physical forces exerted by mastication and brushing. In addition, the maxillary and mandibular third molars demonstrated normal occlusal enamel coverage of the dentin. Because the third molars had only recently erupted (the patient was 19 years old), the decrease in saliva and its buffering power had not yet had sufficient time to demineralize the newly exposed enamel.

I noted only minimal caries despite the decreased protective presence of saliva. It is difficult to understand the reason for this fortuitous circumstance. The patient's excellent oral hygiene practices, combined with his use of fluoride toothpaste and topical fluoride applications, may have been the source of the low caries rate.

Despite the extensive amount of dentin exposure, particularly on the maxillary posterior teeth, the patient had no subjective pain complaints. It is possible that the enamel loss, which occurred over many years, had afforded the patient's secondary dentin the opportunity to form and protect the pulp.^{35,36}

There are only two other conditions in which xerostomia is intense enough to initiate dental damage, and they can be differentiated readily from the oral dryness and dental breakdown created by salivary gland aplasia. Sjögren's syndrome, a systemic disease, can cause an oral dryness sufficient enough to lead to extensive caries. Xerophthalmia and xerostomia are classic symp-

oms of Sjögren's syndrome, and often a systemic autoimmune disease is present. The second condition, radiation caries, results from external beam radiation for the treatment of an oral malignancy. A radiation sialadenitis and xerostomia with rampant caries can be the inevitable end product.

TREATMENT

Clinicians can alleviate complaints about xerostomia to some extent by suggesting that patients use salivary substitutes. If residual salivary function from the sublingual or minor salivary glands is present, salivary production can be increased with the use of sialogogic agents such as pilocarpine

or cevimeline. Salivary flow also can be amplified with sugarless chewing gum or sour candy. Hopefully, the increased volume will be sufficient for saliva to carry out its protective functions.

Dental health can be maintained with energetic preventive care.²¹ Use of fluorides in the form of toothpastes, mouthwashes, sealants and topical applications should be encouraged. Oral hygiene must be thorough. Alkaline mouthwashes are helpful.^{32,33,37} Dietary instruction regarding a noncariogenic diet and instruction in the proper method of brushing with a soft-bristled toothbrush are required. Restorations should be placed as deemed necessary by the dentist.

CONCLUSIONS

I have presented a case report of a patient who had a congenital absence of his two parotid glands and his two submandibular salivary glands. The history and clinical evidence of xerostomia, the unusual pattern of dental breakdown best described as "chipping," the absence of duct orifices and the results of a radioisotope study all served to confirm that the patient had salivary gland aplasia. The case is unique in that no other anatomical structures were involved and there was no indication of a similar condition in any family member. ■

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