Early Double Switch With Lecompte Maneuver for Life-Threatening Airway Obstruction
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A 6-month-old boy with congenitally corrected transposition of the great arteries, ventricular septal defect, and mild pulmonary valve stenosis presented with acute obstructive respiratory life-threatening events. Airways obstruction was caused by aneurysmal dilation of the branch pulmonary arteries compressing the carina and bronchi. The child was ventilator dependent. A double switch operation with Lecompte maneuver was performed, bringing the pulmonary arteries forward and relieving the airway compression. The child made a complete recovery and remains well 9 months later with no clinical or radiologic signs of airway compression.

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Congenitally corrected transposition of great arteries (ccTGA) is a rare congenital heart disease, with a reported incidence of 1 in 33,000 live births [1]. Clinical presentation is usually related to the associated lesions such as ventricular septal defect, pulmonary stenosis, tricuspid regurgitation, or abnormality of conduction system [1, 2] and depends on the severity of these associated lesions. This article reports the unique case of an infant with ccTGA, unrestrictive ventricular septal defect (VSD), and mild pulmonary stenosis who presented with several episodes of obstructive respiratory life-threatening events. The cause was airway compression caused by aneurysmal dilation of the branch pulmonary arteries (PAs), which has not previously been reported in the setting of ccTGA. He was successfully treated with a double switch operation associated with a Lecompte maneuver.

The patient, a 6-month-old boy, was antenatally diagnosed with ccTGA, VSD, and pulmonary valve stenosis. An echocardiogram done at birth confirmed the diagnosis, showing situs solitus with mesocardia, discordant atrioventricular and ventriculoarterial connections, with balanced ventricles, large muscular inlet VSD, and mild pulmonary valve stenosis with good size confluent pulmonary arteries (Fig 1). The child was initially asymptomatic, with oxygen saturations of 100% in room air. Hemodynamics were considered to be well balanced for the combination of VSD and moderate pulmonary valve stenosis, and for that reason, the patient was followed up regularly, with a plan for future consideration for anatomical repair.

From 6 weeks of age (weighing 6 kg), the infant had progressive acute episodes of respiratory distress with cough, wheeze, tachypnea, and severe desaturation. He was admitted to the emergency department during one of these respiratory events and underwent further investigation. During the hospital stay, he had a prolonged obstructive respiratory event requiring intubation and mechanical ventilation. After intubation, he continued to deteriorate with repeated airways obstruction if positive pressure was reduced. A laryngotraceobronchoscopy showed that airways could not be opened even with a positive end-expiratory pressure of 30 mm Hg, and there was extrinsic compression of the carina (Fig 2). Subsequent chest computed tomography scan showed aneurysmal branch PAs compressing the carina with a complete occlusion of proximal left bronchus and partial occlusion of right bronchus (Fig 3). Indications were that the degree of pulmonary valve stenosis was not as severe as previously thought, and that the high flow and hypertensive PAs were obstructing the airway, exacerbated by their posterior position in the mediastinum.

The surgical repair consisted of a double switch operation with Lecompte maneuver. The atrium was opened as for a Senning procedure [3]. Then the VSD was closed transatrially with a Dacron (C. R. Bard, Haverhill, PA) patch, and the arterial switch was performed as routinely done for TGA with particular attention to the Lecompte maneuver. Then, the Senning procedure was completed using a small patch of pulmonary homograft to augment the outer layer [3]. The patient was easily weaned from cardiopulmonary bypass, and the chest was left open for 48 hours. There was mild neoaoic stenosis, with a peak Doppler velocity of just under 2 ms⁻². He made a straightforward postoperative recovery without obstructive respiratory events, and postoperative laryngotraceobronchoscopy demonstrated widely patent airways with no signs of compression (Fig 2C). He was discharged home 20 days after the operation in good clinical condition, saturating 100% in room air with no respiratory symptoms. Nine months after the operation, the patient is stable, growing well, and free of respiratory problems. His last echocardiogram showed good biventricular function with no stenosis of the Senning pathways and mild neoaoic stenosis.

Comment
This presentation of ccTGA with obstructive airways compression has not previously been reported. Initially it was thought that the cardiac physiology and circulation were well balanced owing to the association between an unrestrictive VSD and moderate pulmonary stenosis. However, the high flow resulted in dilation of the branch PAs, resulting in airways compression in a similar clinical presentation usually associated absent pulmonary valve and aneurysmal dilation of pulmonary arteries [4]. The condition was exacerbated in this case by the unrecognized high pressure in the branch PAs and

Accepted for publication Nov 19, 2012.

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http://dx.doi.org/10.1016/j.athoracsur.2012.11.060
their proximity to the airways due to the transposition arrangement of the great vessels. The trachea and main bronchi appeared to be of normal caliber and morphology, and as soon as the extrinsic compression was released, they reverted to normal function.

The Lecompte maneuver has been very successful in relieving airway compression in the setting of absent pulmonary valve syndrome [4]. Thus, the attraction of the double switch operation in this setting was that it could achieve complete repair of the patient’s congenital heart disease.
defect and relieve the airways obstruction at the same time. The Lecompte maneuver with or without PA plication or competent right ventricular outflow tract conduit implantation represents the treatment for relief of airway compression due to dilated pulmonary arteries [4]. An alternative approach could have been to perform the Lecompte maneuver and place a limiting left ventricle to PA conduit, but that would have sacrificed the pulmonary (neoaortic) valve and required a ventriculotomy. Although the double switch is a complex procedure, we have previously shown that it can be performed in infants and in high-risk situations such as this with an actuarial survival at 10 years of 83.9% [5]. Similar results for double switch operation are reported from other centers, and it can be considered an optimal procedure to treat symptomatic ccTGA [6, 7]. This case report describes a very rare association of anomalies for ccTGA and shows the benefit of the Lecompte maneuver in bringing aneurysmal PAs away from the major airways in infants with extrinsic compression.

References

Fatty Infiltration of an Aortic Valve
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Adipose tissue is a normal constituent of the heart, but not a normal anatomic finding of cardiac valves. Fatty infiltration of the aortic valve is rare, with unknown significance on valve function. We report a case of fatty infiltration and replacement of the spongiosa layer in an incompetent aortic valve. The mechanism of fat infiltration is unknown, but may be explained by differentiation of preexisting valve interstitial cells secondary to valvular injury.

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Accepted for publication Dec 31, 2012.
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