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Delayed Repair of Congenital Oesophageal Atresia with Tracheoesophageal Fistula: A Case Report

Chimaobi I. Nwagboso, FWACS, Chidiebere Peter Echieh*, FWACS, Stephen O. Ogbudu, MBBCh, Edwin O. Apreala, MBBS, Nkemakolan J. Eze, MBBS, Anietimfon U. Etiuma FWACS, Okon O. Bassey FWACS

Division of Cardiothoracic/Vascular Surgery, Department of Surgery, University of Calabar/University of Calabar teaching Hospital, Calabar, Nigeria

***Corresponding author:** Chidiebere Peter Echieh
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Abstract | **Case Report**

Congenital Oesophageal Atresia with tracheoesophageal fistula is an uncommon condition. Notable for high mortality due to its pulmonary complications of aspiration and pneumonitis, survival rates have increased markedly with improved neonatal intensive care, surgical technique of repair and nutritional support. We present a neonate who was referred to us at 21 days of age following an inability to pass a nasogastric tube, with excessive drooling of saliva, choking on feeds, fever and weight loss. The patient was diagnosed with oesophageal atresia, distal tracheoesophageal fistula and pneumonitis following a confirmatory chest radiograph. Other anomalies were ruled out and the baby had an initial gastrostomy followed by a right posterolateral thoracotomy, complete disconnection of the fistula, closure of trachea ostium and end-to-end anastomosis of the oesophagus 5 weeks later. Post-operative anastomotic leak occurred and was managed conservatively. Early diagnosis remains invaluable in the management of this condition. This is because aspiration of feeds and chemical pneumonitis is avoided. However, with adequate support and treatment, delayed management may still be successful in the management of these patients.
Keywords: Oesophageal atresia, pneumonitis, tracheoesophageal fistula.

INTRODUCTION

The earliest cases of oesophageal atresia (OA) with tracheoesophageal fistula (TOF) were first described by Durston and Gibson in 1670 and 1697 respectively [1]. Mortality from the management of patients with Oesophageal Atresia has improved remarkably. Oesophageal atresia is reported to have an average rate of 2.4 per 10000 births [2]. In this case report, we present the clinical presentation, findings and management of a patient with Oesophageal Atresia, distal tracheoesophageal fistula and pneumonitis of the right lung.

CASE REPORT

Baby AT, a female, was born via caesarean section to a 37 years old mother at a general hospital. After birth, it was noticed that the baby had laboured breathing, was drooling saliva excessively, and choking on feeds. For these, she was referred to a tertiary care sick baby unit. Patient was initially diagnosed with transient tachypnoea of the new born secondary to Congenital Pneumonia with a differential of meconium aspiration syndrome. Patient was placed on intravenous fluids, broad spectrum antibiotics and nil by mouth. However, patient deteriorated with development of continuous high grade fever, increasing dyspnoea and weight loss but no cyanosis. There was abdominal distension despite having passed meconium. With failure of Nasogastric intubation, a tracheoesophageal fistula was suspected and a chest X-ray was requested, which showed the nasogastric tube in the upper part of the oesophagus, radiological features of pneumonia of the right lung and gas in the gastrointestinal tract suggestive of an Oesophageal Atresia with distal tracheoesophageal fistula. The Cardiothoracic surgery unit was then invited. Patient's birth weight was 2.95kg. Antenatal period was uneventful. Caesarean section was on account of previous myomectomy. Abdominal ultrasound scans done twice during pregnancy showed normal findings. Patient appeared acutely ill, weighed 2.35kg, was febrile with a swelling on the left side of the neck which bulged on swallowing. There were bronchial breath sounds and basal crepitation over the right chest wall. Abdomen was full and soft. No other congenital anomalies were noted. A dilute barium oesophagogram done showed the blind ended upper pouch consistent with Oesophageal atresia (see Figure 1).

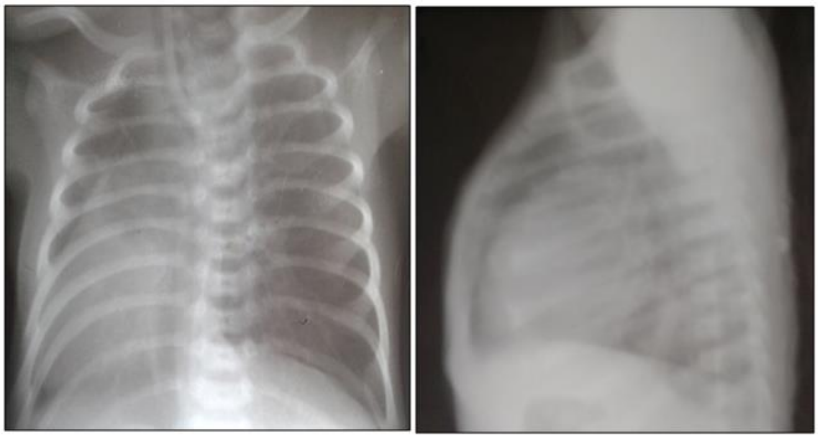

Fig-1: Chest radiographs (Anteroposterior and lateral) done while instilling dilute barium, showing curved nasogastric tube in blind ended upper pouch

Baby was nursed head up, NG tube left in upper pouch and suctioned intermittently, a gastrostomy was done for feeding, intravenous antibiotics continued based on blood culture and sensitivity and baby was kept nil by mouth. An Echocardiography and chest CT scan ruled out any other congenital abnormality. However, baby failed to gain substantial weight despite adequate feeding on account of continuous pulmonary soiling. The baby was scheduled for a Right postero-lateral Thoracotomy and closure of fistula 38 days after presentation to prevent further soiling of the lungs and enable the neonate thrive. Laboratory investigations including Full blood count and electrolytes were within normal limits. Intraoperative findings were: dilated proximal oesophageal pouch (see Figure 2), a distal tracheoesophageal fistula just above the carina and a partial collapse of the lower lobe of the right lung.

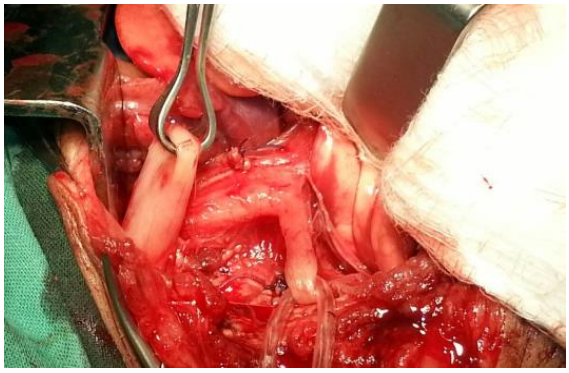

Fig-2: Proximal oesophageal pouch held with Babcock`s forceps, and distal trachea-oesophageal fistula

The fistula was divided and over sewn with pleura. The upper pouch was mobilized and the single layer anastomosis of the oesophagus was done without tension (See Figure 3).

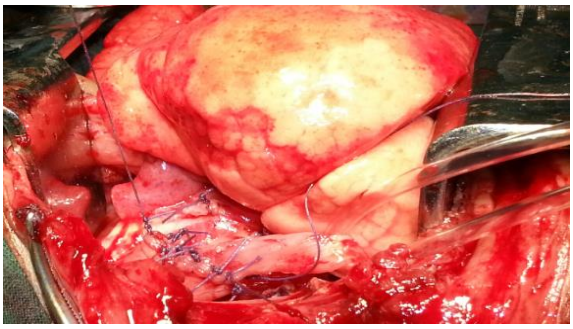

Fig-3: Single layer anastomosis of oesophagus

A chest drain placed before closure. On the 5th day post op, frothy saliva was noticed in the chest tube. An oesophagogram (Figure 4) confirmed an anastomotic leaked which was managed conservatively.

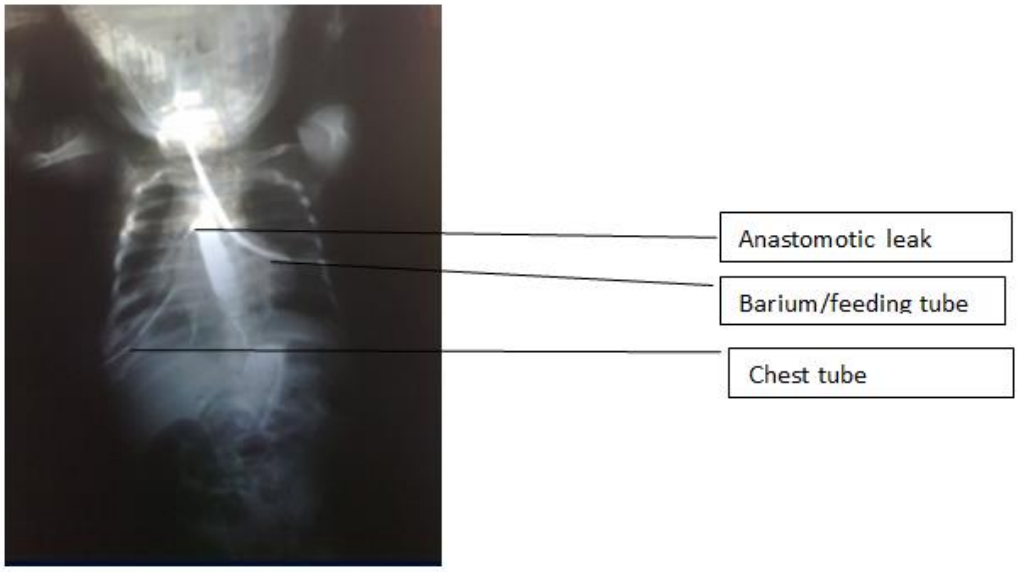

Fig-4: Post-operative chest x-ray showing anastomotic leak

Weight gain was slow but progressive. By the 34th day post op, the leak had ceased, oral feeding was initiated and patient discharged three days later for follow up as an outpatient. Patient has been seen subsequently at the surgical outpatient and has achieved adequate weight gain and developmental milestones for age.

DISCUSSION

Oesophageal atresia (OA) with Distal Tracheoesophageal (TOF) is the commonest form of OA with TOF. It accounts for about 86.5-88.4% of known anatomical variations of this condition [3, 4]. It occurs with a male-to-female ratio of 1.26, an increased risk with the first pregnancy, and a slight increase in rate with rising maternal age [5]. Associated anomalies occur in 50% to 70% of infants with oesophageal atresia [5].

The aetiology of OA is not completely understood. Classically, the embryological origin of OA with TOF results from failure of fusion of mesodermal ridges which separate the Oesophagus from the Trachea. Kluth and colleagues [6], propose that the trachea and oesophagus develop and divide from cranial, ventral, and dorsal folds of the foregut and that OA with distal TOF is due to excessive ingrowth of the dorsal fold.

There are five distinct anatomical variations of this condition (see Table One below): OA without TOF, OA with proximal TOF, OA with double TOF, OA with distal TOF, TOF without OA [4]. However, it is better to describe the anatomy of the malformation. Our patient had OA with a proximal blind pouch and a distal TOF between the oesophagus and the membranous part of the trachea just above the carina.

Table-1: Anatomical Variations of Oesophageal Atresia [4]

Type	Description	%
A	Isolated Oesophageal atresia without tracheoesophageal fistula	5.4
B	Oesophageal atresia with proximal tracheoesophageal fistula	0.8
C	Oesophageal atresia with distal tracheoesophageal fistula	88.4
D	Oesophageal atresia with proximal and distal tracheoesophageal fistula	0.8
E	Isolated tracheoesophageal fistula	4.6

Congenital OA may present in utero, in the neonatal period or later in life. It is usually diagnosed early in neonatal life. It may be diagnosed in-utero with features of small for gestational age pregnancy and polyhydramnios on obstetric ultrasound scan [5]. These features were not seen in the case of our patient or were missed as she had two obstetric scans during pregnancy.

The Neonate usually presents with excessive salivation, choking on feeds despite willingness to suck, difficulty with breathing and in some cases fever when the lungs are soiled [5]. This was the case with our patient who was not diagnosed before feeding was initiated, with the feeds soiling the lungs.

The adult usually presents with recurrent respiratory tract infections from childhood, characterised by mucopurulent sputum, cough, haemoptysis and sometimes, food particles in the sputum. Malek Hosseini *et al*. [7] have reported a congenital broncho-oesophageal fistula in a 49 year old male.

Diagnosis is usually confirmed with a chest x-ray which may show an NG tube held up in the proximal pouch, presence or not of gas in the intestine, size of the heart and vertebral anomaly. A water soluble oesophagogram will outline the proximal pouch and delineate a proximal fistula. The necessity of this has been questioned as proximal fistulae are rare and are usually searched for during surgery or bronchoscopy [5].

Associated congenital anomalies (ACA) are often multiple with a frequency of about 50% -70% [5, 8, 9]. This is because organogenesis occurs simultaneously between the third to eight weeks of intrauterine life. The anomalies occur as the VATER or VACTERL constellation [5]. Associated anomalies are sought via clinical examination, CT scan and echocardiography. No associated anomaly was found in our patient.

The presence of low birth weight, pneumonia and associated anomalies have been found to adversely affect outcome [5]. Waterston *et al*. [10] classified patients into three groups based on the presence or absence of these factors. Group A were patients with birth weight >2500g and no ACA, Group B were patients with birth weight between 1800g and 2500g with no ACA, such as our patient, while Group C patients had ACA with birth weight less than 1800g. The chances of survival decreased from A to C. The presence of pneumonia moved a patient into the next group with worse prognosis.

Waterston's classification advocates immediate repair for group A patients, delayed repair for group B patients after emergent preoperative treatment and a staged repair for patients with the worst prognosis, group C. Preoperative treatment includes suction of proximal pouch with a replogle sump tube, nutritional support and treatment of pneumonia.

However, with earlier diagnosis, improved neonatal perioperative care and nutritional support, treatment has moved to immediate primary anastomosis minimising the usefulness of Waterston's risk classification[11–13].

The aim of treatment is discontinuation of the fistula and primary anastomosis of the oesophagus. Tyson *et al*. [11] reported primary repair of 22 neonates of whom 12 were high risk, of which only three had gastrostomy. There were only 2 mortalities.

Surgical treatment is usually via open surgery but Thoracoscopic repair is gaining ground. A multi-institutional study found a slight benefit of thoracoscopic over open surgery [14]. However, most of the operations were performed by surgeons with greater than 10 years' experience with the technique. Thoracoscopic approach is also more difficult in babies less than 2kg, or with pneumonia [14]. There is also a steep learning curve associated with the procedure before excellent results are attained [14].

Complications of anastomosis may be early or late and include: leakage, aspiration, stricture, gastroesophageal reflux, tracheomalacia and recurrence [4,5]. These patients are thus followed up for life. Manning et al reported an 8.5% anastomotic leak rate, 17% for single layer repair and 6.2% for double layer repair. Our patient had an anastomotic leak which was managed conservatively.

In conclusion, we report the management of a case of OA with distal TOF and pneumonitis diagnosed 21 days after birth. Though immediate primary repair is the trend in management of these conditions, when diagnosis is late and patient unstable, delayed primary repair may be a necessary mode of management.

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