

## Best Therapeutic Approach for Riga-Fede Disease in Dental setting—A Scoping Review

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### Abstract

**Introduction:** Riga-Fede disease (RFD) is a benign pathological condition characterized by the presence of ulcers on the tongue surface, lips, palate, mucous membrane of the vestibule and the floor of the oral cavity. The lesions are caused by frequent traumatic injuries due to the sharp nature of the lower incisors. **Aims & objective:** To find the best therapeutic approach for riga fede disease in dental setting by means of scoping review.

**Materials & methods:** Comprehensive electronic and manual searches were performed by two independent authors for relevant available articles focused on the topic, published in the last 14 years (between January 2011 and December 2024) accessible on the most important Internet databases (MEDLINE/ PubMed, Embase and Google Scholar). This process was carried out during the month of January 2024. The search strategy was carefully adapted for each database to identify the appropriate studies. The search terms chosen, MeSH or free-text terms, keywords, and Boolean operators, alone or in combination. The full-text version of potentially relevant articles was obtained if the inclusion criteria were fulfilled and if the article contained relevant clinical information.

**Results:** Total of 105 articles screened, out of which 9 articles selected for qualitative data synthesis of scoping review.

**Conclusion:** Most of the natal/ neonatal teeth are prematurely erupted primary teeth. Based on this systematic review, it is concluded that the tooth maintenance and elimination of irritation by conservative means would be an appropriate treatment of choice if the tooth is not severely mobile.

### 1. Introduction

Riga-Fede disease (RFD) is a benign pathological condition characterized by the presence of ulcers on the tongue surface (>50%), lips, palate, mucous membrane of the vestibule and the floor of the oral cavity [1]. The lesions are caused by frequent traumatic injuries due to the sharp action of the lower incisors [2]. In 1881 an Italian doctor named Riga identified this for the first time, but Fede in 1890 conducted the histological studies and described the lesion.<sup>2</sup> Ulcerous lesions involve the maternal breast due to traumatic breastfeeding in many cases [3].

Natal teeth are defined as teeth present at birth and neonatal teeth those that erupt within the first month of life [4]. The incidence is estimated respectively in 1:2000 and 1:3500.<sup>5</sup> Mostly, these teeth are not supernumerary, but in 90–99% of cases they belong to the deciduous dentition [6]. Though the etiology of this condition is not well understood, many hypotheses have been proposed regarding the development of natal and neonatal teeth. Some studies report the autosomal dominant transmission of an unidentified genetic trait [5]. Some authors proposed the possible role of endocrine disorders, especially originated from gonads, pituitary and thyroid glands [5]. Another thesis about the physio-pathological mechanism of natal and neonatal teeth

considers the role of excessive resorption of the overlying bone that would induce an early eruption of the teeth [5].

According to other authors, natal teeth seem to be related more to an accelerated or premature dental development pattern than a superficial positioning of dental germs in relation to the overlying bone [7]. From an anatomical and structural view, natal and neonatal teeth appear underdeveloped and the root formation is frequently incomplete [8]. This leads to high crown mobility which might cause possible accidental avulsion, with the risk of inhalation or ingestion of the tooth [8,9]. The crowns are generally normal both in shape and size. Histologically, they appear hypo mature with well-organized hard tissues [9].

From a histopathological point of view, Riga- Fede disease is characterized by ulcerated mucosal areas, with granulation tissue and mixed inflammatory infiltrate, rich in lymphocytes, mast cells, macrophages and high levels of eosinophils [11]. Thus histologically, Riga- Fede disease is Traumatic Ulcerative Granuloma with Stromal Eosinophilia (TUGSE). A TUGSE lesion often seen in infants (Riga-Fede disease) occurs within 1-week to 1-year [11]. The diagnostic approach consists a clinical evaluation of the ulcerous lesions, considering size and anatomical site, and the research of the pathogenic stimuli linked

to the presence of natal and condition that involves tongue and oral mucosa ulcerations, such as fungal or bacterial infections, allergies or immunological disease, neoplasms (traumatic neuroma, granular cells myoblastoma, lymphoma, lymphangioma, salivary gland tumors or metastatic tumors), primary syphilis, tuberculosis, agranulocytosis, genetic disease [1,2,10]. Syndromes associated with natal teeth are Down syndrome, Carpenter's syndrome, Aicardi–Goutier syndrome, Ellis–van Creveld syndrome, pachyonychia congenita, Hallermann–Streif syndrome, Pierre–Robin sequence, cleft lip and palate, Pfeiffer syndrome, ectodermal dysplasia, craniofacial dysostosis, Sotos syndrome, epidermolysis-bullosa simplex, Van der Woude syndrome, Down syndrome and Walker–Warburg syndrome.<sup>16</sup> It is difficult to diagnose the ulcer typically in infants, because of the appearance of the ulcer which mimics malignant ulcer. Thus, the article is written with the intention to clarify the facts and features of the Riga- Fede disease which will help the clinician for diagnosis and management.

Treatment of the lesion is elimination of the causative factors, varies from the extraction of the natal or neonatal tooth, smoothing of sharp incisal edges of teeth or placing glass ionomer cement over the teeth [12,13].

The aim of this work is to analyze and discuss the different therapeutical approaches to Riga- Fede disease described in the literature by means of a scoping review.

## 2. Methods

The scoping review of the literature was carried out according to the PRISMA (Preferred Reporting Items for Systematic Reviews and Meta-Analyses) statement checklist. Initially, a protocol including all aspects related to review methodology was developed: (1) problem formulation through the design of a well-structured research question; (2) definition of inclusion and exclusion criteria in order to identify relevant studies; (3) literature search and reference list screening; (4) selection and retrieval of studies relevant to the research question; (5) charting of data from selected studies, and finally (6) collating, summarizing and reporting the results from the most important articles on Riga-Fede disease in children. This process completed with the discussion and interpretation of the collected clinical findings of the included studies.

### 2.1. Justification

The significance of the present review was to identify the current therapeutic protocol regarding the general fundamentals of the Riga-Fede disease. This aim may be achieved by providing a detailed synthesis of evidence from available and relevant published studies. We are sure that this review will contribute to the existing literature regarding the topic.

### 2.2. Research question:

The following focused research question was structured prior to conducting the literature search, according to the PICO (Patient, Intervention, Comparison, and Outcome) format: For children with Riga-Fede disease, what is the best management approach in dental clinical setting?

### 2.3. Article selection criteria:

(a) Inclusion criteria:

- Studies where Riga- Fede diseases associated with natal/ neonatal teeth with the treatment mentioned.
- Observational studies on Riga- Fede disease and its management.

(b) Exclusion criteria:

- Articles where the treatment had not been described.

### 2.3. Literature search strategy:

Comprehensive electronic and manual searches were performed by two independent authors for relevant available articles focused on the topic, published in the last 14 years (between January 2011 and December 2024) accessible on the most important Internet databases (MEDLINE/ PubMed, Embase and Google Scholar). This process was carried out during the month of January 2024. The search strategy was carefully adapted for each database to identify the appropriate studies. The search terms chosen, MeSH or free-text terms, keywords, and Boolean operators, alone or in combination, were as follows: “Riga-Fede disease”; “natal tooth”; “neonatal tooth”; “children”; “paediatric dentistry” and “dentistry for children”. The final search algorithm was: (“Riga- Fede disease”[All Fields] AND [“natal tooth” OR “neonatal tooth”[All Fields] AND “children”[All Fields] AND “paediatric dentistry” OR “dentistry for children”[All Fields]). From each database, retrieved article-title lists and abstracts were fully screened. The full-text version of potentially relevant articles was obtained if the inclusion criteria were fulfilled and if the article contained relevant clinical information. A complementary hand search was also performed on the reference list of each selected article.

### 2.4. Article data extraction and assessment:

From each pre-included publication, the following data were collected and put in a spreadsheet: first author/country/year, type of the study, teeth associated and treatment provided. Full-text articles were assessed by independent reviewers in order to judge whether findings from the included articles were relevant to the search question. All the reviewers were previously calibrated, with a Cohen’s kappa interrater agreement score of 0.80. Any discrepancy was resolved by discussion between reviewers for mutual agreement, and if necessary with the intervention of a third reviewer. Finally, descriptive tabulating for included studies was performed.

### 2.5. Charting of data:

For all included studies, specific relevant clinical information concerning the management of children with Riga- Fede disease was collected, including diagnosis methods, age/gender, site of the lesion, healing period, follow up period and special medical considerations (including potential side effects).

## 3. Results

**3.1. Search outcomes:** Figure 1 displays the flowchart of articles screened through the article evaluation and selection processes. The preliminary search generated in total 118 potentially relevant titles/abstracts. After careful screening, only 15 studies were considered eligible and were thoroughly reviewed. Finally, publications fulfilled the selection criteria and were included in the systematic review and the principal characteristics and findings of these studies are described in Table 1.

**Table 1.** Main characteristics of the included study for scoping review.

First author/country/year	Study design	Relevant clinical information
Volpato LE <sup>12</sup> / Brazil/2015	Case report	Two natal teeth (mandibular) Reported with Sublingual ulceration Age/sex: 1 month old/ female Healing period: 15 days Follow up: 1 year Treatment: placement of glass ionomer followed by extraction of teeth as the mobility increased Natal teeth was a part of primary dentition
Shivpuri A <sup>14</sup> / India/2021	Retrospective analysis	12 patients who reported to the dental center between 2012 and 2015 with natal or neonatal teeth was carried out. The age group ranged from 4 to 27 days in which there were eight male and four female infants. All the teeth were white to yellowish white in color, grade III mobile, and were devoid of roots. Only one had ulcer in tongue. All of them were treated with extraction except one case due to lack of consent from parent.
Bulut G <sup>15</sup> /Turkey/2019	Retrospective study	27 infants out of 17,829 infants (2005-2011) 1:660 incidence Periapical radiographs showed that all these teeth were of normal complement of primary incisors. None of the patients exhibited sublingual ulceration. All teeth showed grade 2 mobility except 2 children. Strong family history Treated with extraction under LA due to the risk of aspiration.
Samuel SS <sup>16</sup> /India/2018	Retrospective study	33 babies with a total of 52 teeth were included, of which 28 teeth were natal and 24 teeth were neonatal. All the teeth were located in the mandibular primary incisor region and majority were in pairs. A positive family history was present in eight cases. Extractions were carried out only in cases where the teeth were found to be extremely loose or interfering with feeding. The only local complication noted in this study was Riga-Fede disease. (2012-2014). All the teeth were noted to be prematurely erupted primary teeth rather than supernumerary teeth
Moura LF <sup>17</sup> /USA/2014	Case series	23 cases of children with natal or neonatal teeth. natal teeth (83 percent) mild degree of mobility (64 percent) Tooth maintenance was the most common treatment of choice (64 percent), and only two patients presented Riga-Fede disease (nine percent)
Yen VA <sup>18</sup> / India/2017	Prospective pilot study	Prospective study for a period of seven months. Out of 4,341 children four neonates were born with natal teeth. Our study showed female preponderance and most commonly erupted teeth were mandibular anteriors. The type of delivery had no significant relation with the presence or absence of natal teeth.
Jamani NA <sup>19</sup> /Malaysia/2018	Case report	Age/sex: 1 month old/female Findings: Nipple pain/sublingual ulcer in tongue Treatment: Extraction of neonatal teeth. Grade 2 mobility was noted
Khandelwal V <sup>20</sup> / India/2013	Case report	Age/sex: 13 days old/female Clinical findings: Neonatal tooth in mandibular anterior region. Grade 2 mobility was noted. Extraction was done No follow up is mentioned.
Van der Meij	Literature	Age/sex: 6 months old/ male
EH <sup>21</sup> /Netherlands/2012	Review with	Clinical findings: Sublingual ulcers due to prematurely
	case report	erupted primary lower incisors
		Treatment: Extraction
		Healing period: 15 days
		follow up: 6 months

### 3.2. PRISMA chart:

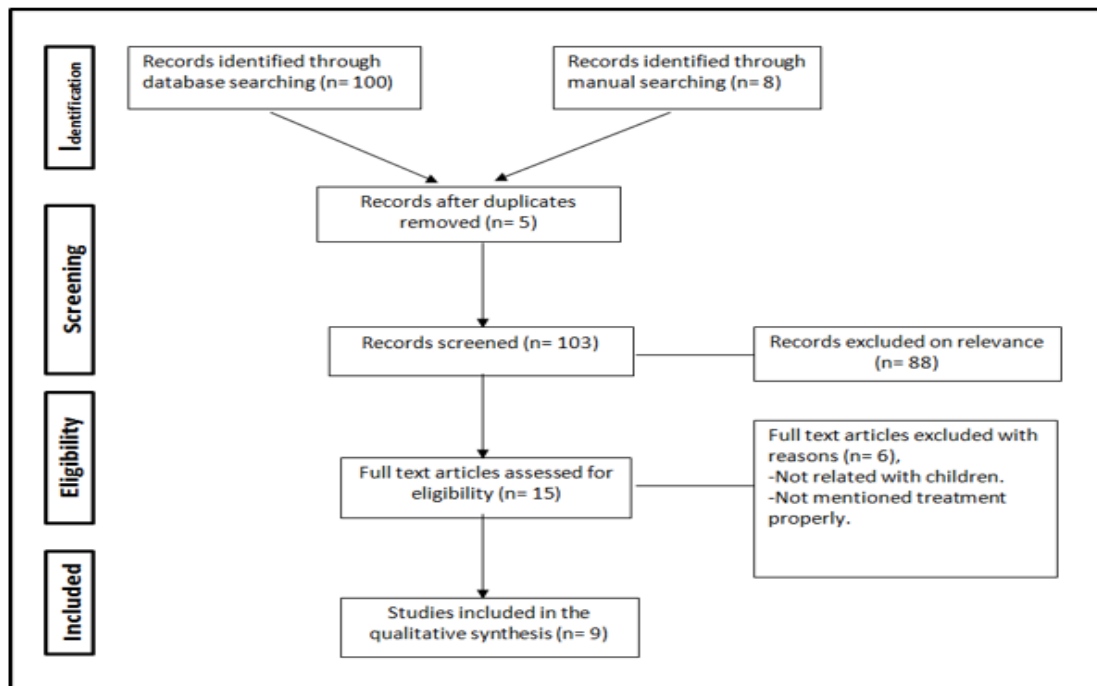


Fig.1. PRISMA flow diagram of literature search

### 4. Discussion

After reviewing the main findings of the included studies, discussion is divided into two parts to make the review reader friendly. 1. Prevalence of natal or neonatal tooth, 2. Therapeutic approach of Riga- Fede disease.

#### 4.1. Prevalence of natal or neonatal tooth:

Bulut G15 et al found that the prevalence of natal or neonatal teeth in children were 1:660. The examination exhibited no evidence of systemic diseases, congenital abnormalities or any syndromes. male: female ratio was found to be 3:2. There was strong family history associated with the development of natal or neonatal teeth. When assessing the location of the erupted teeth, all teeth were observed to be in the mandibular anterior region (100%). More than half of the population had rudimentary roots.

SS Samuel16 et al., reported that all the natal or neonatal teeth were present in the mandibular anterior region and with female predilection. Neonatal teeth are more common than the prevalence of natal teeth. Moura LF17 et al., found the natal teeth was more prevalent and present in mandibular anterior region. 64% of the natal teeth showed mobility and only 4 patients developed Riga-Fede disease.

Retrospective analysis done by Shivpuri A14 et al., showed male predominance of natal teeth. Yen VA18 et al., demonstrated the incidence of natal teeth in their study was 1:1085 with the female prevalence in prospective pilot study. The findings of the study need to be validated by further prospective studies to measure the incidence of natal/ neonatal teeth.

#### 4.2. Therapeutic approach of Riga- Fede disease:

Majority of the studies mentioned extraction of the natal/neonatal teeth as treatment of choice (Shivpuri A14, Bulut G15, Jamani NA19, Khandelwal V20, van der Meij EH21). All the patients showed satisfactory healing after the extraction of natal or neonatal teeth. The reason put forth by them for the

extraction is the mobility of the natal or neonatal teeth which may lead to aspiration as most of the teeth have rudimentary or underdeveloped roots. But all the studies mentioned that the extracted natal or neonatal teeth were the part of primary dentition only.

Only two studies had mentioned maintenance of natal /neonatal teeth as a treatment in which teeth are not severely mobile (Samuel SS16, Moura LF17). Moura LF17 stated that 64% of the children was treated with tooth maintenance in their study and there was no complication like aspiration was reported in follow up. Thus, the tooth can be maintained in the oral cavity by conservative approach to prevent the loss of a component of primary dentition.

Baldiwala M22 et al., reported as case of Riga- Fede disease treated with mouth guard as a conservative approach successfully in a child. Volpato LE12 et al in their article described a case of Riga- Fede disease by two approaches. The child was treated by conservative means initially with placement of glass ionomer cement over the sharp incisal edges, but the tooth was extracted in the follow up as it became mobile.

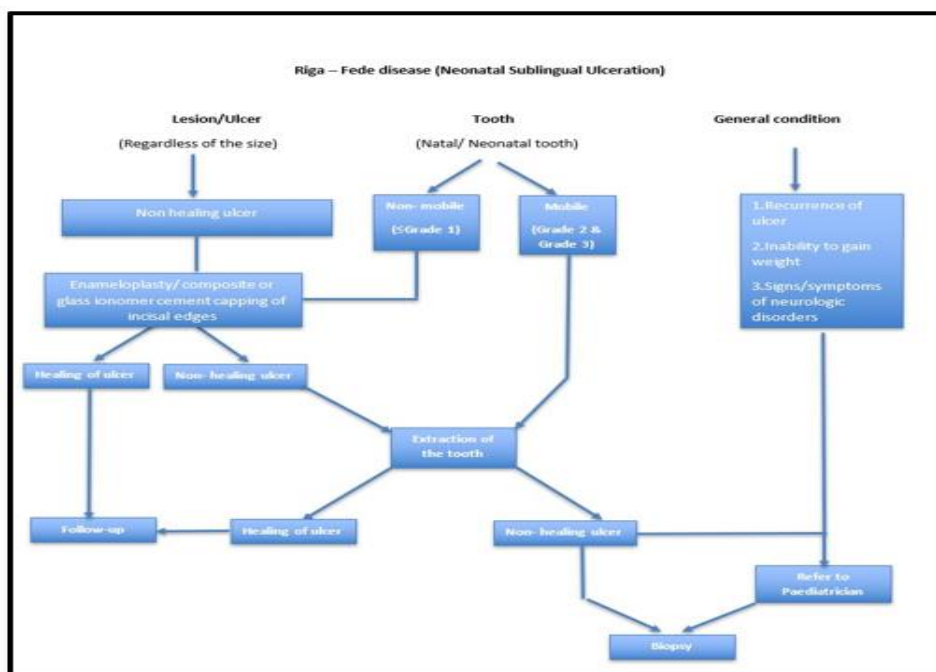
Jamani NA19 et al., stated that the natal teeth were associated with the nipple pain in mother which was stressful for breastfeeding. The proper history regarding the pain or ulcer in breast of the mother should be kept in mind before making the treatment plan. Gardnier23 reported space loss in nine cases after extraction of natal teeth in their study and SS Samuel et al., also stated about space loss in two children treated with extraction of natal teeth. M Sridhar et al24., reported a case of accidental displacement of primary tooth buds during the extraction of neonatal teeth. Newborns have the tendency to bleed more as they have low vitamin K level in blood (Vitamin K Deficient Bleeding)25. So, the extraction should be avoided in newborns who are not administered with vitamin K injection. Such kind of complications should be avoided and kept in mind while making treatment plan.

**Table. 2:** Neurological disorders associated with Riga-Fede disease [26.27.28.29]

Neurological disorders associated with Riga- Fede disease	Manifestation
Familial dysautonomia (Riley- Day syndrome)	Poor sucking and swallowing, choking inspirations, pneumonia, red blotching of skin, diminished/no fungiform papillae of the tongue. Absence of overflow tears with emotional crying can be detected only after 7 months of age.
CIPA syndrome (Congenital Insensitivity to Pain and Anhydrosis)	Episodes of hyperpyrexia due to lack of perspiration. Nerve biopsy reveals total absence of unmyelinated nerve fibers that convey impulses of pain, temperature and autonomic functions.
Hereditary Sensory and Autonomic Neuropathy- Type 3 (HSAN syndrome)	hypotonia, feeding difficulties, poor growth, lack of tears, frequent lung infections, and difficulty maintaining body temperature. Other features include alterations of the taste and the perception of pain, heat, and cold
Lesch- Nyhan Syndrome	impaired kidney function, hyperuricemia, acute gouty arthritis, and self-mutilating behaviors such as lip and finger biting and/or head banging. Additional symptoms include involuntary muscle movements, and neurological impairment.
Tourette syndrome	characterized by sudden, repetitive, rapid, and unwanted movements or vocal sounds called tics. Commonly associated with Attention Deficit Hyper activity Disorder (ADHD).

Neonatal sublingual ulceration is also found in patients with underlying neurological conditions (Listed in Table 2). It might be an early manifestation of the neurological disease in children. Thus, the proper evaluation of child and opinion from the paediatrician are mandatory, if the ulcer is recurring even after

the treatment. Nevertheless, biopsy of the lesion is indicated in nonhealing ulcers which is refractory for the therapeutic interventions. The proposed therapeutic strategies based on the current scientific evidence for the Riga- Fede disease are illustrated in the schematic flow diagram (fig. 2).



**Fig.2.** schematic illustration of management of Riga-Fede disease

## 5. Conclusion

Riga- Fede disease is a benign pathological condition of the oral cavity of newborns due to the sharp incisal edges of the natal or neonatal teeth. Most of the natal/ neonatal teeth are prematurely erupted primary teeth. Based on this systematic review, it is concluded that the tooth maintenance and elimination of irritation by conservative means would be an appropriate treatment of choice if the tooth is not severely mobile. Though the extraction of the mobile natal/ neonatal tooth prevents the

accidental aspiration of tooth, it should be performed in mobile teeth.

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