Left pulmonary artery sling and congenital tracheal stenosis: to slide or not to slide?

Michele Torre

Units of Pediatric Surgery and Airway Team, Istituto Giannina Gaslini, Genova, Italy

Correspondence to: Michele Torre. Unit of Pediatric Surgery and Airway Team, Istituto Giannina Gaslini, Via Gaslini 5, Genoa 16147, Italy. Email: micheletorre@gaslini.org.

Provenance: This is an invited Editorial commissioned by Section Editor Dr. Raffaele Giordano (Department of Advanced Biomedical Sciences, Adult and Pediatric Cardiac Surgery, University of Naples Federico II, Naples, Italy).


doi: 10.21037/jtd.2017.11.76

View this article at: http://dx.doi.org/10.21037/jtd.2017.11.76

The article published on Int J Clin Exp Med (1), raises very interesting issues regarding the surgical treatment of left pulmonary artery sling (LPAS) associated with congenital tracheal stenoses (CTS). CTS are rare anomalies characterized by the presence of complete cartilaginous rings along a variable length of trachea. In most cases, the complete rings extend for more than half of tracheal length. LPAS can be associated with CTS (the so-called ring-sling complex), with an incidence reported of about 50% of cases with CTS (2). Conversely, complete rings have been described in 50–65% of the patients with LPAS, as reported by old reports cited by the authors (3,4). This is the first issue for discussion, the real association of CTS in patients with LPAS. While ruling out a LPAS in a CTS patient is very simple with an angio CT scan, to be sure that a LPAS patient does not have any complete ring is more demanding. We cannot assume that the reported incidence of 50–65%, reported more than 30 and 40 years respectively (3,4), is correct. In those years, the diagnosis of CTS was probably based more on clinical symptoms than on bronchoscopy, so in our opinion this should be studied more precisely. We suspect that the incidence of CTS in patients with LPAS is underestimated. In fact, the diagnosis of CTS can be difficult, as symptoms are sometime very mild or absent. The only way to determine exactly in how many LPAS patients CTS are present, is to perform a tracheoscopy in all patient with LPAS, including asymptomatic, as CT scan is not sufficient to rule out CTS. The endoscopic evaluation should be performed by an expert using a rigid scope, as it is well known that CTS can be easily missed in particular in case of mucosal thickening by edema. Such study has never been performed before at our knowledge.

This fact has important practical implications on treatment. LPAS alone can be fixed with a surgical repair that is usually straightforward and can be performed even without cardiopulmonary bypass, while LPAS with CTS requires usually a tracheoplasty and cardiopulmonary bypass. The authors of the paper observed a better outcome in patients treated for LPAS but not undergoing tracheoplasty, than in those in whom tracheoplasty or other tracheal procedure were performed. Based on this observation, they propose to treat LPAS alone and then observe if the tracheoplasty will be required. The proposal of the authors could be theoretically justified by two considerations. First, tracheoplasty is a surgical procedure potentially carrying risks, even serious. Secondly, not all isolated CTS require surgery: the surgical indication for isolated CTS depends on the severity of stenosis and patient symptoms. If the patient is doing well and trachea with complete rings grows with the patient, tracheoplasty can be postponed or not performed at all. In our experience and in the literature, there is a percentage of isolated CTS followed conservatively (5,6).

On the other hand, to fix LPAS without treating CTS could be risky, as the post-operative period of a patient with a reduced tracheal lumen who has to be extubated after
a cardiovascular surgical procedure can be troublesome. Moreover, if later on the tracheoplasty will become necessary due to the respiratory worsening, a second mediastinal approach through a redo sternotomy is more demanding. For these reasons, in most reports it is strongly recommended to repair the CTS at the same time as LAPS, with a multidisciplinary surgical team (2,7-10).

In our opinion, it is difficult to predict, before surgery, if respiratory symptoms in infants and children with LAPS and CTS have to be ascribed to one, the other or, most probably, to the combination of both. LAPS is usually reducing the tracheal lumen at a single level, while the tracheal caliber in patients with CTS is reduced along a longer segment of the trachea, sometimes extending to all the trachea. It is therefore impossible to assume that the repair of LPAS alone will be able to resolve the patient condition.

The authors state that patients operated for LPAS without any tracheal procedure who survived were followed for 5 months to 2 years and had a good outcome. Actually, the tracheal diameter, measured with CT scan was very low in this group (2.89 mm), and it is difficult to believe that with such a small trachea they won’t have any respiratory symptoms in the future. The tracheal diameter, measured twice in two patients, tended to grow together with the child, reaching 3.2 and 5.2 mm. respectively. Although it is not reported the age of the patients at the second measurement, our impression is that, again, such size cannot assure a symptoms free life.

The results of the paper show a very bad outcome in patients operated to the trachea, either with slide tracheoplasty and tracheal stent insertion. This data differs substantially from the experience of most Centers with large experience in tracheal surgery (2,7-11). The mortality reported by the authors accounts for 85% of the patients, while the mortality reported after slide tracheoplasty in larger series accounts for around 10% (8,9,11). A possible reason for the bad results of tracheal surgery reported by the authors could be their initial experience, due to the small numbers of the series itself (seven patients only). Tracheal surgery is technically demanding and the management of these patients requires a multidisciplinary team. The failures reported in the paper do not mean that the indication to the slide tracheoplasty was wrong, and that it is advisable to avoid to repair CTS when associated to LPAS, but that it is of paramount importance to manage these complex patients in Centers where a large number of cases is referred (12).

A consideration has to be addressed to the insertion of stents in CTS, that was described by the authors. It is not clear from the paper why the author preferred to insert a tracheal stent in some patients as a treatment for CTS instead of slide tracheoplasty, but the results shown by them are very bad. In the literature there are few reports of CTS managed by stent insertions as primary treatment (13,14), and there are other reports about the insertion of stents in CTS after slide tracheoplasty (9,15). We agree with Monnier, Le Bret, Grillo, and others there are no data supporting stent insertion as a primary approach (2,16,17).

In conclusion, more studies should be performed to establish exactly the percentage of LPAS patients having complete rings; according to the literature, LPAS and CTS should be treated by a combined surgical approach (reimplantation of the left pulmonary artery and slide tracheoplasty in cardiopulmonary bypass) while stents should be avoided as primary treatment. In centers with large experience of tracheal surgery the result of this approach has a good outcome in 90% of cases. The bad results reported by the authors could be due to their initial experience in slide tracheoplasty and to the use of stents in CTS more than to a supposed wrong indication to the slide tracheoplasty in patients with LPAS and CTS.

**Acknowledgements**

None.

**Footnote**

**Conflicts of Interest:** The author has no conflicts of interest to declare.

**References**


