Descriptive Case:-

Syndrome.

Cleft lip and palate patients, cleidocranial dysplasia or Gardner's syndrome is found to have an increased incidence in 5% of the permanent dentition by different authors. A supernumerary tooth is found in 0.1 to 0.8% of primary and 2.1% (1-5%) of the permanent dentition by different authors. A supernumerary tooth is referred as hypodontia excluding third molars and absence of more than six teeth is referred as oligodontia excluding third molars. The supplemental supernumerary may be referred to duplication of the teeth, which assist in function, while a complementary supernumerary is located out of the arch and do not assist in the function.

Supernumerary teeth may resemble the teeth of the group it may belong, ie, molars, premolars, or anterior teeth, or it may bear little resemblance in size and shape to the teeth with which it is associated. Supernumerary teeth may be classified on the basis of its resemblance in size and shape to the teeth with which it belongs, ie, molars, premolars, or anterior teeth, or it may bear little resemblance in size and shape to the teeth with which it is associated. Supernumerary teeth may be classified on the basis of its resemblance in size and shape to the teeth with which it belongs, ie, molars, premolars, or anterior teeth, or it may bear little resemblance in size and shape to the teeth with which it is associated.

The exact etiology of hyperodontia is not well understood. Some hypothesis has been proposed to explain the formation of supernumerary teeth like atavism, dichotomy, hyperactivity of the dental lamina and the concept of multi-factorial inheritance. Similarly, some of the theories proposed on the tooth agenesis like disturbances in the normal course of development of neural crest cells and association with interactions between the epithelial and the mesenchymal cells during the initial stage of tooth development may be responsible for hypo-hyperdontia.

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Supernumerary are found in 0.1 to 0.8% of primary and 2.1% (1-5%) of the permanent dentition by different authors. This particular condition is found to have an increased incidence in cleft lip and palate patients, cleidocranial dysplasia or Gardner's syndrome.

Description of Case:-

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found than central incisors when literature is reviewed.

Many cases are reported on concomitant hypo-hyperdontia (CHH). A literature review on the hypohyperdontia shows prevalence ranges from 0.002% to 3.1%. One epidemiological study on orthodontic population finds incidence of 0.3% in Nonsyndromic CHH. Supernumerary is more common in males while hypodontia is more common in females. CHH has been reported to affect both genders equally. The case reported here is unique case of CCH, where a supernumerary right central incisor is present and the counterpart teeth i.e. lower central incisors are absent.

Hypodontia is inherited as autosomal dominant mode, but occasionally autosomal recessive and X-linked and polygenic/multifactorial models of inheritance have also been reported. There are a large number of genes involved in the odontogenic process, so there are higher chances for mutations to disrupt this process. Tooth agenesis is the most common craniofacial malformation with missing one or more number of teeth (hypodontia, oligodontia or anodontia). Hypodontia can be associated with syndromes or may present as an isolated condition. Hypo-hyperdontia is rare in isolation and has been associated with over 50 syndromes (orodigitofacial dysplasia syndrome, Ellis van Creveld, Downs syndrome, cleft lip and palate etc).1-8

Normal development of the tooth germ is appropriately regulated by molecular signaling pathways, if not, can give rise to supernumerary/supplemental teeth. These pathways include components of the Hedgehog, FGF, Wnt, TNF and BMP families. Two genes have been identified by mutational analysis as the major causes of non-syndromic hypodontia (PAX9 and MSX1). Hypodontia is resulted from haploinsufficiency of any one of these and point mutations can cause some or multiple missing teeth.9

Expression of transcription factors and signaling molecules operating both intracellularly and extracellularly are guiding tooth development throughout the odontogenic process. BMP4 (transforming growth factor-beta family) and the transcription factors PAX9 (pairedbox domain) and MSX1 (homeobox domain), are examples of controlling factors during the odontogenic process. Expression of PAX9 has been found in the sights of tooth development prior to there being any morphological signs of odontogenesis.10

Mandibular central and lateral incisors show lowest incidence of permanent tooth agenesis, also permanent tooth agenesis of maxillary central incisors, maxillary cuspids and maxillary first molars are also rare.11

In a study, frameshift mutation of Pax9 gene (chromosome 14) was identified as responsible for autosomal dominant oligodontia in a large family for four generations. In some of the affected members, maxillary and mandibular second premolars and mandibular central incisors were absent in addition to the lack of permanent molars; although, a normal primary dentition was present.12 Different mutations in the same gene can result in hypodontia or oligodontia, hence these conditions are not fundamentally different. Furthermore, there are evidences to show that more severe the hypodontia, the smaller the mesiodistal width of the teeth formed, conversely, patients with supernumerary teeth tend to have a significantly larger maxillary central and lateral incisors.7 The size of teeth in case of hypo-hyperdontia is not documented in the literature.

Conclusion:-
This case report raises awareness about hypo-hyperdontia of counter teeth. It is a unique case of supplemental maxillary central incisor and missing mandibular central incisors along with missing right lower second molar and all third molars. This type of case is not reported in the literature, affecting counter teeth. In these cases, a wise treatment plan is necessary to alleviate the patient’s problems.

REFERENCES:-