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Severe pulmonary hypoplasia associated with giant cervical teratomas

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Abstract

Background: The use of the ex utero intrapartum treatment (EXIT) procedure has salvaged many fetuses with giant neck masses. Despite an adequate airway, a subset of these patients die from an inability to achieve adequate gas exchange.

Methods: We reviewed our experience with the EXIT procedure from 1996 to 2004. The EXIT was used to deliver 23 fetuses with giant neck masses.

Results: Three fetuses with giant cervical teratomas died of severe pulmonary hypoplasia. On postmortem, these patients had severe airway distortion by the mass. The carina was retracted superiorly to the first or second rib resulting in compression of the lungs in the apices of the chest and pulmonary hypoplasia. Hypoplasia was reflected in the lung weights of 24 vs 38 g and 17 vs 34 g for age-matched normal lung.

Conclusions: Unsuspected obstructive fetal neck masses can be fatal because of an inability to secure an airway. Prenatal ultrasonography can identify fetuses at risk, allowing the fetus to be salvaged using the EXIT procedure. Despite obtaining airway control, a subset of these patients will die because of pulmonary hypoplasia. When counseling patients with large cervical masses it is important to discuss potential pulmonary hypoplasia in these patients.

Ultrasonography has increased the prenatal diagnosis of a number of fetal structural malformations that directly impact the perinatal management of the fetus and subsequent outcome. One such malformation is giant fetal neck masses, which include cervical lymphangiomas and teratomas. These lesions can grow to such large proportions that the fetal airway becomes distorted [1], resulting in an inability to secure an airway at the time of delivery [2]. Current therapy for fetuses diagnosed with a giant neck mass is the use of the ex utero intrapartum treatment (EXIT) procedure to secure the airway. The EXIT procedure has been shown to provide time to secure the airway and salvage these fetuses that otherwise may die from an inadequate airway at delivery [3-5]. The salvage of

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this patient population has brought to light a significant subset of patients who, despite an adequate airway, have died of severe pulmonary hypoplasia and an inability to achieve adequate gas exchange.

1. Methods

To examine this subset of patients with pulmonary hypoplasia, we reviewed our experience with the EXIT procedure in the management of fetuses with giant neck masses referred to the Center for Fetal Diagnosis and Treatment at The Children's Hospital of Philadelphia from 1996 to 2004. This study was performed under a protocol approved by the Children's Hospital of Philadelphia Institutional Review Board. Charts were reviewed for the patient's hospital course, the presence of respiratory failure, and patient survival. In addition, the patient's diagnosis was verified by reviewing the final pathological evaluation of resected specimens and autopsy records.

2. Results

We used the EXIT procedure to deliver 23 fetuses with giant neck masses. There were 11 cervical teratomas and 12 lymphangiomas. Of the 23 patients undergoing the EXIT procedure there were 5 mortalities. Three of these mortalities were in fetuses with giant cervical teratomas that died of severe respiratory insufficiency and an inability to oxygenate these patients despite maximal ventilatory support.

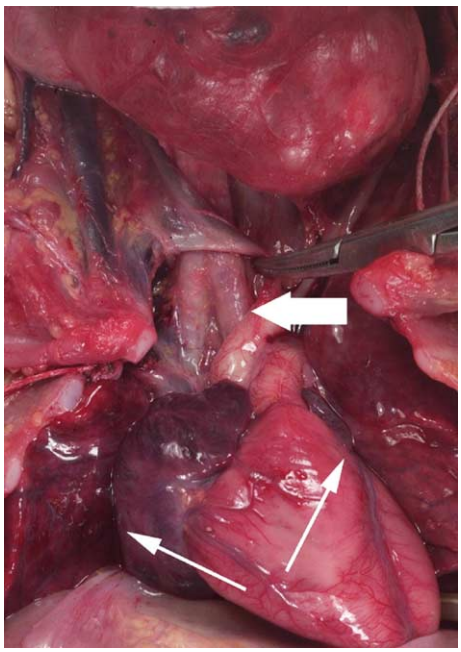

Fig. 1 Postmortem photograph of the open thorax of a neonate with a large cervical teratoma delivered via the EXIT procedure who subsequently died of severe pulmonary hypoplasia. The carina is indicated by the wide arrow and is shown in the patient's neck above the level of the clavicles. The mainstem bronchi are distorted, oriented in a vertical manner and originate in the neck. The lungs are indicated by the narrow arrows and are compressed in the apices of the chest cavity.

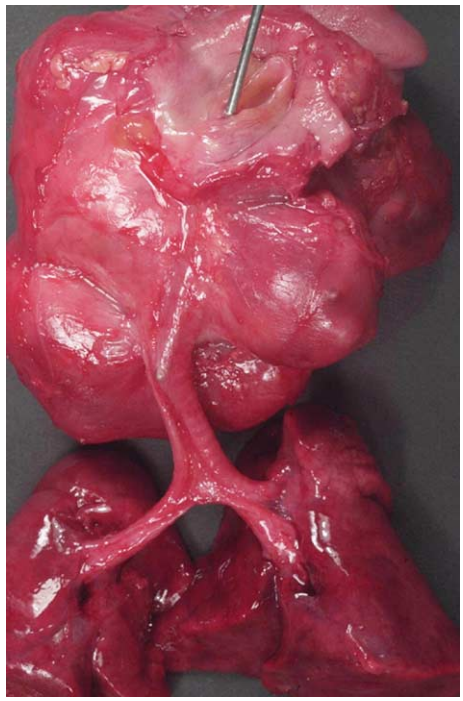

Fig. 2 Postmortem photograph of a large cervical teratoma, airways, and lungs from a neonate delivered via the EXIT procedure who died of severe pulmonary hypoplasia. A probe is seen in the proximal airway demonstrating the airway to be patent. The lungs are small and distorted from being compressed in the apex of the chest cavity.

On review of the postmortem examination, these 3 patients with giant cervical teratomas had severe distortion of the airway by the mass. In these patients, the trachea was stretched out over the mass, retracting the carina superiorly to at least the level of the first or second rib (Fig. 1). This resulted in compression of the lungs in the apices of the chest, a vertical orientation of the mainstem bronchi, and severe pulmonary hypoplasia (Fig. 2). Despite severe airway distortion, the airway remained patent (Fig. 2). The pulmonary hypoplasia seen on gross pathological examination of these patients was also reflected in the total lung weights. One fetus had a total lung weight of 24 vs 38 g for age-matched control normal lungs. The second fetus had a total lung weight of 17 vs 34 g for age-matched control normal lungs. The third patient died of respiratory insufficiency and hypoxemia on day 7 of life. The lungs from this patient were edematous from fluid resuscitation and did not accurately reflect lung weights at birth.

The other 2 mortalities were in seen in patients with giant cervical lymphangiomas. One patient had an extensive lymphangioma involving the face, oropharynx, and larynx