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Case Report: Esophageal Atresia with Congenital Esophageal Stenosis... 18 Years Later

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1. Abstract

We present a patient with Esophageal Atresia (EA) type C and Congenital Esophageal Stenosis (CES) treated in our hospital since birth. The first years she required several dilations and was studied according to her evolution, presenting respiratory complications until she was 7 years old. She discontinued follow-up and returned at age 18 due to digestive symptoms. Previously she felt asymptomatic, although she sleeps semi-sitting and requires drinking water after each swallow. The patient has a good quality of life, despite the fact that motor disorders were found in high-resolution manometry.

2. Keywords: Esophageal atresia;Congenital esophageal stenosis;Esophageal dilation;Esophageal manometry

3. Introduction

Esophageal atresia is the loss of continuity between the upper and lower part of the esophagus and Tracheoesophageal Fistulas (TEF) are an abnormal connection between the trachea and the esophagus. The incidence of EA is approximately 1 per 4,000 live births, with type C being the most common in 85% of cases (EA with distal TEF) [1]. Congenital esophageal stenosis (CES) is a rare pathology characterized by narrowing of the esophageal lumen that is present at birth and may be asymptomatic in newborns. Three types of congenital esophageal stenosis have been described: fibromuscular, membranous, and tracheobronchial rests[2,3].CES may rarely be related to esophageal atresia with an incidence ranging from 3% to 14%[4].

Our objective is to describe the long-term evolution of a patient with EA type C and CES treated in our hospital.

4. Case Presentation

We present the case of a female preterm newborn with appropriate weight for gestational age, prenatally diagnosed with polyhydramnios and esophageal atresia with distal TEF confirmed

at birth.

At 24 hours of age, end-to-end surgical correction is performed at our institution.

On the 7th postoperative day, the patient presented vomiting, and an anastomotic stenosis was diagnosed, leading to dilations with Savary-Guillard bougies with good response (Figure 1). She continued receiving Ranitidine.

At 9 months, at the beginning of complementary feeding, she had vomiting, sialorrhea, and night time cough, with episodes of bronchiolitis and two hospitalizations for pneumonia with atelectasis.

A serial esophagogram showed a notch in the middle esophagus and stenosis in the lower esophagus. Phmetry was also performed: Reflux index 7.1; 5 episodes longer than 5 minutes; longest episode 32.3 min; Total episodes 88 in 24 hours. Upper digestive video endoscopy (UDVE) with stenosis in the distal third of the esophagus. Pathological anatomy: Esophagitis. Up to 18 months, 6 esophageal dilations were performed with Savary-Guillard plugs and a pneumatic balloon with good response (Figure 2).

At 2 years, the esophagogram was normal, and at 3 years, the pHmetry was normal, allowing for discontinuation of ranitidine.

She presented respiratory symptoms, being treated with budesonide and at 4 years the EGDS X-ray showed a delay in esophageal emptying of 2 minutes in the middle and lower third.

Until the age of 7, the patient continued with recurrent respiratory symptoms and without digestive problems. At that age he had an episode of food impaction. In the UDVE, at 27cm of the Upper Dental Arch (UDA) food remains and esophageal stenosis were observed. Two dilations were performed until the caliber was reached according to age.

The patient discontinued follow-up due to moving to another city. During all those years he did not report respiratory events, but she drank water after eating solids to improve esophageal clearance and he slept in a semi-sitting position.

At the age of 18, she developed heartburn and abdominal pain, so she consulted again. Esophagogram X-ray was requested: Esophagus without radiological evidence of anatomical findings. Conventional manometry: Lower Esophageal Sphincter (LES) location 35 cm to 38cm; Pressure: 22mmHg; complete relaxation. Esophageal body: presents simultaneous and repetitive low amplitude waves (13mmHg) (aperistalsis). Upper Esophageal Sphincter (UES): pharyngo-esophageal coordination present, location 17cm. UDVE: Esophagus with normal characteristics Histology: esophageal epithelium with mild acanthosis without significant alterations.

High-resolution esophageal manometry: Esophagogastric junction with mean basal pressures lower than reference values, hypotensive LES with normal relaxation. Esophageal body, in response to the swallowing stimuli offered, no peristaltic contractions were obtained (Figure 3). Normal Ph-impedancemetry.

He received a 2-week treatment of proton pump inhibitors, with disappearance of symptoms.

Currently the patient is asymptomatic, with postural measures and ingestion of water with meals for the proper passage of food.

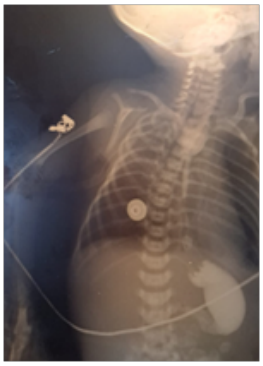
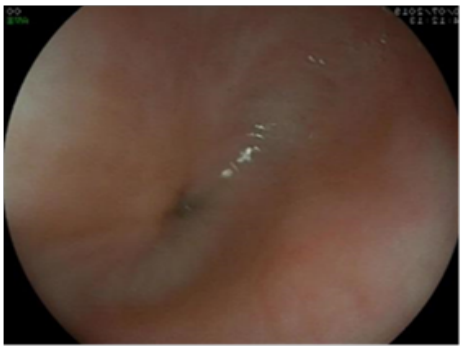


Figure 1: Esophageal stenosis at 7 days of life.



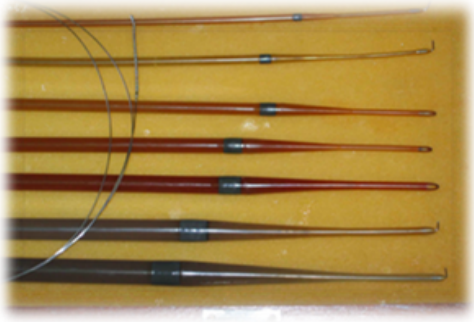


Figure 2: UGIE congenital stenosis. Savary-Guillard dilators.

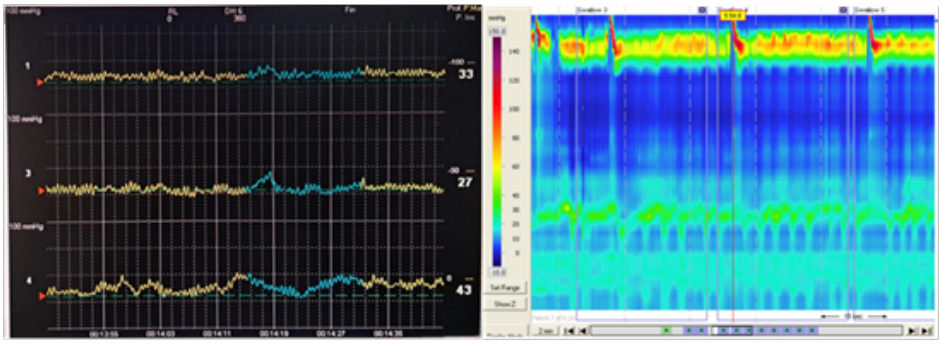


Figure 3: Conventional perfusiomanometry *vs.* High-Resolution Manometry.

5. Discussion

EA is a rare entity, even more when associated with CES.

Reports of the association of EA-TEF and CES have been described in isolated cases and retrospective studies with small numbers of patients.

In our patient, the diagnosis of EA-TEF was made in the neonatal period and dilations were performed in the anastomotic area with good response. Then, she presented recurrent symptoms when starting complementary feeding and the presence of CES was diagnosed. Esophageal dilation was effective in our patient over time, both for the anastomotic area and for the CES, without complications. In the literature, patients are reported who presented esophageal perforation or recurrence of stenosis after treatment as a complication.

Maccann et al, in Canada, reported 477 patients with EA, of which 17 presented the association with CES (incidence 3.6%). The mean age of CES diagnosis was 11.6 months and 15/17 had non-specific symptoms such as regurgitation or dysphagia. CES in 1 patient was diagnosed at the time of surgical repair; it was suspected in 12 patients based on an abnormal esophagogram and in the remaining 4 patients based on endoscopy. Of 17 patients, 11 were treated with dilations and 3 patients presented esophageal perforation[4].

Kawahara et al reported 81 patients with EA-TEF, of which 11 were associated with CES, with a 14% incidence; 5 patients showed histological evidence of CES, (2 patients of fibromuscular type and 3 with tracheobronchial remains). Esophageal dilation was performed as treatment in 9 patients and its effectiveness was temporary except in 2 patients. Esophageal perforation occurred in 4 children [5].

Newman and Bender, at Children's Hospital of Pittsburgh, in 26 years had 225 patients with EA-TEF, of which 18 had CES. The reported incidence was 8% and symptoms were present in 17 patients, difficult feeding was the most common problem in 8 patients, followed by aspiration pneumonitis and foreign body impaction. Esophageal dilation was the treatment of choice in patients and esophageal perforation was the most frequent complication (33%)[6].

In the Spanish work of Jové Blanco, a significant number of patients presented respiratory and digestive comorbidities and lung function progressively worsened. Compared to our clinical case, respiratory symptoms only occurred in childhood, without presenting pulmonary sequelae[7].

The patient had a pathological Phmetry at one year of age, after which the study was repeated at 3 and 5 years due to persistent respiratory symptoms, with normal results, the same happening in the control Ph-impedanciometry at 18 years of age[8].

Romero Manterola et al, from the Santísima Trinidad Children's Hospital in Córdoba, presented 11 patients with CES, who were treated with a favorable response, 4 surgically and 7 with dilations[9].

In the paper of Zubiri, Ramos et al., adolescents with corrected esophageal atresia were studied with high-resolution manometry. Three types of manometric patterns were described [10]. Two patients had complete aperistalsis, 7 patients had distal contraction (distal contractions in the middle or distal third in no less than 2 swallows) and one patient presented panpressurization. Our patient corresponds to the type of complete aperistalsis[10,11,12]..

At the time of our patient's treatment, there were no follow-up protocols for these pathologies. She received ranitidine for the first 3 years, and then she was studied according to the symptoms she presented. Currently, there are ESPGHAN-NASPGHAN Guidelines, which are suitable for standardizing the follow-up of these patients[13].

Our group is the first to present a patient diagnosed with EA-TEF with CES and its long-term evolution. In our patient, despite persisting with esophageal motor disorders, she had good clinical evolution, with improvement in her quality of life.

Early recognition of this association contributes to minimize digestive and extradigestive symptoms, offering appropriate treatment and avoiding complications.

Multidisciplinary management of patients is essential, following international guidelines, with personalized treatment for each particular case[14].

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