

# First successful neonatal primary repair of esophageal atresia with distal tracheoesophageal fistula in Belize

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## Abstract

Esophageal atresia repair represents the epitome of Pediatric Surgery and it is considered as an indicator of the level of quality of any Surgical Neonatal Service. This article describes the first case completely and successfully treated in Belize and it is focused on its management strategies. In the midst of COVID-19 pandemic, a full-term male neonate, adequate for gestational age and with normal birth weight was referred to Karl Heusner Memorial Hospital's Neonatal Intensive Care Unit and diagnosed with esophageal atresia with distal tracheoesophageal fistula. Primary repair was performed through open thoracotomy, closing the tracheoesophageal fistula and completing end to end esophageal anastomosis with minimal tension. A minor anastomotic leak was identified on postoperative day 9, which was initially managed conservatively but required surgical repair 5 days after, using a pedicled intercostal muscle flap over the re-sutured esophagus. The patient recovered completely with no midterm complications. It is feasible to perform complex neonatal surgical repair such as the AE/TEF in small countries like Belize, when both experienced surgical team and adequate neonatal intensive care are available. Proper management of major surgical complications is essential to achieve good results.

## Keywords

Esophageal atresia repair; neonatal surgery; Belize

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## ■ INTRODUCTION

Esophageal atresia (EA) repair represents the epitome of Pediatric Surgery (1) and it is considered as an indicator of the level of quality of any Surgical Neonatal Service. It is a life threatening condition affecting 1/2500 to 1/4000 newborns. (2) First successful primary repair was documented by Cameron Haight in 1941 at the United States. Early repair through a thoracotomy or thoracoscopy is essential to avoid complications such as gastric fluid aspiration to the lung and abdominal over distention by the air passing to the bowel. (2,3)

EA can be divided clinically into isolated EA and syndromic EA, occurring at almost the same rate. The most common

associated anomalies are cardiac, vertebral, anorectal, limb, and renal. They can present isolated or as part of a syndromic associations (VACTERL, CHARGE). The prognosis is influenced mostly by the presence of major associated anomalies. (4)

EA comes with a distal tracheoesophageal fistula (TEF) in 85% of cases (Gross C, Ladd II, Vogt IIIa). Surgical repair demands experience and skills from the team, as well as resources to ensure adequate care for these patients. Hence, only certain institutions are responsible for the complete care of newborns with this condition. (3)

For more than 15 years, a Pediatric Surgeon from the Cuban Medical Brigade has been posted at Karl Heusner Memorial Hospital (KMH), since it is the only Belizean public health institution that has a Neonatal Intensive Care Unit (NICU) and where critical neonatal surgeries are performed. In 2017, the NICU was extended and upgraded, and since 2018, three Neonatologists (one Belizean and two from the Cuban Brigade) have been working there. However,

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neonates diagnosed with EA were sent to the United States of America (USA) for surgical treatment under the World Pediatric Project (WPP). Gastrostomy tube placement was required before the transfer, and sometimes the process took some days to complete. (5)

The COVID-19 pandemic limited the possibility of transferring patients to the USA. Considering the availability of human resources such as Pediatric surgeon, Cardiothoracic surgeon, Neonatologists, Anesthesiologists, and an equipped NICU, the team decided to perform the first attempt of primary repair in August, 2020. Unfortunately, despite a proper closure of the fistula and primary esophageal anastomosis, the patient died five days after surgery because of a major congenital cardiac defect.

Finally, in October 2020, the first successful primary repair was performed at KMH, Belize. This article describes the clinical case focused on management strategies.

### ■ CASE PRESENTATION

A full-term male neonate, adequate for gestational age, with an Apgar 8/9 and birth weight of 2,630 g was referred from the Northern Region of Belize with sialorrhea and impossibility to pass the orogastric tube. A thoraco-abdominal X-ray revealed image suggestive of an esophageal atresia with distal tracheoesophageal fistula.

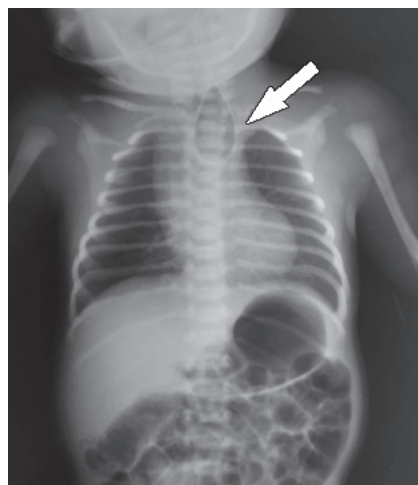


Figure 1. Chest and abdomen plain radiography showing the upper esophageal pouch with the looped tube inside (arrow). It was also relevant the distended stomach and bowel as the result of the passage of air through the tracheoesophageal fistula.

(Figure 1)

The newborn was stable and had no respiratory distress on arrival at KMH's NICU. There was no clinical evidence of associated malformations. Laboratory tests results were within normal limits as well as the preoperative arterial blood gas (ABG). Abdominal ultrasound and echocardiography did not report any renal or cardiac defect. The father was informed about the diagnosis, proposed surgical procedure, risks and potential complications. Consent for surgery was obtained.

### DESCRIPTION OF THE SURGICAL PROCEDURE

Right posterolateral thoracotomy was performed with the patient in left lateral position. Transpleural access to the mediastinum was achieved followed by division of the azygos vein. The tracheoesophageal fistula was identified. Division and closure of the tracheal side of the fistula with interrupted sutures was performed. (Figure 2)

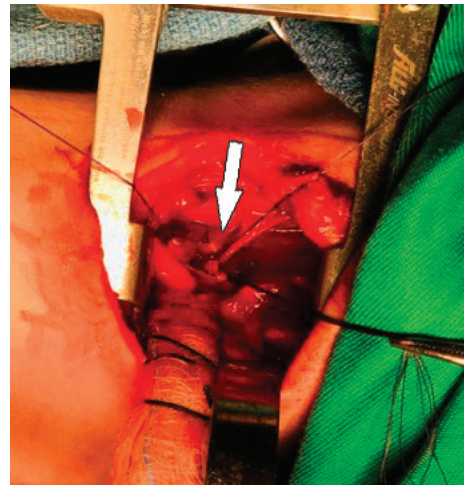


Figure 2. Intraoperative picture showing the tracheoesophageal fistula divided and sutured (arrow).

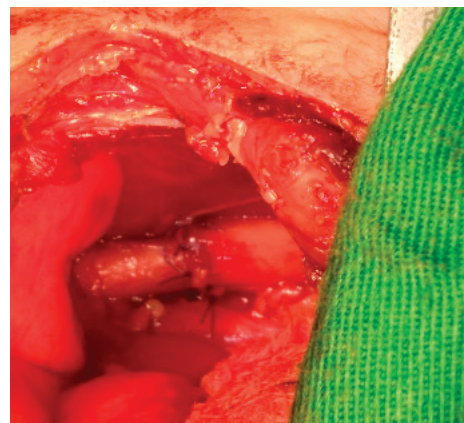


Figure 3. Completed end to end esophageal anastomosis.

After mobilization of the proximal esophageal pouch, one layer end to end esophageal anastomosis was completed leaving an 8Fr transanastomotic tube inside the esophageal lumen.

### POSTOPERATIVE EVOLUTION

The neonate was on mechanical ventilation for 3 days and remained hemodynamically stable with normal laboratories control results in the first week. There was no evidence of saliva passing through the chest tube and no abnormal images on chest X-ray were seen.

Dynamic esophagogram under fluoroscopy using

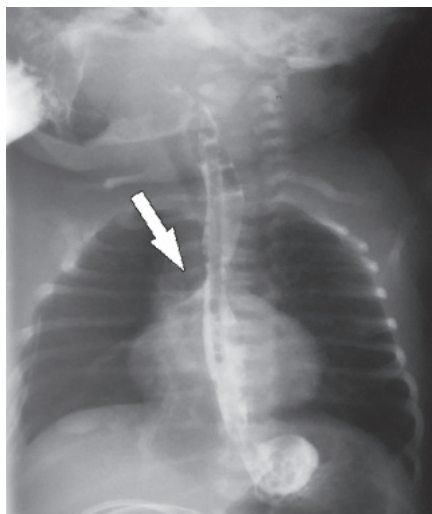


Figure 4. Postoperative esophagogram revealed limited leakage of contrast to the mediastinum (arrow).

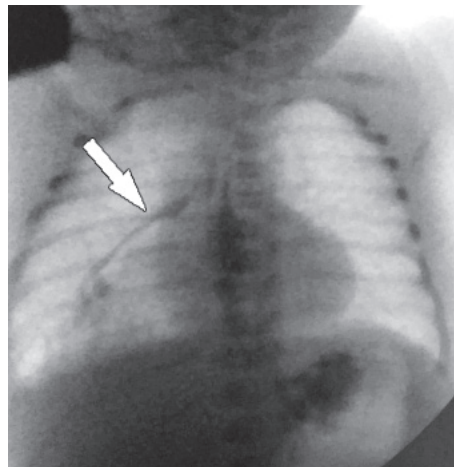


Figure 6. Follow up esophagogram showing persistent anastomotic leak (arrow).

hydrosoluble contrast (postoperative day 9) revealed a limited anastomotic leak (Figure 4). Considering the clinical stability of the patient and the limited contrast spill seen on the radiological study, a conservative management was started. The patient continued with nothing per oral (NPO), with total parenteral nutrition (TPN), and with broad spectrum antibiotics. A central venous line was placed on the right subclavian vein.

Two days after (postoperative day 11), some saliva started to pass from the chest tube. Progressive clinical deterioration started 5 days later; leukocytosis and



Figure 5. Chest X-ray showing opacity of the right lung with a suggestive image of pleural effusion.

thrombocytopenia were observed. Plain chest x ray showed peripheral opacity on the right lung field suggestive of pleuritis vs effusion. (Figure 5) Another esophagogram was performed on postoperative day 15, showing a persistent anastomotic leak. (Figure 6)

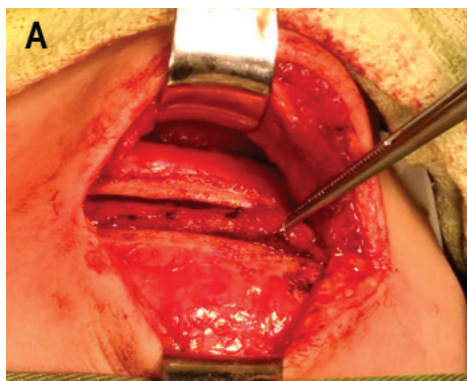


Figure 7. Intercostal muscle dissected (A). Pedicled muscle plasty around the esophagus (B).

## REINTERVENTION

After a discussion involving the team and the parents, the decision of taking the patient for a reintervention was made.

Extensive fibrinopurulent pleuritis and minimal disruption on the posterior aspect of the esophageal anastomosis were found. Decortication and lavage of the pleural cavity with normal saline solution was performed. After re-suturing the esophagus, a plasty with pedicled intercostal muscle was made. (Figure 7)

## OUTCOME

The patient was stable in the immediate postoperative period and was extubated after 2 days. There were clinical and radiological improvement, no passage of saliva from the chest tube. Control esophagogram on day 9 after reoperation showed no esophageal leak or stricture. The chest tube was removed after starting oral feedings on day 10 and the child was discharged from the hospital at 34 days of age.

Follow up at the outpatient clinic was made at months 1, 3, 6, 9 and 12. Neurological and nutritional development were evaluated, showing normal parameters after 3 months of



age. No evidence of dysphagia or respiratory problems were seen.

## ■ DISCUSSION

Despite current technological development and surgical experience in the treatment of patients with AE/TEF, the management of these cases continues to be a challenge. Survival rate of esophageal atresia has increased over the last few decades. Even in developed countries, survival rates are 85% to 95% in cases where esophageal atresia is not associated with severe congenital anomalies. (3,4,6)

The improvement in survival rate is attributed to early diagnosis, better understanding of the disease and neonatal physiology, a properly equipped NICU, an adequate parenteral nutrition, antibiotics, advances in neonatal anesthesia and improved surgical techniques. (5,6) However, it continues to be challenging in resource limited settings, as in many underdeveloped countries.

Nisar et al (6) presented a series of 140 patients treated in Pakistan. A significant high overall mortality of 57.9% was reported. The authors stated that several factors contribute to this, including lack of NICU, non-availability of dedicated and specialized paramedical staff round the clock, low doctor to patient ratio, non-availability of neonatal echocardiography/pediatric cardiologist for screening and management of associated cardiac anomalies and no ventilator support for most of the neonates. The current situation in Belize, and specifically at KMH, is quite better.

The experience of international collaboration for complex surgical problems in small low to middle income countries like Belize, has been implemented for years. It is of great help when local resources are limited, including professional experience. These initiatives have helped to save many children lives. In the case of EA in Belize, the process involves the transfer of the neonate to an institution in the USA, but with prior gastrostomy tube placement for feeding and gastric decompression, and any other measures to minimize airway aspiration while the infant awaits transferal. (5,8)

Major postoperative complications in patients with EA/TEF include anastomotic leak, recurrence of TEF and esophageal stenosis. Especially the first two are life threatening and many times require surgical interventions. An overall leak rate of 3.5–17% has been reported by several authors. Major leaks that require active intervention occur much less frequently. (4,9,10)

Conservative management of minor postoperative leak is widely accepted. Although some authors (11) state that routine esophagram is not necessary after repair of esophageal atresia, it can identify limited leaks in asymptomatic patients. Management strategies include total parenteral nutrition, semi Fowler position of the baby

(15° to 30°), wide spectrum antibiotics and proper drainage of the thoracic cavity with a chest tube (pleural or extrapleural). Sepsis control and clinical stability are essential for good results. Extrapleural repair is mentioned as a good predictor for spontaneous closure of limited anastomotic leaks. (4,10,12,13) In the case presented, an attempt for non-surgical management was initially started but the deterioration of the clinical condition of the patient along with no improvement in radiological images led us to the decision of reintervention.

There are different surgical strategies to treat the anastomotic leak, depending on the clinical situation of the neonate, contamination of the mediastinal/pleural space, and the presence of necrosis around the leaking area, among others. If it is possible, the repair of the disrupted anastomosis should be attempted, to preserve the native esophagus. Re-suture of the esophageal edges can be performed alone or with a pleural patch, using mediastinal fat or intercostal muscle. (4,14) Pedicled intercostal muscle flap was created to cover the sutured disruption of the anastomosis in our case.

Liyang Y et al (15) reported the use of pericardium to repair the anastomotic leak in one case after esophageal atresia surgery. The use of glycopyrrolate has also been reported to repair the anastomotic leak. (16) In a randomized control study conducted by Vaghela MM et al, (17) they concluded that administration of glycopyrrolate in patients of anastomotic leak after primary repair of esophageal atresia resulted in reduced oral secretions, which helped in healing of the anastomotic dehiscence in a significant number of patients.

In the case presented, the leak was controlled after the second surgical procedure. Intensive care management was essential to control the infection, manage fluid balance and provide respiratory support. Ammar S et al (17) reported a series of 58 patients treated in Tunisia, finding prematurity, low birth weight, cardiac malformations and delayed diagnosis as variables associated with higher morbidity. In their series, prolonged tracheal intubation (>5 days) was also associated to short term complications. Especially in those children, midterm follow up is important after hospital discharge.

## ■ CONCLUSIONS

The first successful repair of EA/TEF in Belize is documented. When an experienced surgical team, neonatologists and an equipped neonatal intensive care unit are available, it is feasible to perform complex surgeries such as the AE/TEF repair in small countries like Belize. Proper management of major surgical complications is essential to achieve good results. Other variants of EA requiring stage surgeries or even more complex procedures should be considered for referral to specialized institutions.

## Primera reparación primaria exitosa de un neonato con atresia esofágica y fístula traqueoesofágica en Belice.

### Resumen

La reparación de la atresia esofágica representa el epítome de la Cirugía Pediátrica y es considerada como un indicador de calidad de cualquier Servicio Quirúrgico Neonatal. Este artículo describe el primer caso completamente tratado en Belice, enfocado en las estrategias de tratamiento. Recién nacido masculino a término, con peso normal al nacer, diagnosticado con atresia esofágica con fístula traqueoesofágica distal, fue remitido al Karl Heusner Memorial Hospital, coincidiendo con un pico de COVID-19 en Belice. La reparación primaria se realizó mediante toracotomía abierta, ligadura de la fístula traqueoesofágica y anastomosis esofágica término-terminal con mínima tensión. Se identificó una fuga anastomótica menor el día 9 después de la cirugía, que inicialmente se trató de forma conservadora, pero requirió reparación quirúrgica 5 días después, utilizando un colgajo de músculo intercostal pediculado sobre el esófago re-suturado. El paciente se recuperó completamente sin complicaciones a mediano plazo. Es factible realizar cirugías complejas como la reparación de AE/TEF en países pequeños como Belice, si se dispone de un equipo quirúrgico experimentado y cuidados intensivos neonatales. El manejo adecuado de las complicaciones quirúrgicas mayores es fundamental para lograr buenos resultados.

### Palabras clave

Reparación de atresia esofágica; cirugía neonatal; Belice

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