A NOVEL CASE OF A PEDIATRIC PATIENT WITH A
SOLITARY MEDIAN MANDIBULAR CENTRAL INSICOR
AND A MIDLINE NECK MASS

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INTRODUCTION
The presence of a solitary median maxillary central incisor is a finding that has been well described in the literature and is the hallmark feature of a syndrome by the same name. It is frequently associated with other abnormalities, most commonly midline craniofacial defects. However, the incidence of a solitary median mandibular central incisor has not been well described in the literature. We present the unusual case of a patient with such a finding who also had a midline neck mass.

CASE REPORT
An 11 year-old twin boy presented to the office of the senior author, MMA, for evaluation of a neck mass. The mass was first noticed one month prior to presentation. There were no associated symptoms such as pain, changes in phonation, difficulty breathing, or swallowing. He was otherwise healthy with no other medical problems or developmental abnormalities, and no recent illnesses. There were no known genetic anomalies in the family, and his fraternal twin brother was healthy with no medical problems.

On examination, he was a normal appearing 11 year-old male child. Palpation of his neck revealed a soft, non-tender, midline suprahyoid neck mass, approximately 5 cm in diameter. There were no overlying skin changes. His oral exam demonstrated normal maxillary teeth with a solitary median mandibular central incisor. He had no other craniofacial abnormalities.

An MRI exam revealed a central mass in the floor of mouth located immediately above the mylohyoid muscle. There was no surrounding infiltration suggesting inflammatory change. There was no involvement of the hyoid bone or strap muscles. On T1-weighted images it was isointense to muscle, while on T2-weighted images it was hyperintense to muscle. The MRI also further demonstrated the presence of a solitary median mandibular central incisor.

He underwent a successful surgical excision of the neck mass. Final pathology revealed a 3.5 cm dermoid cyst that was completely excised.

CASES IN THE LITERATURE
There are several reports of congenital absence of bilateral mandibular central incisors:
* Lehman (1976) reported a pair of siblings with congenital bilateral missing mandibular central incisors. There was no other information about associated abnormalities and the status of their parents was unknown.*
* Kurtz and Brownstein (1974) described the absence of the mandibular central incisors in a man and 2 of his 3 children, a son and daughter. Dyslexia appeared to be segregating independently in this family.*

*Pitts (1923) described a family in which the mandibular central incisors were missing in 4 male sons. Again, the status of the parents is unknown.*

Only two reports of a solitary mandibular central incisor were identified:
*Oberoi and Vargervik (2005) described a series of three siblings and their mother with velocardiofacial syndrome. One sibling had a solitary central maxillary incisor, while another sibling had a solitary central mandibular incisor. The third sibling and mother had normal dentition.*
*Miller (1941) reported a solitary central mandibular incisor seen in three generations in a Japanese family.*

SOLITARY MEDIAN MAXILLARY CENTRAL INCISOR SYNDROME
Solitary median maxillary central incisor (SMMCI) syndrome was first described by Scott in 1958. It occurs in approximately 1:50,000 live births and its etiology is unknown. The development of a solitary median maxillary central incisor is initiated by unknown errors in development involving the central incisor tooth germ between gestational days 35-38, and is often associated with developmental anomalies in other midline structures.

A diagnosis of SMMCI syndrome requires the occurrence of a single maxillary central incisor with normal crown width located exactly in the middle of the upper jaw in both the primary and permanent dentition. Other causes of a single central incisor must be excluded including traumatic loss of one tooth, a supernumerary tooth which would develop lateral to midline, or instances in which a normal central incisor fails to develop completely in which case the remaining solitary central incisor would also be lateral to midline.

There are several features that are strongly associated with SMMCI syndrome that involve midline structures. These include choanal atresia, midnasal stenosis, or congenital nasal pyriform aperture stenosis, pituitary gland abnormalities, and holoprosencephaly. Other associated features include short stature and intellectual disability. While none of these features are diagnostic, they are found in high frequencies in patients that have SMMCI syndrome. There are many other case reports of additional anomalies seen in these patients; many of them involve midline structures, and include reports of SMMCI syndrome associated with a cervical dermoid.

CONCLUSION
While a rare syndrome, SMMCI syndrome and its associated features are well described in the literature. This is the first case report of a solitary median mandibular central incisor with a cervical dermoid. The association between these two findings is unclear; however, the presence of a cervical dermoid has been described in patients with SMMCI syndrome. Given the association with midline defects, it is warranted to perform imaging of the head and neck in patients who have a solitary median central incisor, including imaging of the central nervous system for patients in which this central incisor is maxillary. This is an unusual case that may represent a new syndrome and we encourage other practitioners to report similar findings.

REFERENCES