LeCompte Maneuver for Airway Compression Management in Late-presenting Absent Pulmonary Valve Syndrome


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INTRODUCTION
Patients with absent pulmonary valve syndrome (APV) often present early with airway compression, from diffuse aneurysmal dilatation of the pulmonary artery branches. Repair usually includes pulmonary artery reduction plasty to relieve proximal obstruction of the mainstem bronchi. The LeCompte maneuver has been proposed to address this issue, although there is limited data available. This study reviews our recent experience in managing APV syndrome in later presenting children, and surgical techniques used for managing airway compression.

METHODS
This study is a retrospective chart review of all patients who underwent repair of tetralogy of Fallot and APV from 2000 to 2012 at our institution. Patients with clinical evidence of airway compression undergo systematic pre- and post-operative bronchoscopy. The primary endpoints were postoperative bronchoscopic and clinical evidence of persistent airway compression, and need for reinterventions or reoperations on the pulmonary arteries.

RESULTS
19 patients were included during the study period. The mean age at repair was 4.1±3.0 years (range, 10 months – 11 years). 6 patients had associated anomalies: 3 with discontinuous left pulmonary artery from major aorto-pulmonary collaterals (MAPCA), 1 with a MAPCA to the LPA, 1 with a right aortic arch, totally anomalous pulmonary venous return and infradiaphragmatic MAPCA to the right lung, and 1 patient with 22q11 microdeletion. There were no perioperative deaths. 8 patients with respiratory symptoms had preoperative bronchoscopy, which showed airway compression in 7 patients and managed by pulmonary artery reduction plasty in 4 patients, and LeCompte maneuver in 3 patients. In the first group, 2 patients had no postoperative airway compression, 1 patient had improved compression, and 1 patient had unchanged compression. In patients managed with a LeCompte maneuver, 2 patients had no or trivial airway compression and 1 had improved compression (P = 1.0). There were 6 late reinterventions or reoperations on the RV-PA conduit (2/4 in the PA plasty group, 1/3 in the LeCompte group, P = 1.0).

CONCLUSIONS
In patients with APV and airway compression, either pulmonary artery reduction plasty or the LeCompte maneuver can relieve proximal airway compression, without a significantly different risk of pulmonary artery reintervention between techniques.