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CASE REPORT

Esophagus atresia and duodenal obstruction: report of two cases

Júlio Onofre de Oliveira Tavares[1], Mauro Roberto Basso[1], Andrea Morgato Mello Miyasaki[1], Ricardo Silva Parreira[1], Heloisa de Carvalho Mota Menezes[1]

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Abstract

Esophageal atresia, with or without tracheoesophageal fistula, and congenital duodenal obstructions are relatively frequent changes in the digestive tract in pediatric surgery. The combination of both, although described in the literature, is unusual. It is assumed that the early diagnosis with imaging tests and the combined surgical schedule, although still undefined, can reduce the mortality of these children. We report the cases of two newborns with esophageal atresia and congenital duodenal obstruction, concerning diagnosis, surgical treatment and clinical evolution.

[1] State University of Londrina, Department of Pediatrics and Pediatric Surgery of the State University of Londrina, Northern Paraná Regional University Hospital, Londrina, PR, Brazil.

Correspondence to:
Julio Onofre de Oliveira Tavares.
Universidade Estadual de Londrina - Endereço: Av. Robert Koch, nº 60, Centro de Ciências da Saúde - Prédio I - 1° andar, Vila Operária - Londrina/PR, Brazil. CEP: 86039-440 E-mail: julioonofre.qi@gmail.com

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INTRODUCTION

Esophageal atresia (EA), with tracheoesophageal fistula (TEF) is a relatively common congenital malformation, affecting one in 2500 to 4000 live births. In 30-50% of cases, EA/TEF is associated with other abnormalities, most commonly being part of the VACTERL spectrum (vertebral, anorectal, cardiac, tracheoesophageal, renal and limb)[1]. Congenital duodenal obstruction (CDO) affects approximately one in 2500 to 10,000 live births, representing almost half of all cases of neonatal intestinal obstruction. CDO may come from an intrinsic cause, such as atresia, stenosis and duodenal; or extrinsic, by annular pancreas, intestinal malrotation and pre-duodenal portal vein[2]. The association between EA and CDO is a rare entity, not associated with the VACTERL complex and not very much described in the literature.

CASE REPORT

CASE 1

Newborn from 30 weeks of gestation, with birth weight of 1,325 grams. Gestational polyhydramnios and fetal echocardiogram with patent ductus arteriosus and interatrial communication. With Down syndrome and no other apparent malformations. In the delivery room, a nasogastric tube was attempted, but without success. He was referred to intensive care unit due to prematurity. Chest and abdomen radiography showed EA with a probable association with TEF and a double bubble sign consistent with CDO (figure 1). A surgical procedure was performed on the 3rd day of life in which the surgeons corrected the EA/TEF with primary esophageal anastomosis; and repaired his annular pancreatic duodenal obstruction with anastomosis (Diamond shaped); both procedures were carried out in a single surgery. He progressed well, being fed orally on the 7th postoperative day and was discharged from the hospital only after 60 days due to very low weight and the associated cardiac complications. Esophageal dilation sessions were performed due to stenosis in the 12-month period. After one year of life, he underwent bilateral orchiopexy and videolaparoscopic fundoplication to correct gastroesophageal reflux disease. He has been in outpatient follow-up for two years without complications.

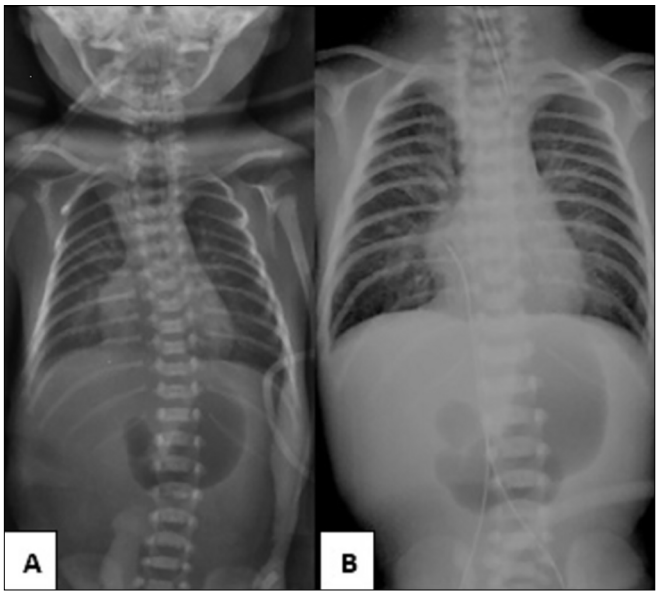


Figura 1. A) Chest x-ray with a nasogastric tube without progression, folding up in the upper esophageal stump. B) Double bubble sign, suggesting duodenal obstruction.

CASE 2

Newborn at 31 weeks of gestation, with birth weight of 1,880 grams, gestational polyhydramnios and no other apparent malformations. In the delivery room, we tried to insert a nasogastric tube unsuccessfully. He was referred to intensive care unit due to gestational age and for clinical investigation. Chest and abdomen radiographies also showed EA with a proximal stump in the cervicothoracic transition, with probable association with TEF and a double bubble sign compatible with CDO (figure 2). A surgical procedure was performed on the 2nd day of life, finding a long-gap EA/TEF (04 vertebral bodies), and we did a cervical esophagostomy with distal fistula ligation, and we treated the annular pancreas-type duodenal obstruction with duodenal-duodenal anastomosis (Diamond shaped), performed in a single surgical procedure, in addition to gastrostomy. He progressed well, being fed on the fifth postoperative day and he was discharged from the hospital after 40 days due to low birth weight and neurological complications (ventricular hemorrhage and convulsive syndrome). The patient has been undergoing outpatient follow-up for two years without complications and in surgical schedule for reconstruction of esophageal transit when reaching ideal age and weight.

DISCUSSION

The association of EA with or without TEF and CDO is known, although rare[3]. A 30-year series published by Ein et

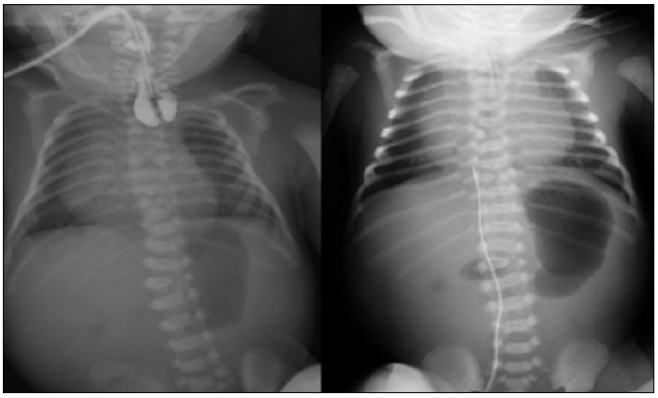


Figura 2. Chest x-ray with contrast progression interrupted in the cervical-thoracic position. B) Double bubble sign.

al. showed a 6% association between EA with or without TEF and CDO.

Adequate management of newborns with EA with or without TEF and CDO lies in the early recognition of these two anomalies and a well-organized approach to their repair. In babies with EA with TEF, the diagnosis of a coexisting CDO can usually be suggested with an abdominal radiography in which a double bubble sign is visualized, as were the two cases described[3]. In cases of EA without TEF, the diagnosis of CDO, in most cases, is delayed until esophageal transit is established or when gastrostomy feeding fails[4], which can lead to an increase in the mortality rate, especially due to bronchoaspiration and respiratory failure[3], which was not described in this reported case series.

The timing of surgical repair is still controversial[5] due to comorbidities and associated malformations. The procedure with prior diagnosis of the two conditions, EA and CDO, seems to reduce mortality by 26%[3]; however, it is not possible to attribute this reduction to this data alone, since it is a phenomenon with multiple variables due to the complexity of the cases.

CONCLUSIONS

EA with or without TEF associated with CDO is a rare entity that deserves attention. Complementary investigation of other congenital malformations of the digestive tract, such as CDO, in addition to the VACTERL spectrum in newborns with EA with or without TEF should always be performed with simple propedeutics and early signs of non-progression of the diet after restoring esophageal transit. The programmed and synchronous treatment of the two malformations (EA and CDO) brings a considerable reduction in mortality, mainly due to the reduction of complications associated with CDO, such as aspiration pneumonia. Therefore, the surgical management of newborns with multiple malformations remains a challenge for the pediatric surgeon who, linking high clinical suspicion with organization and efficiency in treatment brings, in most cases, better outcomes.

REFERENCES

1. Stark Z, Patel N, Clarnette T, Moody A: Triad of tracheoesophageal fi stula-esophageal atresia, pulmonary hypoplasia, and duodenal atresia. J Pediatr Surg 2007; 42: 1146–1148
2. Chen QJ, Gao ZG, Tou JF, Qian YZ, Li MJ, Xiong QX, Shu Q:Congenital duodenal obstruction in neonates: a decade's experience from one center. World J Pediatr. 2014 Aug;10(3):238-44
3. Ein SH, Palder SB, Filler RM: Babies with esophageal and duodenal atresia: a 30-year review of a multifaceted problem. J. Pediatr Surg 2006; 41: 530-532
4. McCook TA, Felman AH: Esophageal atresia, duodenal atresia and gastric distension: experience of 18 patients. Pediatr Surg 1981; 16:4 – 7
5. Karlan M, Thompson J, Clatworthy HW: Congenital atresia of the esophagus with tracheoesophageal fistula and duodenal atresia. Surgery 1957;41:544 - 8