Follow-up of the oral findings and treatment planning in a mother, son and daughter diagnosed with Trichorhinophalangeal Syndrome Type I

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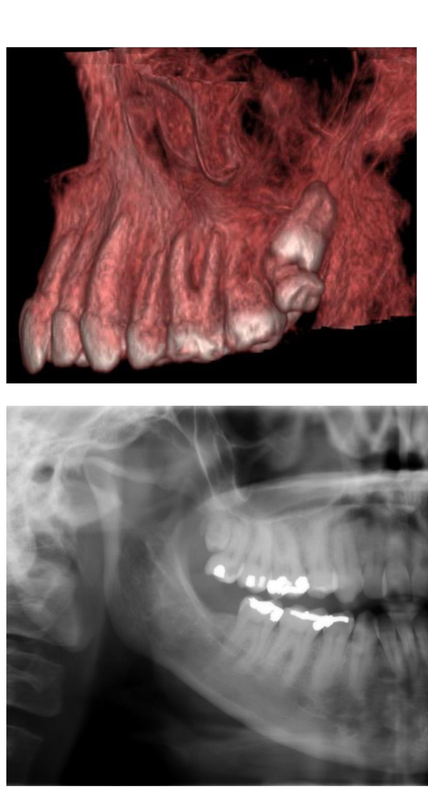
Trichorhinophalangeal syndrome (TRPS) is an autosomal dominant multisystem disorder caused by disruption or changes of TRPS1 gene. Three types of TRPS (I, II and III) are described, all of which exhibit the common features of hair, craniofacial and skeletal abnormalities. Dental anomalies frequently associated with TRPS type I include presence of supernumerary teeth, congenital absence of teeth, malocclusion, delay in tooth development and abnormal tooth shape.

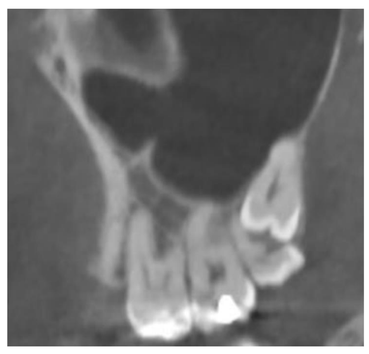
Objective

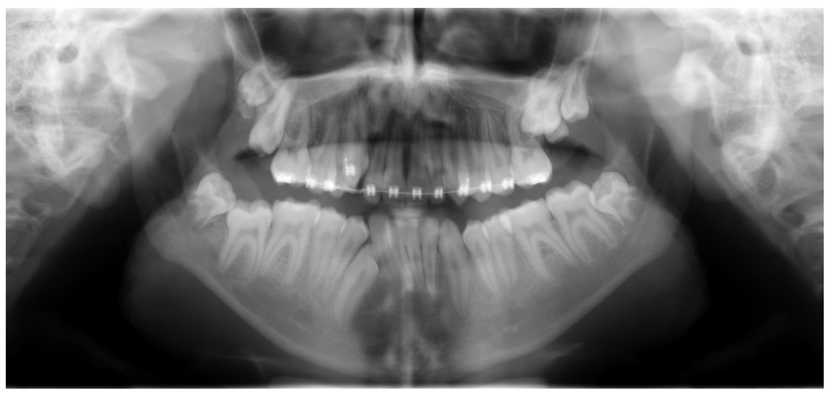
The aim of the study was to present dental anomalies and malocclusion observed in three individuals of the same family with TRPS type I and at the follow-up in order to optimize the treatment plan.

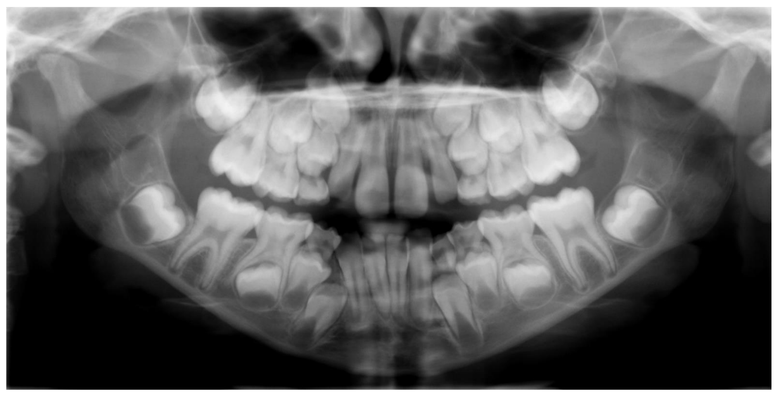
Results

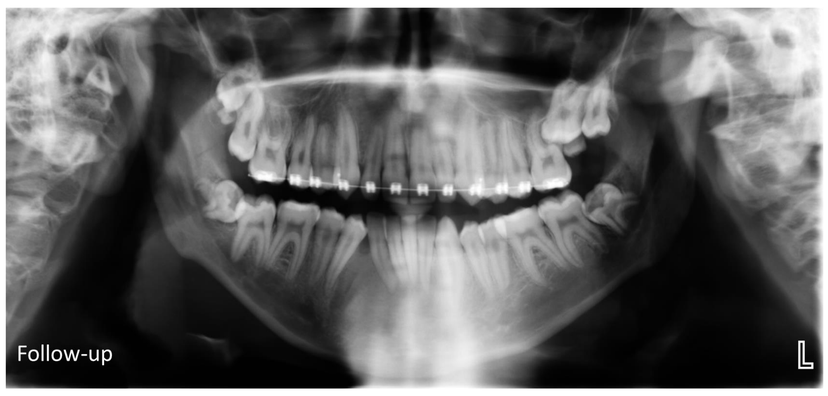
The patients in this study had a similar dentition and the same type of malocclusion, lack of space in both jaws and supernumerary teeth. They were seen again later in a follow-up. All of them are in different stages of occlusal development and the orthodontic treatment planning had been different for the mother and the son. When planning the orthodontic treatment for the daughter, an interceptive approach was considered to prevent an extended orthodontic treatment and worsening of oral health.

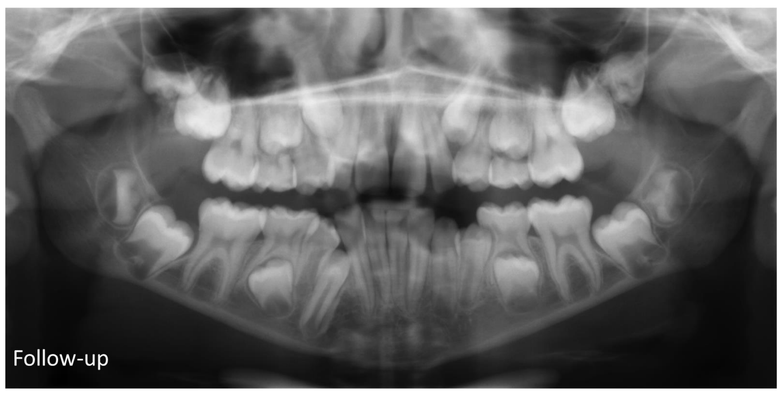












Case 1: Mother, 41-year-old with two maxillary supernumerary molars on each side - one partially erupted and the other one impacted. CBCT shows resorption of the root on tooth 28.
Due to malocclusion, crossbite on the right side with midline shift and lack of space, she received orthodontic treatment with fixed appliances during which all four first premolars were removed. Maxillary second molars have advanced carious lesions.

Case 2: Son, 17-year-old with two impacted maxillary supernumerary molars on each side, one of which is not yet fully developed. He has severe malocclusion with impacted molars and canines, lack of space, crossbite on the left side with midline shift and abnormal shape in certain teeth. He is currently receiving orthodontic treatment with fixed appliances in the maxilla without extractions. Tooth 43 was removed and tooth 13 was placed on the dental arch.

Case 3: Daughter,10-year-old with impacted both maxillary canines and right mandibular canine. Early loss of deciduous canines and severe lack of space in both jaws. Late mixed dentition. The orthodontic treatment plan proposed was: serial extraction of first premolars and observation of the eruption of the second molars. Later, in the permanent dentition, orthodontic treatment and extraction of maxillary supernumerary molars may be necessary.

Conclusion

Early interceptive planning can prevent a long and complicated orthodontic treatment. This study highlights the complex nature of dental anomalies and of malocclusion in individuals with TRPS type I. A careful clinical and radiological examinations of the oral cavity in these individuals is important. Our findings also indicate that a multidisciplinary team may be required to create a proper treatment plan.

References

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