TREATMENT OF ULTRA LONG-GAP ESOPHAGEAL ATRESIA. OWN EXPERIENCE BASED ON COMBINED FOKER AND KIMURA TECHNIQUE

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The aim of the study. Authors present their own experience in the treatment of ultra long-gap esophageal atresia using combined Kimura’s and Foker’s methods, while the latter one to our best knowledge has been used for the first time in Poland.

Material and methods. Multi-stage process of treatment was used in four patients. In three of them, in case of previous spit fistula presence, Foker’s and Kimura’s methods were simultaneously applied. In one of the patients without the spit fistula, only Foker’s method was used.

Results. Only patients, who completed treatment were discussed: in two of them esophageal anastomosis was achieved, however in one case delayed perforation of esophagus occurred during an attempt of intrathoracic correction of the huge hiatal hernia.

Conclusions. In the authors’ opinion Foker’s method, eventually in combination with Kimura’s technique (in cases of previous spit fistula), seems to be a promising option for the treatment of ultra long-gap esophageal atresia. It allows to save the patients’ own esophagus, and in case of the treatment failure it does not preclude the future use of other methods of esophageal substitution.

Key words: ultra long gap esophageal atresia, esophageal lengthening, Foker’s operation, Kimura’s operation, salivary fistula, spit fistula

Ultra long-gap esophageal atresia is congenital malformation of esophagus, in which the primary anastomosis of both esophageal segments is not possible. This definition is not unambiguous and qualification into the group of the ultra-long gap esophageal atresia in borderline situations depends on the patient’s size and surgeon’s experience and skills. Usually the distance of 3-3.5 cm (or alternatively more than 3 vertebrae) between upper and lower segments is considered to be the borderline (1, 2, 3). Presence of the ultra long gap esophageal atresia should be suspected in any case of esophageal atresia without tracheoesophageal fistula (2). Treatment of the ultra long-gap esophageal atresia is long-lasting and multistage. The missing esophageal part can be substituted from other parts of the alimentary tract like stomach, ileum or colon (4-10).

Another utilized method is esophageal lengthening. The original method of staged subcutaneous lengthening of previously created spit fistula of the upper part of the esophagus was presented by Kimura (11, 12). The most recent technique of esophageal lengthening was developed by Foker (2, 13).

Authors present their own initial experience with the treatment of ultra long-gap esophageal atresia using Foker’s method, possibly in combination with Kimura’s technique, in four patients, while the former, to our best knowledge, has been used in Poland for the first time ever.

MATERIAL AND METHODS

According to Foker’s technique esophageal gap is overcome by gradual, multistage esopha-
gus lengthening. Use of a temporal axial traction (internal or external) stimulates the growth of both esophageal segments and allows for their future anastomosis. Usually as the first stage of the treatment, thoracotomy is done, both esophageal segments are identified and sutured to the chest wall. This is called internal traction. Identification of the lower esophageal segment is usually more difficult. It is often small and can be found by making incision of a pleura of the posterior mediastinum above the diaphragm. Further dissection along the vagus nerve helps in the esophagus identification. To control lengthening progress using X-ray both esophageal ends are marked with metal clips. Silicone sheets are placed around esophageal segments, as well as in an open intercostal space, to minimize future adhesions. In the next stages, depending on the quality of the esophageal segments and distance between them, internal or external traction can be used again. In the case of external traction, sutures are brought out through the thoracic wall and skin and kept under constant traction for a period of several days in order to get proper esophageal length (fig. 1). During external traction the patient should be heavily sedated or alternatively intubated and ventilated. In patients with previous spit fistula Foker’s technique was combined with Kimura’s method which details have been provided elsewhere (11, 12).

Clinical material consists of four patients with long-gap esophageal atresia treated with Foker’s method. All patients were initially treated in other institutions and 3 of them had salivary fistulas created. We have presented in details two patients who completed their treatment. The third child is still in the course of therapy, while the fourth one with multiple anomalies died because of circulatory problem not associated with surgery.

CASE REPORT

1. The girl was operated on the first day of life because of esophageal atresia with fistula of the lower segment. Because primary anastomosis was impossible, after dividing of the esophageal-tracheal fistula, the spit fistula on the right side of the neck and gastrostomy were created. The child was operated again after 18 days because of duodenal atresia. The presence of annular pancreas was noticed and duoden-duodenostomy was made. On admission to our center the distance between the spit fistula and the lower esophageal segment was about 12 cm (fig. 2).

During the first operation, performed when the patient was 10 months old, the spit fistula was elongated for about 2 cm. Simultaneously, right-sided thoratocomy was done and internal traction was applied. After the first procedure the distance between both esophageal segments decreased to 6.5 cm (fig. 3). After two months the patient was re-operated and external traction was applied. After a few days because of a traction, lower esophageal segment was lengthened by another 4 cm (fig. 4). In the third stage (after next 8 days) spit fistula was removed, thoracotomy was performed and both esophageal segments were anasto-

Fig. 1. Sutures of the external traction

Fig. 2. X-ray of the stomach and the lower segment of the esophagus
mosed under tension. Postoperative period was complicated by the anastomotic leak, which healed spontaneously after about 2 weeks. Partial oral feeding was introduced and endoscopic dilatations of the esophagus were started (fig. 5).

Because of the presence of large hiatal hernia, two months after esophageal anastomosis, hiatal hernia was corrected with the right thoracotomy. The operation was complicated by esophageal perforation which was initially treated conservatively (with drainage and antibiotic therapy). Finally because of the chronic inflammation of the pleural cavity, lower esophageal segment was closed and spit fistula was created. Presently the lengthening of the upper esophageal segment using Kimura’s and Foker’s methods is planned or alternatively esophageal substitution using intestinal interposition can be applied.

2. Girl was operated on the first day of life because of the ultra long-gap esophageal atresia without tracheal fistula. During the first operation gastrostomy was created. The child was operated again on the second day of her life because of duodenal atresia. On the 9th day of life relaparotomy was performed because of disruption of the duodenal anastomosis. On the 15th day of life left-sided thoracotomy was performed because of coarctation of the aorta. On the 21st day of life the spit fistula was created on the left side of the neck. Patient has also preauricular appendages and agenesis of the left thumb. After referral to our center contrast study of the stomach done via gastrostomy did not show presence of the lower esophageal segment, however endoscopic retrograde examination showed lower esophageal part which was about 2-3 cm long. The distance measured
between Spit fistula on the left side of the neck and lower esophageal segment was about 13 cm. First stage of the operation was performed when the patient was 1.5 years old. First spit fistula was lengthened by about 2 cm, then during the right thoracotomy lower esophageal segment was found and lengthened by about 4.5 cm. The lower segment was put on internal traction. During next operation (after 1.5 months) upper segment was lengthened by about 1 cm and the external esophageal traction was applied. After 2 weeks of traction patient was re-operated, however both esophageal ends were not connected because the distance between them was felt to be too large. Again an external traction was re-applied on the lower segment. After next 2 weeks right-sided thoracotomy was performed, the upper esophageal segment was moved into the pleural cavity by creating the passage between trachea and spine (from left side of the neck to right pleural cavity) and both esophageal segments were anastomosed. Initially esophageal diverticulum developed which disappeared spontaneously after esophageal dilatations (fig. 6, 7). Six months after reconstruction of the esophagus patient is in good overall condition, gains weight and swallows half liquid or mixed food products. Her esophagus is periodically dilated. In future treatment of gastro-esophageal reflux is planned.

DISCUSSION

Treatment of the ultra long gap esophageal atresia still poses a difficult therapeutic problem; various surgical techniques can be utilized, however all of them can lead to many complications. Elongation of the esophagus with bouginage is successful only in the first months of life and with relatively short distance between both ends. Use a small or large intestine conduit can be limited because of anatomic considerations and in the long term can cause kinking of the bowel with food retention in spite of preserved peristalsis (14, 15). Retained food can become infected and later on can cause inflammation and odour from mouth. Large intestine does not get kinked so often as the small intestine and usually has a good blood supply but its mucosa is very sensitive to stomach acid (16). Stomach acid causes colon wall inflammation, scars with stenosis and bleeding ulceration, which can cause acute anemia (6). There is also no evidence of propulsive peristalsis of the isolated large intestine fragment (6). Methods involving the replacement of the esophagus using intestinal fragments require at least three surgical anastomoses of the gastrointestinal tract. Stomach’s transposition requires only one anastomosis, however full displacement of a stomach into the chest may compress the lungs and in some cases even cause tracheal narrowing which needs stenting (17, 18). Additio-

Fig. 6. Esophageal diverticulum

Fig. 7. Status after dilatation. Diverticulum is not visible
nally stomach transposition into the chest requires division of the vagus nerves, which leads to disturbances in the stomach peristalsis with food retention despite pyloroplasty or pyloromyotomy. When stomach is located in the chest, a food can easily get regurgitated into the bronchi with recurrent inflammation of the airways and in extreme cases – asphyxia (19). Foker’s method seems to be a promising alternative for the treatment of the ultra long-gap esophageal atresia. Its major advantage is utilization of the patient’s own esophagus. After the lengthening procedure only one anastomosis is required and the new esophagus has straight passage. This method is relatively uncomplicated and does not need microsurgery. Additionally in case of failure it does not preclude the use of other methods. Presence of esophageal mucosa provides a natural defense barrier against stomach acid. However clear disadvantages of Foker’s method are necessity of two or more thoracotomies, as well as the use of long-lasting anesthesia or sedation during the period of external traction. The first of these problems can be addressed by the use of thoracoscopic minimally invasive techniques which actually has already been described (20).

Two of our patients have got their esophagus reconstructed; with the distance between both esophageal ends at the onset of treatment of 12 and 13 cm. However in one of them because of further complications the final esophageal outcome was unsatisfactory. Perforation of esophagus in the first patient was probably caused by the short period between the two operations, as well as intrathoracic route of the hiatal hernia correction. In both anastomosed patients, because of the previously elsewhere created spit fistula, upper esophageal segment was lengthened using the Kimura’s method. However it needs to be pointed that the spit fistula causes a loss of length of the upper esophageal segment with difficulties to put traction on it, moreover in these cases creating a passage to the chest may be difficult, especially if fistula is located on the left side as was in one of our patients.

CONCLUSIONS

1. Based on own’s initial experience it seems that Foker’s method is a good alternative for the treatment of the ultra long-gap esophageal atresia and is an alternative for other methods of esophageal substitution in comparison with other esophageal replacement techniques.
2. The disadvantage of this method is the necessity of making two or more thoracotomies and technical difficulties and long-term problems which come from that, for instance possibility of the lung injury due to separation of adhesions and eventual chest and spine deformations.

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