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Solitary Median Maxillary Incisor and Holoprosencephaly: A review of the literature

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Case Presentation: Solitary Median Maxillary Incisor and Holoprosencephaly: A review of the literature

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ABSTRACT/INTRODUCTION

A solitary median maxillary incisor (SMMCI) is a rare anomaly that affects approximately 1:50,000 live births. The presence of an SMMCI tooth may be an isolated finding or may be associated with other anomalies such as Holoprosencephaly (HPE). Early recognition of SMMCIS is important given the possibility of other associated findings that may impact pediatric growth and development. The purpose of this clinical case is to report the clinical findings associated with a nine-month-old patient who presented to our clinics with a primary solitary median maxillary incisor and a diagnosis of HPE. This presentation will review the literature regarding the medical and dental findings associated with HPE and will discuss strategies for appropriately diagnosing and managing care for such patients.

CASE REPORT

A 9-month-old male patient presented to Children's Mercy Hospital (CMH) dental clinic for a new patient exam. Medical and dental history was reviewed with child's mother and was significant for Holoprosencephaly (HPE). The medical history reveals an MRI Brain w/o contrast completed previously. Reason for MRI exam included developmental delay, pyriform aperture stenosis, concerns regarding holoprosencephaly. On the published radiology report dental findings included, "there is a solitary median maxillary incisor and congenital nasal piriform aperture stenosis." Figure 2

Mother's chief complaint is, "He has one really big tooth coming into the middle of his mouth and I'm worried that it's an adult tooth because it's so big." Clinical knee lap examination reveals typical eruption pattern for teeth O, P , D and a solitary median maxillary incisor erupting at the midline. Figure 1 Given the history of HPE a tentative clinical diagnosis of solitary median maxillary central incisor syndrome was given. All other dental findings WNL. Given the infants age no dental intervention were indicated at that time. Instructions regarding a healthy diet and proper oral hygiene were given. Anticipatory guidance discussed regarding tooth eruption given. Follow up six months for further evaluation.

Figure 1

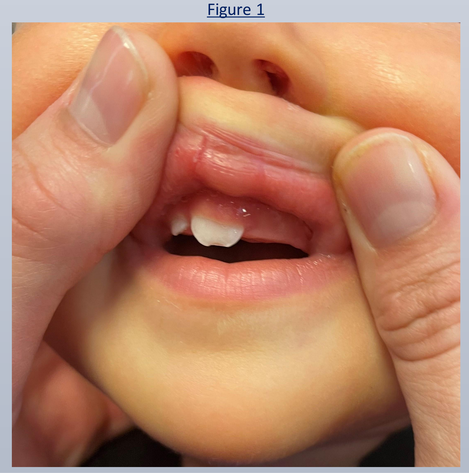
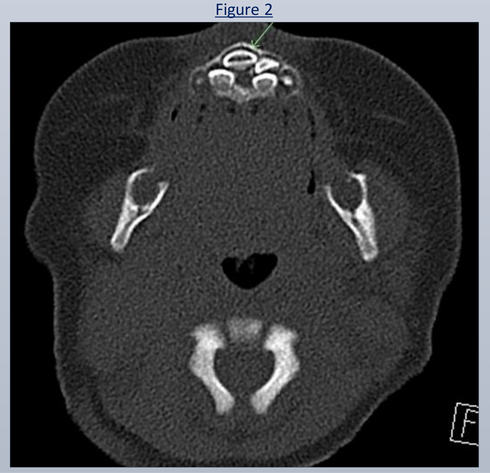


Figure 2



DISCUSSION/CONCLUSION

SMMCI syndrome is a rare developmental anomaly. It was first described by Scott in 1958 and etiology of this syndrome is associated with mutation in gen Sonic Hedgehog (SHH) in chromosome 7q36.3.(3) Holoprosencephaly (HPE) is the most common condition accompanying SMMCI Syndrome. HPE is a developmental defect resulting from incomplete separation of the fore brain during the embryonic period, affecting the development of anterior midline structures.(1) If a SMMCI is present in the primary dentition it's likely to affect the permanent dentition.(2) From a dental perspective the main concern for many parents and patients presenting with a SMMCI is esthetics. Utilizing a multidisciplinary approach is crucial for diagnosis, treatment, and long-term success. At the appropriate age orthodontics, surgical intervention, dental implants, fixed and removable prosthodontics are options to restore function and esthetics In conclusion, patients with HPE and a SMMCI should receive routine dental care to monitor progression through primary and permanent dentition. Dental intervention at the appropriate age should be used to restore function and esthetics.

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