Review Article

The repair of esophageal atresia and major complications—a systematic review and our experience in dealing with the tracheoesophageal fistula

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Abstract: Esophageal atresia (EA) is a rare and complicated congenital malformation. Since it was first reported, it has had a long history. Nowadays, standard treatment options consist of open procedure and thoracoscopic surgery. Each procedure has its benefits and defects, and the relative merits of both methods are still debated. We conducted a systematic literature review on the surgical repair of the EA, from the first descriptions of EA with tracheoesophageal fistula (TOF) appeared in the 19th century to 2018. The opening and the laparoscopic surgery are the two effective methods to deal with the EA, and they all have their benefits and some aspect to be improved. The main complications for the operation were anastomotic leakage, anastomotic stenosis and recurrence of TOF. Results indicate that the patients of opening surgery were more likely to have leakage, but these patients also showed a lower stenosis rate than the thoracoscopy patients. Meanwhile, the rate of recurrence for TOF had no statistical difference between the procedure types. Thoracoscopic surgery was beneficial and promoted a good prognosis in EA patients when compared to open surgery, although there is still more research to be done in this regard.

Keywords: Esophageal atresia (EA); thoracic laparoscopic procedure; open procedure

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Background

The esophageal atresia (EA) is a rare and complicated congenital malformation. It was first described by Thomas Gibson in 1696 (1), and the first descriptions of esophageal atresia with tracheoesophageal fistula (EATEF) appeared in the 19th century (2). At the end of the 19th century, the first cases began to be treated with surgery (3). It was Dr. Haight in 1943 who first ligated the fistula and created a somewhat unusual one-and-a-half-layer anastomosis (4). Moreover, the first thoracoscopic EATEF repair was performed in March, 2000 (5). Both these methods have proven to be effective in treating patients, yet some controversy still remains regarding these operation types. The thoracoscopic procedure can be considered similar or superior to open thoracotomy in that it can avoid the musculoskeletal morbidity usually associated with the open surgery. Another point of contention concerns the cosmetic aspect, scoliosis after open surgery, and the risk of CO_2 hypercarbia.

Pathological classification and prognostic factors

EA and tracheoesophageal fistula (TOF) are generally divided into five pathological types (6): (I) EA without TOF, with usually the two blind atresia ends with being distance >2 vertebrae), accounting for 7.7% of cases; (II) EA with

proximal TOF, accounting for 0.8% of cases; (III) EA with distal TOF, accounting for 86.5% of cases; (IV) EA with proximal and distal TOF, also known as EA plus double TOF, accounting for 1% of cases; (V) no EA but with TOF, also known as H-type TOF, accounting for 4.4% of cases.

In 1994, Spitz *et al.* (7) believed that with the improvement of diagnosis and treatment, weight and pneumonia were no longer important factors affecting the prognosis of EA. It was more reasonable to classify the risk according to weight and the severity of cardiovascular malformations. He classified the risks into three types: (I) type I, body weight >1,500 g, without cardiovascular malformations; (II) type II, body weight <1,500 g, or with congenital cardiovascular malformations; (III) type III, body weight <1,500 g, with severe complicated congenital cardiovascular malformations. Spitz's risk classification was beneficial for predictive use in preoperative assessment and prognosis. The survival rate of type I was above 95%, the rate of type II was about 80%, and the rate of type III was only 30–40%, with the quality of life being poor. His classification was more international.

The operation of the EA

The treatment of EA with TOF depends mainly on its pathological type and general condition. It can be operated on once or in multiple stages.

Most cases of EA with distal TOF (pathological type III) can be operated on at one time with TOF repair and esophageal anastomosis. The surgical approach method is described below.

The open procedure

The open procedure and the minimal invasive (laparoscopy) procedure are the main two methods of the operation. The open procedure always selects the right 4–5 intercostal to do an extrapleural esophageal anastomosis. First, a knife is used to make an incision in the intercostal muscles, and care is taken not to damage the pleura. The pleura is then pushed bluntly. Next, the back wall of the chest is gently pushed with the fingers, so that there is enough space for the placement of a thoracic distractor. The lung tissue is pulled forward and lowered to expose the azygos vein, which is then cut off and knotted. Then, the proximal blind side of the esophagus and the TOF are separated (both ends are monolayer anastomoses). The proximal blind end is cut off like a fish mouth at the end and enlarged via a longitudinal incision at the distal end. The 5-0 absorbable suture is then sutured and knotted. The right thoracic approach can also be used in endoscopic surgery. The left thoracic approach is used in patients with right aortic arch.

The thoracic laparoscopic procedure

The thoracic laparoscopic procedure uses the intercostal space to place the trocars. The first one for the laparoscope is located in the intercostal space under the lower shoulder angle, and the other two for the forceps are placed at the 2nd and the 5th intercostal space at the axillary midline. For placing the trocars, the procedure is mainly the same, but it requires more careful maneuver and more thoracoscopic skills and experience to accomplish the operation.

The two methods of operation are both needed to completely separate the proximal blind end from the trachea to prevent the possibility of proximal fistula. It should be remembered, that for the anastomotic technique, anastomotic tension and suture material are the key factors affecting anastomotic leakage. Most foreign scholars choose PDS and Dexon suture.

Postoperative monitoring

According to the tension of anastomotic stoma during the operation, the child patient should be kept in anesthesia for 3–7 days. Continuous positive pressure ventilation can slow down the movement of the thorax, reduce the tension of the anastomotic stoma, and prevent the occurrence of anastomotic leakage. A postoperative ventilator is routinely used. Respiratory frequency should be maintained at 40–45 times per minute, and sputum aspiration should be conducted every 30–60 minutes, with a negative pressure of <20.27 kPa. Sterile operation should be performed when respiratory tract secretion is sucked. The respiratory ventilator should be withdrawn smoothly and spontaneously. Total parenteral nutrition and antibiotics should be used via the femoral vein or subclavian vein. In children with gastrostomy, milk and breast milk can be injected through the fistula tube 72 hours after the operation to maintain a positive nitrogen balance.

Account of complications

Our retrospective study examined the period between January 2007 and 2017, and statistically analyzed the complications and the prognosis of the patients who

Table 1 Cases of complications in the kinds of EA

Groups	Type I	Type II	Type III	Type IV	Type V	Summary (%)
Anastomotic leakage	3	1	56	3	1	64 (19.22)
Anastomotic stenosis	10	4	78	1	0	93 (27.93)

Table 2 The differences between open *vs*. thoracoscopy in type III patients [case (%)]

Groups	Anastomotic leakage	Anastomotic stenosis	Tracheoesophageal fistula
Open surgery (n=87)	39 (44.82)	9 (10.34)	2 (2.30)
Thoracoscopy surgery (n=198)	17 (8.59)	69 (34.85)	17 (8.59)

P<0.05 meant the difference was statistically significant.

underwent the EA procedures. In this period, our hospital treated 333 infant cases of EA, with 194 being male and 139 being female. The main complications for the operation were anastomotic leakage, anastomotic stenosis, and recurrence of TOF are showing in the *Table 1*. The differences between the two methods in treatment with type III is shown in *Table 2*. The conclusion in our study is that the open surgery patients were found more likely to have leakage; however, the stenosis rate was lower to that found in the thoracoscopy procedure, and the rate of recurrence for TOF showed no statistical difference between the procedure types. We have more and more experience in dealing with the EA with using thoracoscopy procedure this years ,but it seems the rate of the stenosis is still in a relative steady level when contrast with the former study (8).

The treatment for the complications

All the children who had anastomotic leakage after operation did not undergo surgery again, but spontaneously healed with the placement of the thoracic drainage tube, and the provision of antibiotics and parenteral nutrition support. A total of 93 children underwent esophageal dilatation because of anastomotic stenosis. Esophageal dilatation was performed under anesthesia and examined under rigid esophagoscopy or gastroscopy. Before 2011, a rigid probe was used for expansion, and balloon dilatation catheter was used thereafter. The number of expansions ranged from 2 to 25 times per person, and the interval ranged from 1 week to 3 months. There were 2 cases of patients with esophageal leakage due to esophageal dilatation, with 1 of these cases resulting in death. Four cases of esophageal dilatation were cured by reoperation, and the remaining cases were relieved after esophageal dilatation. There were 17 cases of TOF recurrence which were first treated conservatively without success, and then were re-operated to ligate the fistula and subsequently cured.

Discussion

The incidence of anastomotic leakage after surgery reported in different studies is inconsistent, and the overall results of this study were similar. However, for type III EA, which is the most common type, the incidence of anastomotic leakage after open surgery was higher than that of the endoscopic group, with the difference being statistically significant. This may be due to the following reasons: (I) thoracoscopic surgery provides a wider field of vision, which is beneficial for separating the esophagus; (II), as a new technique, it is performed less frequently than the open surgery. Thoracoscopy still requires time to develop and to improve itself, and as the operation technology matures, the occurrence of anastomotic leakage will accordingly decrease. Open surgery does have more complications for different surgical factors. Also, as can be seen in many recent studies from different hospitals, the leakage, stricture, and ligation of the fistula method (5,9-17), of either the thoracoscopic or open surgery cannot avoid these complications. In a recently study by Borruto *et al.* (15), five series that compared open versus thoracoscopic repair were reviewed via a meta-analysis, which revealed no statistically significant differences in the complications or outcomes between the two methods.

In this study, 56 severe cases of anastomotic leakage were treated through fasting, intravenous nutrition support, and gastric tube feeding, resulting in spontaneous healing. Anastomotic stenosis is one of the more common postoperative complications. Studies have shown that anastomotic leakage (18) is a risk factor for anastomotic stenosis and recurrence of TOF. However, in this study, the incidence of anastomotic leakage was higher in the

thoracotomy group, and the incidence of anastomotic stenosis was lower, which may be because the proximal fish mouth incision and the longitudinal incision of the distal esophageal wall enlarged the anastomotic stoma. In the thoracoscopic surgery, proximal deroofing was used to enlarge the diameter of the anastomotic stoma and intermittent inversion suture was used to reduce the incidence of anastomotic stenosis, but the incidence of postoperative stenosis was still high.

Meanwhile, the advantages of thoracoscopic repair are plain to see: the scarring is clearly better in thoracoscopic repair. Other benefits have been established by other researchers. For instance, diminishing musculoskeletal deformity (10,14), that consisted of the winging of the scapula caused by consequent muscle weakness was seen (19). Scoliosis is another problem after open thoracotomy: one key study published by the Helsinki's Medical doctors (20), reported that scoliosis occurred in 11% of open thoracotomy patients, whereas the population prevalence was 2%. A recently study by Lawal *et al.* (21) compared children who underwent a thoracotomy versus a thoracoscopic approach, find and found a very high rate of scoliosis in the open group (54%) versus the thoracoscopy group (10%).

Thoracoscopy has been used in treating EA for just 18 years; it is a relatively new method, and time is needed for its benefits and defects to be fully understood. So in a long time and in the meanwhile, open and thoracoscopic surgery will co-exist to give us different options in helping patients with EA. Thoracoscopy has been considered by some to have a higher prevalence of anastomotic narrowing, yet the anastomotic narrowing required on dilation was at least 20%, and was <10% statistically by the end of the year 2012 (5), meaning that the outcomes have improved with experience and volume. Thus, there is no physiological reason not to pursue this technique.

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