



Case report

Esophagus atresia without tracheoesophageal fistula: a case report

Atresia de esôfago sem fístula traqueoesofágica: um relato de caso

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Abstract

Objective: report the case of a newborn patient who presents esophageal atresia without tracheoesophageal fistula, highlighting reserved conduct and prognosis. **Materials and Methods:** this is a descriptive study of the type report of a case of a newborn patient, male, born in the city of Montes Claros/MG, carrier of congenital malformation atresia of esophagus without tracheoesophageal fistula. The project was submitted to Plataforma Brasil and approved by the Research Ethics Committee, under opinion n. 5.455.159. **Case report:** an infant patient, male, with negative family history, was born with esophageal atresia without tracheoesophageal fistula and received his diagnosis after birth. The anatomical alteration was suspected after the newborn presented abundant upper airway secretion and impossibility of nasogastric tube passage still in the delivery room. Soon the diagnosis was confirmed by contrast chest radiography. The patient underwent nondefinitive surgical procedures such as gastrostomy and Esofagostomia. **Conclusion:** atresia of the esophagus without tracheoesophageal fistula represents a rare involvement which, without the possibility of early attachment of the esophageal and stomach stumps, represents a condition with a reserved prognosis.

Keywords: Esophageal atresia. Tracheoesophageal fistula. Congenital malformation. Diagnosis. Surgical treatment.

Resumo

Objetivo: relatar o caso de um paciente recém-nascido que apresenta atresia de esôfago sem fístula traqueoesofágica, destacando conduta e prognóstico reservado. **Materiais e Métodos:** trata-se de um estudo descritivo do tipo relato de um caso de um paciente recém-nascido, do gênero masculino, nascido na cidade de Montes Claros/MG, portador da malformação congênita atresia de esôfago sem fístula traqueoesofágica. Houve submissão do projeto à Plataforma Brasil e aprovação pelo Comitê de Ética em Pesquisa, sob parecer n. 5.455.159. **Relato de caso:** paciente lactente, do sexo masculino, com história familiar negativa, nasceu com atresia de esôfago sem fístula traqueoesofágica e recebeu seu diagnóstico após o nascimento. Suspeitou-se da alteração anatômica após o recém-nascido apresentar abundante secreção de via aérea superior e impossibilidade de passagem de sonda nasogástrica ainda na sala de parto. Logo o diagnóstico foi confirmado por meio de radiografia de tórax contrastada. O paciente passou por procedimentos cirúrgicos não definitivos, como gastrostomia e esofagostomia. **Conclusão:** a atresia de esôfago sem fístula traqueoesofágica representa acometimento raro o qual, sem a possibilidade de ligação precoce dos cotos do esôfago e estômago, representa condição com prognóstico reservado.

Palavras-chave: Atresia de esôfago. Fístula traqueoesofágica. Malformação congênita. Diagnóstico. Tratamento cirúrgico.

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Introduction

The esophagus is the initial part of the digestive tract, which transports the food bolus from the pharynx to the stomach¹. Its development begins with the formation of the laryngotracheal tube, which occurs in the fourth week of pregnancy. Subsequently, it is divided into esophagus and trachea. When this structure does not divide properly, tracheoesophageal fistula (TEF) and/or esophageal atresia (EA) form².

Esophageal atresia encompasses a group of congenital malformations related to the interruption of the continuity of the esophageal wall, with or without connection to the trachea³. The global prevalence of esophageal atresia corresponds to 2.99 per 10,000 births. 60% of newborns with esophageal atresia have other congenital abnormalities, predominantly, but not limited to vertebral anomalies⁴. As in other malformations, the etiology of esophageal atresia is believed to be multifactorial. However, risk factors have been pointed out in some studies as possible causes of esophageal atresia, such as gestational diabetes mellitus, prenatal infections, drug exposure and vitamin A deficiency⁵.

Fetal diagnosis is possible and is related to better postpartum prognosis. Second-semester prenatal ultrasonography can detect EA and TEF⁶. The classic signs of EA and TEF in ultrasonography are the absence of the gastric bubble and polyhydramnios⁷. Fetuses with esophageal atresia are not able to absorb the amniotic fluid and take it to the intestine, and fluid accumulation occurs⁵.

Clinical signs at birth that lead to the suspicion of EA or TEF are aerated sialorrhea, worsening with oral feeding, impossibility of swallowing, associated with cough and choking, cyanosis and dyspnea^{5,8}. The classic clinical sign of EA is the inaccessibility of nasogastric tube insertion during postnatal care. A chest x-ray shows the sinus tube wavy in the proximal esophagus⁹.

The treatment of EA is surgical. The prognosis of patients treated for EA varies according to the number of comorbidities related to malformation³. Therefore, the objective of this study is to report the case of a male patient with the described malformation, its possible treatments and prognoses.

Case report

Newborn (NB), male, gestational age 35 weeks + 4 days, was born cesarean, due to polyhydramnios with prelabor rupture of membranes in the act, with the presence of yellowish amniotic fluid, APGAR 1'6/5'9.

Mother in second pregnancy, evidenced only change in obstetric ultrasound at 32 weeks without identification of the fetus's stomach and polyhydramnios. Previous ultrasonography described the stomach as partially full. Performed all necessary consultations, adequate vaccination card, vaccine against covid-19 with 12 weeks and second dose with 24 weeks of pregnancy, negative serologies and absence of comorbidities and risk factors.

At birth, NB weighing 2.145kg, respiratory depression, with good response to positive pressure ventilation (PPV), presence of upper airway hypersecretion, performed aspiration of secretion and being requested gastric aspiration, but there was no progression of the passage of the gastric tube; stained, dehydrated light, immediate perfusion, full and symmetrical peripheral pulses, normophonetic heart sounds in 2 times, without breaths, acyanotic, rude vesicular murmur, abdomen excavated, with reduced hydroaerial noises, without visceromegaly.

Contrast chest radiography was performed with evidence of esophageal atresia without tracheoesophageal fistula (Figure 1). The patient was referred to the surgical block in his 3rd day of life for gastrostomy. Admitted to the Neonatal Intensive Care Unit for postoperative follow-up of gastrostomy in good general condition.

Patient performed three neonatal heel prick, in which two were positive for cystic fibrosis, later was submitted to the Sweat Test that presented negative result. Echocardiographic examination performed with 3 days of life was within normal range.

On his 5th day of life, he presented hyponatremia and hyperkalemia, being corrected satisfactorily, later evolved with late sepsis with pulmonary focus, being well treated. On the 6th day of life, total abdomen ultrasonography presented extrarenal pelvis on the left and minimal amount of free fluid in the abdominal cavity.

At thirty-nine days of life was submitted to the measurement of esophageal stump with distance of 3 vertebral bodies between the stumps. The patient was kept hospitalized and new evaluation and surgical approach were reprogramed. He was sent to a specialized service in another location, being submitted to a new evaluation and measurement of the distance between the vertebral stumps, whose result pointed out 5 vertebral bodies of distance.

At 3 months of age, it was necessary to perform Esophagectomy for the drainage of salivary secretion and new gastric approach due to pylorus hypertrophy, which was obstructing milk absorption. He began to feed via parenteral for approximately fifteen days, later this period returned to feeding via gastric tube gradually. Patient remains in ICU-Neonatal. To date, there is no schedule

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for surgery to reconstruct the esophagus and the medical team awaits clinical improvement of the patient.

Figure 1 - Contrast chest X-ray demonstrating the esophagus on a blind background.



Ethical care

The project was submitted to *Plataforma Brasil* and approved by the Research Ethics Committee, under opinion N. 5.455.159.

Discussion

One case of esophageal atresia without tracheoesophageal fistula was reported in a male patient, not diagnosed in prenatal care. Fetal diagnosis may be on second-semester sonographies⁶, but in the case presented, only one ultrasound performed at 32 and 34 weeks of gestation showed that it was not possible to identify the fetus' stomach and the presence of polyhydramnios, thus not

being possible to perform the intrauterine diagnosis. Even with the evolution of ultrasound devices, EA is still a forgotten and little investigated diagnosis in the prenatal¹⁰.

The postnatal diagnosis can be made in the delivery room, when the failure of passage of the probe to perform the aspiration of secretions, or after the first hours of life of the newborn, with the observation of the clinical picture, and the later the diagnosis, the likelihood of developing pneumonia. The diagnosis is confirmed by chest and neck radiography. In chest radiography, it should also be observed if there are findings indicative of pneumonia, atelectasis and other congenital malformations, the most common being cardiac or skeletal malformation¹¹. In the case presented, both the difficulty of gastric probing and the typical radiological changes were observed.

In 93% of patients with esophageal atresia, tracheoesophageal fistula is also present, however, in about 7% of cases, pure esophageal atresia occurs, with no tracheal effection², as the reported case.

This case fits the epidemiological profile of EA, in which the most of those affected are male, according to a study by Matsuura¹², whose diagnoses are performed after birth in most cases due to suspicion, mainly due to the presence of abundant upper area secretion and sialorrhea at birth.

Associated abnormalities are apparently more frequent in patients with pure esophageal atresia². In a study by Alberti *et. al*¹³, it was observed that the most associated malformation with EA is cardiovascular and associated VACTERL syndrome (vertebral abnormalities, anorectal atresia, heart defects, tracheoesophageal fistula/esophageal atresia, renal abnormalities and limb defects), but no other malformations have been identified in the patient until this time of the study. The most frequent complications are infections, especially systemic ones, to which the study patient was affected several times during his hospitalization.

In a study conducted by Sfeir *et al.*¹⁴, it was noticed, in addition to a greater association with VACTERL and cardiac malformations, an important correlation with polyhydramnios, congenital esophageal stenosis and CHARGE (coloboma, heart defects, choanal atresia, growth retardation, genital/urinary defect, ear abnormalities and tracheoesophageal fistula). Moreover, low birth weight, associated cardiac abnormalities, trisomy of 18, inborn birth, prenatal diagnosis and difficult anastomoses were associated with increased in-hospital mortality at 3 months.

Achieved optimizations of general condition and pulmonary clinical conditions, the candidate neonate to surgical repair is enabled, the definitive treatment modality. Ideally this can be

performed early, from 24-72 hours after birth, by performing the echocardiogram. Postponement can cause increased risk of respiratory complications¹⁵.

Esophageal repair is performed in a single intervention or in two stages, according to the extent of atresia and clinical conditions of the patient¹⁵. In cases of esophageal atresia without tracheoesophageal fistula, which is the case reported, gastrostomy is initially performed, a procedure performed on the second day of life of the newborn. The next surgical procedure in most cases is the anastomosis of the esophageal stumps, but this approach can be postponed due to the increased distance between the esophageal stumps or poor general state¹⁶.

The pathophysiology of EA is based on gastrointestinal motility disorder. Inadequate peristalsis seems to be associated primarily with abnormal esophageal innervation, but after surgical treatment, vagus nerve injury may occur, which seems to be associated with peristalsis dysfunction, hiatal hernia, and esophageal shortening in the anastomosis^{17,18}.

Another surgical procedure that can be performed as a palliative form is Cervical Esophagectomy, indicated when found that the distance of the stumps is greater than six vertebral bodies by radiography, or those who developed complications in anastomosis and for whom reconstruction in a single time is not possible, for the drainage of secretions housed in the upper esophagus¹⁹. These secretions can be regurgitated and aspirated by the patient, which may mean possible cases and aspirative pneumonias²⁰. Therefore, the patient in this report underwent the procedure in order to avoid respiratory complications. Some studies compared children who underwent primary anastomosis with those who underwent Esophagostomy, in which the result was that the last group was more prone to atrophies¹⁷.

Finally, the definitive treatment in this patient profile, which was not a candidate for gastroesophagectomy surgery, is the maintenance of gastrostomy and Esophagostomy up to one year of life, to later perform the esophagus replacement, after the colon or stoma can be used²¹.

Conclusion

The study showed a case of a newborn in which an esophageal atresia was diagnosed with absence of fistula with trachea. The form of presentation of the condition, in this case, is rare and shows difficult resolution. The condition may be accompanied by other unidentified malformations up to this stage of the study. The patient was followed up to 7 months of age, in nutrition via gastrostomy and there is no estimate for gastroesophageal anastomosis surgery. Therefore, the presentation of esophageal atresia without tracheoesophageal fistula represents rare involvement

and, without the possibility of early attachment of esophageal and stomach stumps, it represents a condition with a reserved prognosis.

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