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Flexible Retrograde Endoscopy before Reconstructive Surgery in Esophageal Atresia-A Case Report

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Abstract In esophageal atresia multiple surgical techniques were designed to repair the esophageal conduct, the decision of choosing a specific type of intervention being strictly related to the length of esophageal gap which is mandatory for operative planning. We present a case of a newborn male with type III LADD esophageal atresia with an anastomotic leak and severe mediastinitis that complicated the primary surgical procedure and imposed cervical esophagostomy and gastrostomy. At the age of one year the child was admitted again in our unit for the esophageal reconstruction. In order to choose between an end to end suture and a procedure of esophageal replacement we tried to estimate the esophageal gap using flexible retrograde endoscopy. The operative decision was finally the esophageal replacement because the esophageal gap was longer than four vertebral bodies. Depending on endoscopist's skills the fiber optic endoscopy with radio opaque landmark and X-ray might be considered as a useful procedure in measure the gap length and surgical decision.

Keywords: *esophageal atresia, fiber optic endoscopy, esophageal gap, children*

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1. Introduction

Esophageal atresia (EA) is a congenital malformation, with an incidence of 1 in 3500 live births. A multifactorial etiology has been discussed. The disease can occur in families and has an occasional association with trisomy 21, 13, and 18 which suggests genetic determinism. Embryopathologic processes, specific FGF-signaling absences like FGF1 and the III b splice variant of the FGF2R receptor and early gestation teratogenic influences such as twinning and toxin exposure has been discussed [1]. The original classification made by Vogt in 1929 was modified by Ladd and Gross in 1944 [2]. The most common type is EA (esophageal atresia) with distal tracheoesophageal fistula present in approximatively 86% of cases. VACTREL (vertebral, anorectal, cardiac, tracheoesophageal, renal and limb defects) anomaly group is associated in 10% of cases [3].

Imaging studies include prenatal ultrasonography, chest radiography, early renal ultrasonography, echocardiography, limb radiography, spinal ultrasonography and gap-o-gram. Surgical techniques are chosen according to variations in pathologic anatomy and the experience of the operatory team in different procedures.

The short-term prognosis is considerable improved with the advances made in surgery, parenteral nutrition and neonatal intensive care. Long term prognosis is determined by various respiratory and digestive complications like recurrent lower respiratory tract infection, tracheomalacia, gastroesophageal reflux disease and anastomotic strictures [4].

Our main objective is an accurate measurement of the esophageal gap length using retrograde endoscopy and, according to the results, to introduce this method in preoperative evaluation of children with esophageal atresia addressed to our service.

2. Material and Methods

Data was collected from clinical records, endoscopy charts and operative protocol of the patient. Parents signed an informed consent that allows the usage of patient's data for research and teaching purposes. The references were selected after performing a literature review conducted in Medline using the keywords "esophageal atresia", "esophageal gap" combined with "retrograde endoscopy ".

3. Case Report

A one year old male was admitted to the Pediatric Surgery Unit of "St. Mary" Children's Emergency Hospital for preoperative investigation and esophageal reconstruction. He was diagnosed at birth with type III LADD esophageal atresia. In his second day of life he was

operated by an open thoracotomy in the 4th right intercostal space, ligation of the distal trachea-esophageal fistula and esophagus anastomosis. The postoperative evolution was unfavorable with important disunion of the anastomosis and consecutive mediastinitis which imposed cervical esophagostomy and gastrostomy.

Physical examination at admission: the patient was a well-developed, poorly nourished male in no apparent distress, W=9.5 kg, vital signs: temperature 98.4 Fahrenheit degrees, pulse 121 bpm, respiratory rate 20/min, blood pressure 96/67 mmHg without any orthostatic changes; head was normocephalic and atraumatic, extraocular muscles were intact, pupils were equal, round, and reactive to light and accommodation, mouth was well hydrated and without lesions, mucous membranes were moist, normal skin color, cervical esophagostomy, left flank gastrostomy without any sign of inflammation; lungs clear to auscultation; cardiac: regular rate and rhythm without murmur; abdomen: soft, nontender, and nondistended, positive bowel sounds, no hepatosplenomegaly was noted; extremities: warm without clubbing, edema or cyanosis; neurologic: no focal deficits, cranial nerves II through XII are grossly intact. Laboratory data showed only slight hepatic cytolysis and mild iron deficiency. (Table 1)

Table 1. Patient's laboratory data

	Patient's values	References range
HBG	120 g/L	112-165 g/L
WBC	$6x10^9$/L	3.5-12.0 x 10^9/L
PLT	$300x10^9$/L	150-400 x 10^9/L
Urea	4.1 mmol/L	2.9-8.2 mmol/L
Creatinine	67 µmol/L	50-110 µmol/L
ALT	49 U/L	5-35 U/L
AST	56 U/L	7-40 U/L
Serum iron	8 µmol/L	13-31 µmol/L
CRP	<0.6 mg/dL	<0.6 mg/dl
LDH	78 U/L	60-170 U/L

In order to choose the best operative plan between a direct esophago-esophageal anastomosis and a procedure of esophageal replacement it is important to have an accurate measurement of the gap length.

We tried to estimate the esophageal gap using flexible retrograde endoscopy. Under general anesthesia a pediatric flexible video endoscope was inserted through the gastrostomy and slightly advanced into the stomach; after it reached the end of lower esophageal pouch a biopsy forceps was inserted as a radio opaque landmark through the working channel. Another landmark was placed near the cervical esophagostomy and we performed a plain X-ray with a mobile radiology unit (Figure 1, Figure 2). Then we measured the distance between the two landmarks which was more than 4 vertebral bodies so the surgical decision was esophageal replacement by colon interposition with favorable evolution.

Postoperative course was favorable; the patient was dismissed after 15 days with the recommendation of anti-reflux medication. At three month follow-up he was in good general condition with satisfactory weight gain and no signs of esophageal swallowing difficulties or persistent dysphagia. The barium meal also showed a normal aspect of neoesophagus.

Figure 1. X-ray during flexible retrograde endoscopy

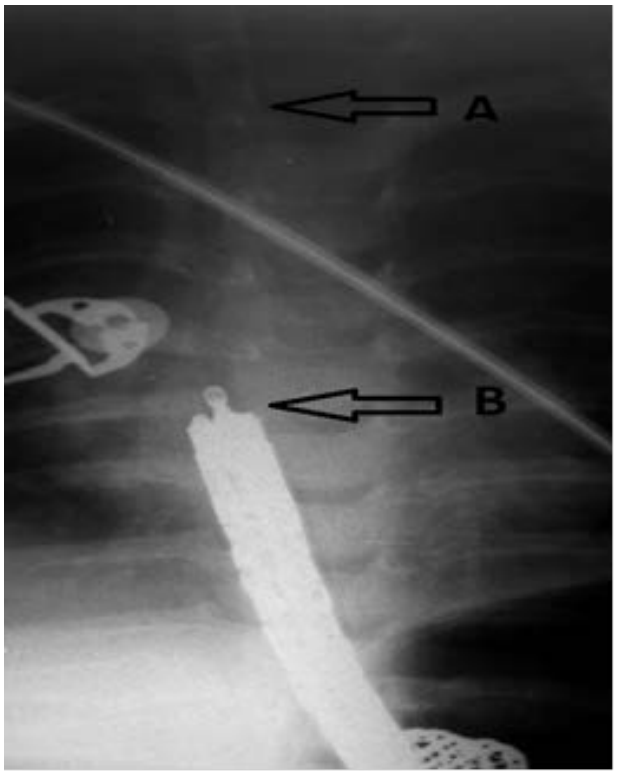

Figure 2. X-ray during fiber optic endoscopy. A-proximal landmark; B-biopsy forceps

4. Discussion

In esophageal atresia the antenatal diagnosis can be difficult; ultrasonography findings include polyhydramnios

(which has a higher positive predictive value in combination with the absence of fetal stomach bubble) and blind-ending upper pouch at the examination of the fetal neck [5]. Magnetic resonance imaging was suggested by some authors [6]. At birth, the impossibility of the nasogastric tube to pass more than 10 centimeters from the lower alveolar ridge together with chest X-ray who visualize the tube coiled into the proximal pouch confirm the diagnosis [2]. The next important step is to determine the type of the esophageal atresia in order to choose the best surgical strategy for the patient. At the initial X-ray, the presence of gas bubbles into the stomach is considered to be a sign for distal tracheoesophageal fistula, which is the most common type of EA encountered in approximatively 86 % of the cases [7]. Besides risk factors like low birth weight, respiratory distress and associated congenital heart disease, the gap length between the proximal and distal end plays a major role in surgical decision and long term prognosis [8]. It is widely accepted that the gap length can be measured in centimeters or vertebral bodies. There are no precise definitions for the gap length; some authors consider an ultra-long gap more than 3.5 cm and a long gap over 3 cm (over 2 cm according to other reports); a short gap is less than 1 cm [8]. In terms of vertebral bodies, a short gap esophageal atresia is considered to be less than 3 vertebral bodies and a long gap EA over 3 vertebral bodies [9]. Esophageal atresia with long gap distance is considered to be a surgical challenge. In order to make the best surgical decision, many techniques are used to provide an accurate measurement, because the gap length is considered an important predictor of the outcome of treatment. A number of tests are available for the measurement of the gap length and for a more accurate investigation of the presence or absence of the trachea-esophageal fistula. Contrast studies are usually used to measure the gap length in the pure esophageal atresia but are rarely used in other types of EA because of aspiration risk; barium meal may offer the best visualization but is well known that extraluminal barium can cause a granulomatous and fibrotic reaction which may lead to fibrous mediastinitis [10]. Recently, high-resolution ultrasonography was used to visualize the upper and lower pouch, and to evaluate the gap length with superior results when compared to plain X-ray. The disadvantage of this non-invasive method is that the trachea-esophageal fistula cannot be identified [11]. Some authors describe an intraoperative endoscopy as an alternative way to measure the length of upper esophagus and to exclude a pouch fistula [5]. The role of the endoscopy in the management of patients with esophageal atresia is complex. A fiber-optic endoscopy with radio-opaque landmark combined with an X-ray might be a useful test to an accurate measurement of the gap length, to identify a fistula and to diagnose other associated anomalies. The drawback of this procedure is that it requires adapted equipment and high skills. Authors report cases in which fiber-optic endoscopy was successfully used to accurate measure the gap length, with no incidents during the evaluation. Certain studies consider this procedure to have a predictive value for the feasibility and timing of delayed primary anastomosis [12,13]. Some authors consider that this is the ideal procedure to assess the esophageal pouch and tissue integrity [14]. A comparison was made between fiber optic endoscopy and other practiced techniques and retrograde esophagoscopy is considered to have better results than distal esophagogram [15]. Some studies strongly recommend systematic follow-up endoscopy and biopsy, in order to evaluate long term gastrointestinal complications, such as gastroesophageal reflux, Barret's esophagus, and esophageal strictures [16]. In our patient the flexible retrograde endoscopy was performed without any incidents and an accurate measurement of the esophageal gap permitted the optimal surgical decision. In Romania the pediatric endoscopy is not a routine procedure and is performed only in a few tertiary centers. In most cases the procedures are only diagnostic due to lack of technical possibilities and inadequate skills. Even the already published data highlights this technique, to our knowledge this is the first report in Romania of using flexible retrograde endoscopy in preoperative evaluation of esophageal reconstruction in children.

5. Conclusion

A multidisciplinary approach in the preoperative management of an esophageal atresia ensures an optimal evaluation and an accurate measurement of the esophageal gap. The fiber optic endoscopy with radio opaque landmark and X-ray might be considered as a useful procedure before surgery, despite the fact that these techniques require high skills and adapted instruments. Pediatric endoscopists should be able to perform this technique in tertiary centers.

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