Solitary Median Maxillary Central Incisor Syndrome – A Case Report

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Purpose

Pediatric dentists are trained in the diagnosis, treatment, and management of craniofacial abnormalities and dysmorphisms as it relates to management of special healthcare needs and those with medical complexities. This case illustrates a rare midline disorder known as Solitary Median Maxillary Central Incisor (SMMCI) syndrome which presented to Children's Wisconsin in 2020. SMMCI syndrome occurs during the 7th fetal week of the embryonic period and has a strong genetic predisposition.

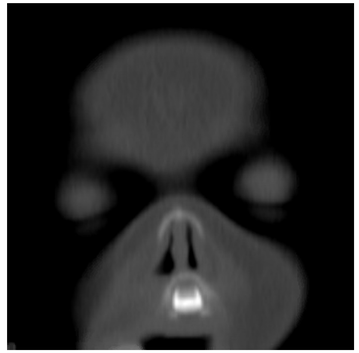
Case Report

A 13-month old female with a history of congenital nasal pyriform aperture stenosis presented to Children's Wisconsin dental clinic after being referred by otolaryngology. Due to young age, extent of treatment needs, and coordination with ENT for additional treatment needs, patient was evaluated and planned for dental treatment in the operating room due to hypoplastic nature of incisor. The parent was presented several treatment options for tooth #E: single anterior strip crown, prefabricated porcelain-faced crown, facial resin shaped and contoured to appear as 2 central incisors. The parent was strongly opposed to the last treatment option and elected to attempt the single anterior strip crown restoration.

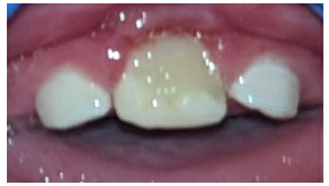
Chromosomal abnormality	Main clinical manifestations
18p deletion	SMMCI, microcephaly, short stature, growth retardation, delayed speech, mild conductive hearing loss
18p deletion	SMMCI, short stature, intellectual disability
18p deletion	SMMCI, anterior nasal stenosis, hypotelorism, growth hormone deficiency, thyroid hormones deficiencies, delayed speech
18p deletion	SMMCI, growth hormone deficiency, pituitary dysplasia
18p deletion; 15p deletion	SMMCI, microcephaly, short stature, frontal lobes dysplasia, small sella turcica, intellectual disability, delayed speech, alopecia universalis, scoliosis
18p11.2 deletion	SMMCI, anterior nasal stenosis, short stature, growth hormone deficiency, ectopic posterior pituitary, delayed speech, absence seizures
18p11 deletion	SMMCI, amblyopia, mild intellectual disability
18p deletion; 4q duplication	SMMCI, short stature, mild intellectual disability, Beckwith-Wiedemann syndrome
18p11.21 deletion	SMMCI, ptosis, protruding ears
ring 18	SMMCI, submucous cleft palate, congenital pyriform aperture stenosis, hypotelorism, microcephaly, short stature, growth hormone deficiency
mosaicism ring 18	SMMCI, deviation of nasal septum/narrow nasal cavity, *columella* dysplasia, hypotelorism, microcephaly, short stature, growth hormone deficiency, frontotemporal atrophy, large cisterna magna, intellectual disability, autistic features, fusion of C2–C3 vertebrae, cryptorchidism, small penis
7q36 deletion	SMMCI, hypotelorism, microcephaly, short stature, growth retardation, intellectual disability
7q36 deletion	SMMCI, hypotelorism, esotropia, microcephaly, short stature, growth retardation, severe intellectual disability, scoliosis
7q36 deletion	SMMCI, microcephaly, growth retardation
7q36 deletion	SMMCI, choanal stenosis, microcephaly, mild intellectual disability
7q deletion	SMMCI, microcephaly, hypertrophy of tonsil, nasal polyp
7q deletion	SMMCI, lumbosacral dysplasia, subcutaneous lumbosacral mass
7q36 deletion; 5q duplication	SMMCI, choanal atresia, hypotelorism, ptosis, microcephaly, short stature, severe intellectual disability, small penis
22q11 deletion	SMMCI, midnasal stenosis, hypotelorism, microcephaly, short stature, Velocardiofacial syndrome (velopharyngeal incompetence)
22q11.2 deletion	SMMCI, deviation of nasal septum/narrow nasal cavity, DiGeorge syndrome
22q11 deletion	SMMCI, Velocardiofacial syndrome, obstructive sleep apnea
22q11 deletion	solitary median mandibular central incisor, cleft palate, Velocardiofacial syndrome
47,XXX	SMMCI, bifid uvula, hypotelorism, intellectual disability, epilepsy, patent ductus arteriosus
1q duplication; 6q deletion	SMMCI, hypertelorism, microcephaly, growth retardation, corpus callosum dysgenesis, intellectual disability, seizures
1p31.3 duplication	SMMCI, deviation of the nasal septum, delayed myelin degeneration, deep sulci in cerebral hemispheres, delayed speech, intellectual disability, epilepsy
2q21.2 deletion; 20p12.1 duplication	SMMCI, hypertelorism, convergent strabismus, short stature, growth hormone deficiency, growth retardation, empty sella, panhypopituitarism, mild intellectual disability, hypothyroidism, absence of puberty, inner genitals dysplasia

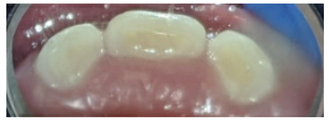
Case Exam and Treatment Photos

CT image from ENT visit (3 m.o)

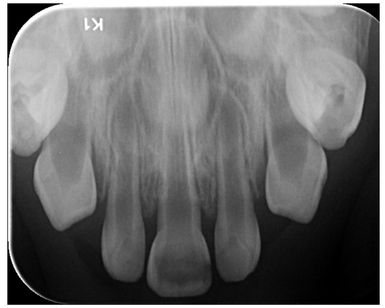


Initial Examination (13 m.o.)

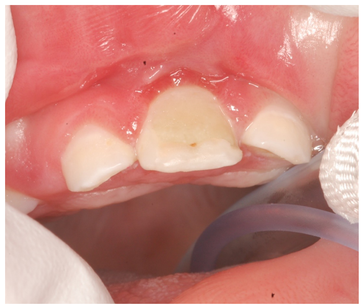




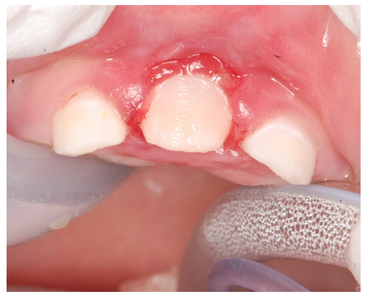
Exam Radiograph



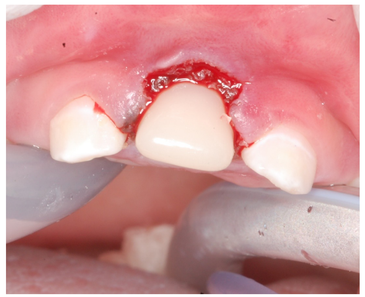
Pre-op Presentation



Tooth Preparation



Tooth Restoration



Treatment options (primary)	Pros/Cons
Anterior Resin Strip Crown	Pros: more esthetic Cons: less resilient than full coverage crown
Prefabricated Porcelain veneered crown	Pros: full coverage Cons: less esthetic, potential for porcelain chipping
Facial resin (Contoured into 2 teeth)	Pros: less invasive Cons: less esthetic ("mini central incisors")
No treatment	Pros: no intervention Cons: hypoplastic nature susceptible to caries

Gene	Nucleotide variation	Main clinical manifestations
SHH	c.331A > T p.I111F	SMMCI, choanal stenosis
SHH	c.331A > T p.I111F	SMMCI, choanal stenosis, slow learner
SHH	c.995T > C p.V332A	SMMCI, choanal stenosis, hypotelorism, microcephaly, patent ductus arteriosus, premaxillary region dysplasia
SHH	c.995T > C p.V332A	SMMCI, cleft palate, hypotelorism, short stature, corpus callosum dysplasia, colpocephaly
SHH	c.420C > G p.H140Q	SMMCI, hypotelorism, microcephaly, neurohypophyseal tumor
SIX3	c.686C > T p.P229L	SMMCI, hypotelorism
SIX3	c.109G > T p.G37C	SMMCI, cleft lip/palate, choanal atresia, ptosis, coloboma, microcephaly, short stature, mild intellectual disability, ventricular septal defect
TGIF1	c.83C > G p.S28C	SMMCI, congenital nasal pyriform aperture stenosis, hypotelorism, microcephaly, growth retardation, corpus callosum dysplasia
COL4A2	c.3896G > A p.G1299E	SMMCI, congenital nasal pyriform aperture stenosis, microcephaly, growth retardation, schizencephaly, dermoid cyst
COL4A2	c.3896G > A p.G1299E	SMMCI, delayed speech, dermoid cyst
DISP1	c.4049delC p.S1350fs	SMMCI, choana stenosis, coloboma of iris and retina, microcephaly, growth hormone deficiency, growth retardation, corpus callosum dysplasia, delayed speech, epilepsy, central diabetes insipidus
ZIC2	c.80C > T p.A27V	
PTCH1	c.109G > T p.G37W	
SIX3	c.514G > A p.A172T	
ASLX1	c.583G > A p.A195T	
SMO	c.1265G > A p.G422E	SMMCI, congenital nasal pyriform aperture stenosis
PLD2	c.956delA p.Q319fs	
P2RY13	c.615G > A p.W205*	

Future Treatment

Due to the nature of primary teeth giving rise to permanent tooth buds, there is close correlation between congenitally missing primary teeth and their permanent successors. Orthodontic treatment may be indicated during mixed dentition to manage missing permanent central incisor. One treatment consideration may include bodily movement of the solitary incisor laterally and restoring neighboring lateral incisor to appear as missing central incisor. Space may then be maintained for future implant/prosthesis to serve as the "missing lateral" that became the "new" central incisor. Growth and maturation of the patient would need to be closely followed in order to properly time successful treatment and placement of future prothesis.

Conclusion

Craniofacial abnormalities with complex sequelae require complex solutions. Interdisciplinary collaboration between specialists (pediatric dentist, orthodontist, prosthodontist, oral surgeon) is recommended for best management of patient's long-term dental needs for those with SMMCI syndrome.

References

de Lima Pedro R, Kchler EC, Primo LG, de Castro Costa M (2017) Solitary Median Maxillary Central Incisor Syndrome: An esthetic solution in a child. J Dent Probl Solut 4(4): 072-075. DOI: 10.17352/2394-8418.000053

Li J, Liu D, Liu Y, Zhang C and Zheng S (2022) Solitary Median Maxillary Central Incisor Syndrome: An Exploration of the Pathogenic Mechanism. *Front. Genet.* 13:780930. doi: 10.3389/fgene.2022.780930