Nigerian Journal of Clinical Practice
March 2008 Vol. 11 (1): 78-81

CONGENITAL BILATERAL FUSION OF THE MAXILLOMANDIBULAR ALVEOLAR PROCESSES WITH CRANIOSYNOSTOSIS: REPORT OF A RARE CASE.

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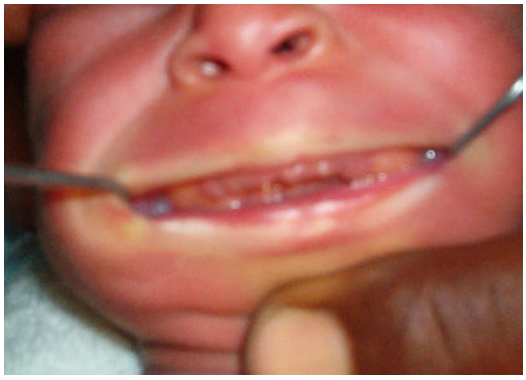
ABSTRACT
A rare congenital anomaly of maxillomandibular fusion with pectus excavatum and craniosynostosis in a neonate is presented. The child was kept alive by nasogastric tube feeding. A modification of classification of syngnathia is proposed. The aetiopathogenesis and difficulty in management in our environment are discussed.

Keywords: Bilateral, alveolar fusion, craniosynostosis, pectus excavatum *(Accepted 14 March 2007)*

INTRODUCTION
The temporo-mandibular joint is a unique and complex joint. Abnormal immobility of the joint could be due to causes within the joint (true ankylosis) or outside the joint (false ankylosis). Very few of this disorder are of congenital origin. Congenital fusion of the jaws may be due to soft tissue (synechiae) or bone (syngnathia) [1] with majority due to bony fusion. Occurrence of ankylosis in neonates and children produces severe micrognathia if bilateral and a deviation of the mandible to the affected side when unilateral [2,3].
In childhood ankylosis, treatment should be instituted as early as possible to restore functional mobility and to stimulate growth of the condyle, thus correcting or minimizing future facial disfigurement. The aim of this paper is to present a case of congenital fusion of the gingival pads with associated craniosynostosis and pectus excavatum and to discuss the problems of management in a rural resource-poor center.

CASE REPORT
A two-day-old female neonate was referred to the Department of Dental Surgery, Federal Medical Centre, Nguru, Nigeria, with a history of inability to open the mouth since birth. The baby was the fourth child of a 25-year-old mother and was delivered at the Maternity wing of the hospital by spontaneous vertex delivery after a full-term of uneventful pregnancy There was no history suggestive of birth trauma, asphyxia, convulsion or spasms. However, there was a maternal history of ingestion of traditional herbal medications during the ante-natal period. Similar presenting symptoms did not occur among other siblings. Clinical examination revealed a full term, ill and hypotonic baby weighing 2.2 kg at presentation.
There was mandibular retrognathia and micrognathia with fusion of the gingival pads (mandible to maxilla) anteriorly. The mouth opening was limited to only a slit of about 2mm,

Figure 1: **Photograph showing maxilliomandibular fusion with a narrow slit (about 2mm) anteriorly**



The anterior fontanelle was absent and a posterior fontanelle was noted measuring 0.5 x 0.5 cm the occipito-frontal circumference was 32 cm. Pectus excavatum was evident

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Figure.2 : **Photograph showing severe micrognathic and retrognathic mandible**

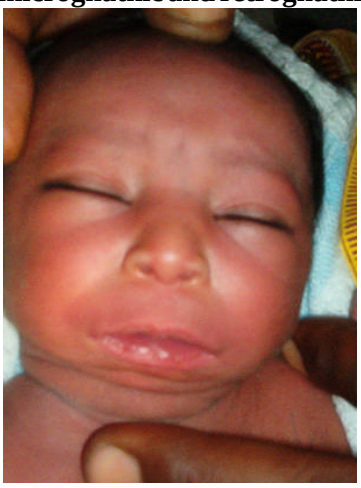
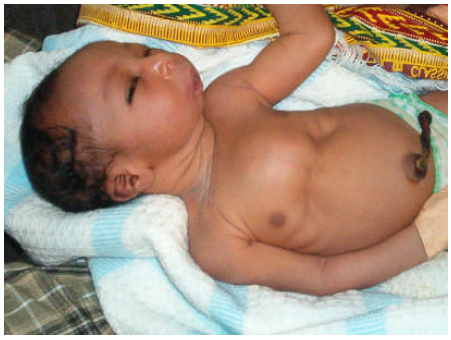


Figure. 3: **photograph showing pectus excavatum**



The palate could not be examined due to limited mouth opening. There were, however, no abnormalities noted in the cardiovascular, urogenital and musculoskeletal systems. Radiographic examination revealed absence of the sutures of the cranial vault and fontanelles, an ill-defined sella turcica with intracranial calcifications. The mandible was irregular and small, with fusion of the alveolar processes of the mandible and the maxilla anteriorly

Figure.4**: True lateral view of the left mandible showing irregular and small mandible with maxillomandibular fusion.**

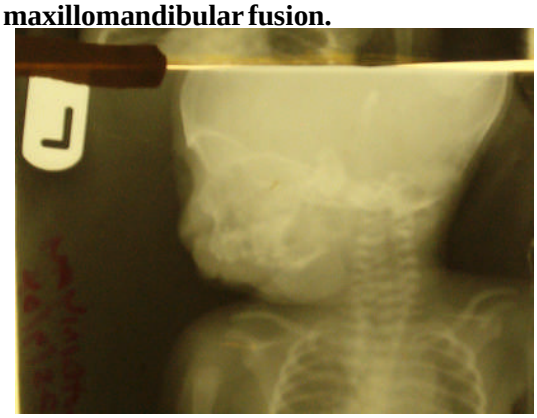
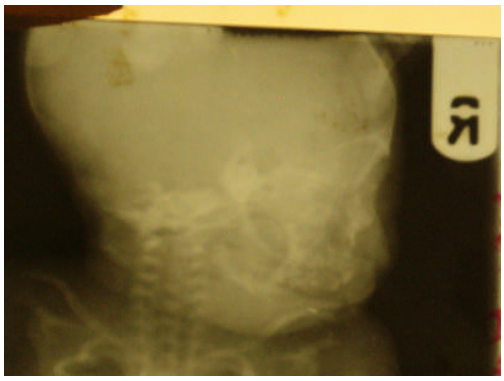


Figure. 5**: True lateral view of the right mandible showing irregular and small mandible with maxillomandibular fusion.**



Based on the history, clinical and radiological findings, a diagnosis of bilateral fusion of the maxillo-mandibular processes associated with craniosynostosis and pectus excavatum was made. Patient was fed via nasogastric tube while preparing patient for surgery. Unfortunately, the patient died one week after birth. Post mortem examination was disallowed by parents for religious reasons.

DISCUSSION

Congenital ankylosis of the jaws is rare. According to Laster et al [4] it may be due to soft tissue (synechiae) or bone (syngnathia). Occurrence in childhood leaves a negative psychosocial effect because of inability to enjoy feeding and poor facial aesthetic. When it occurs bilaterally, there is impaired growth and function of the jaws [5,6]. In a review of the literature, Laster at al [4] reviewed 24 cases of congenital ankylosis to which they added another case. Extensive search of the Internet and the English literature revealed four additional case reports which were not included in the Laster et al's [4] report. Of these [3,6,7,8], one was Nigerian [7]. Analysis of these 29 cases revealed that 20 involved maxillo-mandibular fusion while 9 were of zygomatico-mandibular fusion. Our case reported here had maxillo-mandibular bony fusion, a case of syngnathia thus adding to the previous case report from Nigeria.

Syngnathia has been classified by some authors. Dawson et al. [9] classified syngnathia into 2 main types: Type 1: Simple syngnathia with no other congenital anomalies in the head and neck. Type 2: Complex syngnathia with subgroups: 2a: complex syngnathia with aglossia and subgroup 2b complex syngnathia co-existent with agenesis or hypoplasia of the proximal mandible.

In 2001, Laster et al.[4] proposed another classification of bony syngnathia:

Type 1a: Simple anterior syngnathia characterized

by bony fusion of the alveolar ridges only and without other congenital deformity on the head and neck. Type 1b: Complex anterior syngnathia characterized by bony fusion of the alveolar ridges only and associated with other congenital deformity in the head and neck. Type 2a: Simple zygomatic mandibular syngnathia characterized by bony fusion of the mandible to the zygomatic complex causing only mandibular micrognathia. Type 2b: Complex zygomatic mandibular syngnathia characterized by fusion of the mandible to the zygomatic complex, and associated with clefts or temporomandibular joint ankylosis.

The case presented here is similar to but different from type 1b of Laster et al's [4] classification. Our case has a bony fusion of the alveolar ridges with craniosynostosis and pectus excavatum (Fig 3). We therefore propose a modification of Laster et al's [4] classification of syngnathia by adding a type 3, which will take care of the pectus excavatum observed in this patient.

Type 3: Complex anterior syngnathia characterized by bony fusion of the alveolar ridges only, associated congenital deformity of the head and neck and associated extracranial skeletal deformity.

The aetiopathogenesis and timing of occurrence of congenital jaw ankylosis remains a subject of controversy. Teratogenic agent or trauma [4], anomalous fusion [7], depressed fetal swallowing [10], hypervitaminosis A[11], abnormality of the stapedial artery in the fetus [12], early loss of neural crest cells [13] have been speculated. In this patient we suspect that the aetiopathogenesis is probably related to the ingestion of herbal concoctions during pregnancy. It is possible for the herbs to have some teratogenic effect. In our local environment, it is common practice for ante-natal patients and indeed the generality of our patients to ingest herbal medications because of fetish beliefs, poverty and ignorance. The actual pharmacology of these preparations awaits comprehensive investigations. Demonstration of syngnathia as early as the 23rd week of gestation [4] may be of assistance in elucidating the aetiology and the cause-effect relationship of herbal concoctions in further studies.

In our patient there was microcephaly, an occipitofrontal circumference of 31 cm, which was less than the 10th percentile for age and sex at 36 weeks of fetal gestation. An occipitofrontal circumference of 31 cm is appropriate for a fetus of 34 weeks gestation. The microcephaly observed may be related to the existence of craniosynostosis. While the syndromic presentation of congenital ankylosis of the temporo-mandibular joint, with cranial deformities are noted, the pectus excavatum seen in this patient may be a clinical manifestation of the short sternum often noted in the trisomy heart disease as in Treacher Collins syndrome [14,15] The surgical plan was the excision of the bony fusion but this was not accomplished before the expiration of the patient. The management of ankylosis is challenging particularly in a resource-poor third world country like Nigeria. The problems of management were further aggravated by the rural environment of our hospital. Other problems are poor nutrition, the lack of neonatal anaesthetic facilities such as lack of correct size of neonatal and paediatric anaesthetic/ tracheostomy tube, paediatric fibre-optic laryngoscope, absence of neonatal anesthesiologist and compliance with jaw exercise. The cause of death in this patient remains a speculation of respiratory difficulty, as autopsy was not done. Temporomandibular ankylosis in children usually lead to a compromise of the upper airway and cor pulmonale [16,17].

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