

# Oral manifestations of Parry–Romberg syndrome: A review of literature

Nader A. Al-Aizari, Saleh Nasser Azzeghaiby, Hashem Motahir Al-Shamiri, Shourouk Darwish, Bassel Tarakji

Department of Oral Maxillofacial Sciences, Al-Farabi College of Dentistry and Nursing, Riyadh, Saudi Arabia

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## ABSTRACT

Parry–Romberg syndrome (PRS) or progressive facial hemiatrophy is a developmental craniofacial disorder of unknown etiology characterized by a slowly progressive unilateral facial atrophy. It is associated with different systemic manifestations particularly, maxillofacial, neurologic and ophthalmologic abnormalities. Dentists must be aware of PRS to identify this invalidating disorder. In this article, we review the etiology, clinical features (especially craniofacial and dental manifestations) and treatment of PRS. We searched in PubMed line using specific words such as PRS from 2008 to 2014 (August). We identify 14 papers have described oral manifestations of this syndrome. We excluded all the article papers that did not indicate to oral manifestations of PRS.

**Key words:** Facial hemiatrophy, oral manifestations, Parry–Romberg syndrome

## INTRODUCTION

Parry–Romberg syndrome (PRS), also known as progressive hemifacial atrophy was first described by Parry in 1825 and later by Romberg in 1846.<sup>[1,2]</sup> This syndrome is an uncommon degenerative condition characterized by a slow and progressive but self-limited unilateral atrophy of facial tissues, including muscles, bones, skin and cartilage. It leads to aesthetic troubles as well as functional and psychological problems due to asymmetry of the face.<sup>[3]</sup>

Etiology is unknown; although trauma, viral infections, genetic factors, autoimmunity, endocrine disturbances, peripheral trigeminal neuritis, increase of cervical sympathetic nerve activity and cerebral disturbance of fat metabolism are possible suggested factors playing a role in pathogenesis of this condition.<sup>[4]</sup>

Parry–Romberg syndrome is associated with several developmental and congenital deformities, such as neurologic, ophthalmologic, cardiac, endocrine, autoimmune, cranio-maxillofacial and orthodontic abnormalities.<sup>[5]</sup> Congenital manifestations that might be associated with PRS are contralateral Poland Syndrome,<sup>[6]</sup> congenital lower

limb hypoplasia,<sup>[7]</sup> congenital ipsilateral cerebral atrophy,<sup>[8]</sup> microphthalmia<sup>[9]</sup> and renal malformations.<sup>[9]</sup> Moreover, hypertrophic cardiomyopathy,<sup>[10]</sup> hypothyroidism,<sup>[11]</sup> rheumatoid arthritis,<sup>[12]</sup> lupus erythematosus<sup>[9]</sup> and scleroderma<sup>[13-15]</sup> are some developmental conditions reported in PRS.

Parry–Romberg syndrome, although rare, has been reported in the literature quite abundantly, and has been associated with multiple findings. The treatment options offered in the literature are diverse but not curative.<sup>[16]</sup>

The present review highlights the etiology, clinical oral manifestations and treatment of PRS.

## ETIOLOGY

Despite PRS being recognized for >150 years, the exact etiology and pathogenesis of this condition is not well understood and seems to be heterogeneous. Cerebral disturbance on fat metabolism, local facial trauma, endocrine disturbances, autoimmunity, heredity, hyperactivity or hypoactivity of the sympathetic nervous system, abnormality of the trigeminal nerve and viral infections, including

**Address for correspondence:** Dr. Bassel Tarakji, Department Oral Maxillofacial Sciences, Al-Farabi College of Dentistry and Nursing, Riyadh, Saudi Arabia. E-mail: [denpol@yahoo.co.uk](mailto:denpol@yahoo.co.uk)

*Borrelia burgdorferi*, are believed to be associated with the pathogenesis of this disease.<sup>[3,17,18]</sup> Tang *et al.*, 2014 believed that some inherent relationship between PRS and the disorder of neural crest cell migration may exist and that malformation or disturbed migration of neural crest cells might be relevant.<sup>[5]</sup>

## SYSTEMIC AND ORAL MANIFESTATIONS

Parry–Romberg syndrome is a degenerative condition characterized by a slow and progressive atrophy of facial tissues, generally unilateral, including muscles, bones and skin.<sup>[3,13,19]</sup> Its onset occurs along first two decades of life.<sup>[13,20]</sup> This syndrome seems to have higher incidence among women and affect most often the left side of the face.<sup>[13]</sup> The skin may become thin, dry and hyperpigmented.<sup>[3]</sup> Ocular involvement is common with enophthalmos as a main manifestation.<sup>[21]</sup> Neurological manifestations such as trigeminal neuralgia, epilepsy, facial paresthesia and migraine may be associated with this condition.<sup>[13,22]</sup>

Several oral manifestations could be associated with PRS.<sup>[5,25,26,27,28,29,30,31,32,33,34,35,36,37]</sup> The oral mucosa and tongue can be affected, also jaws, salivary glands and teeth [Table 1]. There is deviation of the mouth and nose toward the affected side.<sup>[3]</sup> Atrophy of superior lip led to exposure of anterior teeth<sup>[3,21]</sup> [Figure 1].

Intraoral soft tissue and muscles of mastication are also involved in PRS but the normal function like speech, deglutition are not disturbed.<sup>[20,21]</sup> Unilateral atrophy of muscle of the tongue is seen with PRS<sup>[3]</sup> [Figure 2]. The condition may be associated with deficiencies of the soft and hard palates in all dimensions, shortness and deficiency of the mandibular body and ramus. Delayed tooth eruption, root atrophy and retarded tooth formation may also be observed<sup>[23,24]</sup> [Figure 3]. However, the affected teeth are normal and vital clinically.<sup>[3,23]</sup> Frequently, there is unilateral posterior crossbite, as a result of jaw hypoplasia and delayed teeth eruption.<sup>[23]</sup>

**Table 1: Oral manifestations of Parry Romberg syndrome**

Soft tissue changes	Osseous and dental findings
Unilateral tongue atrophy <sup>[5, 16-21]</sup>	Jaw hypoplasia <sup>[5, 17, 18, 20-27]</sup>
Wasting of masticatory muscles <sup>[20, 21, 25]</sup>	Short roots on affected side <sup>[17, 18, 20-22, 24, 27]</sup>
Deficiency of soft and hard palate <sup>[5, 21]</sup>	Delayed ipsilateral tooth eruption <sup>[5, 19, 20, 22-24]</sup>
Rare features	Unilateral crossbite <sup>[5, 19, 20, 22-24]</sup>
Angular cheilitis <sup>[20]</sup>	Malocclusion due to dental crowding <sup>[18, 21, 22, 27]</sup>
Absent parotid and submandibular salivary glands <sup>[25]</sup>	Missing teeth <sup>[21, 25, 28]</sup>
	Rare feature
	Upper canine radiculomegaly <sup>[21]</sup>

## DENTAL CONSIDERATIONS

Importantly, the dental characteristics in treatment of patients with PRS need to be considered. Correction of malocclusion can be restored by orthodontic movement of teeth. Biomechanical components of functional orthodontic



**Figure 1:** Marked hypoplasia of the left side of the face with deviation of lips and nose toward left side and notching of lips and nose with exposure of teeth. Alopecia in left eyebrow region (Deshingkar *et al.* 2012)



**Figure 2:** Unilateral atrophy of tongue papillae of left side (Deshingkar *et al.* 2012)



**Figure 3:** Orthopantomograph showing retarded eruption pattern of teeth on left side compared to that of right side. Decreased vertical height of ramus along with loss of gonial angle prominence on the affected side (Deshingkar *et al.* 2012)

appliance may result in clinically significant morphologic changes in the dentoalveolar and skeletal regions during facial growth. Virtually, the stage of root formation and eruption of permanent teeth is compatible with the active stage of hemifacial atrophy. Accordingly, delayed tooth eruption along with deficient root development appears to occur. Therefore, correcting the eruption problems in patients with PRS can be achieved by application of functional appliance. Prosthetic intervention or additional surgery for the jaws can be used also as a mean to recover the dental occlusion.<sup>[36]</sup>

## TREATMENT AND PROGNOSIS

The disease is self-limiting and has no definite cure. The patients affected should have multidisciplinary attendance, involving experts such as dermatologists, dentists and psychologists.<sup>[31]</sup> The treatment is usually based on reposition of adipose tissue that was lost due to atrophy. Autogenously fat grafts, cartilage grafts, silicone injections and prostheses, bovine collagen, inorganic implants and recently cell fat mixed with platelet gel are some alternatives to aesthetic correction of the atrophy.<sup>[38]</sup> Esthetic effects are the usual results from this disorder rather than disability, especially in mild cases. Indeed, the recovery period for overall prognosis of PRS is unpredictable.<sup>[28]</sup>

## CONCLUSION

Parry–Romberg syndrome is an uncommon poorly understood condition characterized by slow progressive atrophy of one side of the face. Its exact etiology is unclear with unknown pathophysiology. More than an aesthetic concern, the condition also causes functional and psychological problems to patients that necessitate a multidisciplinary team approach to identify treatment expectations of these patients.

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