

# Management of patient with NOMID syndrome in the Pediatric Dental Clinic

Praneeti Sodhi, DDS

University of Nevada, Las Vegas - School of Dental Medicine

## INTRODUCTION:

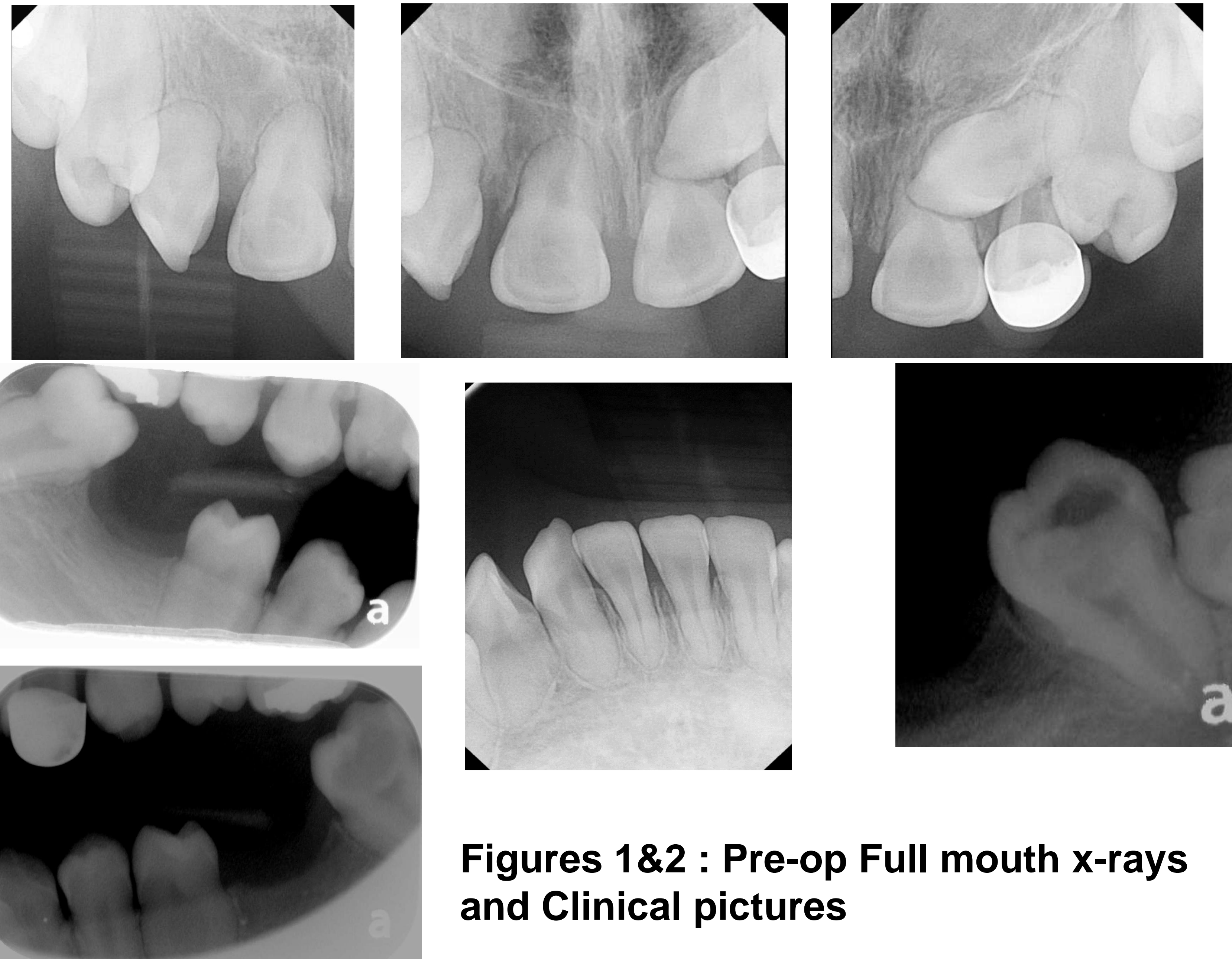
**Neonatal Onset Multisystem Inflammatory disease (NOMID)** is a congenital inflammatory disorder characterized by tissue damage of the nervous system, skin, and joints. It is a subset of hereditary cryopyrinopathies which result due to a mutation in NLRP3 gene. Individuals with NOMID have a skin rash that is present from birth and persists throughout life and can also have chronic aseptic meningitis and a characteristic arthropathy. History of seizures and vomiting resulting due to chronic meningitis present a specific challenge in the dental management of the pediatric patient with NOMID. This is a case report of a 14-year-old female with NOMID who was treated for severe dental caries at UNLV pediatric dentistry clinic. The patient had a positive history for seizures and hence was treated in a comprehensive manner under general anesthesia. This report discusses our management protocol for this case and possibly for a standardized protocol for patients with NOMID. To our knowledge this is the first report of its kind

## CASE REPORT:

A 14-year-old female patient presented to the pediatric dental clinic at University of Nevada, Las Vegas for a Periodic oral exam. Patient had no chief complain at this visit. Patient presented with past medical history of Neonatal-onset Multisystem inflammatory disease, Erythema multiforme, Vasculitis, Hypothyroidism, Dysmorphism, Short stature, Ventriculomegaly, History of hepatitis at birth but no viral infection as present at birth . Upon Clinical and radiographic examination, it was noted that the patient has short and blunt roots of the maxillary anterior teeth. Missing Maxillary lateral incisors and impacted maxillary right canine. Mesocephalic, skeletal class 3 malocclusion, Anterior open bite. Caries present on Maxillary molars and premolars and Mandibular molars both right and left side. Considering patient's age and compliance with dental treatment, Dental treatment under General anesthesia was planned. Before treatment her physician was consulted to ensure that she did not require any pre-medication. Maxillary right and left molar and pre- molars were restored with composite resin restorations. Pulpotomy and stainless-steel crown restorations were performed on the mandibular left molar. Stainless steel restoration was performed for the mandibular right molar. At this moment we decided to not to do any treatment of the impacted canine. Finally, fluoride varnish was applied to prevent progression and occurrence of caries. Successful dental treatment was performed under general anesthesia.

## DISCUSSION:

Neonatal onset multisystem inflammatory disease (NOMID) is a genetic disease, often caused by mutations in the *NLRP3* (also known as *CIAS1*) gene. People with NOMID often experience joint inflammation, swelling, and cartilage overgrowth, causing characteristic prominent knees and other skeletal abnormalities that worsen over time. Joint deformities called contractures may restrict the movement of certain joints. Other features of this disorder include short stature with shortening of the lower legs and forearms, and characteristic facial features, including a prominent forehead and protruding eyes. There is an increased risk for individuals with NOMID to develop abnormal deposits of a protein called amyloid, which may cause kidney damage. Therefore, when dental treatments are performed under general anesthesia, dentists should collaborate with anesthesiologists during the perioperative period.



Figures 1&2 : Pre-op Full mouth x-rays and Clinical pictures



Considering joint anomalies that can lead to impairment of function and deformities, careful positioning of these patients is necessary during dental treatment under general anesthesia. As postoperative pain is less likely to induce alterations in the immune system, postoperative analgesic should be considered. However, there are no reports about the stress adversely effects of general anesthesia on the patient's condition.

## SUMMARY:

In this case, dental treatment under general anesthesia was successfully performed in a patient with NOMID syndrome. Before dental treatment, the medical conditions should be thoroughly examined in a patient with NOMID syndrome. Since elevated IL-1 $\beta$  level is associated with dental caries and periodontal conditions, professional oral prophylaxis and topical fluoride application should be considered through periodic visits.

## References

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