

A Trio Discovery—Cardarelli-Riga-Fede Disease: A Case Report and Review of Literature

Sonal Dinesh Patil, Uma B Dixit

ABSTRACT

The natal and neonatal teeth have been reported to cause ulceration on the ventral surface of the tongue in neonates and infants. These can be presented with mild to severe ulceration which may affect the child's feeding habits. This appearance was described by Cardarelli, Riga and Fede and hence been termed as Cardarelli-Riga-Fede disease. It is important to diagnose the lesion early and treat it by eliminating the cause of trauma to avoid untoward complications. We present a large Riga-Fede lesion in an infant along with the clinical management. The purpose of this report is to review the literature related to neonatal teeth epidemiology and discuss their possible etiology and treatment.

Keywords: Natal teeth, Neonatal teeth, Cardarelli-Riga-Fede's disease, Traumatic sublingual ulceration.

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INTRODUCTION

Child development from conception through the first years of life is marked by many changes. The eruption of the baby's first tooth will bring milestone both in terms of functional and psychological changes in the child's life and in emotional terms for the parents. It is associated with lots of emotions. When teeth are observed at birth or during the first 30 days of life, being denoted as natal and neonatal teeth¹ respectively, it leads to a plethora of reactions which are combined with a lot of interest, curiosity and concern of clinicians are similar to that of parents. Because of its rare occurrence, in the past, this anomaly of eruption was associated with superstition and folklore, being related to good or bad omens. There are various difficulties, like pain on suckling, refusal to feed, faced by the mother and the child due to the natal and neonatal teeth.² One of the modern guiding principles of dentistry is to provide early full infant care during the first year of life.

Traumatic ulceration on the ventral surface of the tongue is most commonly associated with natal or neonatal teeth in newborns.^{2,3} It may also occur in older infants after the eruption of primary lower incisors with repetitive tongue thrusting habits⁴ and in children with familial dysautonomia (in sensitivity to pain).⁵ Typically, the lesion begins as an

ulcerated area on the ventral surface of the tongue with repeated trauma, it may progress to an enlarged, fibrous mass with appearance of an ulcerative granuloma. It may interfere with proper suckling and feeding and put the neonate at risk for nutritional deficiencies. Also due to its mobility, these teeth may swallowed or aspirated by the infant during nursing. In such instances, dental intervention may be required. In view of the above considerations, the objective of the present article was to present a review of literature for natal and neonatal teeth and clinical management techniques for Riga-Fede disease.

CASE REPORT

A 27-day-old male was referred for evaluation of an ulcerated area on the ventral surface of the tongue (Fig. 1). The mother complained of child exhibiting pain during suckling and would not nurse. Oral examination revealed two crowns in the mandibular anterior region, whitish in color and exhibiting grade II mobility in central incisor position. The ventral surface of tongue showed 5 × 10 mm ulceration that extended from anterior border of the tongue to lingual frenum. On palpation, area elicited a pain response from the patient. Examination of the rest of intraoral mucosa revealed no other lesions. Radiographic examination revealed a neonatal tooth, probably, a primary incisor, with well-formed crown but minimal root development (Fig. 2). Based on clinical findings, diagnosis of 'Riga-Fede' disease was made.



Fig. 1: Ulcerated area on the ventral surface of the tongue with two neonatal teeth

Extraction of the teeth was chosen as treatment of choice over more conservative treatment, a pediatrician was consulted and vitamin K (0.5 = 1.0 mg) was administered intramuscularly as a part of immediate medical care to prevent hemorrhage. Extraction was carried out under local anesthesia which patient tolerated well. The extracted teeth had a crown but were devoid of roots (Fig. 3). Patient was reviewed after 10 days and the lesion was fully resolved. Mother informed that infant was feeding normally. At a follow-up appointment, the lesion was fully resolved and infant was feeding normally (Figs 4 and 5).

REVIEW OF LITERATURE

History

Natal and neonatal teeth were reported during Roman times by Titus Livius (59BC) and Gaius Plinius Secundus (23BC) and were described in the cuneiform inscriptions found at Nineveh. The folklore and misconceptions surrounding natal and neonatal teeth vary; in some cultures like Malaysian communities, a natal tooth is believed to herald good fortune.

In England, infants born with natal teeth were considered as famous soldiers⁶ while in China, Poland, India and Africa, the affected children are considered to be monsters and bearers of misfortune. Shakespeare also contributed his thoughts on natal teeth.⁷ Several terms have been used for natal teeth, such as congenital teeth, fetal teeth, predeciduous teeth and precocious dentition as well as dentitia praecox and dens connatalis, have been used to describe these teeth.^{8,9} Currently, the terminologies, 'natal' and 'neonatal,' used by Massler and Savara¹, have been adopted. Even these terms only define the time of eruption and give no consideration to anatomy and histology or whether the tooth is a component of primary dentition or whether it is supernumerary.^{1,7}

The prevalence is 1:700 to 1:30,000 depending on the type of the study. The highest prevalence is found in the only study that relies on personal examination of patients.¹⁰ Other reports reveal it to be around 1 in 2000 to 3500 live births.^{1,9} The reported incidences vary greatly, from those of Mayhall at 1:11 and Gordon at 1:100 through those of



Fig. 2: Radiographic examination showing well-formed crown but minimal root development



Fig. 4: Fully resolved lesion at a follow-up appointment



Fig. 3: Extracted neonatal teeth



Fig. 5: Happy child and mother at follow-up

Massler and Savara, Gardiner, Spouge, all at 1:2000; down to those of Ballantyne at 1 in 6,000 births, and Howkins at 1:10,000.^{1,11,12}

CLASSIFICATION

In 1966, Spoug and Feasby et al classified natal and neonatal teeth according to their degree of maturity.⁶

1. A mature tooth is nearly or fully developed and it has relatively good prognosis.⁷
2. Immature tooth is incomplete or substandard structure with poor prognosis.

CLINICAL APPEARANCE

Reports about significant differences between males and females are conflicting, with females,¹⁵ in general, being more affected; and also Muslim children exhibited more natal/neonatal teeth as compared to Hindu children. Natal teeth are more frequent, approximately three times more common than neonatal teeth,⁷ with the most commonly affected teeth are the mandibular central incisors (85%), followed by maxillary incisors (11%), mandibular cuspids or molars (3%), and then maxillary cuspids or molars (1%).⁷ Natal or neonatal cuspids are extremely rare.¹³ Only 1 to 10% of natal and neonatal teeth are supernumerary.¹⁴⁻¹⁶

Natal teeth resemble to primary teeth in size and shape; however, the teeth are often smaller, conical and yellowish, and have hypoplastic enamel and dentin with poor or absent root development. Most natal and neonatal teeth are mobile.¹⁶ The appearance of each natal tooth into the oral cavity can be classified into four categories.^{6,10}

1. Shell-shaped crown poorly fixed to the alveolus by gingival tissue and absence of a root.
2. Solid crown poorly fixed to the alveolus by gingival tissue and little or no root.
3. Eruption of the incisal margin of the crown through the gingival tissues.
4. Edema of gingival tissue with an unerupted but palpable tooth.

ETIOLOGY

Etiology of natal and neonatal teeth is debatable. The various hypothetical factors reported by investigators include the following:

1. The most acceptable theory is based upon the result of a superficial localization of the dental follicles, probably related to a hereditary factor.^{13,17}
2. *Genetic blueprint*: The rate at which baby's teeth come through will depend on his 'genetic blueprint'. If one or both of his parents were very late or very early teether, there is a strong chance that he too will be a very late or very early teether respectively.^{6,10,16} Bodenhoff and

Gorlin have reported that 15% of children with natal and neonatal teeth had parents, siblings or close relatives with a history of same condition.¹⁴ In 1950, similar condition with hereditary factor has been studied by Massler and Savara.¹

3. *Endocrine disturbances*: It is thought to be because of excessive secretion of the pituitary, thyroid or gonads.
4. Jasmin and Clergeau-Guerithault reported that the eruption of natal and neonatal teeth could be dependent on osteoblastic activity within the area of the tooth germ.^{6,16}
5. *Infection*: For example, congenital syphilis appears to have varying effect; in some cases, it has erupted early, while in others it has been retarded.¹⁶
6. Nutritional deficiency, e.g. hypovitaminosis (which in turn is caused by poor maternal health, endocrine disturbances, febrile episodes, pyelitis during pregnancy and congenital syphilis).^{7,16}
7. *Febrile status*: Fever, exanthemata during pregnancy tend to accelerate eruption as they do in various other processes.
8. Superficial position of the tooth germ.
9. *Environmental factors*: Polychlorinated biphenyls (PCB) and dibenzofurans¹⁸ seem to increase the incidence of natal teeth. These children usually show other associated symptoms, such as dystrophic finger nails, hyperpigmentation, etc.¹⁶
10. It may be associated with and recognized syndromes. These syndromes include Ellis-van Creveld (chondroectodermal dysplasia), pachyonychia congenita (Jadassohn-Lewandowsky), Hallermann-Streiff (oculo-mandibulo dyscephaly with hypotrichosis), Rubinstein-Taybi, steatocystoma multiplex, Pierre-Robin, cyclopia, Pallister-Hall, short rib-polydactyly type II, Wiedemann-Rautenstrauch (neonatal progeria), cleft lip and palate, Pfeiffer, ectodermal dysplasia, craniofacial dysostosis, multiple steacystoma, Sotos, adrenogenital, epidermolysis bullosa simplex including van der Woude and Walker-Warburg syndromes.^{7,9,10,18,19}
11. The literature reflects the association of natal and neonatal teeth with developmental abnormalities, such as reactive fibrous hyperplasia,¹⁰ congenital hydrocephalus with bilateral mandibular hamartomas,²⁰ pyogenic granuloma,²¹ peripheral ossifying fibroma, eruption cyst,¹⁰ gingival fibrous hamartoma²² bifid tongue and deaf mutism.¹¹

HISTOLOGIC AND RADIOGRAPHIC FEATURES

First report on microscopic observation of natal and neonatal teeth was done by Howkins (1932). Histologic investigation has revealed a failure of root formation despite eruption,¹⁴

a large vascular pulp, the alteration in amelogenesis, irregular genesis of dentin and a failure of cementum formation.⁶ The usual radiographic feature of the tooth consists of a hollow calcified cap of enamel and dentin without pulp tissue, rather like a celluloid crown in shape.²

CLINICAL MANAGEMENT TECHNIQUES

The diagnosis is based on a complete history, physical examination, clinical and radiographic findings to rule out them being part of normal dentition or supernumerary so that no indiscriminate extractions would be performed. Dental treatment is depending on size of ulceration, tooth mobility and condition of child (Table 1).

DISCUSSION

For the past 200 years, many reports of infants born with teeth or teeth erupting immediately after birth have appeared in the medical and dental literature.²³ In modern dental literature, most satisfactory terminology natal and neonatal teeth has been defined by Massler and Savara.¹ Neonatal teeth often present with hypoplastic enamel and underdeveloped roots, with resultant mobility. It is necessary to investigate the possible local or systemic factors related to the eruption of teeth, in order to promote a better oral condition. A complete oral examination is recommended in newborns, in order to establish the differential diagnosis.^{13,24}

Major complication from neonatal teeth is an ulceration on the ventral surface of the tongue caused by tooth's sharp incisal edge. It is interfere with proper suckling and feeding and put the neonate at risk for nutritional deficiencies. Many investigators have been reported that neonatal teeth with sharp incisal edge causes ulceration on the nipple of the mother and interference with breast feeding.¹⁷ Many investigators reported that there is no relationship between injury to mother's nipple and the presence of the natal teeth since the tongue is interposed between these teeth and the nipple during breastfeeding.⁶

The lesion was first described by Antonio Riga, an Italian physician in 1881. Histological studies and additional cases were subsequently published by F Fede in 1890.⁷ It has been subsequently been known as 'Riga-Fede disease'.^{6,20}

Recently, small ulceration with membrane formation on the under-surface of the tongue in infants with these teeth is further studied by Antonio Cardarelli. So it has been

known as Cardarelli's aphthae, Cardarelli's disease, Cardarelli's syndrome. Treatment of Riga-Fede disease has varied over the years. Early treatment consist of excision of the lesion. Allwright advocated maintaining the neonatal tooth by smoothening of incisal edge with an abrasive instrument. In cases of mild-to-moderate irritation to the tongue, such treatment may suffice.^{3,6,14} Also removal of natal and neonatal teeth is indicated when they are poorly developed, interfere with feeding, highly mobile, and associated with soft tissue growth.^{2,10} If extraction is carried out, it is necessary to ensure that the underlying dental papilla and Hertwig's epithelial root sheath are removed by gentle curettage as root development can continue if these structures are left *in situ*.^{3,16} Such tooth-like structures have been termed as 'residual natal tooth'.^{25,26} We did not observe any such alterations after a follow-up for 1 year in our case report. Prophylactic administration of vitamin K (0.5-1.0 mg, im) is advocated because of the risk of hemorrhage as the commensal flora of the intestine might not have been established until the child is 10 days old, and since vitamin K is essential for the production of prothrombin in the liver. Other conservative treatment modalities includes a small increment of composite may be bonded to the incisal edges of the teeth.²⁷ As many of these teeth show evidence of hypomineralization, limited surface of enamel for bonding of resin, difficulty of access, inadequate moisture control and etching of enamel surface which leads to poor retention of the resin to teeth.²

Teething symptoms, such as those seen with the eruption of other primary teeth (infantile diarrhea, drooling of saliva, malaise, etc.), though reported by Massler, Savara, and Spouge, were not seen in the present case report.²⁸ One of the reasons for eruption of neonatal teeth is superior placement of the tooth germ.¹⁹ In the present case, it could the reason for teeth eruption with no hereditary influence. Teeth that are stable beyond 4 months have a good prognosis. Esthetically, they are not pleasing due to the discoloration. Pediatric dentists should make every effort to educate parents and the medical community on the preferred treatment for these teeth. If extraction of a natal and neonatal tooth is indicated, then it should be performed by a dentist to avoid unnecessary trauma to the area.²¹ Periodic follow-up by a pediatric dentist to ensure preventive oral health care is very essential.¹⁰

Table 1: Treatment modalities for Riga-Fede disease

Preventive treatment	Surgical treatment
<ol style="list-style-type: none"> 1. Roundening of the incisal edges with an abrasive instrument^{3,6} or 2. A small increment of composite may be bonded to the incisal edges of natal and neonatal teeth 	Extraction of natal and neonatal is indicated in case of supernumerary teeth, poorly developed, interferes with feeding, highly mobile and associated with soft tissue growth

We conclude that infants with prematurely erupted teeth must be carefully examined for further treatment planning, and parent counseling to bring about awareness is also equally important. Longitudinal and more divergent studies are also necessary to confirm the etiology and nature of natal teeth and to determine whether they are deciduous or supernumerary teeth.

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ABOUT THE AUTHORS

Sonal Dinesh Patil (Corresponding Author)

Senior Lecturer, Department of Pedodontics, MGM Dental College, Kamothe, Navi Mumbai, Maharashtra, India, e-mail: drsonupatil@yahoo.com

Uma B Dixit

Professor, Department of Pedodontics, Dr DY Patil Dental College Navi Mumbai, Maharashtra, India