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Prognostic significance of an antenatal magnetic resonance imaging staging system on airway outcomes of fetal craniofacial venolymphatic malformations

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ABSTRACT

Background: The aim of the article was to determine if anatomical findings on fetal magnetic resonance imaging (MRI) of venolymphatic malformations of the face and neck (VLMFN) can be used to create a staging system predictive of airway outcomes.

Methods: We reviewed 13 fetuses evaluated for VLMFN. Stage was assigned based on anatomical findings on fetal MRI. Stage I: no evidence of polyhydramnios with free egress of amniotic fluid and clear visualization of the aryepiglottic folds and larynx. Stage II: lesions of the tongue or epiglottis but with normal aryepiglottic folds without polyhydramnios. Stage III: lesions of the tongue or larynx; nonvisualization of the aryepiglottic folds without free egress of amniotic fluid along with polyhydramnios.

Results: Six met stage I criteria with no airway involvement, nor any subsequent issues. Two met stage II criteria and were managed by ex-utero intrapartum therapy and intubated. One had minimal involvement of the upper airway, was extubated, and had no subsequent issues. Child two had involvement of the tongue and larynx and received a tracheostomy. Five were assigned stage III, delivered by ex-utero intrapartum therapy and intubated. Postnatal evaluation showed involvement of the upper airway by the lesion and was managed with tracheostomy. All treated by tracheostomy remain cannulated because of persistent symptomatic lesions at follow-up (relative risk 4.0; confidence interval 1.2-13.3). Median follow-up was 4 y (range 2-7 y).

Conclusions: Although numbers are small, data suggest anatomical details obtained by antenatal fetal MRI appear to correlate with airway outcomes in children affected by a VLMFN. This information may be useful when counseling expectant families of affected fetuses.

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Introduction

Venolymphatic malformations of the face and neck (VLMFN) are rare congenital lesions, having an incidence of one in 2000-4000 live births.[1]These lesions are believed to result from anomalous growth and obstruction of developing lymphatic channels. Approximately 50% of these benign malformations are of the head and neck.[2] Although benign, involvement of the face and neck causing obstruction or compression of the upper airway can be immediately life-threatening. With continued improvements in image quality along with a broad application to the population, affected fetuses are increasingly likely to be identified prenatally. These images provide the opportunity for antenatal evaluation, family counseling, and delivery planning to best facilitate the management of a potentially involved airway on birth.

Several postnatal staging systems demonstrate some ability to predict prognosis after medical and surgical intervention. In 1995, de Serres *et al.*[3] proposed a staging system based on anatomical location and extent of the lesion. This staging system demonstrates the importance of the anatomical relationship of the lesion in regard to the infrahyoid or suprahyoid positions on outcomes. Wiegard *et al.*[4] proposed a staging system based on involvement of the tongue and floor of the mouth suggesting, with increased involvement of these structures, a poor prognosis was to be expected. In 2013, Berg *et al.*[5] published a review of their experience demonstrating the importance of laryngeal obstruction because of cervical extension of a VLMFN. In this study, stages were applied based on anatomical details and then correlated with outcomes. Stage I was described as having mass effect abutting the larynx but otherwise having grossly normal laryngeal anatomy. Stage II was defined as having involvement of the epiglottis but with limited effacement of the glottis opening. Stage III lesions had more extensive involvement of the epiglottis and aryepiglottic folds. Finally, stage IV lesions had diffuse microcystic involvement of the epiglottis and aryepiglottic folds (Figure).

The goal of this study was to review the airway outcomes of fetuses referred to our center having a VLMFN on antenatal magnetic resonance imaging (MRI). These fetuses were referred because of concerns for potential airway involvement that might affect management of the airway at birth. Fetuses having evidence of an airway obstruction by a VLFMN are managed by ex-utero intrapartum therapy (EXIT) to obtain an appropriate airway before separating the fetus from the placental circulation.[6] Because existing VLMFN staging systems have not included fetuses as subjects, we aimed to correlate airway outcomes with anatomical data generated by fetal MRI using criteria that might be similar to gross anatomical criteria as described by published staging systems.

Methods

Institutional review board approval was obtained for this retrospective study. The records of patients referred to the fetal center at Children's Health/UT Southwestern Medical Center because of concerns for airway involvement by a VLMFN between 2007 and 2015 were reviewed. Imaging interpretations of antenatal MRIs were examined for anatomical detail. An analysis of clinical notes for specific details regarding airway management during the perinatal period and subsequent follow-up was carried out. Specifically, need for tracheotomy, use of interventions both operative and nonoperative, and success in decannulation. Attempts were made to contact parents for follow-up information when appropriate.

Staging points for the study were derived from Berg *et al.*[5] as interpreted by the fetal radiology member of the study team. This was done by correlating the postnatal

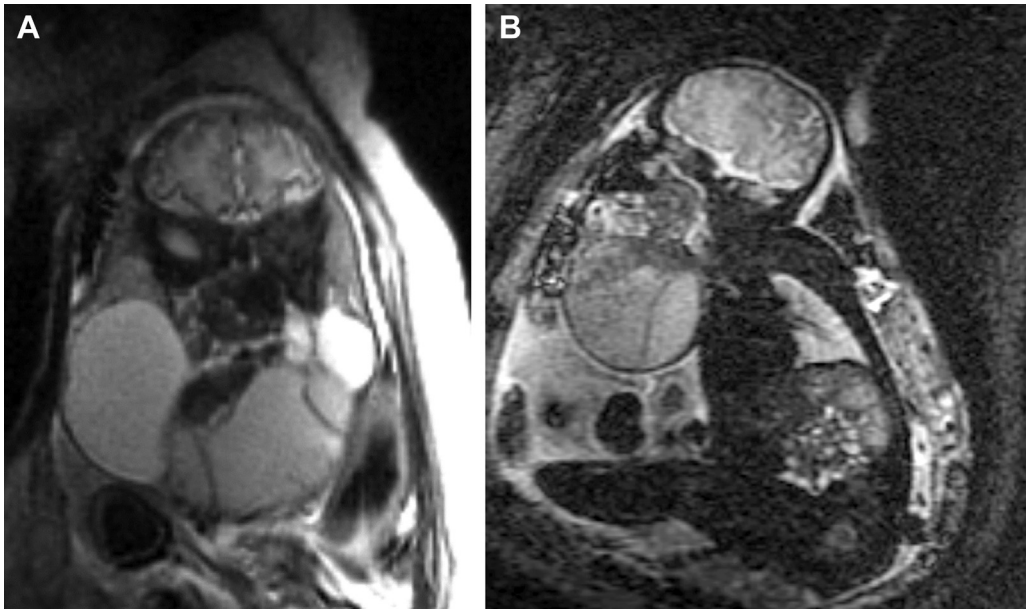

Figure – Coronal (A) and sagittal (B) images of a 32-wk fetus with a grade III venolymphatic malformations.