CASE REPORT

# Familial aggregation of maxillary lateral incisor agenesis (MLIA)

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## SUMMARY

Agenesis of maxillary lateral incisors and mandibular second premolars is the most frequent form of hypodontia. Its prevalence varies across population from 0.8% to 4.5%. Genetic aberrations and environmental factors may cause agenesis of one or more teeth. The management of child having such a problem is very important since diastema in teeth especially in upper anteriors not only affects child's physical appearance but also its psychological development as the child wants to look like other children. In this article is presented a case of non-syndromic agenesis of maxillary lateral incisors (MLIA) and mandibular central incisors in a 10-year-old boy (patient) in permanent dentition with its management along with the radiographic investigations and photographic presentations of the other members of his family affected with this condition.

## BACKGROUND

The condition of congenitally missing teeth has been defined as either absence of 1 or more teeth or total absence of teeth. Absence may take the form of total absence (anodontia) or partial absence (hypodontia, ie, less than 6 missing teeth or oligodontia, ie, more than 6 missing teeth). Its frequency ranges between 2% and 10%.[1] Hypodontia in the primary dentition is uncommon, with a prevalence of 0.1–0.9%.[1] Agenesis of maxillary lateral incisors (MLIA) and mandibular second premolars is the most frequent form of hypodontia; its frequency varies across populations ranging between 0.8% and 4.25% for MLIA in permanent teeth.[2–12] The present article reports the diagnosis of non-syndromic familial aggregation of MLIA agenesis in permanent teeth in a 10-year-old boy (patient), his two sisters and his father along with its management in the boy.

## CASE PRESENTATION

A 10-year-old boy visited the department of Paedodontics and Preventive Dentistry, Career Post Graduate Institute of Dental Sciences, Lucknow with the chief complaint of spacing in his upper front teeth. Upon general physical examination the child appeared physically normal, moderately built and nourished with no underlying systemic conditions. Extraoral examination was normal. Intraorally there was midline diastema in maxillary arch and both maxillary permanent lateral incisors, that is, 12 and 22 were missing (figure 1). In the mandibular arch, the right second deciduous molar 85 was extracted due to caries 1 year previously and left first deciduous molar 74 was grossly carious, which needed extraction. Medical history was non-contributory but family history revealed that there was history of missing teeth and spacing in other family members too. The patient had two siblings, one elder sister and one younger sister. Both had missing teeth as his father informed.

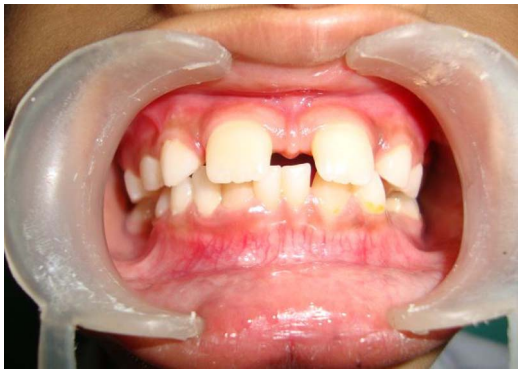

**Figure 1** Intraoral photograph of patient showing midline diastema.

## INVESTIGATIONS

Orthopantomograms (OPGs) were advised for the patient and his father. The patient's father was advised to bring his other children (2 daughters) during his next visit. During the next visit OPGs of the patient's elder and younger sisters were obtained.

(A) Dentition of the patient at the age of 10 years was,
16 55 54 53 11 21 63 64 25 26
46 84 83 42 81 71 32 74 75 36
Congenitally missing teeth were 12, 22, 31 and 41 (figure 2).

(B) Dentition of the patient's father at the age of 42 years was
17 16 15 14 13 11 21 23 24 25 26 27
47 46 45 44 43 42 32 33 34 35 36 37
Congenitally missing teeth were 12, 22, 31 and 41 (figure 3A,B).

(C) Dentition of the patient's elder sister at the age of 12 years was:
16 55 13 53 11 21 23 63 64 65 26
47 46 85 44 83 42 41 31 33 34 75 36
Congenitally missing teeth were 12, 22 and 32 (figure 4A,B).

(D) Dentition of the patient's younger sister at the age of 8 years was:
16 55 53 52 11 61 62 63 64 65 26
46 85 84 83 82 42 41 31 32 73 74 75 36
Congenitally missing teeth were 12 and 22 (figure 5A,B).

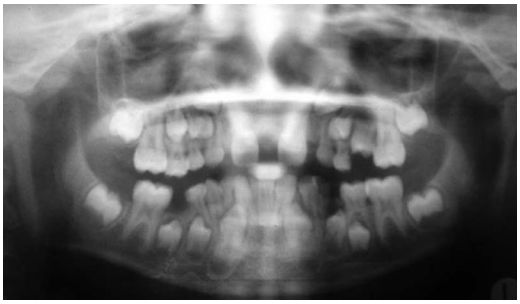

**Figure 2** Orthopantomograph of patient showing congenitally missing 12, 22, 31 and 41.

It was observed that congenital absence of both maxillary lateral incisors (12 and 22) was a common finding in the patient and his family members. Thus, the condition was diagnosed as familial aggregation of MLIA in permanent teeth.

## TREATMENT

Treatment modalities for multiple missing teeth can be removable, fixed or implant prosthesis. Extraction of grossly decayed 74 was done which was followed by placement of lingual arch space maintainer (figure 6). Closure of midline diastema was done by placing orthodontic buttons and elastics (figure 7) followed by replacement of the missing maxillary lateral incisors with prosthesis fabricated chair side using fibre-reinforced composite and acrylic pontic (figures 8 and 9). Definitive treatment was planned after completion of the growth period, and until then a provisional treatment was given to enhance the

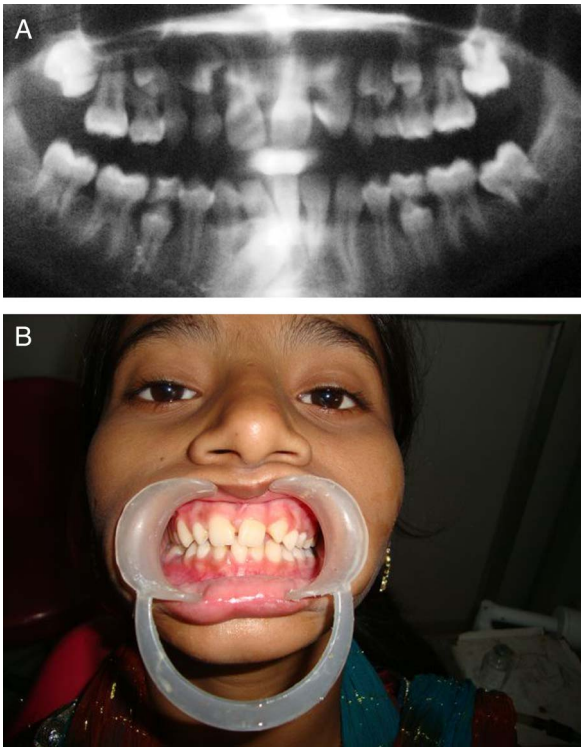


**Figure 4** (A) Orthopantomograph of patient's elder sister showing congenitally missing 12, 22 and 32. (B) Photograph of patient's elder sister.

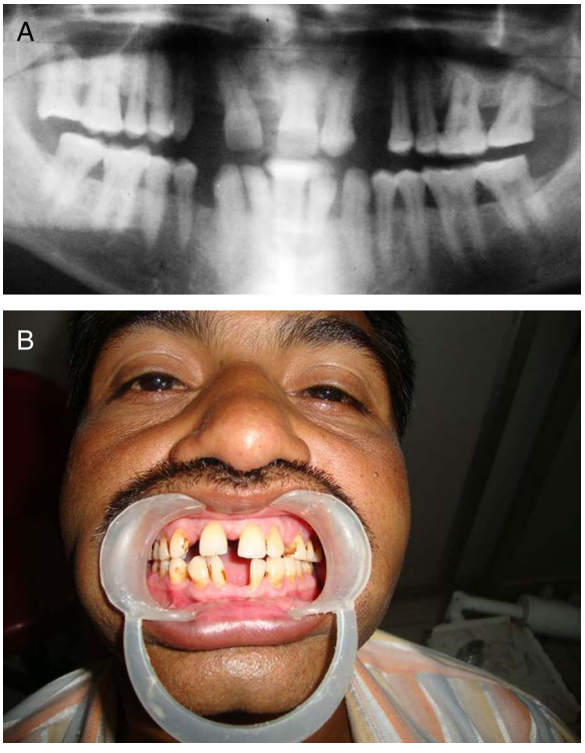


**Figure 3** (A) Orthopantomograph of patient's father showing congenitally missing 12, 22, 31 and 41. (B) Photograph of the patient's father.

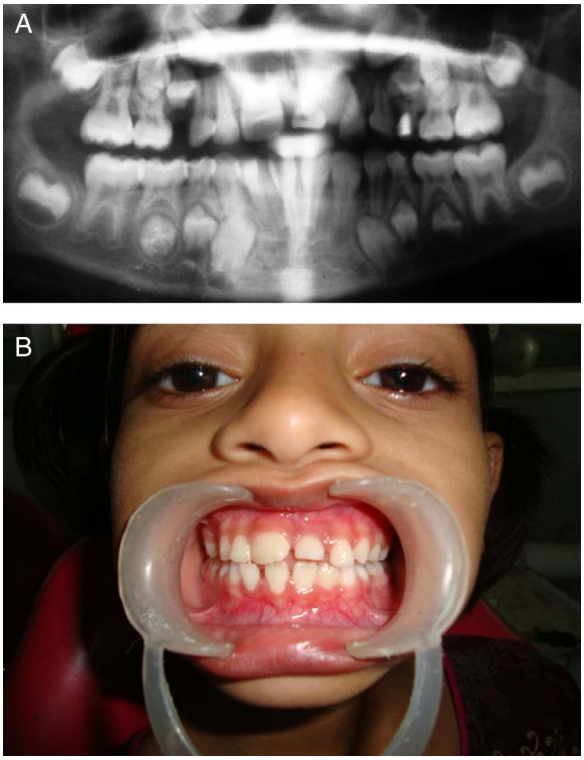


**Figure 5** (A) Orthopantomograph of patient's younger sister showing congenitally missing 12 and 22. (B) Photograph of patient's younger sister.

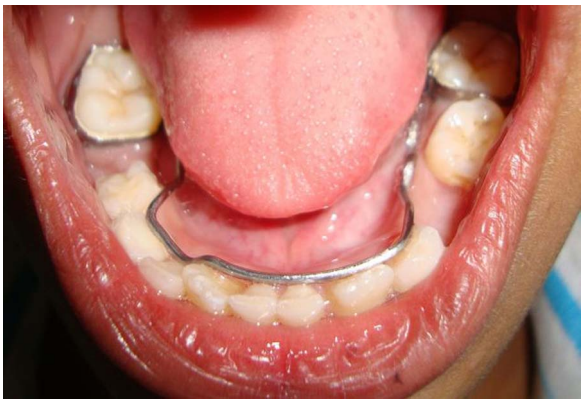

**Figure 6** Intraoral photograph of patient showing lingual arch space maintainer.

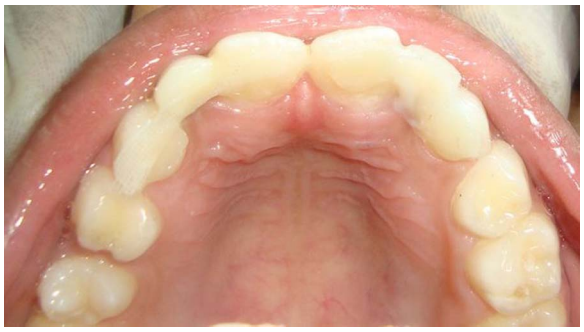

**Figure 8** Intraoral photograph of patient showing palatal view of the prosthesis fabricated chair side using fibre reinforced composite resin and acrylic pontic.

aesthetics and to meet functional and psychological requirements of the patient.

## OUTCOME AND FOLLOW-UP

As the child matures the removable prosthesis needs relining, rebasing or remaking to accommodate growth changes and maintain normal oral functions. However, their prolonged use may increase the risk of caries and periodontal disease, unless there is excellent oral hygiene and usage of topical fluoride. Fixed prosthesis is the preferred definitive treatment for the majority of hypodontia patients. Implant placement usually has to be postponed till the completion of skeletal growth for patients with congenitally missing teeth usually present in childhood. 'Aesthetics for function' should be the mantra when bringing back smiles on children's lovely faces.

## DISCUSSION

Although tooth agenesis is occasionally caused by environmental factors, hypodontia has a genetic basis in the majority of cases. Nevertheless, there is evidence showing that congenital tooth absence can be the result of environmental or hereditary causes, or their interaction.[13] In our case, patient's father and both his siblings manifested hypodontia but its patterns in the siblings were different which might be due to incomplete penetrance and variable expressivity. Neither patient's mother nor his grandparents (maternal side) manifested hypodontia as his history revealed. The patient's grandfather (paternal side) also had less number of teeth as informed by the patient's father. Therefore, it appeared that affecting gene came from the paternal side only. Most of the familial segregation studies suggested that hypodontia showed an autosomal-dominant inheritance in families, with incomplete penetrance and variable expressivity.[2] However, modes of transmission linked to the X-chromosome and polygenic or multifactorial type have also been proposed.[14] In the present case, the hypodontia was supposed to be due to autosomal dominant inheritance as both male and female members of the family manifested it. The patient and his father showed absence of 12, 22, 31 and 41, while 12, 22 and 32 were missing in elder sister and 12 and 22 in younger sister. Thus, it is evident that siblings were not sharing the same patterns of hypodontia with regard to tooth number, region and symmetry, however, absence of 12 and 22, that is, both maxillary lateral incisors was a common finding in all the family members investigated. The similarity of the patterns of hypodontia of our patient with father and dissimilarity with siblings regarding tooth number, region and symmetry might be due to incomplete penetrance and variable expressivity. However, pattern of inheritance cannot be explained in this case, as the population size is very small and sufficient pedigree data of more than three generations is not available.

Female predominance of hypodontia has been reported. But, in most studies, the differences did not reach statistical significance.[8] In the present case, although both of the patient's sisters showed hypodontia, yet no concrete conclusion could be made on this basis only as no vertical and lateral studies were performed and patient was the only son in the family.

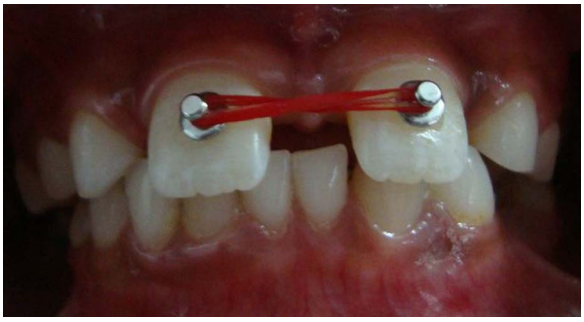

**Figure 7** Intraoral photograph of patient showing brackets and elastics for closure of midline diastema.

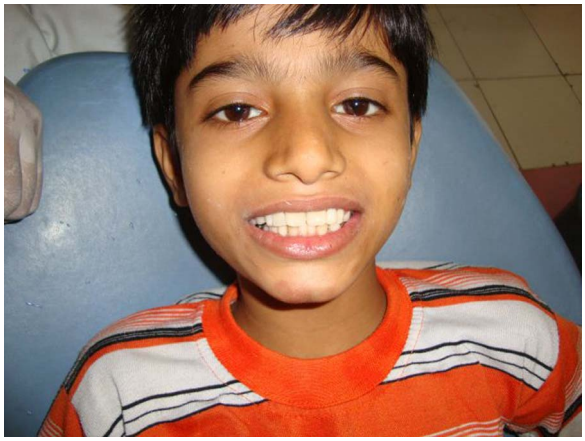

**Figure 9** Photograph of patient showing replacement of the missing maxillary lateral incisors with prosthesis.

## Learning points

- Diastema in teeth especially in upper anteriors not only affects the child's physical appearance but also have an adverse effect on the psychological development leading to emotional disturbance as the patient wants to look like other children.
- Since there is treatment available for diastema closure in children, the treatment should not be postponed to adulthood which may have an adverse effect on personality development of the affected child.
- The treatment of such children gives a healthy smile on the child's face which not only gives satisfaction to the parents and other family members but also promotes them for getting their own treatment in case they are affected with similar conditions.
- In patients with reduced number of teeth, the importance of maintaining the teeth should be emphasised and preventive measures must be taken in this regard.
- The case highlights the importance of proper diagnosis, which can aid in proper treatment planning, which in turn potentially reduces the orthodontic complications in patients with hypodontia.

**Competing interests** None.

**Patient consent** Obtained.

**Provenance and peer review** Not commissioned; externally peer reviewed.

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