

Riga-Fede's Disease

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Abstract

Riga Fede's disease (RFD) is a rare ulcerative lesion of the tongue that can be found in newborns with natal or neonatal teeth. Natal/neonatal teeth are the prematurely erupted teeth which usually are the primary lower incisors. When babies are breastfed or bottle-fed, the sharp edges of natal/neonatal teeth injure the ventral surface of the tongue repetitively. These repetitive traumas cause the sublingual ulcerative lesion. RFD lesions cause pain, eating difficulty and bleeding. Prolong tongue trauma may lead to more severe problems which affect the babies' health including inadequate nutrient intake, dehydration and underweight. The early detection and intervention of RFD is very important to prevent the severe complications or permanent damage such as growth retardation and tongue deformity. This review describes the aetiology, clinical appearance, histology, differential diagnosis, signs, symptoms, complications and treatment of RFD.

Riga-Fede's disease is a rare oral lesion that is found mainly on the ventral surface of the tongue in paediatric patients^{1, 2}. The clinical appearance of RFD is an ulcerated circular erythematous lesion, with an indurated border^{3, 4}. Parents usually bring their child to the dentist because of feeding problems. The child often cries, and refuses to eat due to severe pain in the tongue^{5, 6}.

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Definition

RFD is a term for a sublingual ulcerative lesion that is found in a child less than 2 years of age. If the child is older than 2 years old, the lesion would be called an oral traumatic granuloma or a sublingual ulceration instead. RFD was first reported by Antonio Riga in 1881. Subsequently, the histological studies of this lesion were performed by Francesco Fede in 1890. The term "RFD" was used after these two discoveries⁷. Other terms for RFD are related to their aetiology, histopathology or clinical appearance have been also used. (Table 1)

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1.	Cardarelli's aphthae ¹
2.	Dentitia Praecox
3.	Eosinophilic ulcer of the oral mucosa
4.	Lingual traumatic ulceration
5.	Oral traumatic granuloma
6.	Riga's disease
7.	Sublingual fibrogranuloma
8.	Sublingual granuloma
9.	Sublingual growth in infants
10.	Sublingual ulcer
11.	Traumatic atrophic glossitis
12.	Traumatic eosinophilic ulceration of the tongue
13.	Traumatic granuloma of the tongue
14.	Traumatic sublingual ulceration
15.	Traumatic ulcerative granuloma with stromal eosinophilia (TUGSE) ^{8, 9}

Table 1. Other Terms for RFD.

Aetiology

The exact cause of RFD is unknown, but the main assumption claims to be the repetitive trauma of the tongue over the mandibular teeth¹⁰. RFD is mostly found in new-born children after the eruption of natal or neonatal teeth¹⁰. The traction movement of the tongue over the natal teeth cause chronic trauma to the ventral mucosal tissue. In older children, lesions were found to be associated with the eruption of mandibular incisors and a special group of children, as was concluded in Table 2. These systemic conditions are related to mental or neurological disorders, causing an uncontrollable movement of the tongue and orofacial muscles, which contributes to an ulcerated lesion on the tongue and surrounding tissue^{5, 7, 11}.

1.	Cerebral palsy ¹⁴
2.	Congenital indifference to pain ¹⁵
3.	Familial dysautonomia (Riley-Day syndrome) ^{16, 17}
4.	Lesch-Nyhan syndrome (self-mutilation) ¹¹
5.	Tourette's syndrome
6.	Motor disorders

Table 2. Systemic Diseases or Conditions Associated with RFD.

Tang and et al. studied repetitive trauma in rats. They proposed that trauma is not the main cause of RFD. A chronic trauma allowed the viral and toxic agent to penetrate into the submucosal tissue, leading to an inflammatory reaction, causing a sublingual ulceration¹². Therefore, in cases of immunocompromised children, the potential for infection will be higher and cause more severe and complicated lesions in RFD patients¹³.

Clinical appearance

RFD is usually presented as a circular or ovoid ulcerated area with a raised border on the ventral surface of the tongue. The location of RFD is mostly found on the ventral surface of the tongue, but also on the dorsal surface and tip of the tongue, labial mucosa, buccal mucosa, lingual frenum, and floor of the mouth have been

reported¹⁸⁻²². The size of lesions range from as small as 2 mm. to one which is 30 mm. in diameter^{23, 24}. The repetitive trauma can cause active bleeding from the lesion^{25, 26} and may become larger, or turn into a ulcerative, granulomatous or a fibrous mass lesion^{7, 27}. Lesions with secondary infections may develop into granulation tissue, showing an ulcerated area covered with yellowish white slough in the middle²⁸.

Prevalence

There is no report of RFD prevalence itself. The available data was from natal and neonatal teeth reported in the literature which ranges from 1:1,118 to 1:300,000 depending on the sample groups²⁹. Not all natal-neonatal teeth cause sublingual ulcerations. Therefore, the prevalence of RFD itself should be lower than natal teeth prevalence. On the other hand, RFD has been reported to be associated with some systemic diseases or medical conditions, especially in mental and or neurological disorders^{5, 7}. This may lead to a higher prevalence of RFD in this specific pediatric group.

The distribution of RFD from 46 case reports, show no gender-specific. Most lesions occur in the first year of life due to the premature eruption of the primary lower incisors, natal and neonatal teeth. RFD can occur in children older than 1, especially with systemic conditions such as cerebral palsy, Down syndrome and epilepsy^{23, 30}. Any race including Asian, Latin, European and North America children have been found to have RFD (Table 3).

Age	Nationality	Sex (cases)	
		Male	Female
Newborn-1 month	Indian, Brazilian, Nepalese	6	6
>1-12 months	Japanese, Indian, Syrian, Italian, Korean, Canadian, American, Iranian, British, Dutch	8	10
>12-24 months	Turkey, India, Syria, American	2	2
>24 months	Chinese, Thai	1	1

Table 3. Distribution of Age and Gender in RFD from 46 Case Reports.

Histology

Microscopic findings of RFD primarily show an inflammatory infiltration, consisting of numerous eosinophils, macrophages, lymphocytes and mast cells²⁸. The history and clinical features are most often so typical, that there is seldom a need for additional histopathological examination⁶.

Diagnosis

An early diagnosis of RFD is essential to avoid the extension or progression of a lesion. RFD needs to be differentiated from other oral ulcerations caused from neurological and hereditary disorders. The diagnosis of RFD is usually based on signs, symptoms, clinical appearances and history of premature eruption of the anterior mandibular teeth. Haematological and biopsy examinations are seldom performed due to the very young age and size of the patients^{5, 19, 20, 31}. Evidence of RFD from numerous case reports showed that haematological and biopsy examinations are not routinely performed for diagnosis, unless the lesions are large, not healed after treatment, or signs and symptoms of a systemic infection are present^{19, 31}.

Differential diagnosis

An early accurate diagnosis is essential to differentiate RFD from neurological and hereditary disorders, which presents similar oral ulcerations. These can be excluded by disease history, clinical appearances, laboratory data and biopsy findings. The list of diseases which should be considered, are in Table 4.

1. Agranulocytosis
2. Bacterial infection
3. Electrical or Chemical trauma ³
4. Fungal infection
5. Granular cell tumour
6. Lymphoma
7. Mucosal chancre
8. Myofibroma
9. Primary syphilitic lesion
10. Pyogenic granuloma
11. Sarcoma
12. Tuberculosis
13. Ulcerative candidiasis

Table 4. Differential Diagnosis of RFD.

Signs, Symptoms and Complications

Parents often bring their child with RFD to a physician or a dentist, due to refusal or inability to consume milk, and/or because of natal or neonatal teeth can cause injury to mother's nipples²³. Ulcerations often cause intense pain during feeding, some can be asymptomatic^{4, 31, 32}, and active bleeding can also be found^{13, 25}. Prolonged feeding problems in infants may lead to dehydration, being underweight and growth retardation⁷. Extensive lesions from recurrent trauma and/or untreated ones, can cause permanent deformities to the tongue²¹.

Treatment

There are two main methods for treating RFD, a conservative method and a radical method (Table 5). The treatment decision criteria of RFD are summarized in figure 1. The conservative method consists of tooth grinding, composite placement, formocresol pulpotomy, topical agent applications (protective agent, oral disinfectant or a topical corticosteroid), feeding behaviour modifications, teething rings, or soft splints.

Conservative methods
1. Tooth grinding
2. Composite placement
3. Topical protective agent
4. Behaviour modification (Feeding)
6. Oral disinfectant
7. Teething ring
8. Topical steroid
9. Splint
Radical methods
1. Extraction

Table 5. Summarize of Treatment Options for RFD.

Tooth grinding is performed by using a finishing stone bur or other composite polishing burs to smooth the sharp incisal edges of the

mandibular primary incisors²⁸. The babies are laid on a dental chair, and their movements are controlled by the parents and dentist while the short procedure is carried out, causing no pain to the infant. Composite placements are also used to get rid of sharp incisal edges on the lower incisor teeth, and without destroying the tooth structure. The premature eruption of the primary incisors frequently showed that hypomineralization or hypoplasia enamel are limited areas of bonding and resulted in the poor bond strength of restoration. Moisture control in a new born child is very difficult to achieve, due to a limited access of the oral cavity and uncooperative behaviour of the child³³. The method of composite placement is quite challenging because of being able to achieve an accurate restoration.

Various topical agents have been used to treat RFD. A topical protective agent is odontogenic cream (methylvinylether/maleic acid) which is used as a protective shield on the tongue, to prevent repetitive trauma from opposing teeth. Oral disinfectant (0.2% chlorhexidine) is usually used in conjunction with other methods aimed to reduce bacterial infection of the ulcerated lesion. Corticosteroid in oral base such as triamcinolone acetonide is the topical agent with an anti-inflammatory property, which has been used to reduce inflammation of the lesion. Topical recombinant bovine basic fibroblast growth factor (rb-bFGF) gel³⁴ is another topical agent which has been reported in RFD treatment, that improves the quality and shortens the time to heal the wound.

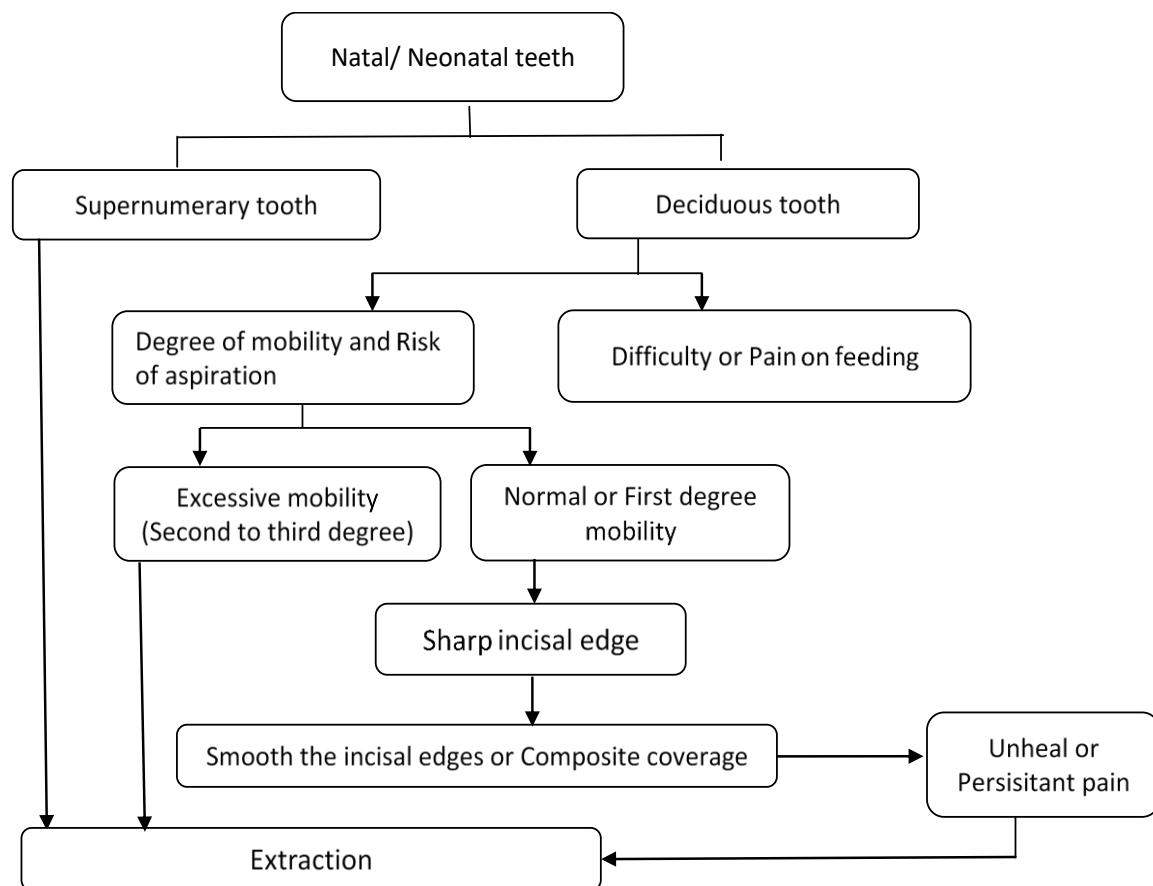


Figure 1. Decision Tree for Treatment of Riga Fede's Disease due to Natal or Neonatal Teeth.

Other methods include using teething rings, splints and feeding behaviour modification have been reported as combination treatments with other methods. Teething rings may be used as a barrier to prevent lesion trauma from the offending teeth³⁵. Teething rings have been used

in very few cases, because only babies older than four to eight months, have developed enough hand-eye coordination to hold one in their hands, and guide it to their mouth. Occlusal splints or mouthguards made from flexible denture base material, soft elastic foil or hard

acrylic resin, were used as a protective shield to prevent injury from the premature erupted lower anterior teeth to the nearby soft tissue. Mouthguards were recommended to be used when babies were fed, and the wearing time gradually extended to reduce traumatic injury and promote wound healing. Unlike other treatment methods, mouthguards are difficult for babies to accept since they are usually uncooperative. Gagging, aspiration and airway obstruction are a cause for concern, due to babies' limited ability to spit out foreign objects. These may happen during the taking of an arch impression or routine wearing at home. Some practitioners modify mouthguards with extended parts to prevent dislodgement or aspiration, using dental floss or medical tape attached to the extraoral tissue, to hold them in place²⁷. Feeding behaviour modifications aim to reduce trauma to the tongue. A bottle with a large nipple hole or spoon feeding may help to reduce the sucking force when babies are fed³⁶.

Extraction of natal or neonatal teeth to eliminate the source of trauma is an aggressive method for treating RFD. This method is indicated when first, natal or neonatal teeth have an excessive mobility (2nd to 3rd degree mobility) which causes a high risk of aspiration⁶, second, large ulcerative lesions and/or patients who have difficulty with drinking or eating^{19, 37}. When conservative treatments do not provide a proper healing of the injury, the extraction of natal teeth may be required. Most RFDs show a rapid resolution within a few weeks^{13, 30, 31}. In new-borns, the excessive mobile natal teeth can be easily removed with extracted forceps or hamostat, since most of them are poorly developed³⁸. Topical anaesthetic gel and/or local anaesthesia can be applied to reduce discomfort during the procedure^{31, 39}. The amount of anaesthetic has to be calculated to avoid an overdose toxicity, due to new-borns being very small. Before performing the extraction on new-born babies, the dentist should consult a paediatrician to avoid any complications of bleeding, infection and delayed wound healing. The blood test screening for haemoglobin, haematocrit, platelet, white blood cell, prothrombin time need to be considered. In the past, some clinicians recommended the extraction of neonatal/natal teeth be deferred until the baby is 10 days old, to allow the vitamin K production by the commensal flora from the

intestine⁴⁰. Nowadays, new-born babies in many countries receive a single injected dose of vitamin K intramuscularly within six hours of birth, to prevent vitamin K deficiency bleeding (VKDV). Therefore, the risk of bleeding complications from hypoprothrombinaemia due to Vitamin K deficiency, should be low. After the extraction, curettage of the socket is recommended to prevent the development of residual dental papilla cells⁴¹.

After the sublingual ulcerative lesions were treated, the babies should be followed up continuously for the next few months. They should have no pain or inflammation and could eat regularly. Routine appointment for dental visits should be scheduled in 3-6-month intervals. An assessment for the abnormal gingival growth and radiographic examination of the lower mandibular area will be performed. Occlusal topographic view of the lower mandibular area will provide the number of remaining primary anterior teeth which may affect tooth alignment and occlusion.

Conclusions

Riga Fede's disease or sublingual ulceration is a traumatized oral soft tissue lesion which is usually found in new-born babies presented with natal/neonatal teeth. The lesions can cause bleeding, pain, fever and difficult feeding, leading to inadequate nutrient intake. Early detection and intervention are essential to prevent serious complications including dehydration, weight loss, infection and tongue deformity. Delayed sublingual ulceration can be found in older children with conditions that are manifested with uncontrollable movement such as cerebral palsy and seizures.

Declaration of Interest

The authors report no conflict of interest.

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