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Research Letter

# Oral malformation with complete maxillomandibular bone fusion (Congenital Syngnathia)

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*Dear Editor,*

We present an unusual case of fetal facial malformation in a 28 year old woman with no significant family or personal medical history. The patient came to the hospital in the week 35 + 1 with premature rupturing of the membrane. The gestation was terminated with a cesarean section because of the risk of fetal suffering. On delivery facial malformation was observed including fused lips, the presence of bilateral mouth corners and respiratory difficulty. An incision was made in the fused lips which revealed complete maxillomandibular bone fusion. A laryngoscope was inserted and oxygen therapy was carried out without success. As a last therapeutic option a tracheotomy was performed but the newborn was already in cardiac arrest (Fig. 1). A necropsy was requested which confirmed complete maxillomandibular bone fusion (syngnathia), wide, low and flattened base to nose, and a forked tongue. Agenesis of the corpus callosum was identified in the central nervous system and at the level of the base of the skull there was widening of the sella turcica. In the musculoskeletal system widening and flattening of the cervical vertebra could be observed and hidden spina bifida in the lumbar-sacral region (Table 1).

The cytogenetic study gave an XX sex determination chromosome formula and disomy for the chromosomes 13, 18 and 21.

Congenital syngnathia is a rare malformation that may form part of a genetic syndrome or exist in isolation. Varying degrees of severity have been described depending on the extent of the fusion, ranging from a slight fusion of mucosa (synechia) to the complete fusion of both bones (synostosis). Synostosis occurs less frequently than synechia [1,2].

The etiology is unknown but an examination of the literature associates 8 cases with the consumption of immunosuppressive drugs [2].

In the reported clinical cases, we put forward the sagittal plane of the face as pointing to congenital syngnathia resulting from bone

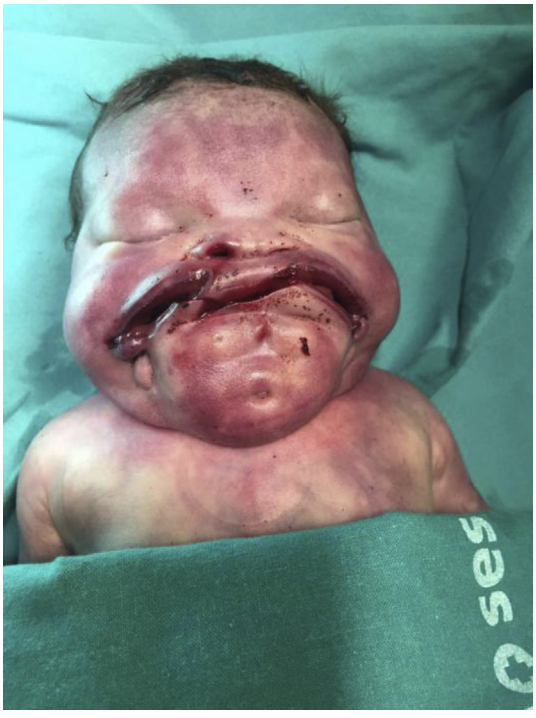

**Fig. 1.** Neonate died after respiratory resuscitation maneuvers. It is possible to appreciate fused lips, the presence of bilateral mouth corners and respiratory difficulty. An incision was made in the fused lips which revealed complete maxillomandibular bone fusion.

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**Table 1**
Necropsy of fetal malformation.

| |
|---|
| Complete maxillomandibular bone fusion (syngnathia). |
| Low and flattened base to nose. |
| Forked tongue. |
| Agenesis of the corpus callosum. |
| Hidden spine bifide in the lumbar sacral region. |

**Table 2**
Classification of syngnathia according to Laster.

| | | |
|---|---|---|
| 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. |

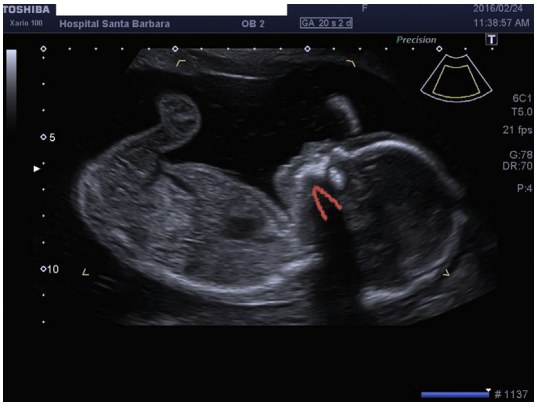


**Fig. 2.** Sagittal plane of fetus where maxillomandibular bone fusion can be observed syngnathid 2b.

fusion accompanied by posterior acoustic shadowing. It is usually accompanied by other genetic defects. On examination of 9 autopsies widening of the sella turcica was present in 9 out of 9 cases, hypertelorism in 8 out of 9 cases, agenesis of the corpus callosum in 7 out of 9 cases, and forked tongue in 5 out of 9 cases [2]. In our case it was also associated with hidden spina bifida. Amongst the syndromes associated with syngnathia are Van der Houde, cleft palate, alveolar synechial and oromandibular limb hypogenesis syndrome [3].

In 2001, Laster et al. classified the types of syngnathia according to the degree of fusion of the mandible to the zygomatic complex and maxillary tuberosity [4] (Table 2).

The case reported would correspond to the type 2b or the most severe degree of the classification. The prognosis of syngnathia type 2b is poor. No case of survival of syngnathia type 2b has been reported.

We propose an ultrasound examination via a sagittal plane (Fig. 2) of the face to achieve an antenatal diagnosis, as well as its association with other malformations such as agenesis of corpus callosum. Multidisciplinary intervention at the birth is important to ensure a correct airway.

## Conflict of interests

The authors declare that no conflict of interests has arisen.

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