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Congenital absence of cartilaginous tracheal rings associated with esophageal atresia and trifurcated carina: a novel anomaly?

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Abstract Tracheomalacia associated with esophageal atresia (EA) is a well-known condition. However, complete absence of tracheal rings (TRs) is extremely rare. Our aim is to describe a novel triad of conditions and to discuss the best treatment. An expremature male operated for EA presented with severe respiratory distress. The diagnosis of absent cartilage rings, suspected on bronchoscopy, was confirmed by optical coherence tomography. The absence of TRs was localized to a short tracheal segment, and the carina trifurcated into right upper lobe, right intermediate, and left main bronchus. The patient was treated with resection and anastomosis with a completely satisfactory course. Absence of TRs was previously reported by us in 2 other cases, both with associated EA and trifurcation of the carina. One child was treated with tracheostomy and the other with a stent, but the outcome was far from optimal. The patient with tracheostomy eventually underwent resection and anastomosis with tracheostomy closure. Congenital absence of TRs is extremely rare. Although localized, it is responsible for severe symptoms owing to complete tracheal collapse and may be misdiagnosed as tracheomalacia. In our experience, it has been associated with EA and trifurcated carina. Our limited experience suggests resection of the abnormal segment and tracheal anastomosis as the best treatment.

Normal tracheal rings (TRs) are C-shaped structures with an anterior cartilaginous component and a posterior gap, which is completed by the pars membranosum.

Various congenital anomalies of TR or pars membranosum have been described in infants (Fig. 1). The most frequent is tracheomalacia (TM) [1]. Tracheomalacia refers to a weakness of the tracheal wall, sometimes associated with other conditions such as esophageal atresia (EA) with tracheoesophageal fistula (TEF) or vascular rings. In TM, the TRs appear flattened, collapsing during expiration and associated with a pars membranosum, which often bulges forward, obstructing the lumen. Other uncommon anomalies

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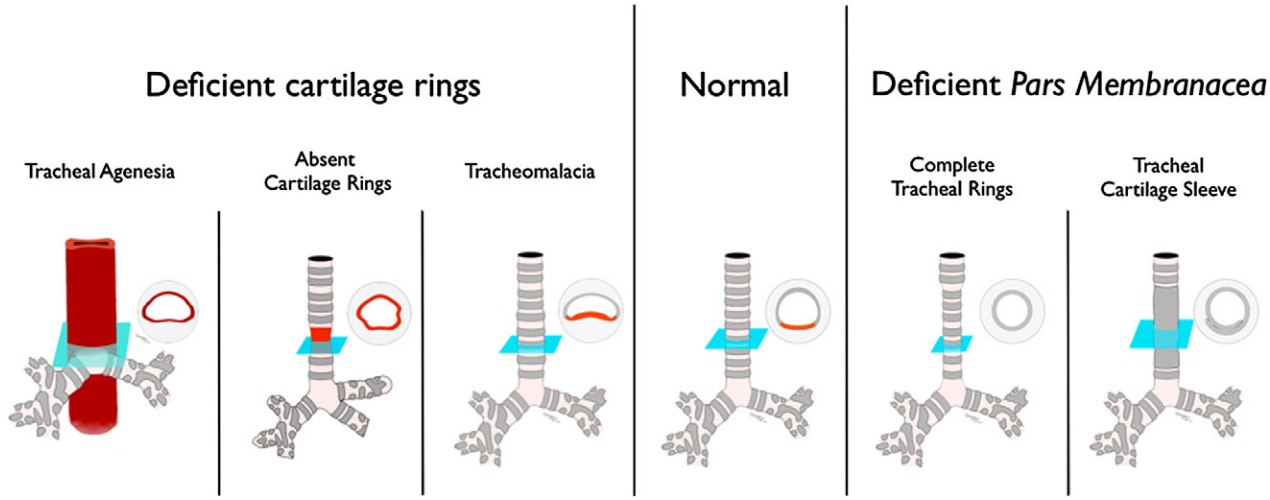

Fig. 1 Congenital anomalies of the trachea can be because of a cartilage rings or pars membranacea defect. In the center of the figure, the normal relationship between cartilage (colored in gray in the section) and pars membranacea (in red). On the left, the conditions with deficient cartilage rings are shown; on the right, the conditions with deficient pars membranacea. Notice the trifurcation of the carina in the absent ring condition.

of TR are the presence of complete "O" shaped rings in congenital tracheal stenosis and, even more rare, the tracheal cartilaginous sleeve, described in association with craniosynostosis [2]. In the tracheal agenesis spectrum, a part or the entire trachea is absent, and there is a primitive foregut without TR forming a communication between some part of the respiratory and the alimentary tracts [3].

In the past, our group described segmental absence of TRs in 2 patients with EA [4]. The aim of this article is to present the clinical features of this congenital anomaly, to discuss the diagnostic tools and the treatment options.

1. Case report

We have observed segmental absence of TRs in 3 patients since 2001. The first 2 cases have been previously reported [4]; we present a new case and the update of the evolution of the clinical history of the previously described cases.

A male patient born at 31 weeks gestation with EA and distal TEF with associated anorectal malformation underwent EA repair. The repair (in another institution) was complicated by an anastomotic dehiscence and mediastinitis, which was managed conservatively. He underwent aortopexy and fundoplication with gastrostomy in 2 separate surgeries, without improvement of his recurrent respiratory symptoms. He was then referred to our institution at 4 years of age. Bronchography confirmed the severe collapse of the middle/distal trachea, which terminated in a trifurcated carina (Figs. 2 and 3). The area of the tracheal anomaly extended for 23 mm just above the origin of the right upper lobe bronchus from the trachea. The diameter of the trachea at this level was 3 mm without any positive pressure and increased to 6 mm at 15 cm H_2O of pressure. No TRs were identified at endoscopy (Fig. 4). Optical coherence tomography (OCT) confirmed the absence of TR in this segment (Fig. 5) and the presence of normal rings in the rest of the trachea. He underwent tracheal resection and direct end-to-end anastomosis, under cardiopulmonary bypass. The postoperative course was uneventful. The histopathology

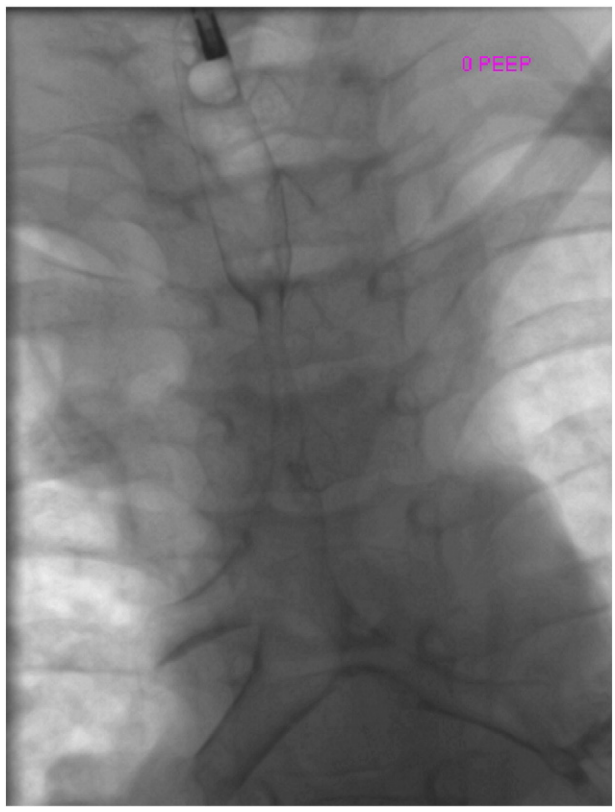

Fig. 2 The bronchography shows the localized collapse of the trachea without a positive end expiration pressure (PEEP 0). The carina is trifurcated.