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Primary Repair of Esophageal Atresia with Distal Tracheoesophageal Fistula in a Low–Birth-Weight Neonate

Düşük Doğum Ağırlıklı Yenidoğan da Özofageal Atrezi ve Distal Trakeoözofageal Fistülün Primer Onarımı

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ABSTRACT

Esophageal atresia is complex congenital anomaly of respiratory tract occuring often associated with anomalies of other systems and chromosomal abnormalities. This congenital anomaly is a surgical emergency. The management mode of Esophageal atresia/ Tracheoesophageal Fistula in the premature, low birth weight neonate remains controversial. Traditionally, treatment has been based on a staged approach. The primary repair of Esophageal atresia and fistula closure is achievable in Low Birth Weight infants and offer a good treatment in the absence of additional severe malformations. Treatment of these neonates should not be limited to tertiary pediatric surgical centers, contrarily it is also achievable in secondary heath care centers. Near cooperation with neonatal intensivist and the pediatric surgeon is essential for good survival.

Key words: Low Birth Weight Neonate, Primary Repair of Esophageal Atresia, Distal Tracheoesophageal Fistula, Esophageal stricture

ÖZET

Özofageal atrezi sıklıkla diğer sistem ve kromozal anormalileri ile ilişkili oluşan solunum yolunun kompleks konjenital bir anomalisidir. Bu konjenital anomali bir cerrahi acildir. Erken doğan, düşük doğum ağırlıklı bebeklerde özofageal atrezi/trakeoözofageal füstül tedavisi hala tartışmalıdır. Geleneksel tedavi evreye göre yaklaşıma dayanmaktadır. Özofageal atrezinin primer onarımı ve fistülün kapatılması düşük doğum ağırlıklı bebeklerde başarılabilir ve ilave ciddi bir malformasyon yoksa iyi bir tedavi sağlanabilir. Bu bebeklerin tedavileri 3.düzey pediatrik cerrrahi merkezleri ile sınırlandırılmaması gerekir. Çünkü aynı zamanda bu tedavi 2.basamak sağlık bakım merkezlerinde de başarıyla yapılabilmektedir. Yenidoğan yoğun bakım ve pediatrik cerrahinin yakın uyumu çok önemlidir.

Anahtar kelimeler: Düşük doğum ağırlıklı yenidoğan, Özofageal atrezinin primer onarımı, Distal trakeoözofageal fistül, Özofageal yapışıklık

INTRODUCTION

Esophageal atresia (EA) with or without tracheosophageal fistula (TEF) is the most common congenital anomaly of the esophagus[1].

Esophageal atresia is complex congenital anomaly of respiratory tract occuring with a frequency of 1/3500 live births and often associated with anomalies of other systems and chromosomal

abnormalities[2]. Neonates with EA/TEF are typically symptomatic immediately after birth, presenting with drooling, choking, cyanosis, respiratory distress, and feeding difficulties[3]. This congenital anomaly is a surgical emergency. The disease is usually complicated with the aspiration of gastric contents leading to pneumonia and respiratory distress. Although certain advances have been made in the treatment of esophageal atresia, there are still many problems we have to deal with. The management mode of EA/TEF in the premature, low birth weight neonate remains controversial. Traditionally, treatment has been based on a staged approach, which consist of an initial ligation of the fistula, combined with feeding gastrostomy, and later it is followed by esophageal repair, after allowing a period of weight gain. Primary surgical repair may be successful even in these high-risk patients[4]. We report a low birth weight, premature, high-risk infant who survived after primary repair of an EA and TEF.

CASE REPORT

A 31-week gestation age low-birth-weight (1300 g) baby girl was born to a 22-year-old G1 healthy mother via spontaneous vaginal delivery after premature rupture of membranes. The prenatal history was uncomplicated except two prenatal ultrasound examinations demonstrating mild polyhdramnios. Antenatal steroid treatment was not given predelivery. At birth she required brief suctioning and positive-pressure ventilation before being transferred to the neonatal intensive care unit on contiuos positive airway pressure (CPAP). The Apgar scores at 1st and 5th minutes were 6 and 8, respectively. A gastric tube could not be passed through the eosohagus and a plain abdominal radiograph without a gas in the stomach confirmed diagnosis of EA with TEF. We did not perform contrasted upper gastrointestinal series.

The baby was nursed with the chest in the upright position and oropharynx and upper pouch were suctioned repeatedly. Total parenteral nutrition was initiated on day 1 of the life and was well tolerated. The oxygen supplementation by hood and broad-spectrum antibiotics were started. We evaluated chest radiographs carefully for skeletal abnormalities, cardiovascular malformations, pneumonia, diaphragmatic hernia and a right aortic arch. Intracranial and renal ultrasound scannings were normal, an echocardiogram showed patent foramen ovale. A complete VACTERL (vertebral, anorectal, cardiac, tracheoesophageal, renal and limbs abnormalities) workup was negative, except right postaxial polydactyly The baby operated after proper evaluation and stabilization on day 3 of life through a right posterolateral thoracotomy. There was a 2 cm distance between the two segments. Through right transpleural thoracotomy, without an extrapleural approach, the fistula ligated with 6/0 prolene and the esophageal ends, lying in near apposition, were anastomosed using 6''0'' PDS in a single layer. The infant was extubated rapidly and placed on high flow nasal canulle, with a caffeine citrate infusion to treat intermittent desaturations. Total parenteral nutrition (TPN) therapy was instituted. Routine medical therapy with prokinetic agents, histamine H2 receptor blockers was started for gastroesophageal reflux. Post-opertively baby was nursed in a TEF chair, so that elevation of the head end was maintained always at an angle of 30-45°. The baby was fed on the 2 nd postoperative day via a trans-anastomotic nagogastric tube which has been inserted at the time of surgery.

He had respiratory distress on tenth day, and CPAP treatment was reconstituted. The ultrasound examination of thorax showed pleural effusion. A chest tube was inserted for treatment. The appereance of the pleural fluid was chylous, triglyceride level was 490 mg/dl and it was rich of lymphocytes so the chylothorax diagnosis was made. The enteral feeding was stopped and he was fed parenterally for several days. After seventeen days, he was fed enterally with the

medium chained fatty acid containing formula. Later this formula was replaced by premature formula and no problem was detected.

She was fed full enterally by day 32 but her progress was compicated by an episode of late neonatal sepsis and intermitant pneumonia attacks. She was evaluated for recurrent pneumonia attacks. A recurrence of TEF has been excluded by contrast studies but the barium examination showed esophageal stricture. The latter treatment was diffucult because of severe esophageal stricture (Figure 1). She required three dilatation procedures in 14 weeks. She tolerated the procedures well and was discharged at 105th day of life. She required long-term follow-up at pediatric surgery and gastroenterology clinics.

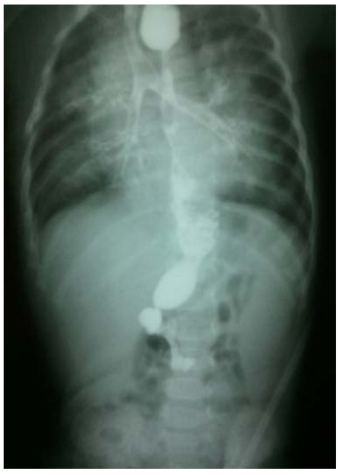

Figure 1. Esophageal stricture was seen in the barium graphy

DISCUSSION

In 1941, the first succesful repair of an infant with EA was reported by Cameron Height5. For many years, the Waterston classification served as a basis for the selection of surgical management. This classification, which was established in 1962, described three different groups of infants with EA and offered a prognostic classification for survival depending on three major factors: birth weight, pulmonary status and severity of congenital malformations[6]. Birth weight is no longer considered as a major risk factor. Therefore, the Montreal classification was established, based on two main prognostic factors, which are preoperative ventilator dependence and associated anomalies[7]. The survival rate of low birth weight infants is affected by the presence of pneumonia or other major anomalies, the Cardiac anomalies are the main cause of the death in these infants. Improvements in neonatal intensive care, including anesthesia and nutritional therapy have facilitated and increased survival in the premature, high-risk infant. In addition, prematurity, low birth weight, and congenital anomalies no longer present the same risk for operative intervention. Increasingly, it is suggested that the patient's clinical condition, rather than the birth weight alone should be the criteria for selecting the technique of surgical repair, with low birth weight alone no longer a contraindication to primary repair[8]. In a review of the literature from 1981 to 2004, only very few infants with birth weight < 1400 g and EA were reported, mostly with distal fistulas. The average survival rate varied between 35 and 41 % in infants > 1700 g in the 1950s. No infant with a birth weight <1700 g survived[8]. In the another group from England, survival rate was 25% in children weighing 1000-1500 g between 1952 and 1981[9]. Choudhury et al. found that the survival of patients with EA had improved to 83% and that low birth weight (<1500 g) did not affect survival (survival rate 67-72%)[10]. This patient was the first VLBW infant (< 1500 g) with EA who was operated in our hospital. We performed a primary anastomosis within the first 3 days of life. We think that both the operation and the supportive treatment was very succesfull despite relative impossiblities of our hospital.

Different approaches have been used for the repair of EA, such as immediate, delayed and staged repair. The management of EA with TEF in the premature, low birth weight neonate remains controversial. Traditionally, treatment has been based on a staged approach, in which initial ligation of the fistula combined with feeding gastrostomy, is followed by esophageal repair,

after allowing a period for weight gain[4]. Primary surgical repair may be successful even in these high-risk patients. Primary anastomosis in all patients who have esophageal atresia was proposed in 1972 by Abrahamson and Shandling[6]. They suggested a significant advantage, and thus improved survival, was achieved by avoiding staged repair in high-risk premature infants. The benefits of primary repair include a reduction in aspiration from the upper esophageal pouch and less esophageal, and hence feeding dysfunction[11]. However, extensive mobilization of the esophagus, and in particular the distal segment, to facilitate a primary anastomosis may be associated with an increased incidence of anastomotic leakage and stricture formation[12], and should be performed with great care. In our patient, the esophageal ends were lying in the near apposition and hence was provided for a pimary repair. Subsequent barium, examination showed stricture. She was required baloon dilatation treatment.

In conclusion, primary repair of EA and fistula closure is achievable in VLBW infants and offer a good treatment in the absence of additional severe malformations. Treatment of these neonates should not be limited to tertiary pediatric surgical centers, contrarily it is also achievable in secondary health care centers. Close cooperation with neonatal intensivist and the pediatric surgeon is essential for good survival. Furthermore postoperative complications may lead to common gastroenterological disorders that require long-term follow-up.

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