

Poster EP09.27

Prenatal sonographic features and EXIT procedure outcomes: case series of nasopharyngeal tumor, complete heart block and CHAOS

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INTRODUCTION

Ex-utero Intrapartum Treatment (EXIT) remains an indispensable means to save fetal life. This case series describes three applications of the ex-utero intrapartum treatment procedure, demonstrating its expanding role in fetal therapy.

NASOPHARYNGEAL TERATOMA

The first case involved a fetus with a large nasopharyngeal mass causing airway obstruction. Ultrasound revealed a 5 x 5 cm polypoidal mass from the right nasal base, filling the cavity and obstructing the left nostril. Fetal MRI showed no intracranial or nasopharyngeal involvement. An emergency EXIT was performed at 32 weeks due to preterm labor. After a lower segment uterine incision, the head was delivered and intubated by the pediatric ENT team, with further stabilization by pediatrics. The tumor was resected postnatally at two months, and histopathology confirmed a mixed germ cell tumor.

This is a rare cause of congenital fetal airway malformation, which is diagnosed prenatally. It enables seamless management from diagnosis to definitive surgical treatment through the EXIT maneuver.

CONGENITAL HEART BLOCK

The second case was a 35-week fetus with severe bradycardia due to complete heart block. The case was referred from another tertiary center due to worsening bradycardia ranging from 27 to 32 bpm despite four antenatal cycles of intravenous Immunoglobulin infusions (IVIg), oral terbutaline, and oral dexamethasone. No failure features were seen on the scan. Subcutaneous terbutaline was administered from admission until just before delivery, and an intravenous terbutaline 0.5 mg was given just before uterine incision. A lower segment uterine incision was made, the baby's head was delivered and intubated by the pediatric anesthetic team, followed by delivery of the rest of the body. Subsequently, the baby was dried, and a cutaneous pacemaker was placed by the pediatric cardiologist on the front and back of the baby's trunk before the cord was clamped and cut. The heart rate was paced at 130 bpm using the transcutaneous pacemaker.

The use of EXIT to Ventricular Pacing is a highly specialized procedure pioneered by Dr. Jay Pruetz to bridge the gap between life-threatening fetal heart block and normal ventricular pacing using novel micropacemaker systems. This transforms a critical neonatal emergency into a highly controlled, synchronized transition.

Bar-Cohen, Yaniv et al. "Preclinical testing and optimization of a novel fetal micropacemaker." Heart rhythm vol. 12,7 (2015): 1683-90. doi:10.1016/j.hrthm.2015.03.022

DEFLEXED HEAD WITH INS MUTATION

The third case: scan and MRI showed a fetus with a severely deflexed head and polyhydramnios. Amniocentesis revealed an INS mutation. She was planned for an EXIT in view of brow presentation and airway obstruction. However, she presented with preterm labor at 31 weeks and delivered spontaneously while waiting for the operating theatre. The baby was intubated ex-utero by the pediatric team.

Current literature identifies INS mutations as a cause of permanent neonatal Diabetes Mellitus, typically characterized by intrauterine growth restriction and hyperglycemia, rather than airway obstruction. However, the fetus was persistently in a deflexed position, necessitating an EXIT procedure.

Hernández, Johana Andrea Botero et al. "Permanent Neonatal Diabetes with High Insulin Requirements due to a New Variant in the INS Gene." Journal of clinical research in pediatric endocrinology vol. 18,Suppl 1 (2026): 45-49. doi:10.4274/jcrpe.galenos.2024.2024-7-11

CONCLUSION

The EXIT procedure provides a controlled, safe environment for securing the airway and establishing emergency cardiac pacing to ensure a smooth transition to ex-utero survival. However, teams must remain dynamic, as unexpected spontaneous labor can prompt immediate ex utero resuscitation.



Figure 1: Mixed germ cell tumour of the fetal nose

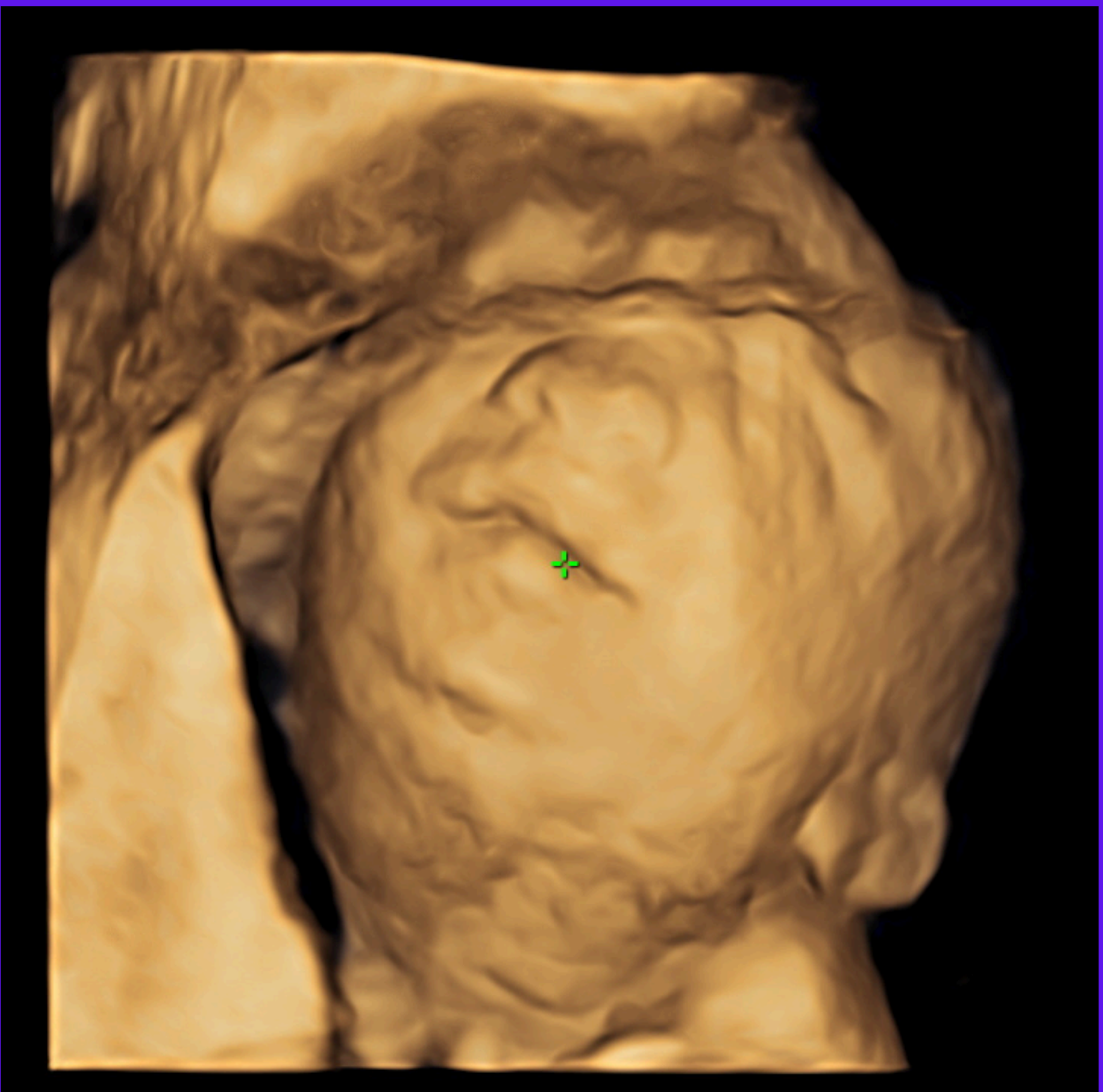


Figure 2: Severely deflexed fetal head causing congenital high airway obstruction (CHAOS)

KEYWORDS: FETAL TUMOR, CONGENITAL HEART DEFECTS, CRANIOFACIAL, POLYHYDRAMNIOS.