Review Article

Radiological Diagnosis of Maxillomandibular Fusion

Fahd Nasser Al Qahtani

Department, Radiology, Faculty of Medicine, Al-Baha University, KINGDOM OF SAUDI ARABIA.
Email: fahdnasseralqahtani@gmail.com

Abstract

Congenital maxillomandibular fusion of the jaws is a rare disorder, presenting as fusion of the soft (synechiae) or bone (syngnathia) tissue. Scant radiologic literature is available on radiologic findings of maxillomandibular fusion, almost all of which were postnatal imaging findings, such as computed tomography (CT). Ultrasound examination via a sagittal plane of the face can achieve an antenatal diagnosis. In this review, we present the prenatal sonographic and magnetic resonance imaging (MRI) findings and with postnatal CT imaging, and review the existing literature.

Key Word: Congenital maxillomandibular fusion, syngnathia, prenatal sonography, CT, MRI

Address for Correspondence
Dr. Fahd Nasser Al Qahtani, Address: Radiology Department, Faculty of Medicine, Al-Baha University, KINGDOM OF SAUDI ARABIA.
Email: fahdnasseralqahtani@gmail.com
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INTRODUCTION

Congenital maxillomandibular fusion of the jaws is a rare disorder, presenting as fusion of the soft (synechiae) or bone (syngnathia) tissue.[1] It may be isolated or as part of a genetic syndrome like Van der Houde, cleft palate, alveolar synechial and oromandibular limb hypogenesis syndrome.[2] The etiology is unknown but some cases were associated with the consumption of immunosuppressive drugs[3,4] Laster *et al*.[5] classified the types of syngnathia according to the degree of fusion of the mandible to the zygomatic complex and maxillary tuberosity into four types which are:

- 1a Simple anterior syngnathia: Fusion of the anterior region without other malformations in face or neck.
- 1b Complex anterior syngnathia: Fusion of the anterior region with malformations in face and neck.
- 2a Simple mandibulozygomatic syngnathia: Mandibular and zygomatic fusion causing mandibular micrognathia.
- 2b Complex mandibulozygomatic syngnathia: Fusion of the mandibular and zygomatic bones associated with cleft palate or ankylosis.

Problems associated with syngnathia include issues with the maintenance and protection of the airway, feeding difficulties and problems with anaesthesia management. Facial growth and development are also compromised. Surgical treatment has been described, but there is no established standard technique. Treatment is individualized, in terms of jaw function and outcome; it may be problematic incomplex cases. The prognosis of syngnathia type 2b is poor. No case of survival of syngnathia type 2b has been reported.[1] Maxillomandibular fusion is diagnosed after birth when it is discovered that the infant cannot open his or her mouth. This unanticipated diagnosis can potentially result in major complications with airway management and feeding at birth. Thus, prenatal diagnosis in suspected cases can have great clinical implications in that necessary preparations for the tailored care of affected infants can be made before delivery to reduce the chances of infant morbidity and mortality. Multidisciplinary intervention at the birth is important to ensure a correct airway.[6] Scant radiologic

literature is available on radiologic findings of maxillomandibular fusion, almost all of which were postnatal imaging findings, such as computed tomography (CT).[6] Ultrasound examination via a sagittal plane (Fig. 1) of the face can achieve an antenatal diagnosis, as well as its association with other malformations such as agenesis of corpus callosum.[7] Laster *et al*[5] showed one prenatal sonographic image depicting the absence of mouth opening and a contiguous appearance of the upper and lower jaws, although these findings did not lead to a prospective prenatal diagnosis in their case.[5] In this report, we present the prenatal sonographic and magnetic resonance imaging (MRI) findings and with postnatal CT imaging, and review the existing literature. We discuss these findings with a focus on particular signs that may suggest the diagnosis of maxillomandibular fusion prenatally and thus allow appropriate prenatal planning to manage any potential complications at time of delivery.

RADIOLOGICAL FINDINGS:

1. **Prenatal ultrasonography:** On sagittal sonographic images of the face, the lips are intact but protuberant, and the chin is small (Figure 2A). The sonographic images in unaffected fetuses of the same gestational age show a substantial gap or space between the maxilla and mandible (Figure 1, B and C). In contrast, the expected gap between the maxilla and mandible are virtually nonexistent on the sagittal image of the face in the fetus with maxillomandibular fusion (Figure 2A).[6]
2. **Fetal MRI:** The fetal MRI also shows a small chin, a characteristically protruding jaw, and prominent, protruding lips (Figure 3). fetal MRI showed the maxillary and mandibular tooth buds to be closely apposed without the intervening space seen in unaffected fetuses.[6]
3. **Neonatal plain radiographs:** Plain radiographs of the face and head obtained on the day of birth, confirm profound micrognathia.
4. **Neonatal CT craniofacial skeleton:** CT scan with 3D reconstruction reveals bilateral bony fusion of the mandibular and maxillary alveolar processes, including fusion of the proximal aspects of the mandible to the posterior maxilla and zygoma. The CT scan reveals normal zygomatic processes, mandibular condyles, and temporomandibular joints bilaterally (Figure 4).[6]
5.

Figure 1: Sagittal plane of fetus where maxillomandibular bone fusion can be observed syngnathid 2b.

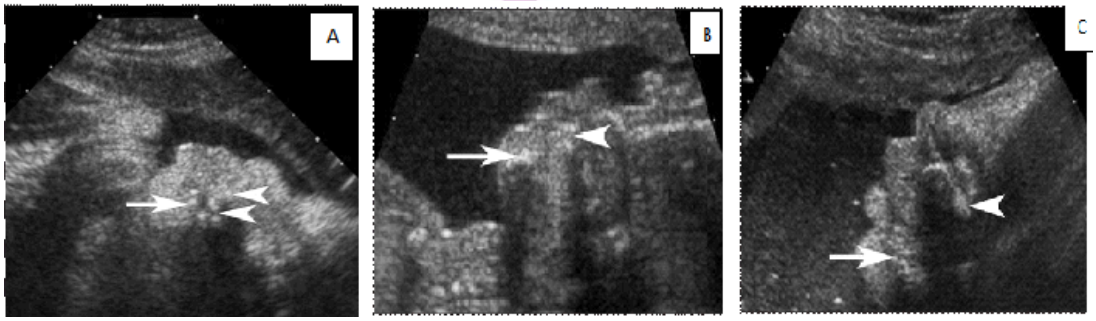

Figure 2: A, Sonogram of a fetus with maxillomandibular fusion showing lack of a normal gap between the mandible (arrow) and maxilla (arrowheads). Also note the small chin and protuberant lips in comparison with **B** and **C. B** and **C**, Sonograms of unaffected fetuses of the same gestational age with closed mouths. Note the substantial gap between the mandible (arrows) and maxilla (arrowheads) in both cases compared with A.

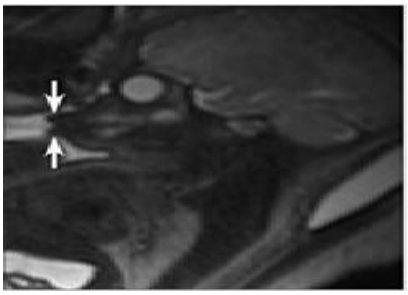

Figure 3: The fetal MRI showing a small chin, a characteristically protruding jaw, and prominent, protruding lips

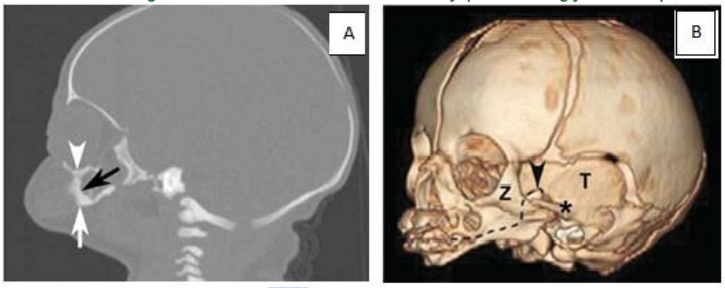

Figure 4: Postnatal CT of the affected neonate. **A,** Postnatal off midline sagittal head CT showing fusion (black arrow) of the mandible (white arrow) and maxilla (arrowhead). Fusion was seen on multiple slices, more easily visualized on the 3D CT reconstruction (B). **B**, Postnatal 3D CT reconstruction of facial bones. Sagittal view depicts fusion of the maxilla and the mandible. The fusion in this case involved bilateral bony fusion of the mandibular and maxillary alveolar processes, with a small anterior slit remaining. The fusion also included fusion of the proximal parts of the mandibles, the ascending rami, to the posterior portions of the maxillas and to the zygomas bilaterally. However, the ascending rami of the mandibles were not fused to the zygomatic processes (asterisk) on either side. The coronoid processes of the mandible (arrowhead) are visualized, pointing to the most proximal aspect of the mandible. T indicates temporal bone; and Z, zygomatic bone. The dotted line represents the expected area of separation between the mandible and the maxilla.

DISCUSSION

Maxillomandibular fusion has always been an unanticipated diagnosis made at birth. Given that congenital oral cavity deformities have potential for causing difficult airway and respiratory impairment, prenatal diagnosis would allow appropriate preparation of staff for airway management at the time of delivery. Given the advanced imaging techniques available today, such as real-time sonography and fast sequences for fetal MRI, maxillomandibular fusion can potentially be detected prenatally.[6] The reported sonographic findings in a case of maxillomandibular fusion, were an absence of mouth opening and a contiguous appearance of the upper and lower jaws. In normal fetus, there is normally a gap between the maxilla and mandible on sonography. Various studies have reported real-time sonography as a reliable method for the evaluation of fetal movement, particularly fetal facial expressions, in the second and third trimesters.[8] These studies reported mouth opening in the context of swallowing, suckling, chewing, and even yawning detected by real-time sonography in fetuses without abnormalities. According to these studies, continued observation by real-time sonography (for anywhere between 15 minutes and 2 hours, depending on the study) is a reliable and reproducible method for visualizing fetal mouth opening. One of the studies even reported regular mouth openings during fetal quiet and active sleep states.[9] Because fetuses with maxillomandibular fusion are incapable of opening their mouths, real-time sonography can a definite method of excluding this rare disorder in suspected cases. Real-time sonography would be best performed during the second trimester for optimal visualization of the fetus. 3D and 4-dimensional (4D) sonography can also be valuable in suggesting or diagnosing maxillomandibular fusion prenatally. Numerous recently published articles emphasized that the interpretation of the fetal image, especially of the fetal face, is easier and faster with 3D images than with 2-dimensional (2D) images.[10,11] Three dimensional sonography has the capability of showing planes of a section that cannot be obtained with 2D sonography and thus allows for a comprehensive evaluation of facial anatomy. Kurjak *et al*[11] commented on the specific advantages of the assessment of the maxilla and mandible for the diagnosis of micrognathia and retrognathia. Surface-mode rendering, in which the surface within the volume of interest (generally the skin) can be seen without the underlying tissue, has proven very useful in the evaluation of facial abnormalities.[10] In the case presented here, the facial characteristics visualized on the The fetal MRI shows the maxillary and mandibular tooth buds to be closely apposed without the intervening space seen in

unaffected fetuses. MRI revealed an open mouth on most sequences in all fetuses without maxillomandibular fusion.[5,6] MRI would likely have been identified on 3D sonography if performed during the second trimester. The maximum-rendering mode highlights the maximal echo (bone) information of a volume data set and is an ideal tool for the 3D reconstruction of bony structures. Generally, cranial bones, the ribs, and other curvilinear bones, which cannot be properly visualized in a single 2D plane, are better assessed in a maximum- mode projection.[11] Selectively imaging the bones on sonography can undoubtedly add to the potential of sonography for suggesting or diagnosing bony maxillomandibular fusion. Finally, 4D sonography, by adding the temporal component (as a virtually live 3D image) in the surface-rendered mode, has been described as advantageous in accurate visualization of subtle and fast facial expressions.[11] The use of 4D sonography to monitor small or fast jaw and mouth movements (complementary to real-time conventional 2D sonography) would also be of potential value in the evaluation of suspected cases of maxillomandibular fusion. Both the management and outcome of patients with bony maxillomandibular fusion have varied. The rarity of the condition makes standardization of treatment difficult.

CONCLUSION

The diagnosis of maxillomandibular fusion should be kept in mind as a possibility if a fetal oral deformity is detected. This is especially true if the fetus has micrognathia because most of the reported cases of maxillomandibular fusion have been associated with some degree of micrognathia. The presence of a closed mouth on prenatal sonography, fetal MRI, or both, especially along with some of the characteristic facial deformities explained above, should suggest the diagnosis of possible maxillomandibular fusion and should prompt further evaluation, such as with real-time sonography, best performed during the second trimester.

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