Introduc\text{\'}on

Primary pulmonary vein stenosis (PVS) is considered a progressive disease caused by neoproliferation of myofibroblasts induced by local or peripheral signaling. However, advances in surgical and interventional treatment, there is still long-term morbidity and mortality. We present radical surgery as a treatment option.

Materials and methods

A seven year follow up of three patients with PVS who were treated with lobectomy or pneumonectomy between 2007-2008.

Results

Patient one: Term female with a birth weight of 2980g, diagnosed at 3 months of age with a stenosed upper right pulmonary vein. She had complicating interstitial pulmonary disease. Mean pulmonary pressure was 29 mmHg at diagnosis. She was initially stented, but 3 months later treated with bi
dectomy of the upper and middle right lobe because of severe re-stenosis. Mean pulmonary pressure was 60 mmHg at surgery. Follow up catheterization demonstrated reduced pressure. However, a progressive narrowing of the left lower pulmonary vein was identified and the patient died at 2 years of age from a respiratory tract infection.

Patient two: Preterm female born at 28 weeks gestational age with a birth weight of 868g, diagnosed at two months of age with two atretic right sided pulmonary veins. She was treated with right-sided pneumonectomy. At 7 year follow-up she is doing well with height and weight for age just below the 3rd percentile.

Patient three: Preterm female born at 25 weeks gestational age with a birth weight of 525g, diagnosed at 7 months of age with an atretic left upper vein and a narrowed left lower vein. Mean pulmonary pressure at diagnosis was 51 mmHg. She underwent lobectomy of the left upper lobe. Her mean pulmonary pressure was reduced to 26 mmHg after three weeks and 22 mmHg after one year. Today no signs of re-stenosis are seen. She is healthy with weight for age on the 10th percentile and length for age on the 25th percentile.

Conclusion

PVS still carries a high rate of morbidity and mortality. Radical treatment with lobectomy or pneumonectomy of tissue containing severely stenosed veins might slow or halt disease progression.