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Pre- and post-operative visualization of neonatal esophageal atresia/tracheoesophageal fistula via magnetic resonance imaging

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ABSTRACT

Esophageal atresia (EA) is a relatively uncommon congenital anomaly, often observed in conjunction with tracheoesophageal fistula (TEF). Surgical repair in neonates typically takes place with little information about the pre-existing EA/TEF structure because there are currently no acceptable tools for evaluating EA/TEF anatomy prior to repair; chest x-ray radiograph does not identify malformation sub-type or gap length, while x-ray computed tomography (CT) demonstrates an unacceptably high exposure to ionizing radiation. There is a need for safe imaging methods to evaluate pre-operative EA/TEF anatomy, which would add value in surgical planning; this need may be met with high-resolution structural MRI.

We report three cases of Type-C EA/TEF in neonates. Patients were imaged prior to surgical repair using high-resolution ultrashort echo time (UTE) magnetic resonance imaging (MRI) to visualize tracheoesophageal anatomy and allow for informed surgical planning and risk management. One of the three patients was imaged post-repair to evaluate surgical efficacy and evolution of the tracheoesophageal anatomy.

1. Introduction

Esophageal atresia (EA) is a relatively uncommon congenital anomaly that affects approximately 1 in 2500–4000 live births [1]. EA is most often observed in conjunction with a tracheaoesophageal fistula (TEF), typically between the trachea and distal esophageal pouch (Type-C); other classification subtypes include Type-A (EA without TEF), Type-B (EA with proximal TEF), Type-D (EA with proximal and distal TEF), and Type-E (TEF without EA) [2,3]. EA/TEF is considered a surgical urgency, involving ligation of the fistula and anastomosis of the esophagus. Repair is required to avoid aspiration, reflux, pneumonitis, gaseous-distension of the stomach, and ultimately to establish a normal feeding mechanism [4].

Currently, there are no acceptable tools for evaluating EA/TEF anatomy prior to repair. In most patients, the EA/TEF diagnosis is confirmed only with a plain chest x-ray showing a coiled feeding tube within the upper esophageal pouch. Importantly, this approach to "confirmation" cannot determine the anatomic subtype of EA/TEF, the number or location of TEFs, the size of the gap between proximal and distal esophagus, or the presence of tracheomalacia. A few studies have evaluated computerized tomography (CT) scanning in pre-surgical evaluation of EA/TEF and found that CT could frequently identify the origin of the fistula and the size of the EA gap [5,6]. Although these studies suggested a benefit from preoperative CT scans improving surgical planning in 40% of cases, they also demonstrated a significant and unacceptably high exposure to ionizing radiation. Thus, there is a need for safe, non-invasive, non-ionizing imaging methods for visualizing and assessing pre-surgical EA/TEF anatomy, which would add value in surgical planning; this need may be met with high-resolution structural MRI. Such imaging may help identify infants at risk for complications and offer clinical teams an opportunity to reduce these risks. Additionally, post-operative imaging may provide an opportunity to evaluate surgical efficacy and developing function during post-surgical healing within the repaired anatomy.

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Abbreviations

CT	x-ray computed tomography
DOL	day of life
EA/TEF	esophageal atresia/tracheoesophageal fistula
MRI	magnetic resonance imaging
UTE	ultrashort echo time

2. Description of case reports

2.1. Case 1

A 38 3/7 week gestation male neonate (birth weight = 2520 g) presented to Cincinnati Children's Hospital on day of life (DOL) 0 with cleft palate, increased secretions, and an inability to pass nasal gastric tube. Family history was significant with a sibling also diagnosed with EA/TEF and Pierre Robin sequence. A chest radiograph on DOL 0 demonstrated coiling of the feeding tube in the upper esophagus, suggesting EA with possible TEF; the chest radiograph also suggested a long proximal esophagus and a short esophageal gap length (Fig. 1A).

The patient was imaged on DOL 1 on a 1.5 T neonatal-sized MRI scanner [7–9] on room air without sedation using an ultrashort echo time (UTE) sequence [10–12] under an IRB-approved research protocol and with informed parental consent. Images had a 3D isotropic resolution of 0.7 mm (Fig. 1B). Tracheoesophageal anatomy was semi-automatically segmented from UTE MR images (ITK-SNAP 3.6.0, 3D structure segmentation, University of Pennsylvania and University of Utah [13]), and 3D surface renderings of tracheoesophageal anatomy were visualized with volume-rendered MR images (ParaView 5.4.0, data analysis and visualization) (Fig. 1C). MRI results indicated the patient had a Type-C EA/TEF with an esophageal gap length of approximately 1 cm, consistent with the chest radiograph. MRI also indicated a long, narrow fistula connecting the distal trachea to a caudally displaced distal esophagus, potentially increasing the effective esophageal gap length and increasing the surgical difficulty in connecting the distal and proximal esophagus. MRI also demonstrated a flattening of the trachea where it crosses the innominate artery, suggestive of a degree of tracheomalacia.

Due to the expected repair difficulty presented by the long, narrow fistula observed on MRI imaging, the surgical plan was changed from thoracoscopic to open approach, and the EA/TEF was repaired on DOL 2 by ligation of the TEF and primary anastomosis under mild tension. Bronchoscopy at the time of repair confirmed a TEF originating immediately proximal to the carina. The patient remained hospitalized until 3 months of age due to complications associated with his Pierre Robin sequence and his gastric tube. At the time of discharge he had feeding problems related to micrognathia, but no esophageal leak or esophageal stricture.

2.2. Case 2

A 38 3/7 week gestation female neonate (birth weight = 2450 g) presented at 0 DOL with respiratory distress, imperforate anus, and inability to pass a nasal gastric catheter past 10 cm. Radiograph demonstrated the catheter in the upper esophagus suggesting EA/TEF (Fig. 2A).

MRI was performed on DOL 1 with parameters and image processing identical to Case 1 (Fig. 2B and 2C). MRI results indicated the patient had a Type-C EA/TEF with an esophageal gap length of approximately 2 cm. Furthermore, MRI results indicated that the trachea curved posteriorly below the large proximal esophageal pouch, and a significant narrowing of this curved tracheal section (approximately 1 mm^2 cross-sectional area) was indicative of severe static tracheomalacia. Bronchoscopy at the time of repair confirmed curved tracheal anatomy and significant tracheomalacia.

Open repair on DOL 2 demonstrated an esophageal gap of approximately 2.5 cm. The TEF was ligated but primary esophageal anastomosis was not possible due to tension and the long esophageal gap. EA was repaired on DOL 47. The post-surgical clinical course was complicated by esophageal anastomotic leak, an esophageal stricture requiring multiple dilations, mechanical ventilation for the first 5 months of life, and two operations to repair tracheal defects that were noted on MRI and bronchoscopy.

2.3. Case 3

A 37 0/7 week gestation female neonate (birth weight = 3260 g) presented at 5 DOL with poor feeding and 600 g weight loss since birth. Chest radiograph on DOL 5 demonstrated coiling of the feeding tube in the upper esophagus, suggesting EA/TEF (Fig. 3A). Bronchoscopy prior to repair indicated a Type-C EA/TEF. MRI was performed on DOL 6 with parameters and image processing identical to Case 1 (Fig. 3B and 3C). MRI results also indicated the patient had a Type-C EA/TEF with a short esophageal gap length of approximately 1 cm.

Open repair of the EA/TEF was performed on DOL 9. MRI at 12 days after repair (DOL 21) (Fig. 3E and 3F) demonstrated an intact esophageal repair with a non-dilated proximal esophagus, and revealed a compression of the upper intrathoracic trachea by the innominate vein anteriorly and the proximal esophagus posteriorly, compatible with a degree of tracheomalacia. There were no detectable esophageal leaks or strictures prior to discharge, but the infant did develop an esophageal stricture at 2 months of life that required dilation. Despite the tracheomalacia observed on MRI, respiratory support was not required,

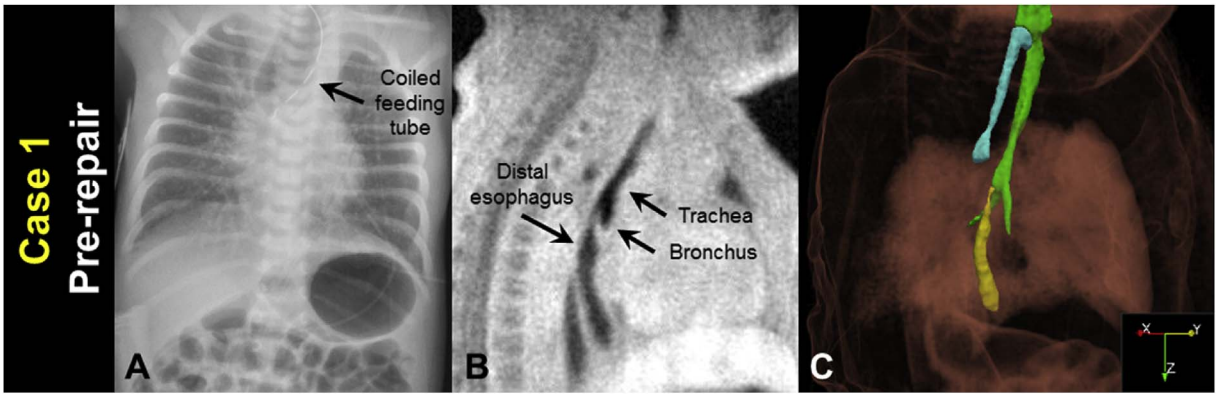

Fig. 1. Case 1 before surgical repair: A) a clinical chest radiograph, demonstrating a coiled feeding tube; B) a sagittal MRI slice (3D isotropic resolution of 0.7 mm); and C) oblique view of a MRI-based 3D surface rendering of EA/TEF anatomy (airways in green, proximal esophagus in cyan, distal esophagus in yellow), shown within a volume rendering of the body MR image. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)