Dental Anomalies in Parry-Romberg Syndrome

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BACKGROUND

Parry-Romberg Syndrome (PRS) is a rare craniofacial disorder characterized by progressive, inflammatory, **unilateral atrophy** of the skin, tissue, muscle, cartilage, and bone. Estimated incidence is 1:700,000[1].

It is a **self-limiting disease** that typically onsets in the **first two decades of life**.

The etiology of Parry-Romberg Syndrome is unknown. Potential causes include trauma, viral infections, genetics, and fat metabolism disturbances[2]. There are oral health concerns in PRS, but they have been reported primarily in small case reports[3,4].

The aim of this study was to **review the dental findings in patients with Parry-Romberg Syndrome** treated at Boston Children's Hospital **over the past 40 years**.

Additionally, another purpose of this study was to **inform multidisciplinary care** for this patient population.

METHODS

A retrospective review was performed of patients with Parry-Romberg Syndrome who were treated at Boston Children's Hospital between 1980-2020.

All records reporting dental findings were reviewed, including:

- Clinical notes
- Photographs
- Radiographs
- CBCTs

Imaging of patients with documented tooth hypoplasia was reviewed.

When available, CBCT imaging was assessed with 3D measurements of the hypoplastic teeth and compared to the same tooth in the arch on the contralateral side.

RESULTS

61 patients with PRS were treated during the study period.

17 patients (27.8%) were referred for dental evaluation, 9 (14.8%) of which had dental anomalies.

Dental anomalies included:

- tooth hypoplasia (n=5)
- tooth agenesis (n = 5)
- delayed eruption of maxillary canine (n = 4)
- mulberry teeth (n=2)

Demographics	**Dental Referrals** (n=17)
Female (%)	12 (70.6%)
Male (%)	5 (29.4%)
Side of Face Affected	
Left Side Atrophy	9 (52.9%)
Right Side Atrophy	8 (47.1%)
Site of Atrophy	
Cheek	12 (70.6%)
Lips	8 (47.1%)
Zygoma	6 (35.3%)
Nose	4 (17.6%)
Chin	3 (17.6%)

Small numbers limited the ability to statistically compare differences between affected and unaffected sides.

Patient	Agenesis		Hypoplasia		Delayed Eruption		Mulberry Molar	
	Affected Side	Unaffected Side	Affected Side	Unaffected Side	Affected Side	Unaffected Side	Affected Side	Unaffected Side
1			Mandibular lateral incisor, Mandibular canine, Maxillary central incisor					
2	Mandibular second premolar		Mandibular first premolar, Maxillary first molar, Mandibular first molar				Mandibular first molar	
3			Maxillary lateral incisor					
4	Maxillary lateral incisor		Maxillary central incisor					
5			Maxillary central incisor					
6		Maxillary first premolar			Maxillary canine			
7	Mandibular second premolar (retained primary)	Mandibular second premolar (retained primary)			Maxillary canine			
8	Maxillary and mandibular second premolar				Maxillary canine		Mandibular first molar	
9						Maxillary canine		

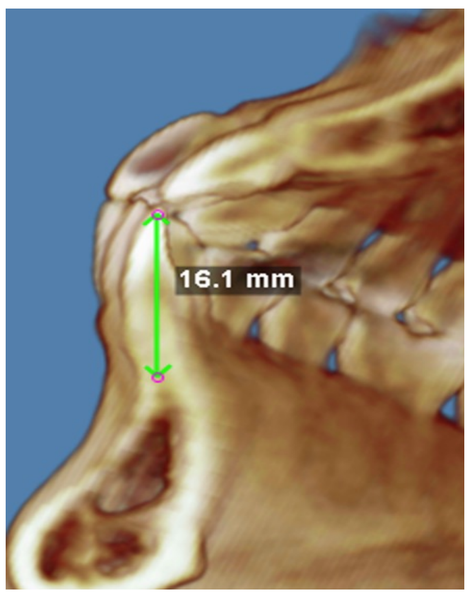
PATIENT CASE

Patient 1 is a 23-year-old with Parry-Romberg Syndrome affecting left cheek, alar base, upper and lower lips and chin.

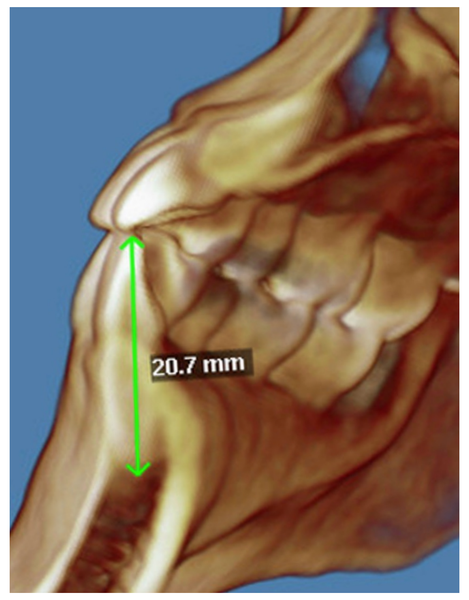
Left upper central incisor tooth was hypoplastic and had been extracted prior to presentation; permanent implant was planned for restoration.

CBCT shows atrophy of alveolus and bone and hypoplasia of left mandibular lateral incisor and canine.

Axial length difference between the mandibular lateral incisor on the affected left side (16.1mm) versus the contralateral side (20.7 mm).



Left mandibular lateral incisor



Right mandibular lateral incisor

CONCLUSIONS

To date, this is **the largest reported series** of dental anomalies in Parry-Romberg Syndrome.

Providers should consider the anomalies found in this study in their evaluation, planning, and treatment of PRS patients.

PRS patients can have **hypoplastic roots with atypical crown morphology**.

Multi-disciplinary care is essential for PRS patients:

- **Dentists** can play a role in identifying **high-risk teeth** and help preserve them with proper care.
- **Orthodontic treatment** may include using **appliances to induce growth of the atrophic side** during pubertal growth
- Due to the progressive nature of this inflammatory condition, **prompt referral to a craniofacial team, rheumatologist, and dermatologist is emphasized.**

REFERENCES

1. Stone J. Parry-Romberg syndrome. Practical Neurology.2006;6; 185-188.
2. El-Kehdy J, Abbas O, Rubeiz N. A review of Parry-Romberg syndrome. *J Am Acad Dermatol*. 2012;67:769-784.
3. Al-Aizari NA, Azzeghaiby SN, Al-Shamiri HM, Darwish S, Tarakji B. Oral manifestations of Parry-Romberg syndrome: a review of literature. Avicenna *J Med*. 2015;5(2):25-8.
4. Yim S, Yang IH, Baek SH. Characterization of dental phenotypes and treatment modalities in Korean patients with Parry-Romberg syndrome. *Korean J Orthod*. 2020;50(6):407-417.