

Tracheal Agenesis Type II of Floyd's Classification in Premature Patient in a Third-Level Hospital: Case Report

Guillermo Yanowsky Reyes, MD^{1,2}, Rafael Santana Ortíz, MD^{1,2}, Manuela Gómez Reyes, MD¹, Jesús Antonio Aguilar Mata, MD^{1,2}, Carlos Oswaldo Yanowsky González, MD^{1,2}, Beda P Terrazas Moreno, MD³, Arely Jaqueline Pérez Padilla, MPSS, MD^{1,2}

¹División de Pediatría, Hospital Civil de Guadalajara "Fray Antonio Alcalde", México

²Servicio de Cirugía Pediátrica, Hospital Civil de Guadalajara "Fray Antonio Alcalde", México

³División de Pediatría, Hospital General de Occidente, México

*Corresponding Author: Guillermo Yanowsky Reyes, División de Pediatría, Hospital Civil de Guadalajara "Fray Antonio Alcalde", Guadalajara, Jalisco, México

ABSTRACT

Introduction: Tracheal agenesis is congenital malformation with incidence of 1:50 thousand births. Data to suspect it are cyanosis, respiratory difficulty, absence of air in auscultation and audible crying and difficult endotracheal intubation. Association with malformations in 90%. **Objective:** Show case of tracheal agenesis and its complications during handling. **Case report:** Male RN of 31.2 SDG. Basic resuscitation VPP cycles without showing response to cyanosis and SDR data at the expense of marked stre-subcostal retraction requiring intubation, during laryngoscopy show findings so that the esophagus is cannulated. Subsequently, due to clinical deterioration, he goes to the operating room, dying during the secondary transsurgical to multiple organ failure and respiratory failure. **Conclusion:** The value of this case lies in the demonstration of clinical problems and severity, with suspicion and approach being key for early diagnosis and treatment.

Keywords: Agenesis, Trachea, Premature, VACTERL

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INTRODUCTION

Tracheal agenesis is a congenital malformation incompatible with life, a reported incidence of 1 in 50,000 births. The data to suspect in this pathology are cyanosis, severe respiratory difficulty, no air movement in the auscultation, absence of audible crying and difficult or impossible endotracheal intubation. They are associated with multiple associated malformations are present in 90% of cases, more frequently affecting the cardiovascular, gastrointestinal and genitourinary systems. There are cases replenished to the VACTERL association In the presence of trachea or bronchoesophageal fistula it is a favorable factor providing adequate oxygenation and the possibility of life, since most die within a few hours or the first days of birth due to other congenital malformations, difficulties in diagnosing and lack of management of the airways,

considering that prematurity and respiratory difficulty syndrome can be the cause of mortality and high morbidity. The purpose of our report is to show a case of tracheal agenesis and its complications during handling(1,2).

CASE REPORT

Male newborn of 31.2 weeks of gestation. Family history of a 21-year-old breast, apparently healthy, product of the first pregnancy, prenatal control or history of polyhydramnia is unknown. Obtained by childbirth, weight 1425 grams, size 40 cm, Apgar of 4.5, and 7 at 10 minutes. Basic resuscitation steps are initiated, ventilation cycles with positive pressure without showing a response for generalized cyanosis and respiratory difficulty data at the expense of marked sternal and subcostal retraction, requiring intubation. As a finding during laryngoscopy, small epiglottis, subglottic stenosis, difficulty in observing vocal cords is observed, so the trachea

is not possible to cannulate and the esophagus is decided to canulate, allowing to reach 85% saturations. Physical examination, generalized cyanosis is observed due to the absence of crying and respiratory failure, imperforated anus and scrotal edema, the rest of the normal examination. During the first 24 hours of life it remains in phase III mechanical ventilation, then begins with abdominal distension and

hemodynamic commitment with hypoperfusion in pelvic limbs. Due to the clinical deterioration, it is decided to go to the operating room to perform bronchoscopy, endoscopy and colostomies, dying during the secondary trans-surgery due to persistent respiratory acidosis caused by multiple organ failure and respiratory failure.



Figure 1. *Imperforated anus*

Postnatal approach, an x-ray of the chest and abdomen is taken, finding data compatible with hyaline membrane disease, air meniscus, at the intestinal level absence of distal air.

Autopsy was performed finding as the main finding tracheal agenesis with e Floyd classification esophageal

bronchial fistula type II, bilateral pulmonary atelectasis, mild cerebral edema, extra medullary, premature erythropoiesis, meconium aspiration, diffuse alveolar damage, acute tubular necrosis, splenic congestion and anoperforated.



Figure 2. *Bronchial communication - esophagus*

DISCUSSION

Congenital malformations of the trachea and esophagus are secondary in the absence of the trachoesophageal septum or alteration in vascular irrigation between the third to sixth week of embryological development, inadequate evolution of the embryonic intestine which forms only esophagus not providing sufficient endoderm for the formation of the trachea.

Epidemiologically the presentation cup is 1/ 50,000 newborn, with the male sex being the most frequent in a 3:1 ratio, 34% is associated with polyhydrmania. It is associated with multiple malformations such as complete

heart disease 25%, pulmonary malformations 23%. 132 cases have been reported since the first publication that was made in 1900.

Etiologically, no environmental or genetic factor has been found. The disease can be suspected with prenatal ultrasound at the end of the second trimester due to the absence of gastric chamber or small stomach, polyhydramnios, hyperecogenic lungs, ascites, absence of blood flow at the laryngeal level on Doppler ultrasound. In the postnatal diagnosis, suspect if there is a history of polyhiramnios, and the aforementioned compatible clinic. The diagnosis is confirmed with direct laryngoscopy, helical computed tomography of the respiratory tract,

endoscopy to confirm the presence of fistula in the face of the impossibility of endotracheal intubation. Differential diagnoses are considered laryngeal atresia, gothic membrane or congenital tracheal stenosis.

We have two classifications which are:

Floyd's classification being the most used system that is divided into:

Type I Absence of proximal trachea and distal trachea is connected to the esophagus by a fistula

Type II Complete absence of trachea and the location of the bronchial bifurcation is normal.

Type III The two main bronchi arise independently of the esophagus.

Faro R.'s classification of tracheal agenesis is divided into:

A. Total

B. Tracheal atresia with upper sac bottom, right and left bronchi connect to the esophagus.

C. Tracheal atresia with upper sac bottom and certain vowels, segment of carina fistula to the esophagus.

D. Vocal cords in a blind sack, atresic segment to carina and communicates to the esophagus.

E. Upper segment with sack bottom, tracheal atresia and small cabo with carina and bronchi.

F. Upper sac bottom, tracheal atresia and distal portion are attached to the esophagus by Carina with normal bronchi.

G. Upper segment ends in a sack bottom and an atresia portion joins the carina with normal bronchi.

Our patient belongs to type II of the Floyd classification and group C of the Faro classification.

As a management in tracheal atresia type, it is recommended to intubate the esophagus to ventilate the child in which some surgical medical intervention is performed. In the latest studies they report that reassuring and esoaphagocarinoplastic could have good results in tracheal atresia type. The reconstruction of the neo trachea with esophagus with or without an external splint or placement of endoluminal stent can be considered. Subsequent reconstruction of the digestive tract with gastric ascent or intestinal interposition. Cadaveric tracheal allografts, 3D printed grafts and autologous free tissue reconstruction can provide other reconstructive options once the trachealized esophagus allows somatic growth(1,2,3,4,5).

Other techniques mentioned are selective intubation (permeable airway), placement of balloon probe, Trans-sternal Y probe (ECMO), Stam GastroStomy to promote

feeding, dachron or goretex graft, Tracheo-bronchial anastomosis and ascension, tracheal transplant wrapped in epiplon(6,7).

CONCLUSION

The value of this case lies in the demonstration of the serious clinical problems that a patient with this type of abnormality presents, in them the laryngoscopy and endoscopic evaluation is very useful to establish the diagnosis, likewise the use of radiological studies to evidence the presence of fistula is a very useful auxiliary support. This abnormality should be suspected in premature pediatric patients with multiple congenital malformations, who have poor oxygenation and difficulties for endotracheal intubation.

REFERENCES

1. Densmore JC, Oldham KT, Domínguez KM, Berdan ER, McCormick ME, Beste DJ et al. Neonatal esophageal trachealization and esophagocarinoplasty in the treatment of flow-limited Floyd II tracheal agenesis. *J Thorac Cardiovasc Surg.* 2017; 153: e121-e125
2. Caliskan E. Management of tracheal agenesis with esophageal intubation can save lives: a reminder for anaesthesiologist and neonatologist. *J Compr Ped.* 2018; 9(3): e60388.
3. Grass B, Simma L, Reinehr M, Zimmermann U, Gysin C, Henze G, Cannizzaro V. Two case reports of unexpected tracheal agenesis in the neonate: 3 C's beyond algorithms for difficult airway management. *BMC Pediatr.* 2017 8; 17(1): 49.
4. González, N. M., Torres, O. L., Kim, S. M., Hernández, J. H., & Pérez, A. H. (2010). Tracheal Agenesis. Presentation of a Tracheal agenesis case. Presentation of a case. *DOAJ (DOAJ: Directory Of Open Access Journals)*. <https://doaj.org/article/02c1a9d7755d44a695c6bb90315d735e>.
5. Statz T, Lynch J, Ortmann M, Roth B, Tracheal Agenesis: A case report *Eur J Ped* 1989 149:203-204. Wolfgang P, Metz V, Birnbacher R, Hormann M, Tracheal agenesis evaluation by helical computed tomography *Ped Radiol* 2000 ,30:200-2003.
6. Nakada K, Kitagawa H, Tomotake E, et al, Tracheal agenesis with Bronchoesophageal fistulas a case report *Japanese Journal Surgery Vol 19, no.4* 1989, 494-497. Fraser N, Stewart R, Grant J, Martin P, Gibbin P Tracheal agenesis with unique *Anatomy Journal Ped surgery* 2005 40 e 7-e10.
7. O'Neill D, Morecroft J, Gibson A, Unusual case of tracheal agenesis, *Pediatric and Developmental Pathology* 1999, 2 176-179.