

# Frey Syndrome in Infancy

Carlota Lopes<sup>1</sup>, Laura Martins<sup>1</sup>, Paulo Varela<sup>2</sup>

Port J Pediatr 2023;54(1):43-6

DOI: <https://doi.org/10.25754/pjp.2023.25980>

## Abstract

Frey syndrome is rare in children and often erroneously attributed to food allergy. It is caused by damage to the auriculotemporal nerve, hence the alternative name of the auriculotemporal syndrome. This study describes a case of Frey syndrome in a 2-year-old girl with flushing without sweating on the left cheek after eating tomato and citrus fruit with a medical history of traumatic forceps-assisted delivery.

Recognition of this condition is vital to avoid unnecessary medical costs and procedures and provide reassurance to parents and primary care providers in this benign condition with a self-limited course.

**Keywords:** Child, Preschool; Diagnosis, Differential; Erythema/etiology; Facial Dermatoses/etiology; Mandibular Nerve Injuries; Sweating, Gustatory/complications; Sweating, Gustatory/diagnosis

## Keypoints

### What is known:

- Frey syndrome is a rare condition in children.
- Most reported causes are secondary to a perinatal injury, especially forceps delivery.
- The clinical hallmark is unilateral facial erythema that appears with feeding and resolves within minutes to hours.

### What is added:

- Awareness of this condition is vital to avoid mislabeling the child as food allergic.
- No allergy work-up is necessary for diagnosis in the absence of symptoms beyond localized.
- It is generally a benign self-limiting condition in children and does not require therapy.

## Introduction

Frey syndrome is characterized by recurrent episodes of facial flushing and/or sweating after a salivary stimulus along the distribution of the auriculotemporal nerve which, when injured, shows abnormal regeneration. It is a well-known phenomenon in adults, most often described as a common postoperative complication of parotid gland surgery. In the pediatric population, on the other hand, it is rare. In about half of the reported childhood cases, it is caused by trauma to the parasympathetic branches of the auriculotemporal nerve during forceps-assisted delivery.

This condition is often misdiagnosed as food allergy. Therefore, awareness of this syndrome is essential as its prompt recognition and diagnosis will prevent unnecessary diagnostic testing or exclusion diets.<sup>1-6</sup>

## Case Report

A 2-year-old girl presented to the pediatric clinic for evaluation of an intermittent flushing on the left cheek. This flushing occurred seconds after eating tomato or citrus fruit and each episode would completely resolve after approximately 30 to 40 minutes. The process first became apparent to her parents when she was 12-18 months and started eating that type of solid food. At that time, there was no associated sweating, pruritus, swelling, burning, or painful sensation.

According to her medical history, the girl had been delivered at term with forceps assistance resulting in bilateral malar ecchymosis (Figs. 1 and 2). At 3 weeks of life, she was examined by a pediatric surgeon because of a firm and indurated nodule on the left cheek. She was diagnosed with subcutaneous fat necrosis of the left cheek, and a conservative approach was adopted. The condition resolved at 8 weeks of age.

1. Pediatrics Department, Hospital do Espírito Santo de Évora, Évora, Portugal

2. Dermatology Department, Centro Hospitalar de Vila Nova de Gaia / Espinho, Vila Nova de Gaia, Portugal

### Corresponding Author

Carlota Lopes | E-mail: [carlotalopes@campus.ue.pt](mailto:carlotalopes@campus.ue.pt)

Address: Largo do Senhor da Pobreza, 7000-081 Évora

Received: 30/6/2021 | Accepted: 12/08/2022 | Published online: 10/12/2022 | Published: 01/01/2023

© Author(s) (or their employer(s)) and Portuguese Journal of Pediatrics 2023. Re-use permitted under CC BY-NC. No commercial re-use.



At the 2-year-old follow-up consultation, the parents mentioned the above situation and showed a picture (Fig. 3). It revealed a unilateral distribution of erythematous patches on the left cheek with a clear demarcation at the midline when eating tomato or citrus fruits. This distribution was in the same location as the previous subcutaneous fat necrosis (Figs. 1 and 2).

The case was discussed with a dermatologist. Given the patient's history and physical examination findings, she was diagnosed with Frey syndrome. No further work-up or treatment was considered necessary.

## Discussion

Frey syndrome, also known as auriculotemporal nerve syndrome, is characterized by unilateral facial erythema and flushing along the distribution of the auriculotemporal nerve.<sup>1-4</sup> The most widely accepted



Figure 1. Left facial ecchymosis from forceps assisted delivery.



Figure 2. Right facial ecchymosis from forceps assisted delivery.

mechanism for developing Frey's syndrome is that it results from the lesion of the vegetative fibers from the auriculotemporal nerve, causing an abnormal regeneration. The postganglionic parasympathetic nerve fibers, intended for the parotid, are erroneously connected. They feed the sympathetic nerve fibers innervating the sweat glands and small blood vessels. Therefore, reflex stimulation of parotid-gland secretions does not happen during meals associated with gustatory stimuli. Instead, subcutaneous arterial circulation is vasodilated, and sweat glands are stimulated through sympathetic innervation. Consequently, facial flushing and sweating occur.<sup>1,3-5</sup>

Frey syndrome is more common in adults and usually follows parotid gland surgery. Other potential etiologic factors include traumas / wounds to the parotid region, condylar fracture, herpes zoster, parotitis, diabetes mellitus, central nervous system disorders, such as syringomyelia or encephalitis, cerebellopontine angle tumors, and Pancoast tumors.<sup>5,6</sup>

The disorder is fairly rare in children and mostly caused by secondary to a perinatal injury, especially in forceps delivery.<sup>1-4,7</sup> It is postulated that in children with no history of birth trauma to the parotid area, a congenital aberration of the auriculotemporal nerve pathway exists between parasympathetic and sympathetic fibers. There were reports of bilateral involvement in some cases.<sup>3</sup>

In the literature, in contrast to adults, most children with Frey syndrome exhibit only flushing over the cheek without sweating, possibly owing to the immaturity of their eccrine sweat glands.<sup>7</sup> This has occasionally been referred to as incomplete Frey syndrome or atypical



Figure 3. Left facial flushing along the distribution of the auriculotemporal nerve while eating tomato.

Frey syndrome.<sup>1,2</sup> Many of these cases present when the child is introduced to solid food, as it elicits a stronger stimulation of the parotid gland than does the sucking of formula feeds; therefore, concern for food allergies is common.<sup>3,5,7</sup> Table 1 lists the distinguishing clinical features between food allergy and Frey syndrome.

In Frey syndrome, the presentation tends to be unilateral, along the distribution of the auriculotemporal nerve. It is limited to erythema, lacking sweating, and without itching, angioedema, respiratory, or gastrointestinal symptoms. The erythema typically begins shortly after mastication of the food and lasts 15 to 45 minutes. Food allergies are likely to involve the bilateral face and skin of other anatomic regions and are accompanied by itching, edema, respiratory, or gastrointestinal symptoms. Some commonly reported food triggers of Frey syndrome are citrus, fruits, vegetables, and candy.<sup>3,5,7</sup> Parents often report noting a more vigorous reaction when the child ingests a favorite food item, probably due to more vigorous chewing. Food allergies, on the other hand, are more commonly attributed to cow milk, egg, soy, wheat, peanuts, fish, and shellfish.

Frey syndrome has a benign course in the pediatric age group and is generally self-limited with spontaneous resolution usually occurring within months to a few years. If the diagnosis is correct, it does not require any therapy.<sup>3,4</sup>

In adults, both medical and surgical treatments varying from using anticholinergic agents and intracutaneous injection of botulinum toxin to dorsal sympathectomies and surgical resections of the auriculotemporal nerve have been performed with variable results. Nevertheless, moreover, spontaneous resolution of symptoms has been observed, and most patients are satisfied by an explanation of the condition and reassurance.<sup>5,7</sup>

**Table 1. Differential diagnosis between food allergy and Frey's syndrome**

Food Allergy	Frey's syndrome
Bilateral and nonsegmental distribution	Usually unilateral, along the distribution of the auriculotemporal nerve
Erythema, edema, itch	Erythema only
Extra-facial involvement	Limited to the face
Angioedema might be present.	-
Respiratory / gastrointestinal symptoms might be present.	
Anaphylaxis might be present.	
Triggers: cow milk, egg, soy, nuts, and shellfish	Triggers: green vegetables, citrus fruit, candy, and the child's favorite food

### Author Contributions

LM participated in the study conception or design. CL participated in acquisition of data. CL, LM and PV participated in the analysis or interpretation of data. CL, LM and PV participated in the drafting of the manuscript. LM participated in the critical revision of the manuscript. All authors approved the final manuscript and are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

### Conflicts of Interest

The authors declare that there were no conflicts of interest in conducting this study.

### Funding Sources

There were no external funding sources for the realization of this paper.

### Provenance and peer review

Not commissioned; externally peer reviewed

### Confidentiality of data

The authors declare that they have followed the protocols of their work center on the publication of patient data.

### Consent for publication

Consent for publication was obtained from the legal guardian.

## References

1. Tillman BN, Lesperance MM, Brinkmeier JV. Infantile Frey's syndrome. *Int J Pediatr Otorhinolaryngol* 2015;79:929-31. doi: 10.1016/j.ijporl.2015.03.023.
2. Bourgeois P, Morren MA. Frey's syndrome after herpes zoster virus infection in a 2-year-old girl. *Pediatr Dermatol* 2015;32:e184-5. doi: 10.1111/pde.12604.
3. Listernick R, Legius E, Charrow J. Gustatory flushing (auriculotemporal nerve syndrome) in children with neurofibromatosis type 1 and facial plexiform neurofibromas. *J Pediatr* 2011;158:1034.e1. doi: 10.1016/j.jpeds.2010.12.038.
4. Caulley L, Hong P. Pediatric auriculotemporal nerve (Frey)

syndrome. *CMAJ* 2013;185:504. doi: 10.1503/cmaj.120443.

5. Humphrey J, Black G, Wild L. Facial flushing with food: The auriculotemporal syndrome. *J Gen Intern Med* 2013;28:475-6. doi: 10.1007/s11606-012-2175-5.

6. Lopes T, Barroso L, Mesquita M, Bitoque S, Lopez D, Ferreira A. Síndrome de Frey: Uma visão global do Tema. *Rev Port Estomatol Med Dent Cir Maxilofacial* 2012;54:103-14. doi: 10.1016/j.rpemd.2013.02.002.

7. Hussain N, Dhanarass M, Whitehouse W. Frey's syndrome: A masquerader of food allergy. *Postgrad Med J* 2010;86:62. doi: 10.1136/pgmj.2009.090225.



### Síndrome de Frey na Infância

#### Resumo

A síndrome de Frey é rara em crianças e muitas vezes erroneamente atribuída à alergia alimentar. É causada por dano ao nervo auriculotemporal, daí o nome alternativo de síndrome auriculotemporal. Este estudo descreve um caso de síndrome de Frey em uma menina de dois anos com rubor sem suor na bochecha esquerda após comer tomate e frutas cítricas com histórico médico de parto assistido por fórceps traumático. O reconhecimento dessa

condição é vital para evitar custos e procedimentos médicos desnecessários e para tranquilizar os pais e os prestadores de cuidados primários nessa condição benigna com curso autolimitado.

**Palavras-Chave:** Dermatoses Faciais/etiologia; Diagnóstico Diferencial; Eritema/etiologia; Pré-Escolar; Sudorese Gustativa/complicações; Sudorese Gustativa/diagnóstico; Traumatismos do Nervo Mandibular