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Case Report

Congenital Tracheo-Esophageal Atresia and Fistula in A Neonate with Associated Congenital Right-Hand Anomalies

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Received Date: September 15, 2025; **Accepted Date:** September 25, 2025; **Published Date:** October 08, 2025

Citation: Shamaki Amb, Sule Mb, Umar Au, Gele Ih, Aminu A, Abdullahi A, (2025), Congenital Tracheo-Esophageal Atresia and Fistula in A Neonate with Associated Congenital Right-Hand Anomalies, *Clinical Medical Reviews and Reports*, 7(7); **DOI:10.31579/2690-8794/285**

Abstract

Congenital esophageal atresia and/or tracheoesophageal fistula is a well-known congenital anomaly that consist of esophageal discontinuity with or without connection to the trachea, occurring in about 1 in 3500 live births, and about half of the cases regarded as syndromic due to associated anomalies.

Approximately 92% of patients with esophageal atresia have a tracheoesophageal fistula, which is a congenital fistulous connection between the esophagus and the trachea or a main bronchus. This is a 15-day-old male infant that was referred for plain radiographs of the chest, abdomen and right hand, with a contrast swallow on account of episodes of choking and respiratory difficulty after every feed, loss of weight and right-hand anomalies; fused 4th web space proximally and absent 3rd digit since birth.

The plain radiograph of the chest including the abdomen was done, the chest demonstrated normal findings, the abdomen also showed pockets of lucencies in the right and left hemi-abdomen and pelvic region, no classic marked gaseous distension of the stomach was however demonstrated.

The plain radiograph of the right hand demonstrated partial fusion of the soft tissue of the 4th web space, and absent 3rd digit.

The contrast swallow performed showed a proximal blind ended esophageal pouch, no contrast was demonstrated in the trachea, distal esophagus, stomach and remaining gastrointestinal tract (GIT).

The patient had a successful surgical repair of the EA and TEF following right lateral thoracotomy with fistula ligation and esophageal anastomosis. The syndactyly will be corrected at the age of 24-months.

We present the radiologic findings of esophageal atresia with tracheoesophageal fistula with associated VACTERL anomaly (right hand incomplete syndactyly and oligodactyly) due to its peculiar presentation.

Key words: esophageal atresia; vacterl-anomaly; choking; digit

Introduction

Congenital esophageal atresia and tracheoesophageal fistula is a rare congenital anomaly that consist of esophageal discontinuity with or without connection to the trachea [1,2].

This is a rare condition that occurs in 1 in 3000-3500 live births, patients with congenital esophageal atresia and tracheoesophageal fistula have chronic respiratory and digestive symptoms due to abnormal development of trachea and esophagus during intrauterine life [3,4,5].

Approximately half the cases present with additional malformations forming either a syndrome of known genetic etiology, or a recognized association, the most recognized of which is the VACTERL (Vertebral anomalies, Anal atresia, Cardiac malformations, Tracheo-Esophageal fistula, Renal and Limb malformations) association [6,7].

Approximately 92% of patients with esophageal atresia (EA) have a tracheoesophageal fistula (TEF), which is a congenital fistulous connection between the esophagus and the trachea or a main bronchus and 4% of patients with TEF do not have EA8.

The etiology of EA with or without TEF is unclear, although 10% of patients have chromosomal abnormalities, including Trisomy 21, Trisomy 18 and deletion of 22q [117,8,9].

The syndromic EA/TEF most often arise from a single gene disorder in humans, with microdeletion occurring at 16q24.1. the most common syndromes associated with EA/TEF are the Feingold syndrome, CHARGE syndrome, AEG syndrome and VACTERL-H syndrome6.

Esophageal atresia is characterized by incomplete formation of the esophagus. Many variations of esophageal atresia with or without tracheoesophageal fistula have been described [10,11,12].

The C type is the commonest variant of this anomaly and consists of a blind esophageal pouch with a fistula between the trachea and the distal esophagus which has an estimated occurrence of about 84%8-12. The fistula however often enters the trachea close to the carina.

Most patients with congenital TEF are diagnosed immediately following birth or during infancy with more than 98% of patients having associated esophageal atresia that results in life threatening complications[13].

Patients with EA-TEF often present with respiratory difficulties, the pathology behind which is multifactorial with potential contribution from retained secretions exacerbated by tracheomalacia, recurrent aspiration, persistent diverticulum/pouch at the site of fistula and chest wall deformities [1,14,15].

Esophageal atresia is reported to be more common in the male gender than the female counterpart, this however does not affect the care or outcome of the condition [16,17].

Epidemiologically, with exclusion of chromosomal anomalies, there is no established link between occurrence of TEF and maternal age, with decreasing incidence of TEF in different regions of Europe [18,19].

The diagnosis of EA-TEF is achieved following clinical evaluation, radiologic imaging and bronchoscopy [18,20,21]. Prenatal detection of polyhydramnios with absent stomach bubble; this has a sensitivity and positive predictive value of 42% and 56% respectively for EA diagnosis. Postnatal diagnosis begins clinically, there is often arrest and failed passage with coiling of introduced orogastric catheter in patients with EA, gas distension of the stomach on plain radiography with a distal TEF, water soluble contrast swallow also demonstrates the esophageal pouch in EA and possible TEF.

Surgical repair of EA/TEF is the main treatment, this is aimed at achieving a high success rate, improving quality of patient's life with marked reduction in morbidity. The surgical treatment involves a lateral thoracotomy (more right laterally except in cases of right sided aortic arch), fistula ligation, and a creation of primary esophageal anastomosis [18,22].

Case Report

This is a 15-day old term baby weighing about 2.3kg given birth through a normal vaginal delivery with a good APGAR score. The patient was referred from a neighboring health care center for plain abdominal radiograph and a contrast swallow on account of choking with feeds; breast milk and water since birth.

The prenatal period was uneventful, the mother denied history of febrile illnesses and consumption of hard drugs, herbal concoction and food supplements. She also admitted to intake of folates with the required and prescribed antenatal drugs. No report/record of any obstetric ultrasonogram done in the antennal period was made available during this report. The patient is the first child of the mother who is about 22-years of age.

The patient had episodes of choking and cyanosis with every feed with associated respiratory difficulty. He has a scalloped abdomen on physical examination. Attempts at passing a feeding tube (NG) failed on several occasions.

The patient is conscious and alert and appeared small for his age, with microcephaly below the 3rd percentile (Head circumference: 24 centimeters), mild dehydration, not in obvious respiratory difficulty, not pale, not cyanosed, anicteric, with absent 3rd digit and partially fused 4th web space of the right-hand.

The blood pressure is normal, normal pulse rate, the muscle grade and neuronal reflexes were also normal for the patient's age.

The packed cell volume was about 37%, the white blood count was 8500 per mm^3, the erythrocyte sedimentation rate also normal and about 8mm/hr.

The plain chest radiograph showed normal heart size, with normal lung fields and chest wall, the abdominal radiograph showed pockets of lucencies in the right and left hemi-abdomen, and the region of the pelvic cavity conforming to bowel gas shadows in the gastrointestinal tract most likely from a distal tracheoesophageal fistula (figures 1 &2).

A contrast swallow (using low osmolar contrast medium: scanlux) was performed. This showed a blind ended esophagus with a pool of contrast medium (figures 3&4). The contrast was not demonstrated in the lung field. The distal esophagus and the stomach were also not contrast opacified. The vertebral spine showed normal appearances.

Plain radiograph of the right hand showed absence of the 3rd digit with partial fusion of the 4th web space proximally to the region of the distal ends of the proximal phalanges of the 4th and 5th digits. The remaining demonstrated digits and the metacarpals are within normal limit (figure 6).

Abdominopelvic ultrasound showed normal hepatic and splenic situs with other abdominal organs showing normal appearances.

An echocardiogram was also performed; this should normal findings excluding possibilities of congenital heart defects.

A diagnosis of type C -Tracheoesophageal Fistula comprising of a proximal esophageal atresia with a distal tracheoesophageal fistula with associated VACTERL anomaly: right hand syndactyly and oligodactyly. This belongs to the syndromic form of EA-TEF.

The patient had surgical repair of the EA-TEF in a health care center through a right lateral thoracotomy with fistula ligation and a primary esophageal anastomosis. Before the surgical intervention, he was placed on intravenous fluid containing dextrose, intravenous antibiotics with occasional oxygen administration.

The patient recovered well with no feature to suggest any form of post-surgical complication as at the time of this report, he was discharged three weeks after for follow-up visits and the parents were well educated on the patient's clinical condition and anticipated complications, they were also advised on the possibility of surgical repair of the incomplete syndactyly when the patient reaches 24 months of age.

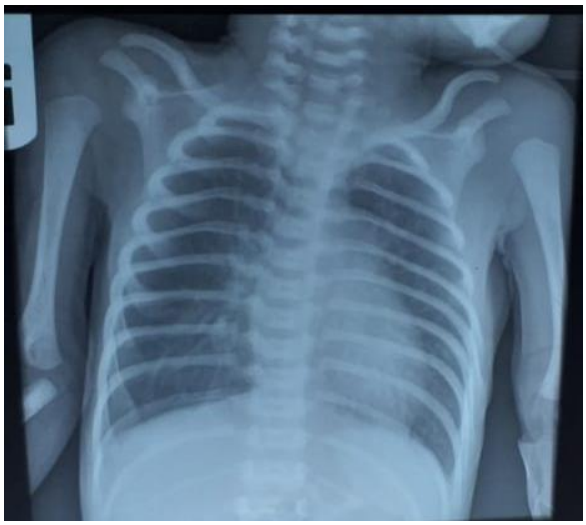

Figure 1: Plain radiograph of the chest; anterior-posterior view demonstrating normal heart size and contour, left-sided aortic arch, normal lung-fields, normal thoracic wall, vertebral spine and normal dmonstrated bones of the upper limbs bilaterally.

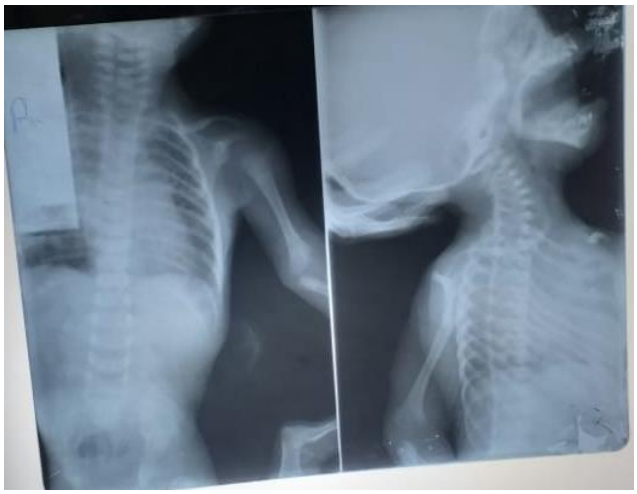

Figure 2: Plain radiographs; left image: anterior-posterior view of chest and abdomen demonstrating pockets of lucencies in the right and left hemi-abdomen and the pelvic region most likely from a distal tracheoesophageal fistula. Normal chest and demostrated vertebral spine. The right image: an attempted laterl demonstrating the cervical, thoracic and part of the lumbar spine, all appear normal. Note a lead blcker on the chest denoting the right aspect of the image.

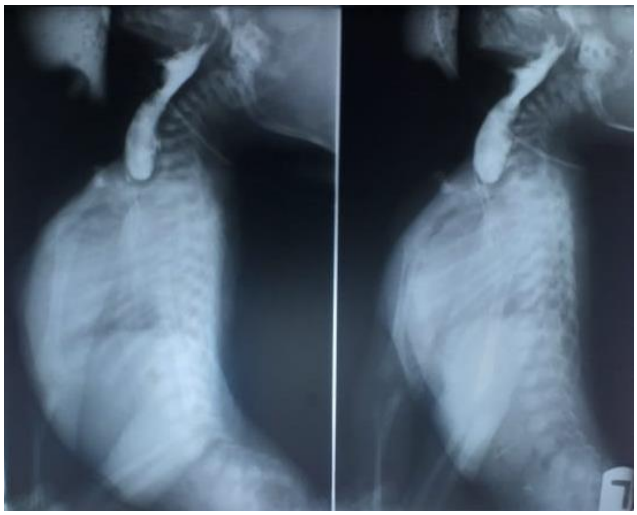

Figure 3: Lateral views of the contrast swallow demonstrating a proximal contrast filled pouch; the proximal esophageal atresia. Normal mediastinal shadow, pocket of lucency in the pelvic region. Normal bones of the thoracic cage and vertebral spine.

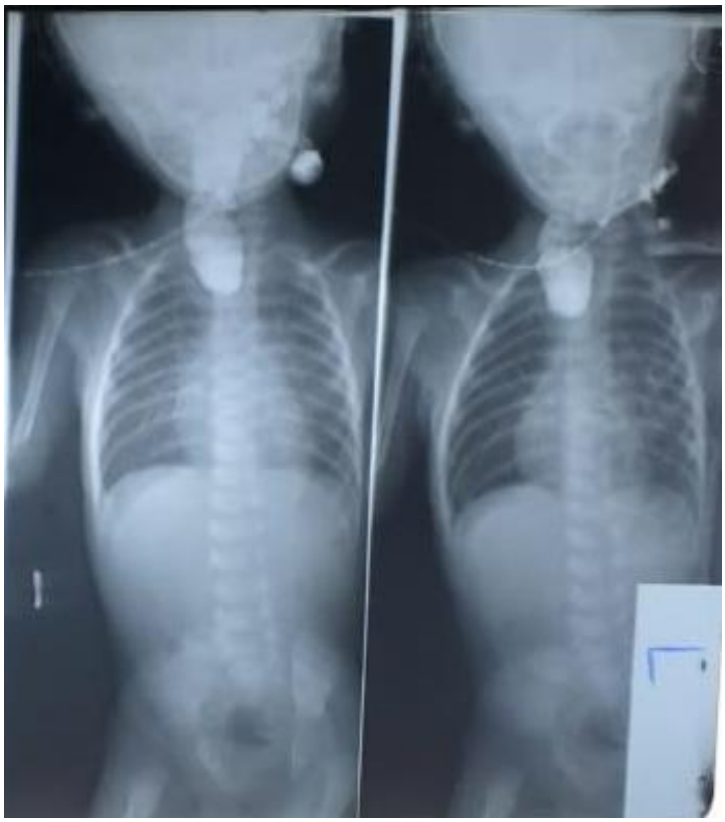

Figure 4: Anterior-posterior views of the contrast swallow demonstrating a proximal contrast filled pouch; the proximal esophageal atresia. Normal heart shadow, lung-fields, pockets of lucencies in the right and left hemi-diaphragms and pelvic region. Normal bones of the thoracic cage, vertebral spine and pelvic bones.

Figure 5: Photograph of the patient's right hand showing partial fusion of the 4th web space and absent 3rd digit. The remaining digits show normal appearances.

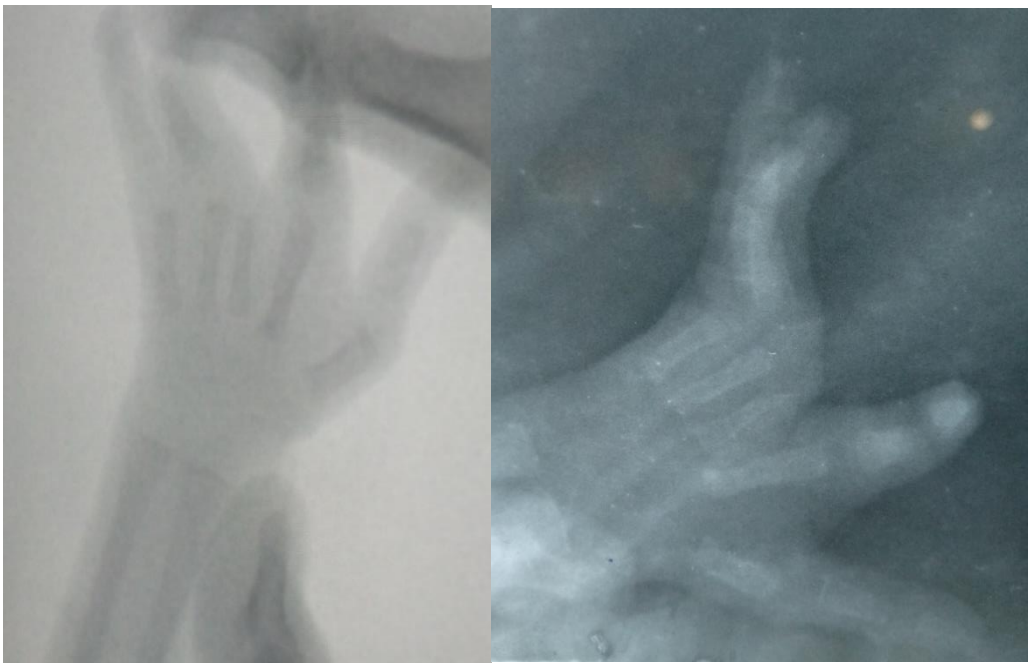

Figure 6: Fluoroscopic and plain radiographic images of the right-hand demonstrating partial fusion of the soft tissues of the 4th web space proximally with associated absence of the 3rd digit. The remaining demonstrated bones appear within normal limit. Note however the digits of the care-giver to stabilize the patient's hand for the imaging.

Discussion

The presence of a blind ending esophagus with a fistulous connection between the trachea and esophagus is a rare entity; the index case had a proximal blind ending esophageal pouch with a fistulous connection between the distal esophageal segment and the trachea classified as type-C, seen in about 84% of cases [8-12].

Patients with EA-TOF present with choking after feeds immediately after birth with varying degree of respiratory distress and difficulties, the pathology behind which is multifactorial with potential contribution from retained secretions exacerbated by tracheomalacia, recurrent aspiration, persistent diverticulum/pouch at the site of fistula and chest wall deformities[1,14,15], the index patient had similar findings of respiratory distress and difficulties following feeds and most likely from the documented pathology, thereby conforming to these literatures.

The etiology of EA-TOF is unknown [7-9], similar finding was observed in this patient, the cause of the condition is unknown, thereby conforming to these literatures.

Esophageal atresia is more common in males than females as documented in most literatures, the index patient happens to be a male conforming to that reported in the literature. Epidemiologically, with exclusion of chromosomal anomalies, there is no established link between occurrence of TEF and maternal age [18,19]. This fact has been observed in the index case, he is the first child of a 22-year-old woman, thereby conforming to these literatures.

Esophageal atresia with TOF may be associated with other congenital anomalies and syndromes, the VACTERL anomaly is an example, the index case presented with right hand anomalies: syndactyly of the 4th web space and absent 3rd digit(oligodactyly) thereby conforming to most literatures.

VACTERL anomalies are common in individuals with EA, this anomaly however does not appear to have a huge impact on overall survival rate. The skeletal defect in the VACTERL anomaly has been reported in about 7.1% and 6.1% of cases [23,24].

The diagnosis of EA-TEF is achieved following clinical evaluation, radiologic imaging and bronchoscopy18,20,21. The index case was not an exception, clinically diagnosis followed history presentation and failure of progression of an orogastric tube. Radiological diagnosis was made from a contrast swallow and plain radiographs of the abdomen, these showed a contrast filled proximal esophageal pouch with pockets of lucencies in the abdominal and pelvic cavities, thereby conforming to these literatures.

Surgical repair of EA/TEF is the main treatment, this is aimed at achieving a high success rate, improving quality of patient's life with marked reduction in morbidity. The surgical treatment involves a lateral thoracotomy (more right laterally except in cases of right sided aortic arch), fistula ligation, and a creation of primary esophageal anastomosis18-22. The index case also had a surgical repair with fistula ligation and esophageal anastomosis through a right lateral thoracotomy, thereby conforming to these literatures.

Conclusion

Episodes of choking immediately after feeds in childhood should be evaluated clinically and investigated by basic radiographic imaging comprising of plain radiography and contrast swallow to rule out esophageal atresia with tracheoesophageal fistula to save the patient's life and improve quality of life.

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DOI:10.31579/2690-8794/283