Editorial

# To do or not to do? The management dilemma of congenital tracheal stenosis in the setting of the ring-sling complex

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Pulmonary sling (PS) is a rare malformation caused by the origin of the left pulmonary artery from the right pulmonary one. In its course to the left hylum the aberrant left pulmonary artery crosses the tracheal carina and right main bronchus determining compression (1). Pulmonary artery sling may be frequently associated with congenital tracheal stenosis (CTS) with complete cartilaginous tracheal rings and this association was defined "ring-sling complex" (2).

Major intracardiac anomalies, abdominal malformations, as well as genetic syndrome are some of the associate lesions often observed in children with CTS. These co-morbidities play an important role on the overall outcomes, even when the surgical repair of the trachea is perfect (3,4). Surgical management of PS is made by the repositioning of left pulmonary artery on the pulmonary trunk and hence anteriorly to the trachea (5).

However, at the moment, the surgical correction of the mild forms of tracheal stenosis (in the case of PS) is still under debate. The concerns arise from the evidence that tracheal reconstruction by himself increase the risk of death or complications. Moreover there is a growing number of reports showing encouraging results with conservative management (6,7).

In the article of Hong and colleagues (7), the authors described their experience and results with conservative and operative management of CTS associated with PS. The median age of the population is 7.5 months and is according with the largest series published in literature. As previously described by Manning and Butler tracheal surgery is particularly done in the first year of life (8,9).

Operative approaches to CTS, in the work of Hong and colleagues (7), were slide tracheoplasty and primary stent placement. Slide tracheoplasty has been introduced by Tsang in 1989 and at the moment has demonstrated to be the treatment of choice in all anatomical patterns of CTS (8,10). Furthermore slide tracheoplasty confers to the reconstructed trachea the possibility of growing (11). This technique is performed safely under cardiopulmonary by-pass thus allowing the simultaneous management of the heart malformations (8,12). On the contrary primary management of CTS by stent implantation has not demonstrated satisfactory results despite the enthusiasm for new biodegradable stents (13,14). Granulations, stent migrations and difficulties to removing are some of the common complications that have limited their use (13-17). Serio and coworkers recently suggested tracheal stent placement only in case of difficulties to the weaning from mechanical ventilation (i.e., for the persistence of tracheo-malacia after surgical correction of PSs) (15). Despite endoscopic procedures are considered nowadays the technique of choice in preoperative evaluation and postoperative management of those children underwent to tracheal surgery (9,16), the series reporting the primary use of stent in CTS are still lacking and the number of

cases can not be compared with the largest ones of surgical experiences (9).

Hong and colleagues, in their article, describe the interesting concept of diameter/length ratio (DLR) as an index to define the severity of the CTS. A significative higher DLR ratio (that means a tracheal anatomy characterized by a short and mild narrowed stenosis) was observed in the group managed conservatively. However computational flow-dynamic studies made on web-like stenosis have demonstrated that the degree of narrowing is the main determinant of the pressure drop across an airway's stenosis instead the length of it and that the pressure drop correlates well with the symptoms (17).

Considering the normal tracheal dimension of an infant below 1 year of age (18), the presence of CTS can reduce up to 75% the normal diameter. This means that infective or inflammatory respiratory disorders, even more reducing the tracheal lumen, could determine the occurrence of apparent life threatening events (ALTE). We experienced a case of emergency tracheostomy in a 6-year-old boy. This child was treated for asthma from birth. During an airway's infective episode the child developed early respiratory difficulties that evolved rapidly in respiratory insufficiency. At arriving to the hospital he was impossible to intubate, requiring hence an emergency tracheostomy. Preoperative instrumental examinations revealed a pulmonary artery sling associate to a funnel shaped tracheal stenosis with complete tracheal rings and an internal diameter of 3 mm.

This value (3 mm) was proposed previously by Huang and coworkers as a "cut-off" diameter above which perform left pulmonary artery repositioning only (6). Anton-Pacheco previously, tailored to 4–6 mm (and minimal clinical symptoms) the criteria to define CTS as mild and to perform conservative management (19). But the question if the narrowed trachea (with complete rings) can grow up to a normal diameter is still under debate (6,7,19,20). Cheng and colleagues (20) have showed a growth of the trachea up to normal values for age. In their work however, the authors underlined the impossibility to define the presence of complete tracheal rings. According with literature, it is certainly that the tracheal rings are the "conditio sine qua non" to define a CTS, for this reason it is difficult to agree with Cheng and coworkers about their findings. Despite previous studies have demonstrated tracheal growth in the presence of complete tracheal rings, the improvement in tracheal diameter was adequate but not appropriate for age in all (6,7,19).

On the other end, the improvements in the surgical management of CTS was impressive in the last two decades. Currently there is a development of pediatric tracheal programs all over the world, exceeding sometimes the real prevalence of the disease. A breakthrough in this field was represented by the introduction of the slide tracheoplasty and the management by dedicated tracheal team (21). Unfortunately, the mortality rate following tracheal surgery still accounts for 10–18% of patients with an incidence of further complications rising up to 40%. Anastomotic leakage is one of the most dramatic complication responsible of an incidence of further tracheal re-interventions from 28% to 48% (8,9).

Furthermore, in the clinical practice, some children scheduled initially for conservative treatment are difficult to wean from mechanical ventilation and for this reason they are re-directed to surgery (6,7). Finally, there are few indications in literature about the methods to evaluate those patients managed conservatively, which instrumental examinations to perform and the timing. An interesting tool may be computational fluid dynamics that offers anatomo-functional informations about the stenotic trachea (17,22). This technique has showed to correlate well with clinical symptoms and degree of stenosis, giving a useful indication to perform surgery or not (22). At the moment the evaluation of CTS is mostly anatomic with bronchoscopy and conventional computed tomography as the gold standard (21,23). The importance of anatomo-functional assessment has been well demonstrated by the role of magnetic resonance imaging in the postoperative evaluation of congenital heart malformations, to the point that nowadays some authors suggested the use of MRI instead of more invasive diagnostic procedures in selected cases (24,25).

In conclusion, association of CTS with complete tracheal rings is the one of the most determinant of the outcomes in children with PS. Surgical management is challenging and a multidisciplinary team approach is unquestionable. Severe forms of CTS require early surgery despite postoperative course may be complicated. Conservative management of mild asymptomatic cases is attractive but a closer surveillance of these patients is required. The anatomic evaluation of CTS should be accompanied by a functional studies to assess the impact of the stenosis on the children's respiratory physiology.

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## Footnote

*Conflicts of Interest*: The authors have no conflicts of interest to declare.

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