Solitary Median Maxillary Central Incisor due to Nasal Pyriform Aperture Stenosis in Fetus: The First Prenatal Ultrasound Case Report

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CASE REPORT

Solitary Median Maxillary Central Incisor due to Nasal Pyriform Aperture Stenosis in Fetus: The First Prenatal Ultrasound Case Report

Florent Fuchs[1], Manon Chadelle[2], Guillaume Captier[3], Olivier Prodhomme[4], Jean Michel Faure[5]

Abstract

Background: The single median incisor is a rare dental abnormality that could be isolated or could be part of many different syndromes or syndromic association with poor prognosis.

Case description: We report the first prenatal ultrasound description of a 31-year-old patient, gravida 1, para 0, whose male fetus was diagnosed at 25 weeks' gestation with a single median incisor suggestive of nasal pyriform aperture stenosis in Montpellier University Hospital (France). A fetal magnetic resonance imaging (MRI) performed at 30 weeks' gestation retrieved no intracranial midline cerebral anomalies and confirm nasal pyriform aperture stenosis suspicion. Amniocentesis, performed at 31 weeks, found a normal fetal karyotype (46XY) and a normal comparative genomic hybridization (CGH) array. After term vaginal delivery, clinical and radiological examination confirmed the diagnosis of an isolated single median maxillary central incisor linked to nasal pyriform aperture stenosis.

Conclusion: Prenatal diagnosis of a single median incisor due to nasal pyriform aperture stenosis is feasible and enables close postnatal follow-up.

Keywords: Congenital nasal pyriform aperture stenosis, Holoprosencephaly, Prenatal diagnosis, Solitary median maxillary central incisor, Syndromic association.

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Introduction

The single median incisor is a rare dental abnormality that could be isolated or could be part of many different syndromes or syndromic association such as the solitary median maxillary central incisor (SMMCI) syndrome, CHARGE, VACTERL, or velocardiofacial syndrome.[1–3] Solitary median maxillary central incisor syndrome, described by Hall in 1997,[4] is a rare complex syndrome that consists of multiple developmental defects mainly affecting the midline, including the cranial bones, the maxillary and its dentition (specifically the roots of the central incisors), the nasal airways (choanal atresia, midnasal stenosis, or congenital pyriform aperture stenosis),[5–8] and sometimes the brain (holoprosencephaly), associated with other midline structures of the body (cleft lip or palate, esophageal atresia, abnormalities of the external genital organs).[2] Its prevalence is estimated at 1 per 50,000 births.

We report the first case of antenatal ultrasound diagnosis of an isolated single median incisor linked to the nasal pyriform aperture stenosis with favorable postnatal evolution.

Case Description

A 31-year-old patient, gravida 1, para 0, of Romanian origin, with no medical or surgical history, was followed in the Obstetrical Department at Montpellier University Hospital (France). She was not related to her husband. After discontinuing an oral contraceptive, she rapidly became pregnant with a singleton pregnancy with a normal first-trimester ultrasound scan and a low risk for Down syndrome prenatal screening (1/10,000) with a PAPP-A at 1.42 MoM, freeB-HCG at 1.07 MoM. She was referred to the prenatal diagnosis unit because of an unusual imaging of the fetal profile during second-trimester ultrasound imaging. A specialized ultrasound scan, performed at 25 weeks, confirmed the presence of a eutrophic male fetus with a single medial incisor and a narrow superior maxilla possibly ogival, without posterior palatal cleft (Figs 1A to C). Fetal brain was normal with no sign of holoprosencephaly. Additionally, a flattened nasal sulcus and a flat profile were also mentioned (Fig. 1D). The nose aspect and medial central incisor were suggestive of nasal pyriform aperture stenosis.

A fetal magnetic resonance imaging (MRI) was then performed at 30 weeks' gestation, confirming upper jaw dysmorphism of triangular appearance, with a single median incisor and nasal pyriform aperture stenosis (Fig. 2). The morphological assessment of the median line of the brain did not reveal any malformation, especially no holoprosencephaly, no hypertelorism, no choanal atresia, no cerebral lobes agenesis, and no microcephaly. After genetic counseling, an amniocentesis was performed retrieving

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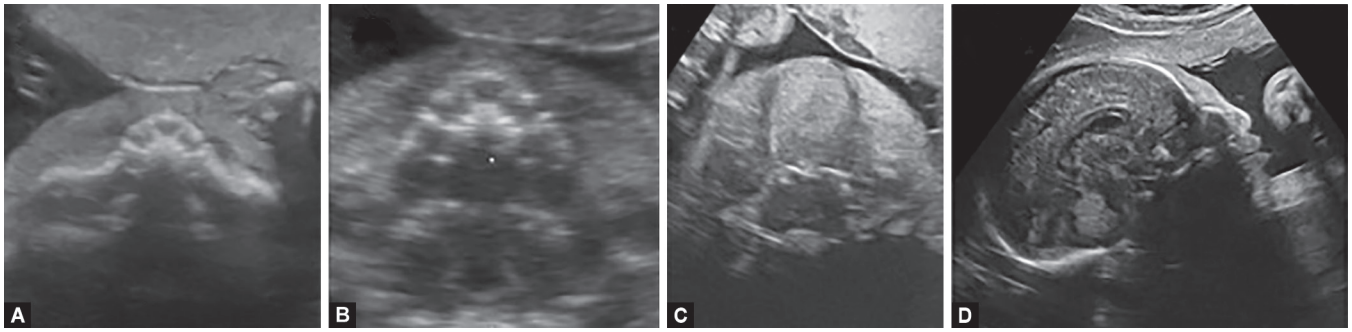
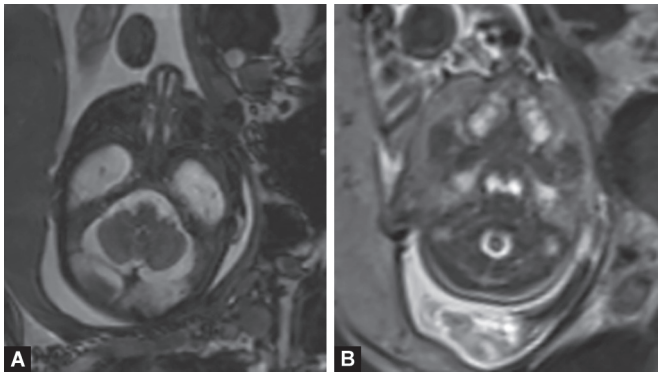
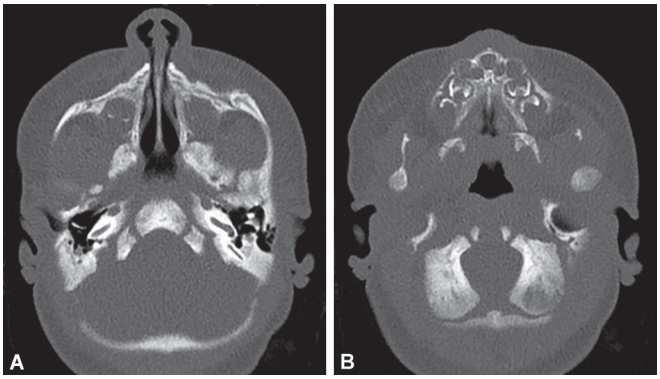


Figs 1A to D: Ultrasound axial transverse view of fetal maxillary and palate showing: (A) A single medial incisor; (B) A narrow superior maxilla possibly ogival; (C) An intact posterior palate; (D) Ultrasound sagittal view of the fetus showing a flattened nasal sulcus and a flat profile



Figs 2A and B: Fetal MRI showing: (A) An aspect of nasal pyriform aperture stenosis; (B) Upper jaw dysmorphism of triangular appearance



Figs 3A and B: Postnatal 3D facial CT scan showing: (A) A nasal pyriform aperture stenosis; (B) A single median incisor

a normal fetal karyotype (46XY) and a normal CGH array. In the absence of cerebral abnormality, the couple was informed not only of the good prognosis of the malformation but also for the need for a specialized consultation with a pediatric otolaryngologist/pediatric plastic surgeon. Depending on the size of the nasal pyriform aperture stenosis, the newborn could experience rapidly after birth or within a few weeks a respiratory distress syndrome, or, he could also tolerate very well the overall hypoplasia. The delivery was therefore organized in a type III maternity for postnatal close survey of the child.

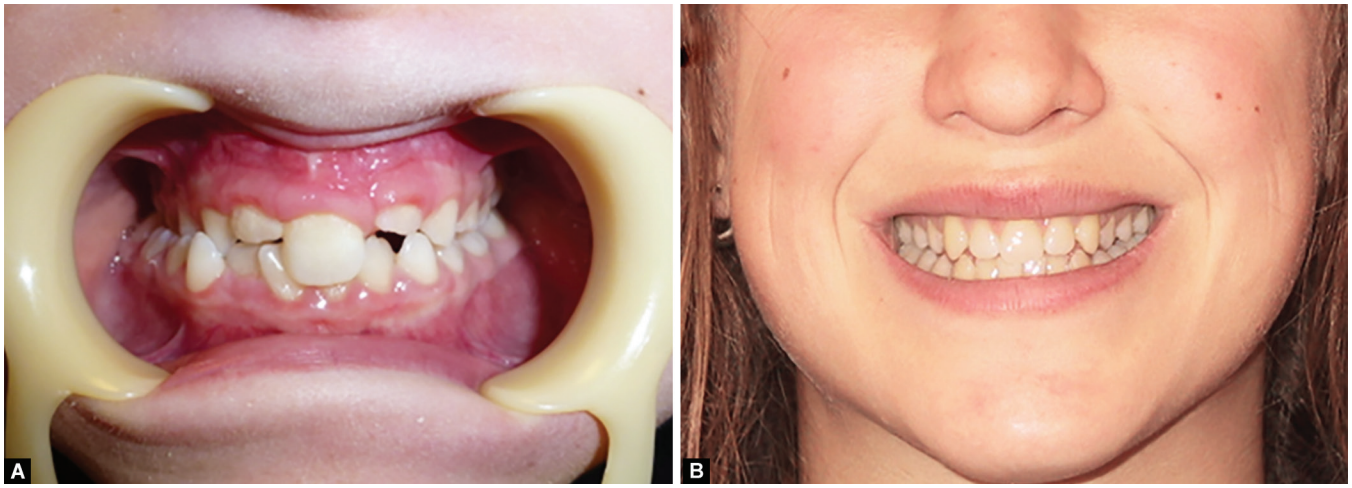
The women delivered vaginally, at 39 weeks' gestation of a baby boy weighing 3085 g (−0.8DS) measuring 46 cm (−1.7DS) with a head circumference of 33 cm (−1DS). He initially adapted very well in the delivery room with an Apgar score of 10 at 5 minutes, venous pH at 7.26, and arterial pH at 7.17. He did not experience immediate transient respiratory distress, and a routine neonatal intensive care unit survey for respiratory disorders was established. The newborn remained in spontaneous ventilation with an air humidifier and no desaturation below 90% was observed despite obstructive dyspnea associated with nasal pyriform aperture stenosis and mid-nasal stenosis confirmed by nasofibroscopy. Neurological clinical examination, child behavior, and paraclinic examinations were noted as normal, especially transfontanellar ultrasound, cerebral MRI screening on day 4, auditory screening, and funduscopic examination. The hormonal balance of the hypothalamic-pituitary axis was also normal.

At 5 days of life, a facial computed tomography (CT) scan confirmed a nasal pyriform aperture stenosis measured at a maximum transverse diameter of 5.4 mm, associated with an overall hypoplastic aspect of the anterior half of the nasal fossae, which had a triangular morphology. The nasal septum, although medial, had a thickened aspect (measured at 4.5 mm). Choanal atresia was not associated and the single median incisor diagnosed prenatally was confirmed (Fig. 3). Due to good respiratory tolerance, the baby boy was discharged at home at 12 days of life.

Discussion

We report the first case of antenatal ultrasound diagnosis of a single median incisor linked to nasal pyriform aperture stenosis without any other associated anomaly. The diagnosis, suspected in the second-trimester ultrasound, was quickly confirmed by ultrasound and then by MRI. This anomaly, when isolated without cerebral midline anomaly, is of good prognosis, besides respiratory risks at birth.

The oral and craniofacial abnormalities probably result from a developmental field defect of the forebrain, arising from unknown events occurring between the 35th and 38th days *in utero*.[4] As mentioned in the introduction part, the single median incisor is a rare dental abnormality that could be isolated or could be part of many different syndromes or syndromic association such as the SMMCI syndrome, CHARGE, VACTERL, or velocardiofacial syndrome.[1–3] Solitary median maxillary central incisor syndrome is a rare complex syndrome that consists of multiple developmental defects mainly affecting the midline, including the cranial bones, the maxillary and its dentition (specifically the roots of the central incisors), the nasal airways (choanal atresia, midnasal stenosis, or congenital pyriform aperture stenosis),[5–8] and sometimes the brain



Figs 4A and B: Photography showing the aspect of untreated single median incisor in a (A) 6 years old boy and in a (B) 25 years old girl

(holoprosencephaly), associated with other midline structures of the body (cleft lip or palate, esophageal atresia, abnormalities of the external genital organs).[2] These children are also usually small in size, often born preterm, hypotrophic, or born from a diabetic mother. Apart from these defects, an acquisition delay or even an intellectual delay can be associated with this syndrome, as well as hypopituitarism.[9] In our case, the child was not hypotrophic (birth weight at −0.8 SD) and the mother did not have preexisting or even gestational diabetes. Among the etiologies of the SMMCI syndrome, there are some chromosomal or genetic abnormalities that have been described postnatally in the literature. Deletions on chromosomes 7 and 18 (7q36.1 and 18p-) that are in the chromosomal regions that carry genes of holoprosencephaly have been reported.[10–12] Children's phenotypes of these deletions were mental retardation, microcephaly, hypotelorism, and a small size but normal levels of plasma growth hormone. At the genetic level, the perturbation of the genes involved in the pathogenesis of holoprosencephaly (SHH, ZIC2, SIX3, TGIF, and DKK1) could also be found in the SMMCI syndrome. A false sense mutation in the SHH gene (I111F) in 7q36 could be specific to the SMMCI phenotype, after a molecular study on 13 cases of SMMCI without holoprosencephaly.[2]

In case of an apparently isolated single median incisor, it is mandatory to perform a fetal MRI in order not only to exclude the presence of holoprosencephaly or complex brain malformations such as semi-lobar holoprosencephaly[13] but also to help in the visualization of the pyriform aperture. It is essential for prognosis and for postnatal management to assess the presence of choanal atresia and to evaluate the nasal pyriform aperture stenosis. Such detection would enable the women to deliver in a center with a neonatal pediatrician present at birth and a pediatric otolaryngologist available. If choanal atresia or severe midnasal stenosis is present, a surgical "sound" will be passed through the bony obstruction and a nasopharyngeal tube (or tubes) will be inserted. If nasal pyriform aperture stenosis is present, a plastic surgeon may be required to enlarge the anterior nares and place a "stent." After birth, 3D cerebral CT is recommended as a useful tool for the postnatal diagnosis of congenital nasal pyriform aperture stenosis.[13]

Finally, the single median incisor is mainly an esthetic problem that most often requires both orthodontic and surgical treatment[14] but may also not be treated (Fig. 4). From the oral care point of view, prevention with an establishment of an oral health program, the preservation of dental capital, and the multidisciplinary management of these dental abnormalities are essential elements of successful treatment.

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