Pulmonary artery sling: associated anomalies, surgical management and follow-up. A single center experience.

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Introduction: Left Pulmonary Artery Sling (LPAS) can be associated with other cardiac abnormalities and tracheal compression and/or stenosis. We describe our experience in the management of this condition.

Methods: In our centre between April 1996 and September 2014, 11 infants (5 male, 6 female) underwent surgical repair of LPAS. Mean age at the time of surgery was 14 months (range 22 days to 5 years and 9 m). In 5 patients LPAS coexisted with other cardiac abnormalities, 3 cases underwent an initial cardiac operation, without sling correction, in one case for critical neonatal COA, in the other two cases for misdiagnosis. The presence of LPAS was suspected by clinical presentation and features on chest X-ray and echocardiography. Confirmation of LPAS was established using angiography (6 patients) or CT (5 patients). Tracheal compression was confirmed by bronchoscopy, in 7 patients we found a tracheal stenosis with complete rings in 5 of them, tracheomalacia in 9 patients. Five patients required surgical LPA reimplantation with slide tracheoplasty (ST) (45%), four patients isolated LPA reimplantation (36%), 2 patients LPA reimplantation with repair of intracardiac anomalies (18%).

Results: Two deaths occurred in the first 30 days after surgery, one of them after ST. Another patient requiring ECMO after ST, is still followed-up under ventilator after two tracheal balloon dilations. In the 8 long-term survivors, LPA patency was assessed using echocardiography, angiography, CT or MR; only 1 patient required a balloon angioplasty (PTBA). Bronchoscopy was performed in all patients who underwent a ST, and 1 ≥ balloon tracheal dilation was necessary in 3 patients.

Conclusions: LPAS has a low operative mortality and excellent long-term patency of LPA. The simultaneous presence of other cardiac abnormalities and/or tracheal stenosis complicates the short- and long-term prognosis. In our center we adopt a multidisciplinary approach. The role of imaging is crucial: in the diagnostic phase the gold standard is Bronchoscopy and CT. A Bronchoscopy is performed at 1,3,6 and 12 mounts post-surgical correction, within cardiology follow up. If tracheal stenosis or pulmonary artery obstruction are suspected CT or MR are performed. Angiography is performed in presence of severe pulmonary artery obstruction.