Detail correction for Gross classification of esophageal atresia based on 434 cases in China

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To the Editor: Esophageal atresia (EA) is the most common congenital abnormality of the esophagus. In 70% to 90% of those born with EA, a tracheoesophageal fistula (TEF) co-occurs. Since Vogt¹ recognized and classified the types of anomalies in 1929, many other classification systems have been proposed, including the Ladd, Gross, Swenson, and Kluth classification systems. However, the Gross classification of anatomical patterns of EA/TEF proposed in 1953 is one of the most widely accepted and used classification systems[¹] and has been cited in Pediatric Surgery.² While surgical and imaging techniques have evolved considerably over recent decades, 434 consecutive cases of EA/TEF in China were reviewed. Through closely examining the imaging studies and the pathoanatomical configurations of these patients, we identified certain “defects” and details in the illustrations that need modification.

In the current research, 434 consecutive cases of EA/TEF receiving surgery were reviewed. The study protocol was approved by the Medical Ethics Committee of Beijing Children’s Hospital, Capital Medical University, National Center for Children’s Health (No. 2019-k-333) in accordance with the Declaration of Helsinki. The requirement for patient informed consent for this retrospective study was waived.

A form for data extraction was predesigned by a senior attending surgeon and an epidemiologist. Data extracted from medical records included patients’ general information, imaging results, and operative reports. All the data were extracted and managed by two residents under the supervision of the senior attending surgeons. Two experienced radiologists (one with 8 years of experience and the other with 3 years of experience in pediatric radiography) measured the diameter of the proximal esophageal pouch at the widest point on esophagograms independently (images of each type of EA/TEF were randomly selected for five cases). The intra-class correlation coefficient (ICC) was used to evaluate the consistency between the two radiologists’ measurements. Data were imported for analysis into SPSS (version 22.0, IBM Corp., Armonk, NY, USA). Continuous variables with normal distribution are presented as the mean and standard deviation, and variables with an abnormal distribution are presented as the median (Q1, Q3). Categorical variables are reported as counts and percentages. Finally, the illustration of the anatomical pattern of different types of EA/TEF was modified by close comparison with the original images from 434 patients with EA/TEF.

A total of 434 patients with EA/TEF underwent surgical intervention at Beijing Children’s Hospital from January 2007 to January 2020 (n = 264) and The Affiliated Children’s Hospital of Nanchang University from January 2013 to January 2020 (n = 170). Further demographic and clinical characteristics are depicted in Supplementary

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Diameters of proximal esophageal pouch at the widest point were measured in five cases for each type of esophagograms. The parameters measured by two radiologists showed good consistency (ICC = 0.89).

Referring to the typical esophagograms and intra-operative findings, the following modifications were made to the illustrations of the Gross classification in Pediatric Surgery [Figure 1].

**Corrigendum A:** Gross type A patients typically present with a notably distended proximal blind-ending pouch. At the distal end, the underdeveloped esophageal remnant leads off a stomach that is small (approximately 5 mL) compared to a normal neonatal gastric volume of 20 to 30 mL.[5] The gap length (the distance between the two esophageal pouches) ranges from 5 to 8 vertebrae.

**Corrigendum B:** The proximal blind-ending pouch in type B appears less dilated than that in type A. Instead of horizontally, the fistula is found to be ascendingly connected to the posterior tracheal wall, and the gap length between the blind pouches measures roughly the same as that in type A. Similar to type A, in type B, the underdeveloped esophageal remnant leads off a small stomach (approximately 5 mL).

**Corrigendum C:** Similar to type A, in type C, the upper esophageal pouch is hypertrophied and dilated. An inflated stomach and a shortened gap length (approximately two vertebrae) can be observed. In addition, the diameter of the proximal esophageal pouch (usually 1.5–2.0 cm) differs significantly from that of the distal tracheal esophageal fistula (often 0.5 cm).

**Corrigendum D:** Affected children with type D EA/TEF are identified with a narrow upper pouch with a blind end, as well as a gas-filled stomach. Similar to type B, in type D, the fistula ascends from the anterior esophageal wall to the posterior of the trachea.

**Corrigendum E:** Congenital pure TEFs (type E) are usually located at the first thoracic vertebral level (level of the superior thoracic aperture).

**Corrigendum F:** The site of stricture favors the lower third of the esophagus.

In the present study, we closely observed the esophagograms of 434 consecutive cases of congenital EA/TEF, which were reviewed in comparison with the classic illustrations of the Gross classification in Pediatric Surgery. Some inconsistencies between them were found. These “defects” have also been noticed by other scholars, and the Gross classification illustrations have been adjusted accordingly in previous reports.[2] However, various details, such as the size of the stomach in type A or B patients, the location of proximal and distal TEFs, and the size of the proximal pouch, have hardly been covered comprehensively. Thus, it is time to modify the

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**Table 1:** http://links.lww.com/CM9/A706. Diameters of proximal esophageal pouch at the widest point were measured in five cases for each type of esophagograms. The parameters measured by two radiologists showed good consistency (ICC = 0.89).
First, both the objective findings and explicable pathogenic mechanisms lend support to our proposed changes. In Gross type A cases, the hypertrophied proximal pouch arises from the effort of the fetus to swallow amniotic fluid and the blind end’s failure in drawing the fluid downward. In contrast, no fluid enters through the underdeveloped esophageal end, and therefore, the stomach is reduced sharply in volume. The stomach volume in newborns weighing 2000 to 2500 g and diagnosed with type A or B EA/TEF could be as small as 5 mL measured via contrast radiography after pyloric antral ligation compared to a normal neonatal gastric volume of 20 to 30 mL.\(^5\) In neonates with Gross type B EA/TEF, a small amount of amniotic fluid is drained to the airway through the TEF at the proximal end, which helps decompress the upper esophageal pouch and makes it appear less hypertrophied than in type A. Meanwhile, the blind distal esophageal end allows no passage of fluid or air, so the stomach in type B patients is gasless and atrophied. Similar to type A cases, in type C cases, the upper esophageal pouch is hypertrophied and dilated secondary to the buildup of amniotic fluid, while air flowing through the distal TEF leads to gastric distention. With both proximal and distal fistulas, air or fluid can move in and out freely. Hence, type D cases are characterized by both a decompressed (narrow) upper pouch and an aerated stomach.

According to our study, whether the proximal esophageal pouch is dilated may provide physicians with valuable insight into the diagnosis. In type A and C patients, the proximal pouch appears dilated with accumulated ingested fluid, as opposed to the narrow-shaped remnant in type B and D patients. In this case, subsequent investigations, including esophageal endoscopy, bronchoscopy, or three-dimensional computed tomography, are recommended to detect the fistulous tract. Therefore, the proximal esophageal end should be handled with extreme care, especially when it is not dilated (as in types B and D), that is, to ligate the TEF in a timely manner and prevent choking, cyanosis, respiratory distress, or chronic pneumonitis caused by aspiration. Pre-operative detection of the fistulous tract is critically important for pediatric surgeons so that it can be addressed adequately during repair.

In addition, the timing of esophageal anastomosis played a key role in surgical outcomes, especially in long-gap EA cases (types A and B). With the adequate application of extra-thoracic esophageal elongation, the more expansive the proximal esophageal pouch is, the less anastomotic tension there is, and the better prognosis becomes. In Figure 1, we have marked the gap between the proximal and distal pouches with vertebrae as an important reference for bedside decision-making.

Next, the proximal fistulas in types B and D branch from the lateral esophagus and reach upward to the trachea posteriorly, contradicting the horizontally joined fistula depicted in the original illustration. Secondary ligation of the proximal fistula in the absence of severe pulmonary infection promises lower risks of complications associated with anastomosis, and therefore, is suggested for patients with smaller proximal fistulas along with primary distal fistula ligation and esophageal anastomosis. Moreover, in patients with long-gap EA/TEF, the esophageal elongation technique can be safely applied since proximal fistulas connect laterally rather than perforate directly into the trachea. To prevent pneumonia, a nasogastric tube should be placed simultaneously above the level of the proximal fistula.

In addition, an accurate depiction of the gap length reveals why gastroscopy is preferred over primary anastomosis in type A and B patients. Because the distal esophageal remnant barely extends beyond the hiatus of the diaphragm, the gap between the pouches is too wide to be covered by one-stage reconstruction. Crucial to staged esophageal anastomosis, the site of the opening should be carefully decided during the gastrostomy procedure. As our research shows, the opening should be made just below the inferior edge of the rib at the midline in type A and B cases.

To summarize, we hope our efforts in rectifying the deviations from the actual findings in the definitive publication in Pediatric Surgery can serve as fruitful guidance to pediatric surgeons-in-training upon encountering EA/TEF. Apart from that, for healthcare providers in underserved areas, including China and other developing countries, a correctly illustrated, state-of-the-art reference is as meaningful as an explicit roadmap for both clinical practice and academic endeavors in their future careers.

**Conflicts of interest**

None.

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