

Riga – Fede Disease: a Challenging Disease for Neonates and Parents – an up-to-date Review of the Literature

SUMMARY

Background/Aim: Riga-Fede disease (RFD) is a severe complication of natal and neonatal teeth. It is a benign traumatic granuloma that causes several problems to both the nursing mother and the infant. However, the disease may appear to older children as well. The aim of this study is to investigate the epidemiological and clinical features of Riga-Fede disease, as well as the modern approaches regarding its management. **Material and Methods:** A thorough investigation of modern literature was conducted, using the scientific databases PubMed and Google Scholar. Published articles of case reports, literature and systematic reviews were collected based on the criterion of publication date. An article was considered eligible for our study when published within the last decade (2015–2024). **Results:** RFD is usually developed on the ventral surface of the infants' tongue due to repetitive trauma from natal or neonatal teeth. The histopathological evaluation of RFD's lesions reveals severe eosinophilic inflammatory infiltration with necrotic layers. However, its clinical appearance may be mistaken and lead to a false diagnosis, given its similarity with oral neoplasms, congenital diseases, infection lesions and other pathological conditions of the oral cavity. RFD usually causes pain during breastfeeding. So, there is a risk of insufficient child nutrition, along with dehydration and aspiration danger, as natal and neonatal teeth are usually unsteady. RFD's management depends on the characteristics of each clinical case and includes monitoring, conservative approach or extraction in cases of high health risks for the child. **Conclusions:** RFD can be considered a challenging condition that should be immediately dealt with, in order to avoid the previously stated problems. The severity of its clinical signs and symptoms reveal the need for enhanced education of modern dentists, pediatricians, obstetricians, midwives and any other health specialist responsible for women's and infants' oral health.

Key-words: Eosinophilic Granuloma, Natal Teeth, Neonatal Nursing, Riga-Fede Disease

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Introduction

Riga-Fede disease (RFD) is a benign lingual ulceration attributed to repetitive trauma^{1,2}. The disease was named after the Italian physician Antonio Riga, who initially described it in 1881, and later Francesco Fede, who made the first histological analyses of the disease¹⁻⁴. The most usual etiological factor of RFD is the sharp

incisal edge of one or more newly erupted teeth in the child's oral cavity^{1,2,5}. Teeth that are present at the time of birth are called natal teeth, and these whose eruption is completed within the first 30 days of a neonatal's life are called neonatal teeth^{6,7}. Such types of teeth are a quite rare condition that leads to increased parents' concern about their children's health^{6,8}. Natal or neonatal teeth require immediate care, in order to prevent complications to both the child and the nursing mother^{8,9}.

Material and Methods

The present review of the literature is based on published case reports, literature and systematic reviews from databases PubMed and Google Scholar. A publication was considered as eligible based on its publication type, abstract, publication date, references and scientific credibility. The key-words used in the process of choosing publications for inclusion were “Riga-Fede disease”, “neonatal teeth”, “natal teeth”, “eosinophilic granuloma”, and the publication date range was decided to be 2015 – 2024. In total, 803 and 332 publications were available for consideration in PubMed and Google Scholar accordingly. After thorough evaluation of all 1135 publications and the exclusion of textbooks, letters to the editor and other types of publication except for articles, 21 papers met the inclusion criteria and were included in our literature review.

Discussion

Throughout this review, all co-authors agreed on the multidimensional nature of Riga-Fede disease. Essential up-to-date knowledge is provided by the included publications regarding several aspects of RFD. Thus, it was decided to approach each aspect separately by gathering updated knowledge for each of them. More specifically, RFD's epidemiological and histopathological characteristics, etiology, clinical features, complications, differential diagnosis, as well as management choices were examined as follows.

What is Riga-Fede disease?

Riga-Fede disease is described as a chronic traumatic ulceration, most usually detected on the anterior ventral surface of the tongue in newborns and infants^{4,10}. Other possible oral structures affected may be the child's lips, the buccal mucosa, the dorsal tongue surface, the palate, the mouth floor, or even the gingival tissue⁴. The literature is uncertain about whether RFD presents a significant male or female predominance^{11,12}.

It is mainly associated with natal, neonatal or deciduous lower incisor teeth^{10,13}. Natal teeth have already erupted by the time of birth, whereas neonatal teeth erupt within 1 month since birth⁷. The incidence of natal and neonatal teeth ranges from 1:1000 to 1:30,000 and natal teeth seem to be more usually detected than neonatal teeth^{6,11,14-16}. Other names are congenital teeth, fetal teeth, predeciduous teeth, dentitia praecox and dens connatalis¹¹. Their structure includes dentin and hypoplastic enamel, but the presence of pulp and a normal root is not guaranteed, and their shape may be irregular or sharp^{6,10,14}. Usually, they erupt on the mandibular anterior

region of the oral cavity, but they are smaller, more conical and yellowish in comparison to primary lower incisors^{6,14,15}.

Etiology

The reason of natal or neonatal teeth formation and eruption still remains unknown, but various theories have been proposed^{4,7,11}. These include a possible infection and prolonged febrile incidents during pregnancy, trauma, inadequate nutrition or poor health of the pregnant woman, a superficial position of the tooth germ in the upper or lower jaw of the infant, hyperthyroidism of the mother, or even exposure to environmental harmful factors^{4,6,7,10,11,14,15}. The hereditary nature of natal and neonatal teeth has also been suggested, according to the autosomal dominant trait^{4,7,10,11}. Natal teeth are also known to be present in cases of severe syndromes, such as Pierre-Robin syndrome, Hallermann-Streiff syndrome, Lesch-Nyhan syndrome, Ellis-van Creveld syndrome, Tourette's syndrome, Soto's syndrome, ectodermal dysplasia and congenital pachyonychia^{4,11,13-15}. On the other hand, even though it was stated in the past, no etiological correlation between infant's weight and natal teeth development has been proven⁶.

In contrast, the etiology of Riga-Fede disease is clearly justified, as the main factor is trauma. Repetitive trauma caused by natal or neonatal teeth leads to sublingual, usually, or other soft tissue ulceration lesions in the oral cavity^{2,10,17}. Risk factors that increase the chance of developing Riga-Fede disease include the sucking reflex, uncontrolled tongue movements, macroglossy and neurological disorders^{4,13,17,18}. So, RFD can also be seen during early childhood, even without the presence of natal or neonatal teeth after birth^{1,2}.

Clinical signs & symptoms – Complications – Differential diagnosis

Sometimes, natal and neonatal teeth appear to be quite unsteady into the newborn's oral cavity, surrounded only by gingival tissue without any osseous support^{10,14}. So, they are likely to make breastfeeding challenging for both the infant and the nursing mother¹⁴. Riga-Fede disease is the most severe complication provoked by those teeth¹⁵. Initially, it is a benign ulceration, which may progress to a fibrous mass, as breastfeeding continues^{2,3,18}. Histopathologically, the RFD lesion presents inflammatory infiltration with necrotic superficial layers^{2,3,10}. This is why in the literature one may find the disease named traumatic tongue granuloma, sublingual ulcer, sublingual granuloma, eosinophilic tongue granuloma, traumatic atrophic glossitis or traumatic ulcerative granuloma with stromal eosinophilia (TUGSE)^{2,3,5,19}. However, the diagnosis is possible based on a proper medical history and clinical examination of the child's oral lesions without the need for a biopsy^{17,18}.

Other issues arising from their presence include child's discomfort during suckling, risk of aspiration and pain to the mother's breast^{4,6,9,11,14,15,20}.

It is not unusual for infants with natal or neonatal teeth to refuse breastfeeding due to the irritation and pain they feel, mainly attributed to the lesions of Riga-Fede disease^{2,9,11,14}. As a result, they experience inadequate nutrition and dehydration²⁻⁴. In cases of Riga-Fede disease in older children, the pain level is lower and the mastication process is less discomforting, but pronunciation difficulties are possible². So, if RFD is not diagnosed and managed early, lingual deformity, malnutrition and growth retardation may occur, even though the disease's lesions usually heal without any intervention^{1-3,11}. Also, in case of increased mobility, the natal or neonatal tooth may provoke gingival fibrous dysplasia¹¹.

One should keep in mind that oral mucosa ulcerations may be attributed to several pathological conditions. So, bearing in mind that Riga-Fede disease lesions mimic malignancy, its differential diagnosis includes squamous cell carcinoma, allergic reactions, ulcerative candidiasis, salivary gland tumors, traumatic neuroma, fungal and bacterial infections, tuberculosis, lymphoma, sarcoma, or even agranulocytosis, congenital syndromes, and primary syphilis^{1,2,5,10}.

Some parents firmly believe in myths associated with natal and neonatal teeth. This depends on the culture they belong to, as some communities consider such teeth a good omen (i.e. Malaysia, England etc) and other communities consider them a bad omen (i.e. India, China etc)⁶. This affects the natural expression of parental love and leads superstitious parents to the demand of immediate removal of these teeth¹¹.

Treatment

The management of natal and neonatal teeth and their consequences depends on the special characteristics of each clinical case⁹. Before taking a final decision about the proper management technique, a thorough clinical and radiological examination is essential¹⁴.

In case of no pathological symptomatology, monitoring is proposed for both the baby's teeth and the nursing mother^{9,14}. Certainly, teeth affecting breastfeeding process with pain, ulcers and developmental consequences have to be managed early. The primary aim is to eliminate the traumatic factor^{2,4}. The first choice of treatment is the conservative methodology, which can be applied on steady natal and neonatal teeth causing slight or no irritation to the child and the mother^{4,6}. This method includes modification of feeding habits, use of protective device to cover the infant's teeth or smoothing the sharp incisal edge of these teeth with the use of dental handpieces^{2,3,5}. The use of a mouthwash or a corticosteroid ointment for temporary relief is also suggested in such cases, although their effectiveness is

controversial². Obstetricians and midwives should request a close follow-up visit, in order to guarantee sufficient nutritional intake of the infant through breastfeeding and check the healing of the ulcer¹⁷.

When natal or neonatal teeth are very loopy or are supernumerary or have provoked the formation of painful ulceration (Riga-Fede disease) and there is inadequate nutrition, the conservative approach is not effective^{3,4,6,10}. Extraction of natal or neonatal teeth is suggested and sometimes, when the ulcers do not show any signs of healing within 2 weeks since the extraction, a biopsy may be required for further investigation^{2,10,14,17}. Nevertheless, extraction should be avoided when the natal or neonatal tooth belongs to the normal deciduous dentition¹⁰. There is no risk of bleeding when vitamin K is administered at birth for normal clot formation⁶. Otherwise, the extraction should occur after 10 days since birth, so that normal bacterial flora is well established in the intestine to produced vitamin K^{11,15}. Tooth extraction is also the only proper treatment in case of children with Riga-Fede disease suffering from neuro-developmental abnormalities¹⁸. After the tooth extraction, recurrence of Riga-Fede disease is not expected, but a usual unpleasant outcome is the delayed eruption of primary teeth at the anatomical region of the previously present natal or neonatal teeth⁵.

Midwives' role

Most nursing women are expected to face painful symptoms at the beginning of breastfeeding. So, it is essential to prepare them by teaching pain management and prevention techniques. This goal is possible with training sessions either in well-organized institutions or by a medical professional (obstetrician or midwife) responsible for the woman's pregnancy²¹. Taking into consideration the pain felt by natal and neonatal teeth when breastfeeding, the role of midwives is more than essential. Their support and advices are precious for mothers, so that they continue providing their babies with normal nutrition and overcome psychological inhibitions related to superstitions¹⁴. Otherwise, nursing mothers as well as their husbands think of breastfeeding and healthy child development as an unachievable goal.

Conclusions

Riga-Fede disease can be considered a challenging condition that should be immediately dealt with, in order to avoid all associated complications and difficulties. It may be a rare disease, but its clinical features are upsetting parents and young patients. So, the severity of its clinical signs and symptoms reveal the need for enhanced education of modern dentists, pediatricians,

obstetricians, midwives and any other health specialist responsible for women's and infants' oral health.

References

1. Yürekli A, Dinçer D (2019). "Successfully Treated Riga-Fede Disease". *Dermatol Pract Concept*. **9** (3): 11. doi: 10.5826/dpc.0903a11. PMID: 31384499.
2. Li J, Zhang YY, Wang NN, Bhandari R, Liu QQ (2016). "Riga-Fede disease in a child". *Clin Exp Dermatol*. **41**: 285-286. doi: 10.1111/ced.12728. PMID: 26307375.
3. Khandelwal J, Bargale S, Dave B, Dharmatti S (2021). "Novel management of Riga-Fede disease associated with early infancy tooth in 4-month-old infant". *Int J Med Dent Case Reports*. **12**: 1-3. doi: 10.15713/ins.ijmdcr.171.
4. Kumari A, Singh PK (2019). "Diagnosis of Riga-Fede Disease". *Indian J Pediatr*. **86**: 191. doi: 10.1007/s12098-018-2776-z.
5. Lakshmanan S, Venkataraman S, Singh U (2021). "Riga-Fede disease: a mimicker of malignancy". *BMJ Case Rep*. **14**: e235976. doi: 10.1136/bcr-2020-235976. PMID: 33737272.
6. Shivpuri An, Mitra R, Saxeva V, Shivpuri Ab (2018). "Natal and neonatal teeth: Clinically relevant findings in a retrospective analysis". *Med J Armed Forces India*. **77** (2): 154-157. doi: 10.1016/j.mjafi.2018.07.001. PMID: 33867630.
7. Gouédard C, de Vries P, Darbin-Luxcey C, Foray H, d'Arbonne F (2016). "Natal and neonatal teeth: Update on current knowledge and treatments". *Arch Pediatr*. **23** (9): 990-995. doi: 10.1016/j.arcped.2016.06.007. PMID: 27451381.
8. Anusha Durairaj B, Gajula Shivashankarappa P, Muthukrishnan K, Saraswat Y (2023). "Natal and neonatal teeth: early diagnosis and management". *BMJ Case Rep*. **16** (11): e256249. doi: 10.1136/bcr-2023-256249. PMID: 37989324.
9. DeSeta M, Holden E, Siddik D, Bhujel N (2022). "Natal and neonatal teeth: a review and case series". *Br Dent J*. **232** (7): 449-453. doi: 10.1038/s41415-022-4091-3. PMID: 35396420.
10. Iandolo A, Amato A, Sangiovanni G, Argentino S, Pisano M (2021). "Riga-Fede disease: A systematic review and report of two cases". *Eur J Paediatr Dent*. **22** (4): 323-331. doi: 10.23804/ejpd.2021.22.04.11. PMID: 35034468.
11. Newadkar UR, Chaudhari L, Khalekar YK (2016). "Natal and neonatal teeth: Terminologies with diverse superstitions!!" *J Family Med Prim Care*. **5**: 184-185. doi: 10.4103/2249-4863.184663. PMID: 27453872.
12. Mondal M, Biswas B, Roy A, Kumar P, Bose S, Biswas T, et al. (2014). "Excessive crying due to erupting neonatal tooth: A case report". *Asian J Med Sci*. **6** (4): 87-88. doi:10.3126/ajms.v6i4.11241.
13. Mathew MG (2022). "Riga Fede disease in twins". *BMJ Case Rep*. **15** (11): e252226. doi: 10.1136/bcr-2022-252226. PMID: 36357103.
14. Khandelwal V, Dubey A, Khandelwal S (2015). "Nurses' experience and role in infants with natal teeth". *J Nurs Midwifery Res*. **20**: 409. PMID: 26120344.
15. Aljohar A, Alwakeel H, Palma A (2021). "Multiple natal Teeth in a one-week-old baby: A Case report". *Clin Case Rep*. **9**: 1292-1294. doi: 10.1002/ccr3.3750. PMID: 33768829.
16. Vitali FC, Santos PS, Massignan C, Cardoso M, Maia LC, Paiva SM et al (2023). "Worldwide prevalence of natal and neonatal teeth: Systematic review and meta-analysis". *J Am Dent Assoc*. **154** (10): 910-921.e4. doi: 10.1016/j.adaj.2023.06.017. PMID: 37598330.
17. Lee JJ, Sarangam M, Feldman KW, Tieder JS (2021). "Riga-Fede Disease: A Case of Sublingual Trauma Not Associated With Abuse". *Pediatr Emerg Care*. **37** (12): e1735-e1737. doi: 10.1097/PEC.0000000000001922. PMID: 31714447.
18. Polat Ekinci A, Kılıç S, Babuna Kobaner G (2019). "Early-onset and persistent traumatic granuloma of the tongue (Riga-Fede disease) associated with neonatal teeth and Down syndrome". *J Eur Acad Dermatol Venereol*. **33** (3): e131-e132. doi: 10.1111/jdv.15336. PMID: 30422352.
19. Sarangarajan R, Vaishnavi Vedam VK, Sivadas G, Sarangarajan A, Meera S (2015). "Traumatic ulcerative granuloma with stromal eosinophilia - Mystery of pathogenesis revisited". *J Pharm Bioallied Sci*. **7** (Suppl2): S420-S423. doi: 10.4103/0975-7406.163474. PMID: 26538890.
20. Kérourédan O, Smirani R, Thébaud NB, Devillard R (2018). "Diagnosis and Management of Natal and Neonatal Teeth: Case Report of Three Newborns". *J Dent Child (Chic)*. **85**(2):79-82. PMID: 30345958.
21. Lucas R, McGrath JM (2016). "Clinical Assessment and Management of Breastfeeding Pain". *Topics in Pain Management*. **32** (3): 1-11. doi: 10.1097/01.TPM.0000502820.55789.3a.

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