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Clinical Finding of Atresia Esophagus in Rural Hospital Kaimana, West Papua: A Case Report

Muhammad Za'im Al Rasyid[1*], Herman Wambrauw[1], Dewa Putu Satria Juristiasa[2], Agung Danan Jaya[3]

[1]Internship doctor, Kaimana Hospital, West Papua, Indonesia
[2]General Surgeon, Kaimana Hospital, West Papua, Indonesia
[3]Resident of Pediatrics, Faculty of Medicine, Udayana University, Bali, Indonesia

E-mail: zaimalrasyid99@gmail.com; hermanwambrauw4@gmail.com; dpe.satrija13@yahoo.com; agunkdj11@gmail.com

*Corresponding Author: Muhammad Za'im Al Rasyid; zaimalrasyid99@gmail.com

ABSTRACT

Background: Esophageal atresia is a relatively rare condition, this complex anomaly is still a challenging problem in pediatric surgery, with a low incidence of 1 in 2500 to 1 in 4500 births. Diagnosis and management are very challenging, especially in remote areas such as Kaimana, West Papua. Delays in diagnosis and management can worsen clinical outcomes due to complications. ***Case presentation:*** A 1-day-old male infant was consulted to the surgical department with complaints of vomiting milk every time he was given a drink, the patient was diagnosed with type A esophageal atresia with aspiration pneumonia. The patient was managed with gastrostomy with the Stamm procedure, the aim of the procedure was to provide nutrition for the patient and decompression. After surgery, the baby showed improvement, but on the 3rd day after surgery the symptoms of aspiration pneumonia continued to worsen and finally, the baby died of respiratory failure and sepsis. ***Conclusion:*** Esophageal atresia is a rare condition, and early diagnosis, especially prenatally, is very important so that the baby can be managed quickly and appropriately. Surgical intervention is important to be done as early as possible to prevent serious complications in infants.

Keywords: esophageal atresia; gastrostomy; stamm procedure; kaimana; west papua.

INTRODUCTION

Esophageal atresia is a relatively rare condition, but this complex anomaly is still a challenging problem in pediatric surgery. Esophageal atresia includes a group of congenital abnormalities consisting of a break in the continuity of the esophagus with or without a connection to the trachea [1,2]. The incidence of esophageal atresia is very low, with estimates of worldwide prevalence varying from 1 in 2500 to 1 in 4500 births [3]. In another study, it was stated that esophageal atresia has an incidence of around 2.43 cases per 10,000 births [4]. Tracheoesophageal fistula (TEF) is thought to occur during embryogenesis in the 4th week of gestational age (GA) when the separation of the primitive trachea and esophagus happens [5,6].

Classification of esophageal atresia anomalies is determined by the location of the atresia and the presence of a fistula connecting the esophagus to the trachea. In this case, five different variants have been described clinically. The first classification was published by Vogt in 1929 and modified by Gross in 1953. Therefore, these two classifications are used today. The main types of congenital esophageal atresia are esophageal atresia with distal tracheoesophageal fistula (85%, Vogt IIIb, Gross C), isolated esophageal atresia without tracheoesophageal fistula (8%, Vogt II, Gross A), tracheoesophageal fistula without atresia or tracheoesophageal fistula type H (4%, Gross E), esophageal atresia with proximal tracheoesophageal fistula (3%, Vogt III, Gross B) and esophageal atresia with proximal and distal tracheoesophageal fistula (<1%, Vogt IIIa, Gross) [7,8].

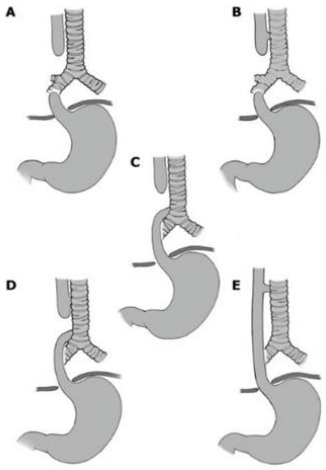

FIGURE 1: Classification of esophageal atresia [1].

The diagnosis of esophageal atresia can be made prenatally because the formation of the esophagus has occurred at 4 weeks of gestation. The main treatment for TEF is surgery which can be performed by thoracotomy or bronchoscopy, and endoscopy [5]. Preserving the native esophagus is the best option for patients with esophageal atresia. However, both delayed anastomosis and esophageal replacement are associated with high rates of postoperative complications, such as anastomotic stenosis and leakage, dysphagia, respiratory tract infections, growth retardation, and especially, gastroesophageal reflux disease (GERD) [9].

In Indonesia, especially in remote areas of Kaimana, West Papua, the geographical conditions are a challenge in providing health services. This condition can trigger delays in diagnosis and referral of patients. Delays in diagnosis and referral will make the management and prognosis of infants with esophageal atresia worse. Delays in referral will cause the infant to come in critical condition and after that, there will be a delay in surgery due to initial stabilization, and after surgery, postoperative healing will be poor. This is due to sepsis, anastomotic leakage, pneumonia, and postoperative pneumothorax.

CASE REPORT

A 1-day-old male infant was consulted to the surgical department with complaints of vomiting milk that was drunk every time he was given a drink. Complaints were accompanied by the child being short of breath and appearing weak. The child also had not defecated since birth. The baby was born normally in the hospital, at term gestational age without any history of previous antenatal care. The APGAR score at birth was 8 and 9 at 5 minutes after birth. The child also had a fever and appeared bluish. The parents admitted that the child always coughed every time he was given milk and vomited.

Physical examination showed that the baby appeared weak, short of breath, and bluish. The eyes were sunken, and the turgor returned slowly. Then, a thoracic examination was performed and severe subcostal retraction, use of accessory respiratory muscles, increased respiratory rate, and rhonchi in both lung fields were found. On abdominal examination, the baby's bowel sounds were not heard. The patient had a normal anus and no other anatomical abnormalities. After that, NGT was installed and the NGT tube could not enter, then Baby gram was performed. The results of the baby gram photo are as follows:

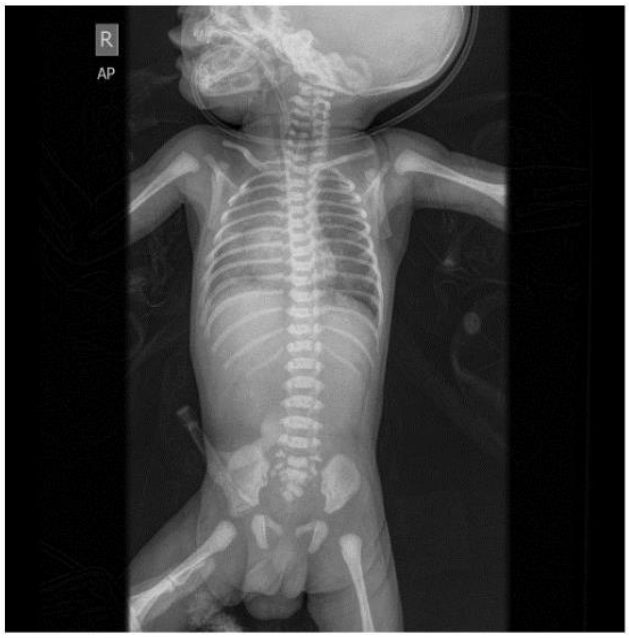

FIGURE 2: Increased bronchovascular markings are visible, no intestinal air is visible, and Coiling sign (+).

From the baby gram photo, there is a coiling image of the ngt tube which indicates total obstruction of the esophagus. In addition, in the thorax, there is an increase in bronchovascular patterns throughout the lung fields indicating aspiration pneumonia due to the absence of breast milk entering the gastrointestinal tract. Then, in the abdominal region, there is no air in the intestines and stomach. In blood tests, only leukocytosis is found, which possibly indicates inflammation of the lung tissue due to aspiration.

From the results of anamnesis, physical examination, and supporting examinations, the patient was finally diagnosed with Esophageal Atresia type A. After the patient's diagnosis was confirmed, we provided management to improve the patient's general condition and performed surgical procedures for rescue with the target of providing nutrition for the patient and decompression. The procedure was performed by making a tube from the stomach to the outside with a tube when the baby was 3 days old, a delay was made to improve the baby's general condition and wait for the anesthesiologist's approval.

The procedure was performed using the Stamm technique. This technique begins by making an incision between the umbilicus and the xyphoid process. After that, an exploration of the abdominal area was carried out to find the position of the stomach. A point in the anterior stomach was selected to allow the stomach to be brought to the anterior abdominal wall without tension.

Furthermore, the omentum was separated from the greater curvature to reduce downward tension. After that, concentric purse-string sutures were made with thick permanent sutures at the selected point in the stomach for gastrostomy, and the gastrostomy was created. Then, an 8 fr catheter tube was installed, the end of the catheter tube was inflated to fix the position of the catheter and the outside of the catheter was fixed with tagle sutures.

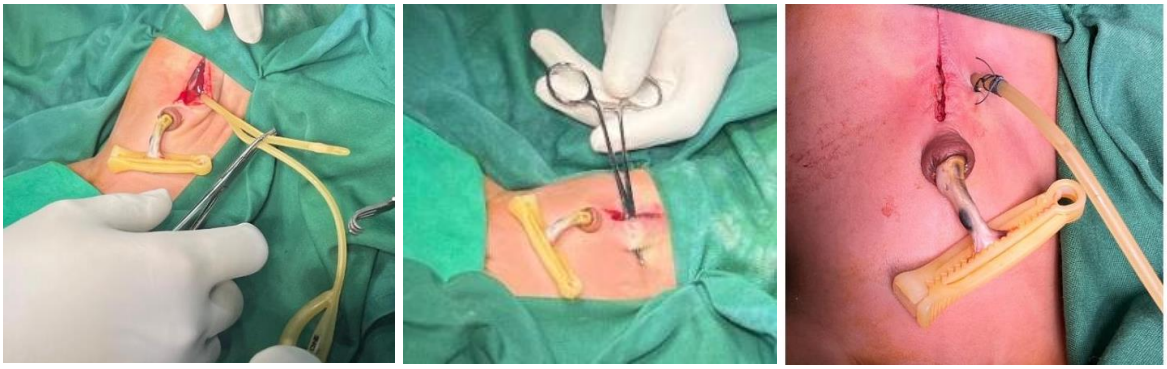

FIGURE 3: Stamm Gastrostomy feeding procedure in infants.

Non-drug therapy from a pediatrician is to fast the baby, elevate the head 30 degrees, and perform periodic suction to prevent and reduce aspiration. Drug therapy from a pediatrician is to fulfill daily fluid needs with D10% and administer antibiotics, namely erythromycin 10 mg/kgbb.

After gastrostomy is performed, a repeat baby gram is performed and intestinal air is obtained, indicating the success of the action taken.

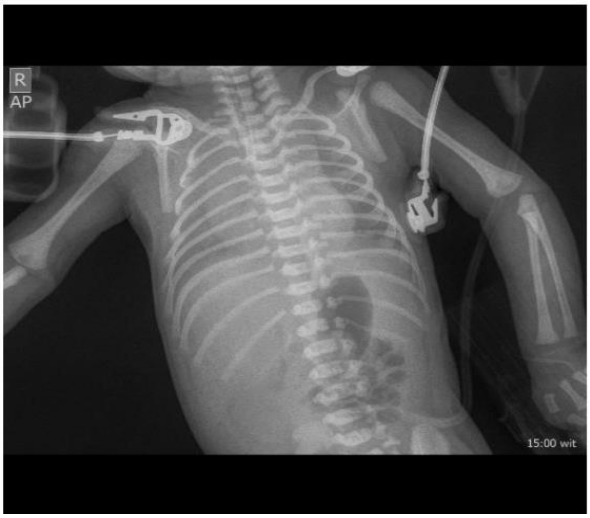

FIGURE 4: Post-operative baby gram shows air has filled the gastrointestinal cavity.

On the first day after surgery, the baby showed improvement, where bowel sounds were heard, but it was difficult to do a weaning ventilator because of the baby's poor lung condition, plus the baby had difficulty breathing spontaneously. On the second day, lung sounds worsened, and blood tests showed leukocytosis. On the 3rd day, symptoms continued to worsen and finally, the baby died of respiratory failure and sepsis.

DISCUSSION

Often patients with esophageal atresia are followed by other congenital anomalies. In a previous study, it was stated that the frequency of anomalies associated with esophageal atresia was more than 50%. Patients with isolated esophageal atresia without TEF showed anomalies in 65% of cases, while a much lower frequency was observed in patients with TEF without atresia, only 10%. Cardiac anomalies are the most common in patients with esophageal atresia. In this case, no other congenital anomalies were found in the patient [10,11].

In this case, there was actually a delay in establishing the diagnosis of esophageal atresia. Because the patient was only diagnosed after birth. Based on previous research, it was explained that the condition of esophageal atresia can be detected at a gestational age of between 16-20 weeks through ultrasound. This patient never had a pregnancy check-up, so the diagnosis of esophageal atresia could not be made. When the diagnosis was established, the patient had already experienced aspiration pneumonia which worsened the patient's condition, although efforts to improve the general condition and surgery had been carried out [7]. In another study, it was explained that antenatal diagnosis provides benefits in facilitating consultation with parents before delivery to discuss clinical management and delivery planning near or in a tertiary pediatric facility [12].

Clinical signs of esophageal atresia are often evident after birth and include excessive salivation; regurgitation, gagging, and coughing at first feed; cyanosis; and respiratory distress [13]. The diagnosis of esophageal atresia is usually made by the presence of respiratory distress due to an overflow of secretions from the proximal esophageal pouch that is blocked by the adjacent tracheal tract.

This can also occur in conjunction with the presence of a distal tracheoesophageal fistula due to the reflux of gastric acid into the bronchopulmonary tree, which is consistent with what was found in the patient [6,14].

In general, the choice of surgery depends on the patient's condition and the suitability of the esophagus; even now that advanced neonatal care and surgical techniques are available, premature infants with severe pneumonia who are unstable for thoracotomy are now undergoing thoracotomy for fistula closure. However, the mortality rate from thoracotomy in infants with poor condition is very high, therefore, the use of guidelines regarding who may benefit from gastrostomy for delayed or staged surgery, rather than performing thoracotomy for emergency fistula closure in the first step, is helpful [15].

The choice of surgical management taken in this case is appropriate. Because the patient is in an unstable condition, as well as limited equipment and difficulty in referring the patient, the target of the management that we can do is only to ensure the patient is stable. Based on previous research, it was explained that the use of primary gastrostomy, even in small fistulas during mechanical ventilation, helps gastric evacuation and further reflux, so this can be done with the aim of saving before we can refer the patient to a more complete health facility and improve the general condition of the patient who is not good [15,16].

After surgery, the patient did show improvement, especially since the main goal of saving the baby's gastrointestinal tract was achieved, but the baby's condition was indeed not good, where the infection process was still ongoing due to aspiration pneumonia, coupled with the surgical procedure that had been performed still had a risk of complications, infection. Patients can still fall into sepsis at any time which can increase the risk of mortality.

CONCLUSIONS

Esophageal atresia is a rare condition, and early diagnosis, especially prenatally, is essential so that the baby can be managed quickly and appropriately. Diagnosis of esophageal atresia requires the doctor's ability to assess the symptoms that appear in the baby because a delay in diagnosis will delay referral and delay management in the baby which will worsen the prognosis of esophageal atresia. Surgical efforts are important to be carried out as early as possible to prevent serious complications in the baby.

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