Predictive Factors for Perinatal Outcomes of Infants Diagnosed With Micrognathia Antenatally

Sok Yan Tay, MD¹, Rekha Krishnasarma, MD², Deepak Mehta, MD¹, Amy Mehollin-Ray, MD², and Binoy Chandy, MD¹

Abstract

Introduction: Advances in fetal imaging have allowed us to identify abnormalities previously not appreciated. With this study, we hope to identify factors predicting a difficult airway at birth and review the perinatal outcomes of these patients. Methods: Sixteen patients with antenatally diagnosed micrognathia were reviewed from a tertiary care hospital database from 2011 to 2016. Jaw index (JI), amniotic fluid index (AFI), glossoptosis, gastric size, and oropharynx obliteration were assessed. The airway support required at birth, specialist team involvement, and outcomes were evaluated. Results: Nine (56.3%) of 16 patients had JI <5th percentile, 3 (33.3%) of 9 had difficult intubation, 2 (22.2%) of 9 needed an emergency tracheostomy, and 1 (11.1%) of 9 died. Seven patients had polyhydramnios, 2 (28.6%) of 7 had difficult intubation, 2 (28.6%) of 7 required tracheostomy, and 1 (14.3%) of 7 died. Twelve patients had either JI <5th percentile or abnormal AFI, 5 (41.7%) of 12 had difficult intubation, 2 (16.7%) of 12 required tracheostomy, and 1 (8.33%) of 12 died. For the group without otolaryngology consultation, 8 (50%) of 16, 1 (12.5%) of 8 had difficult intubation and 1 (12.5%) of 8 died because airway was not secured after 45 minutes of resuscitation. Conclusion: Jaw index <5th percentile or abnormal AFI predicts a difficult airway. A multidisciplinary approach with otolaryngology involvement for airway intervention may be required at birth.

Keywords: micrognathia, retrognathia, fetal imaging, airway, perinatal, EXIT

Introduction

Advances in fetal imaging has allowed us to identify abnormalities previously not appreciated.¹ Fetal ultrasonography (US) and magnetic resonance imaging (MRI) are important imaging modalities used in the assessment of fetuses with potential airway obstruction.²

Following the diagnosis of difficult airway prenatally, the most challenging decision to make is to decide if an ex utero intrapartum treatment (EXIT) is required. The benefits of keeping the baby on uteroplacental circulation while securing the airway had to be weighed against the maternal complications that can arise.³

An EXIT procedure involves securing the fetal airway before complete delivery of the fetus. The EXIT procedure was initially used for removal of fetal tracheal clips used in the treatment of congenital diaphragmatic hernia.⁴ Over the years, it is being increasingly performed for fetuses suspected to have airway compromise at birth. The most common indication is for prenatally diagnosed cervical mass resulting in fetal airway obstruction. Severe micrognathia is a relatively new indication for the EXIT procedure.⁵,⁶

The jaw index (JI) is reported to have 100% sensitivity and 98.1% specificity in detecting micrognathia. It is calculated by taking the anterior–posterior (AP) mandibular length divided by the biparietal diameter (BPD) × 100. The presence of polyhydramnios was often associated with upper gastrointestinal tract obstruction in utero due to micrognathia and glossoptosis and can be used as a surrogate marker to predict airway difficulty at delivery.⁷,⁸

To date, most of the studies were case reports or small case series with few patients.⁹–¹¹ With this study, we hope to identify factors that predict a difficult airway at birth and review the perinatal outcomes of patients with antenatally diagnosed micrognathia on fetal imaging.

¹ Department of Pediatric Otolaryngology, Texas Children’s Hospital, Houston, TX, USA
² Department of Radiology, Texas Children’s Hospital, Houston, TX, USA
Received: March 11, 2019; revised: May 11, 2019; accepted: May 16, 2019

Corresponding Author:
Sok Yan Tay, MD, Department of Pediatric Otolaryngology, Texas Children’s Hospital, 6701 Fannin Street, Suite 640, Houston, TX 77030, USA.
Email: sok_yan_tay@nuhs.edu.sg
Methods

The study was performed after obtaining approval from the institutional review board. Fetuses with micrognathia and retrognathia were identified from a large patient database maintained by Texas Children’s Fetal Center from November 2011 to June 2016. All pregnant women referred for a diagnosis of micrognathia and retrognathia with both fetal US and MRI available for review were included in the study. Patients without one of the 2 imaging studies, who did not deliver at the hospital, had chosen to terminate the pregnancy, had fetal demise, and had not delivered at the time the paper was published were excluded.

The imaging studies were retrospectively reviewed by a single radiologist attending for findings and measurements helpful in the prediction of airway difficulty at delivery. The parameters measured included the amniotic fluid index (AFI), JI, glossoptosis, gastric size, and oropharynx obliteration.

The AFI was measured on US and corrected for gestational age. The JI was measured by using AP mandibular length/BPD × 100. The AP length of the mandible was measured on transverse T2-weighted MR image. A line (a) was drawn from a line connecting the posterior border of the masseter muscle at the mandibular insertion (b), connecting presumed mandibular angle (Figures 1 and 2). Biparietal diameter (c) was measured as the distance between 2 internal tables of the skull at the level of the temporal horns of the lateral ventricles on coronal T2-weighted MR image (Figure 3). Jaw index cutoff was set at 23 (2 standard deviations below the mean). Glossoptosis was defined as posterior displacement of the tongue seen on sagittal view of MRI. Gastric size was assessed subjectively using the MRI images. Oropharynx obliteration was defined as absence of T2 signal within the oropharynx on MRI. The antenatal course was reviewed and information on amnioreduction was recorded.

The degree of airway support required at birth, specialist team involved in the airway management, and outcomes were evaluated. The level of airway difficulty was divided into 5 categories: minimal support, easy intubation, difficult intubation, tracheostomy, and died. The definition of minimal airway support referred to the use of nasal airway or blowby oxygen to maintain saturations. Easy intubation was defined as intubation with direct laryngoscopy using a Miller’s blade by an anesthesiologist or neonatologist. Difficult intubation refers to the use of an anterior commissure scope, C-MAC videolaryngoscope blade (Karl Storz, Tuttlingen, Germany), rigid telescope, or flexible laryngoscope for intubation. The group that died refers to the patients who died because of inability to get an airway.

Continuous variables were compared via Wilcoxon rank sum tests and categorical variables were compared via Fisher exact tests.

Results

There were 32 fetal imaging performed. We excluded 16 patients from our study. Six of them delivered at an outside hospital, 3 of them chose to terminate their pregnancy, 5 patients had intrauterine fetal demise, 1 patient had not delivered at the time the publication was submitted, and 1 patient
was excluded because parents opted for perinatal palliative care. We reviewed the peripartum airway management of the remaining 16 patients and correlated it with the fetal imaging findings (Table 1).

We had 9 patients with JI <5th percentile. Of these 9 patients, 3 (33.3%) of 9 patients had difficult intubation, 2 (22.2%) of 9 patients needed an emergency tracheostomy, and 1 (11.1%) of 9 patients died. The remaining patients, 3 (33.3%) of 9 were managed by minimal support using nasal airway, oxygen supplementation, or were easy intubation. Of the 7 patients with JI >5th percentile, 2 (28.6%) of 7 patients were difficult intubation but the other 5 (71.4%) of 7 patients were managed with minimal support or were easy intubation. There were 2 (28.6%) of 7 patients who required oxygen supplementation, 1 (14.3%) of 7 patients who needed nasal airway, and 2 (28.6%) of 7 patients who were easy intubation performed by anesthesiologist or neonatologist (Table 2).

Of the 16 patients, 7 had polyhydramnios and 9 had normal AFI. In the group of patients with polyhydramnios, 2 (28.6%) of 7 patients had difficult intubation, 2 (28.6%) of 7 patients required tracheostomy surgery, and 1 (14.3%) of 7 patients died. In the group with normal AFI, 3 (33.3%) of 9 patients had difficult intubation. The remaining 6 (66.7%) of 9 patients were managed with minimal support or were easy intubation (Table 2).

Twelve patients had either JI <5th percentile or abnormal AFI. In this group, 5 (41.7%) of 12 patients had difficult intubation, 2 (16.7%) of 12 patients required tracheostomy surgery, and 1 (8.3%) of 12 patients died. Four patients in our study had neither JI <5th percentile nor polyhydramnios. They were all successfully managed with either minimal support or were an easy intubation (Table 2).

Otolaryngology was consulted for 8 patients. Four (50%) of the 8 patients had difficult intubation and 2 (25%) of 8 required a tracheostomy. For the remaining 8 patients who did not have an otolaryngology consult, 1 (12.5%) of 8 had difficult intubation and 1 (12.5%) of 8 died because airway was not secured after 45 minutes of resuscitation (Table 2).

**Discussion**

Micrognathia by definition refers to an abnormally small mandible, while retrognathia refers to an abnormally positioned mandible.9 These 2 findings often coexist and can

### Table 1. Summary of Fetal Imaging Findings and Airway Management in the Antenatally Diagnosed Micrognathia Patients.

| Patient | JI < 5th | Raised AFI | Glossoptosis | Pharynx Obliteration | Small Gastric Bubble | ENT Present | Airway |
|---------|----------|------------|--------------|----------------------|----------------------|-------------|--------|
| 1       | Yes      | Yes        | No           | Yes                  | Yes                  | Yes         | Trach  |
| 2       | No       | Yes        | Yes          | No                   | No                   | Yes         | Min    |
| 3       | Yes      | No         | Yes          | No                   | No                   | No          | Difficult |
| 4       | Yes      | Yes        | Yes          | No                   | No                   | Yes         | Trach  |
| 5       | Yes      | No         | Yes          | No                   | No                   | Yes         | Min    |
| 6       | Yes      | No         | Yes          | No                   | No                   | Yes         | Difficult |
| 7       | No       | Yes        | Yes          | Yes                  | Yes                  | Yes         | Min    |
| 8       | Yes      | No         | Yes          | Yes                  | Yes                  | No          | Difficult |
| 9       | No       | No         | No           | No                   | No                   | No          | Min    |
| 10      | No       | No         | No           | No                   | No                   | No          | Min    |
| 11      | No       | No         | No           | No                   | No                   | No          | Easy   |
| 12      | No       | Yes        | No           | No                   | No                   | Yes         | Difficult |
| 13      | Yes      | Yes        | No           | No                   | No                   | No          | Death  |
| 14      | Yes      | Yes        | Yes          | No                   | No                   | No          | Easy   |
| 15      | Yes      | No         | Yes          | No                   | No                   | No          | Min    |
| 16      | No       | No         | Yes          | No                   | No                   | No          | Easy   |

Abbreviations: AFI, amniotic fluid index; ENT, Ear Nose Throat; EXIT, ex utero intrapartum treatment; JI, Jaw index; Min support, minimal support; Trach, tracheostomy.

### Table 2. Summary of the Perinatal Outcomes.

| Airway Difficulty | JI < 5th | JI > 5th | Raised AFI | Normal AFI | JI < 5th or Raised AFI | JI > 5th and Normal AFI | ENT Present | ENT Absent |
|-------------------|---------|---------|------------|------------|------------------------|-------------------------|-------------|------------|
| Min Support       | 2 (22.2%) | 3 (42.9%) | 1 (14.3%) | 4 (44.4%) | 3 (25.0%) | 2 (50.0%) | 2 (25.0%) | 3 (37.5%) |
| Easy              | 1 (11.1%) | 2 (28.6%) | 1 (14.3%) | 2 (22.2%) | 1 (8.3%) | 2 (50.0%) | 0 (0%) | 3 (37.5%) |
| Difficult         | 3 (33.3%) | 2 (28.6%) | 2 (28.6%) | 3 (33.3%) | 5 (41.7%) | 0 (0%) | 4 (50.0%) | 1 (12.5%) |
| Trach             | 2 (22.2%) | 0 (0%) | 2 (28.6%) | 0 (0%) | 2 (16.7%) | 0 (0%) | 2 (25.0%) | 0 (0%) |
| Died              | 1 (11.1%) | 0 (0%) | 1 (14.3%) | 0 (0%) | 1 (8.3%) | 0 (0%) | 0 (0%) | 1 (12.5%) |

Abbreviations: AFI, amniotic fluid index; Difficult, difficult intubation; Easy, easy intubation; ENT, Ear Nose Throat; JI, jaw index; Min support, minimal support; Trach, tracheostomy.
present as a spectrum of severity. They can occur in isolation or as part of a sequence such as Pierre Robin sequence or syndromes such as Cornelia de Lange, Stickler, or Treacher-Collins.10

The airway management associated with micrognathia depends on the severity of obstruction. In mild cases, it can be managed with positional therapy and a nasal airway. However, in severe cases, an EXIT procedure may have to be considered. Failure to do so can result in hypoxic/ischemic encephalopathy and death of the newborn.12

The diagnosis of micrognathia can be made prenatally using fetal US or MRI. Fetal US (Figure 4) is an inexpensive and useful screening tool that can be incorporated in the routine antenatal visits for evaluation of fetal mandible and calculation of JI. However, it is user dependent and is inferior compared to the MRI in visualization of the larynx and trachea for assessment of airway compression.3 Fetal MRI (Figure 5) should be performed when fetal micrognathia is suspected as it provides more detailed information and enables more accurate assessment of the fetal anomalies. In our study, all patients obtained a fetal MRI as a protocol. This information is useful for the team to identify patients who may be at risk of airway compromise at birth.5 One limitation of both imaging studies is the inability to predict the mobility of the jaw. In rare cases, such as arthrogryposis multiplex congenital, severe micrognathia is associated with mandibular condyle hypoplasia and temporomandibular joint disorders. The jaw immobility in such cases is often the cause of failed intubation and airway emergency.13

Previous literature had reported 100% sensitivity and 98.1% specificity of using the JI in diagnosing micrognathia. Aero-digestive obstruction as measured by using the AFI had been shown to have a positive correlation with airway obstruction.14 Our study results were consistent with the literature findings. Patients with JI <5th percentile and polyhydramnios seemed to be associated with difficult airway at birth. All of the 4 patients who had neither JI <5th percentile nor polyhydramnios were successfully managed with minimal support or were easy intubation.

One of the most challenging issues in the management of patients with antenatally diagnosed micrognathia would be the selection criteria for an EXIT procedure. The benefits of avoiding an unplanned emergency airway intervention had to be weighed against the harm done in an unnecessary EXIT procedure. An EXIT procedure can provide up to 150 minutes of placental support to establish an airway, which is extremely important when the airway is predicted to be critical.15 Two patients in our study required emergency tracheostomy. In both patients, the JI was <5th percentile and AFI were raised. One of them had 3 amnioreductions. These 2 patients could have

Figure 4. Fetal ultrasonography (US).

Figure 5. Fetal magnetic resonance imaging (MRI).

Figure 6. Algorithm for airway management of patients with antenatally diagnosed micrognathia.
benefited from an EXIT procedure to avoid an emergency tracheostomy, which is associated with increased morbidity and mortality.

In this study, only 8 (50%) of 16 of patients with antenatally diagnosed micrognathia had an otolaryngology consult. One patient died after resuscitative measures failed. Although the cause of death may be multifactorial, this child has arthrogryposis multiplex congenita which predisposes her to more difficult airway, the inability to get a secured airway likely contributed to the demise of the patient. We proposed that an otolaryngologist, with their expertise in pediatric airway management and experience with airway adjuncts, should be called upon to assist in the airway management in patients with antenatally diagnosed craniofacial anomalies and difficult airway.

In Texas Children’s Hospital, all fetal US suggestive of micrognathia will be followed with a fetal MRI. Currently, if there are radiological findings suggestive of airway obstruction such as (1) JI <5th percentile, (2) raised AFI, (3) reduced IFA, (4) glossoptosis, (5) pharynx obliteration, and (6) small gastric bubble and a history of polyhydramnios requiring amnioreduction, an otolaryngology consult will be obtained. Depending on the severity of the obstruction predicted, the otolaryngologist may be there to standby for airway management if prior intubation by neonotologist and anesthetist was unsuccessful. In cases with severe obstruction, an EXIT procedure may be planned with weekly meeting on fetus progression and EXIT rehearsal prior to actual expected date of delivery (Figure 6).

The limitations of this study were the small patient sample size and it being a retrospective study. However, given the small number of patients with micrognathia diagnosed antenatally, it would be difficult to perform a prospective study. This would serve as a good pilot study for future multi-institutional collaboration.

**Conclusion**

From our study findings and literature review, we recommend that micrognathia diagnosed antenatally should be managed via a multidisciplinary team approach and a shared decision-making process. The presence of JI <5th percentile or abnormal AFI predicts a difficult airway and otolaryngology involvement and airway intervention may be required at birth.

**Declaration of Conflicting Interests**

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

**Funding**

The author(s) received no financial support for the research, authorship, and/or publication of this article.

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