

# Modified EXIT procedure for fetal macroglossia: a challenging case of maintaining uteroplacental perfusion without the use of uterine hypotonic agents.

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## Introduction

Ex-utero intrapartum treatment, the EXIT procedure, is a technique that allows for life-saving fetal interventions while maintaining utero-placental perfusion. It often involves cases of fetal airway obstruction that are detected on pre-natal imaging. Historically, many of these cases would incur a high degree of morbidity and mortality following delivery. By maintaining utero-placental perfusion, these infants can obtain life-saving interventions while avoiding hypoxia. The technique involves partial delivery of the fetus in order to perform an intervention while remaining on placental support. In order to obtain access to the fetus without interrupting utero-placental perfusion, a high degree of maternal uterine relaxation is required. While optimal for the fetus, decreased uterine tone is not without risk to the mother. The EXIT procedure confers the possibility of large volume blood loss for the parturient.

## Case Description

### Chief Complaint:

"Fetal obstructive macroglossia detected on prenatal imaging; concern for morbidity and mortality due to respiratory failure at time of birth."

### History of present illness:

37 Week fetus with polyhydramnios and fetal macroglossia detected on antenatal US, at risk for fetal airway obstruction and/or asphyxia at time of delivery. Fetal MRI demonstrates severe narrowing of the oropharyngeal airway secondary to fetal macroglossia, with absence of 1.5cm hypopharynx, suggestive of severe narrowing and/or occlusion.

### Clinical dilemma:

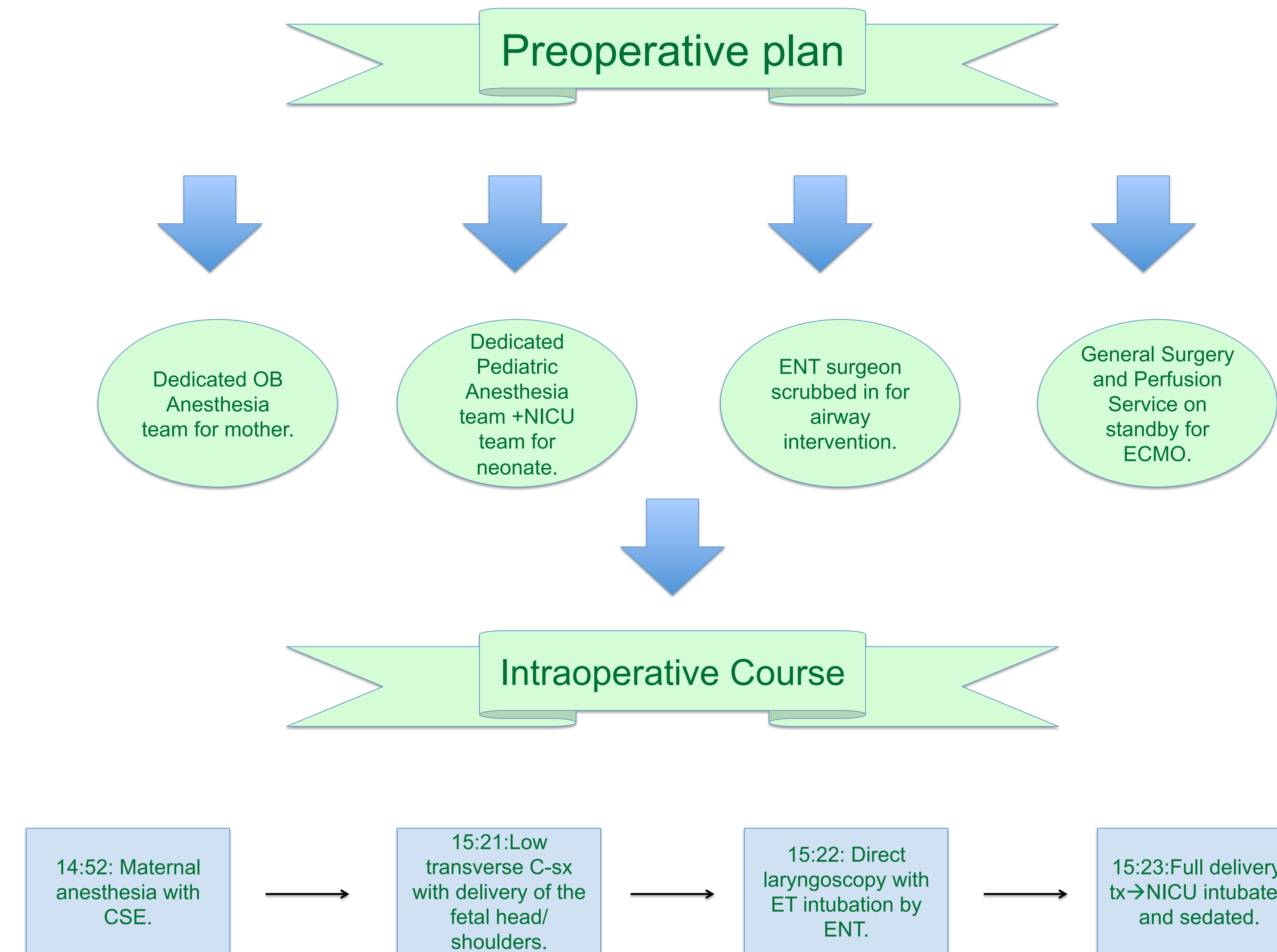
The patient is the unborn fetus of a Jehovah's witness who refuses to accept blood products due to religious beliefs. The mother of the patient declines traditional EXIT procedure due to her increased risk of morbidity and mortality from large volume blood loss with traditional uterine relaxation requirements for EXIT procedure.

### Antenatal hospital course:

- Multispecialty consultations with patient's mother : Ethics, NICU, ENT, General Surgery, OB anesthesia.
- Court order to obtain consent for life-saving blood product administration to neonate if needed upon delivery.
- Multi-specialty team meetings for delivery planning.: Nursing, OB anesthesia, Pediatric anesthesia, Neonatology, ENT, General surgery, Ethics, and Perfusion services.

### Delivery:

- Maternal surgical anesthesia with CSE and IV opioid analgesia; no uterine hypotonics.
- Low transverse cesarean performed with delivery of fetal head and shoulders, with maintenance of uteroplacental perfusion.
- ENT DL with successful endotracheal intubation of the fetus.
- Delivery of the fetus and immediate transfer to the NICU intubated and sedated.



| Maternal Anesthetics   | Maternal Uterine Relaxants                               | Maternal EBL   |
|--|--|--|
| <ul style="list-style-type: none"> <li>• Intrathecal Fentanyl: 15mcg</li> <li>• Intrathecal Bupivacaine: 11.25mg</li> <li>• Intrathecal Morphine 200mcg</li> <li>• Epidural Fentanyl 85mcg</li> <li>• Intravenous Fentanyl 100mcg</li> <li>• Oxytocin 5mg bolus/18u hourly infusion</li> </ul> | <ul style="list-style-type: none"> <li>• None</li> </ul> | <ul style="list-style-type: none"> <li>• Estimated at 1 liter</li> </ul> |

## Discussion

We describe successful fetal airway intervention while maintaining utero-placental perfusion without the use of maternal hypotonic agents, due to maternal refusal of blood products. The patient's mother is a Jehovah's witness who did not wish to accept increased risk of morbidity and mortality from potential blood loss, even if this increased the risk of morbidity and mortality in our patient. Through careful multidisciplinary preoperative preparation, we were able to coordinate a plan to allow for operation on placental support, with minimal increase in risk to the parturient.

## Conclusions

- A short period of maintained uteroplacental perfusion without uterine hypotonics conveyed minimal increased maternal risk compared to traditional C-sx in a patient who refuses blood product administration.
- Successful exposure of the fetal airway can be obtained without uterine relaxation.
- Delayed cord-clamping can successfully maintain utero-placental perfusion in traditional C-sx for short time intervals.
- Careful preoperative planning, and thoughtful multi-specialty teamwork is pivotal to success in these challenging scenarios.

## References

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