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"Solitary Median Maxillary Central Incisor: Case Report And Review Of Literature"

Arpana Bansal, Priyanka Khadatkar, Babita Niranjan, Kartik Choudhary, Prachi Sijeria

ABSTRACT
Solitary median maxillary central incisor (SMMCI) is a rare anomaly that may be associated with a cluster of developmental anomalies. It is sometimes associated with background systemic features, but it could also be a separate localised anatomical aberration. It is estimated to occur in 1:50,000 live births. He presence of SMMCI tooth can predict associated anomalies and in particular the holoprosencephaly. Management depend upon the severity and extend of involvement of syndrome. SMMCI tooth mainly poses an aesthetic problem. However, when it is associated with other defects, the management becomes multidisciplinary.
The objective of this study is to discuss two reported cases of SMMCI syndrome and also review the various reported cases and their management in past.
KEYWORDS*: SMMCI, Developmental anomaly, Narrow nasal aperture, Delayed growth, Deviated nasal septum*

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I. INTRODUCTION

Solitary median maxillary central incisor (SMMCI) syndrome is a rare development disorder affecting the maxillary central tooth germ.[1] It is also associated with multiple development defects of midline structure including the craniofacial bone, nasal cavity airway (choanal atresia and nasal pyriform aperture stenosis), and developing brain (holoprosencephaly (HPE),along with an increased risk of pituitary malformation and malfunction.[2] Although it is autosomal dominant genetic disease ,with isolated trait ,it can also seen as autosomal dominant when associated with holoprosencephaly [HPE] spectrum].[3] HPE is a spectrum of disorder in which brain does not separate into distinct hemispheres and is associated with neurologic impairment dysmorphism of brain and face [4,5].

The presence of SMMCI was first reported in 1958 by Scott DC.[1] "Monosuperoincisivodontic dwarfism'' was the term coined by Rappaport et al, who found its association with short stature. Along with short stature Hall et al found its association also with choanal atresia / mid nasal stenosis syndrome and termed it as ''solitary median maxillary central incisor syndrome or SMMCI syndrome.

SMMCI has on occurrence rate of 1:50,000 live birth with higher incidence on women [1,3]. The aetiology of SMMCI in unknown. It is believed that unknown event occurring between the 35th and 38th days in utero involving midline structure of the head including the cranial bones, the maxilla, its dentition specifically central incisor, the nasal airway, sometime the brain. Li J[3] et al has found 12 genes that were associated with this disease in chromosome region, including TGIF1.[3]

Oral finding is characterized by the presence of symmetric maxillary central incisor, the absence of labial frenum and incisive papilla with narrow nose, v-shaped palate along with unusual narrow ridge at midpalate suture, extending to posterior border of the hard palate.[1]

In past there are only few cases of reported of SMMCI. SMMCI often report to dentist with aesthetic problem. Early diagnosis of SMMCI is important because it may pose just an aesthetic problem to patient but may be sign of other severe congenital abnormalities. Proper diagnosis and comprehensive systematic treatment planning along with multidisciplinary team is must for successful outcome of treatment.

The present case reports describe two cases of SMMCI from the same region. And also review the various cases reported in the past.

Case report 1

A 10-year-old male child was reported to the paediatric department with his main chief complaint of the unsightly appearance of the single mega-incisor in the upper front region. He was the only child born to nonconsanguineous parents. Due to abnormal growth, convex profile, and small stature, a classic syndromic pattern was seen during extraoral testing, also there was limb length discrepancy as one leg was longer than the other leg. The patient also complained of short breath while playing. In order to accomplish a precise

craniofacial and orthodontic diagnosis, X-ray studies were required. (Intraoral periapical radiograph, orthopantomogram, lateral cephalogram, hand-wrist radiograph, CT-PNS)

Medical history

Patients mother underwent a routine delivery and it was uneventful. After birth, the infant weighed 2 kg.

Clinical and X-ray examination

At the clinical extraoral examination the facial profile appeared to be convex. The lips were incompetent because of the mega central incisor (fig 1). Also, an abnormal tooth eruption pattern was noted. There were no signs of temporomandibular joint problems. A solitary upper central incisor and mixed dentition were discovered during the intraoral examination. During the intraoral examination, it was discovered that the patient had a 7-8mm overjet, bilateral angle's class malocclusion, lower anterior crowding, and no crossbite. Only a few teeth showed pre-shedding mobility; no signs of dental caries were discovered. It also demonstrated indistinct philtrum. There was no maxillary labial frenum. Prominent incisive fossa was seen. A very noticeable midpalatal ridge was also observed (fig 2,3).

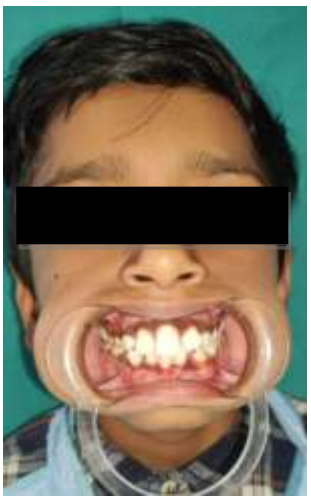


FIGURE 1
(Intra-oral figure showing a mega central incisor in upper front tooth region)

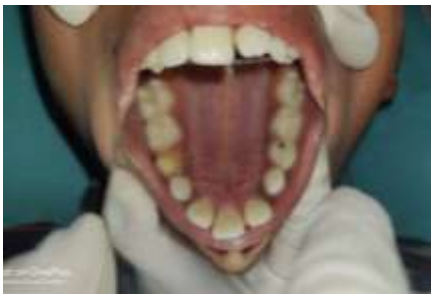


FIGURE 2
(Intra-oral figure showing maxillary arch with single central incisor in the mid with adjacent tooth, and midpalatal ridge can be evidently seen)

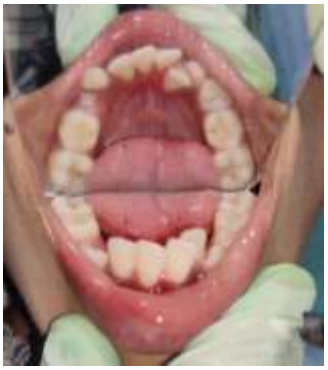


FIGURE 3
(Intra-oral figure showing mandibular arch having minimal crowding in front tooth region and a missing tooth is visible on the right back tooth region)

- HAND WRIST RADIOHRAPH

A hand-wrist radiograph and lateral cephalogram revealed no abnormalities (fig 4, 5).

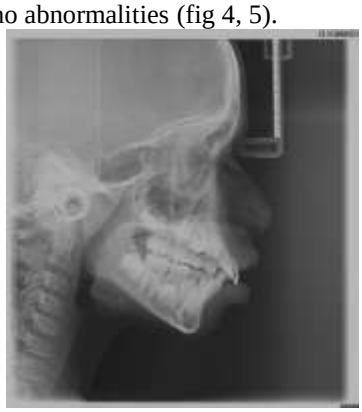


FIGURE 4 **FIGURE 5**
(Both the figure shows no abnormality related to delayed bone growth)

- ORTHOPANTOMOGRAM

Orthopantomography, demonstrated a complete asymmetric eruption of teeth (fig 6). The radiograph also showed agenesis of the central incisor (not determined if 11 or 21). The permanent first molars that had properly erupted and the first signs of second molar root development were seen. A deviated nasal septum was also seen radiographically, prompting the need for additional investigations such as CT PNS.

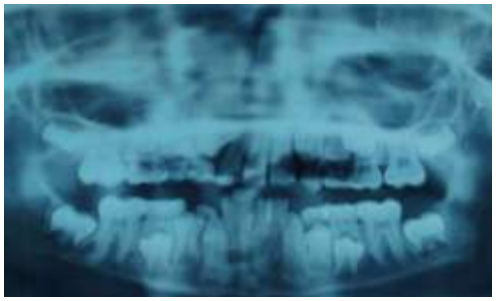


FIGURE 6
(Radiographically asymmetry is visible due to irregular tooth eruption pattern also deviated nasal septum is observed.)

- COMPUTARISED TOMOGRAPHY OF PARANASAL SINUS

CT-PNS scans revealed a relative narrowing of the nasal pyriform aperture. The presence of a midline vomerine ridge and a central maxillary incisor were also noted. The posterior choana appeared normal. Frontal sinuses were well pneumatised and appeared normal. The drainage channels of the bilateral maxillary sinuses were also clear. The sir cells of the ethmoid sinus appear normal. Both sides' osteoeomeatal units appeared normal, with no evidence of any significant variation. The sphenoethmoidal and sphenoid sinuses appear normal. A mildly deviated nasal septum on the left side was observed.

- PRE-OPERATIVE RADIOGRAPH OF PELVIS AND BOTH HIPS (ANTEROPOSTERIOR)

Other than this investigation, a pre-operative radiograph of the pelvis and both hips anteroposterior (PBH AP) was taken to analyse the Risser sign. The Risser sign is used to assess the growth potential of a child. It describes the ossification of the iliac apophysis. Normally, the iliac apophysis ossifies from lateral to medial. No ossification of apophysis in the given PBH-AP X-ray was seen, hence the age is probably around 10-11 yrs. The femoral neck-shaft angle is more, and the pelvis is narrower, so it tells us that it is an x-ray of a male (fig 7).

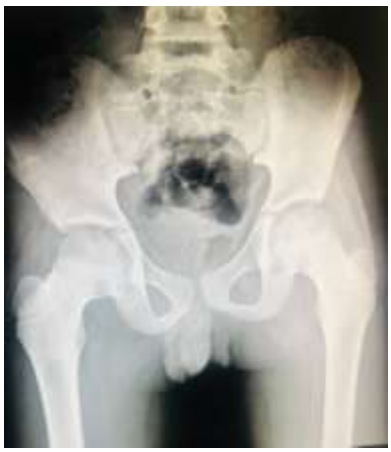


FIGURE 7
(No ossification of apophysis is observed in the given figure)

Case report 2

A normal stature nine year two-month-old patient reported to Department of Paediatric & Preventive Dentistry with chief complain of unerupted upper front tooth. Clinical examination showed only one maxillary central incisor situated exactly in the midline (Fig 8). The central incisor was bigger in size as comparative to the other permanent teeth. Labial frenum and incisive papilla was absent. Philtrum was indistinct (Fig:8). The patient was in mixed dentition phase and had prominent midpalatal ridge. The patient had no history of loss of tooth due to trauma or extraction of any of the anterior teeth.

Patient's parents had nonconsanguineous marriage. Intraoral periapical radiograph showed one central incisor with single root canal (Fig :9). Occlusal radiographs revealed one single maxillary central incisor located exactly in the midline. (Fig :10). A hand-wrist radiograph revels no abnormalities. Orthopantomogram confirmed a solitary maxillary permanent central incisor and all other permanent teeth were developing normally. Further, the patient was referred for complete medical evaluation to rule out other possible systemic anomalies. The diagnosis was confirming after thorough medical check-up by paediatrician and geneticist.

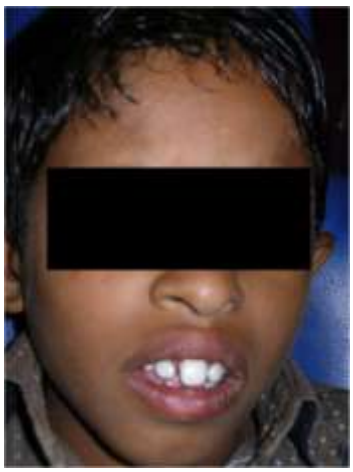


FIGURE 8
(Image shows a big single central incisor present in the upper front tooth region)

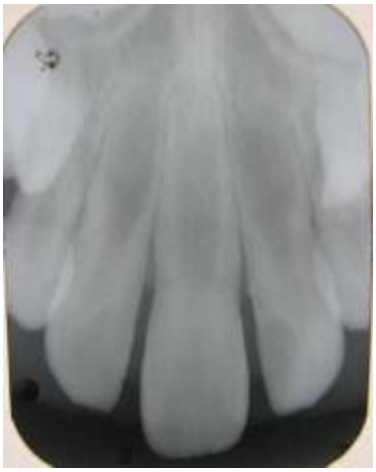


FIGURE 9
(Intraoral periapical radiograph shows single central incisor with apex open)

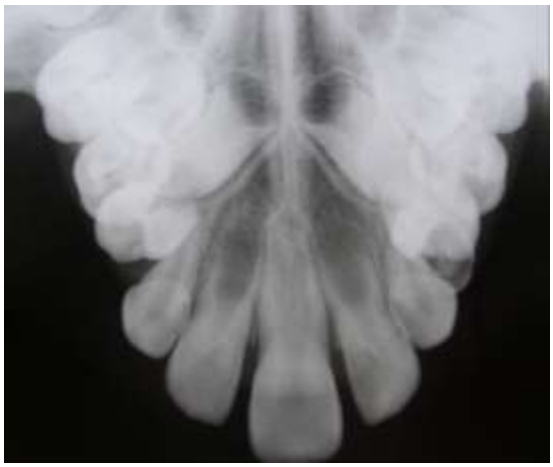


FIGURE 10
(Occlusal radiograph also shows the presence of the single central incisor present in between lateral incisors and congenital missing of another central incisor)

Systematic review
Systematic reviews are routinely relied upon to qualitatively synthesize current knowledge in a subject area. These reviews are based upon following method.

Methodology
This systematic review was conducted to following PRISMA guidelines principles.

Focused questions
PICO ANALYSIS- Children with solitary median maxillary central incisor were made up the study's target population."Whether there is gender dominance in the children having a single median maxillary central incisor," was the analysis of the results.

Eligibility criteria
Studies with a single central incisor in the region of the front teeth were considered.
Studies that were released between January 2012 and February 2023 were included.
The articles were written and published in English.

Information criteria
For identification of studies included or considered for this case report detailed computerized literature search strategies were carried out on following databases. PubMed, PubMed Advanced Search, Google Scholar, articles published between January 2012- February 2023 were included in screening process.

Study selection

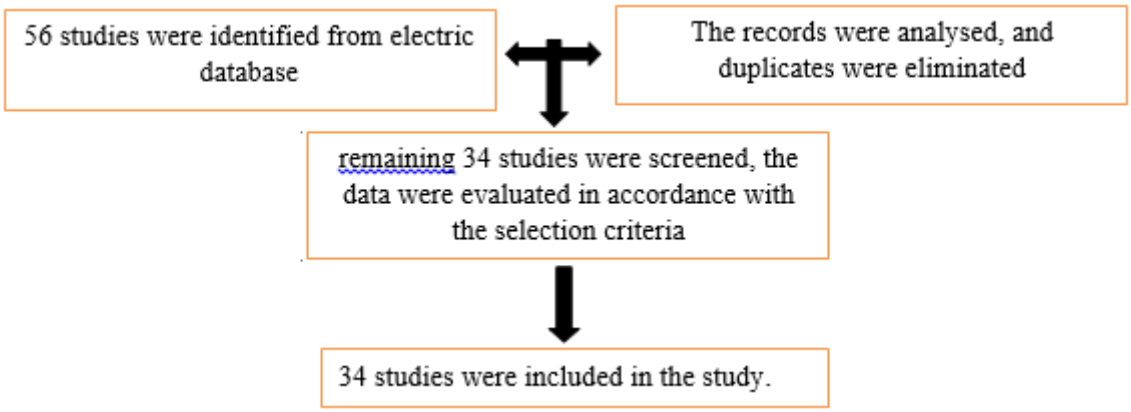
A total of 56 studies were found after searching electronic databases. After reading the titles and abstracts, the records were analysed, and duplicates were eliminated. After 34 studies were excluded, the remaining 34 studies were screened, the data were evaluated in accordance with the selection criteria, and the remaining 34 studies were included in the study.

Data items

Children with a solitary median maxillary central incisor and the presence of gender dominance in this syndrome are variables included in the study.

Outcome and prioritization

Studies were only incorporated if the children had solitary median maxillary central incisor the data were evaluated in accordance with the selection criteria



Review of studies related to the solitary median maxillary central incisor syndrome

Sr. no	Age	Sex	Year	Author	Clinical Features	Treatment Done
01	5 YEARS	F	2012	G. SHILPA et al[6]	-Single symmetrical primary maxillary incisor -absence of labial frenulum, -indistinct philtrum - prominent midpalatal ridge.	-
02	5 YEARS	F	2012	SZAKSZON K et al[7]	- growth retardation -mild intellectual disability -absence of puberty	-
03	4 YEARS	M	2013	LYGIDAKIS NN et al.[8]	- nasal puriform aperture stenosis -respiratory distress	Orthodontic treatment followed by maxillary expansion and then Maryland ceramic bridge was placed.
04	11 YEARS	F	2014	Bernhard C. Pseiner[9]	-presumed microform of holoprosencephaly	Orthodontic space opening A mandibular second premolar was transplanted to replace the missing incisor.
05	9 MONTHS	F	2016	YANG S et al[10]	- congenital nasal pyriform aperture stenosis	Underwent PA surgical repair.
06	6 MONTHS	M	2019	GARCIA RODRIGUEZ R et al[11]	- short stature, corpus callosum anomalies and a microform of holoprosencephaly (HPE), diabetes insipidus, and	Managed by a multidisciplinary team.

					neurodevelopmental delay	
07	12 YEARS	F	2019	MUSTAFA MM et al[5]	- mild nasal stenosis	Extraction of all carious teeth, followed by orthodontic treatment.
08	9 YEARS	F	2020	NEGI A et al[4]	- labial frenulum of the upper lip and incisive papilla were absent -marked midpalatal vomerine ridge	Orthodontic expansion appliance followed by modified removable orthodontic retainer with artificial central incisor.
09	11 YEARS	F	2021	NOTA A et al[12]	Panhypopituitarism - rectilinear eyelid line - reduced interocular distance	-
10	9 YEARS	F	2021	NALAWADE TM et al[13]	- no growth deficiency or any other systemic involvement	Follow-up every 6 months followed by paediatrician and genetic consultant.
11	10 YEARS	F	2022	CHOUCHENE F et al[14]	-swallowing disorder -deviationof the nasal septum to left side	Swallowing rehabilitation combined with palatal expansion and replacement for maxillary central incisor with implant/resin bonded bridge
12	8 YEARS	F	2022	KERBRAT JB et al[1]	- Angle class II occlusion -maxillary transverse deficiency	Osteogenic maxillary distraction followed by fixed orthodontic treatment.

II. Discussion

SMMCI is a rare anomaly that can appear as a standalone finding or in conjunction with other systemic abnormalities. Intraoral examination revealed the presence of a single medially positioned upper central incisor with mixed dentition. According to medical examinations, this patient had delayed growth, limb length discrepancy, narrow nasal pyriform aperture, and mild left-sided deviated nasal septum.

Hall et al.[15] hypothesized that a critical absence or deficiency of lateral growth from the midline – a deficiency caused by a lack of normal cell division at the critical time for midline structures, on or around gestation day 37 or 38 – results in premature fusion of the spreading dental lamina from the left and right sides of the maxilla, preventing the formation of two complete and separate central incisor teeth. Instead, one tooth develops from the inductive epithelium and mesenchymal condensations of these two tooth germs, consisting of two normal distal halves of the central incisors. **Scott** was the first to report the involvement of SMMCI, describing a girl with a single median maxillary central incisor as an isolated finding. **Fulstow** described a case of SMMCI in which the patient had short stature, congenital heart disease, microcephaly, and scoliosis in addition to the single central incisor. There has been a research that has found no correlation between SMMCI and systemic changes.[16] **Wesley et al.** described two cases of SMMCI in patients of normal stature, while **Cho and Drummond** described three cases of SMMCI with no systemic involvement and growth deficiencies. SMMCI has clinical significance because of its association with HPE, which is a fatal disorder when fully expressed. Because SMMCI is thought to be the mildest form of HPE, the children of patients with isolated SMMCI are at risk for HPE.[16,17] **Arlis and Ward** [18] examined six patients with nasal pyriform aperture stenosis and discovered that four of them had SMMCI. **Lo et al**. [19] discovered that 63% of patients with congenital stenosis in the nasal pyriform aperture also had SMMCI, whereas **Hall et al.**[15] discovered that all 21 SMMCI patients had a history of nasal congenital obstruction, 7 had choanal atresia, and 8 had intranasal stenosis.

Until now, very few cases of SMMCI as an isolated anomaly have been reported in the literature, possibly because patients with SMMCI and systemic abnormalities have been reported by both medical and dental professionals, whereas isolated SMMCI has been reported primarily by dentists. Early detection of SMMCI is critical because it may be a sign of more severe congenital abnormalities. SMMCI dental treatment

requires comprehensive treatment planning and a multidisciplinary team of paediatric dentists, prosthodontists, and orthodontists.

The presence of a single medially positioned upper central incisor with mixed dentition was confirmed by intraoral examination. According to the medical examinations, the patient exhibited delayed growth, limb length discrepancy, narrow nasal pyriform aperture, and mild left-sided deviated nasal septum.

III. Conclusion

A case of SMMCI should raise the clinician's awareness of the potential associations. Any patient with SMMCI should be evaluated thoroughly, and the dental and other coexisting developmental anomalies should be diagnosed and managed as soon as possible.

IV. Patient Consent Declaration

The authors attest that they obtained all necessary patient consent forms. The patient(s) parents has/have given his/her/their consent in the form for his/her/their images and other clinical information to be published in the journal. The patients understand that their names and initials will not be published and that while every effort will be made to conceal their identities, anonymity cannot be guaranteed.

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Nil.

Potential conflicts of interest

There are no competing interests.

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