Bronchial decompression following repair of absent pulmonary valve: Fine-tuning by procedural fiberoptic bronchoscopy

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INTRODUCTION
Tetralogy of Fallot is a relatively common congenital cardiac lesion and approximately 3% to 6% of these patients have an absent pulmonary valve (APV). Patients with tetralogy of Fallot and an APV in utero, develop marked aneurysmal dilation of the main pulmonary artery and its branches resulting in the extrinsic compression of the trachea and bronchi leading to tracheomalacia and bronchomalacia. These patients require in addition to an intracardiac repair of the tetralogy of Fallot, additional surgical strategies for reduction of bronchial obstruction like downsizing of pulmonary arteries, translocation of the pulmonary arteries anterior to the aorta and away from the tracheobronchial tree, excision of nearly all aneurysmal pulmonary arteries and placement of a pulmonary homograft/valved conduit. In this case report we explore what other procedures may help to manage the problem of airway compromise persisting despite the standard methods already described. An infant with 1q21.1 chromosomal microdeletion presented with a tetralogy of Fallot complicated by absence of the pulmonary valve. The vital role played by real time intraoperative fiberoptic bronchoscopy in the successful achievement of airway decompression that otherwise would have contributed to postoperative respiratory morbidity is highlighted.

Real time fiberoptic bronchoscopy initially identified the residual right bronchial compression after separation from cardiopulmonary bypass. Then bronchoscopy displayed the limited extent to which right bronchial decompression was achieved following the pulmonary arteriopexy. Subsequently when aortopexy was done, bronchoscopy

ABSTRACT
Marked aneurysmal dilation of the central and branch pulmonary arteries in utero in patients with tetralogy of Fallot with absent pulmonary valve can often exhibit extrinsic compression of the trachea and bronchi. The major morbidity in these patients remains postoperative ventilation issues. This case report highlights the role of intraoperative bronchoscopy in providing guidance for obtaining optimal bronchial decompression that was achieved by an initial pulmonary arteriopexy followed by an aortopexy.

Keywords: Airway management, bronchoscopy, congenital heart disease, infant pulmonary valve insufficiency, tetralogy of Fallot

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confirmed the achievement of the optimal bronchial decompression. The institutional ethical committee approval [SRC#CR6#2020] as well as informed consent from the parents was obtained to publish the case report.

**CASE HISTORY**

A four-month-old boy [weight: 6 kgs; height: 64 cms; Body Surface Area: 0.3 m²], known to have tetralogy of Fallot with APV in combination with large branch pulmonary arteries obstructing the right and left bronchi was referred to the authors institution for urgent surgery. He had recurrent hospital admissions with shortness of breath and stridor. He was known to have 1q21.1 chromosomal microdeletion.

The preoperative transthoracic echocardiography showed an ostium secundum atrial septal defect [5 mm], a large malalignment-type ventricular septal defect [7 mm] with overriding of aorta and with a hypoplastic conal septum, dilated right ventricle with heavily trabeculated outflow tract and an APV. The peak pressure gradient across the right ventricular outflow tract was 80 mmHg and there was severe pulmonary regurgitation. The pulmonary arteries were grossly dilated [main pulmonary artery: 20 mm; right pulmonary artery: 15 mm and left pulmonary artery: 10 mm] with no patent ductus arteriosus. A computerized tomography of the chest showed significant dilatation of main pulmonary arteries, significant narrowing of bilateral main, lobar, and segmental bronchi at the level of the hilum [Figure 1a]. The child was posted for an urgent surgery.

Keeping in mind the possibility that anesthesia introduction could result in airway compromise, the heart lung machine was primed and kept ready to institute emergency cardiopulmonary bypass support in case a life-threatening situation due to inability to ventilate/oxygenate the patient arose. After administration of general anesthesia without any airway compromise, a fiberoptic bronchoscopy was done that showed bilateral bronchial narrowing [80% pulsatile narrowing in the right and 50% in the left bronchus during systole]. The child underwent intracardiac repair on hypothermic cardiopulmonary bypass. The ventricular septal defect was closed with a bovine pericardial patch [Edwards Lifesciences Corp. Irvine, CA, USA]. A 14 mm glutaraldehyde‑preserved valve‑containing bovine jugular vein graft [Contegra, Medtronic Inc., Minneapolis MN, USA] was used to reconstruct the right ventricular outflow tract along with pulmonary artery reduction arterioplasty. The child was separated from cardiopulmonary bypass and a fiberoptic bronchoscopy was done using a 2.8 mm intubation fiberscope [11301 AA1, Karl Storz, Tuttlingen, Germany]. The lower trachea and the left main bronchus appeared wide with no external compression. The right bronchus appeared narrowed due to a pulsatil external mass [Figure 2a]. A right pulmonary arteriopexy was done by suturing the anterior aspect of the right pulmonary artery to the aorta to lift the pulmonary artery anteriorly reducing the external compression on the right bronchus [Figure 2b and Videoclip 1]. As it was felt that the bronchial decompression was not optimal, an aortopexy was done under bronchoscopy guidance by suturing the aorta to the undersurface of the sternum. This helped further open up the right bronchus [Figure 2c]. The patient was shifted to the intensive care unit for further hemodynamic and ventilatory management. The child’s trachea was extubated on the 5th postoperative day successfully and he was discharged home on the 12th postoperative day. A computerized tomography done during the 6th week following surgery revealed a complete relief of the previously noted narrowing of the distal trachea as well as narrowing of the proximal right and left mainstem bronchi [Figure 1b]. The child was doing well with no airway compromise at the 6 months follow up.

**DISCUSSION**

The successful management of an infant with tetralogy of Fallot and an APV is described. The child had repeated hospitalizations due to airway problems and resolving the airway-related issues was one of the top priorities in this child in addition to the surgical repair of the lesions. Real time intraoperative fiberoptic bronchoscopy played a major role in achieving this goal by providing the surgeon constant guidance. Initially the bronchoscopy could display the extent of right bronchial compression after separation of the child from cardiopulmonary bypass.

Bronchoscopy readily displayed the bronchial lumen opening up after the first stage pulmonary arteriopexy. Subsequently it was felt that the bronchial lumen could further be decompressed and hence the aorta was attached.
to the undersurface of the sternum in the form of an aortopexy. Bronchoscopy guided the surgeon to achieve the best possible lumen with the optimal tension to be exerted on the sutures used for arteriopexy and the aortopexy thereby avoiding hemodynamic compromise. Computerized tomography that was done a month post-operatively demonstrated that the technique is effective and the right bronchus showed adequate patency thereby avoiding any surgical or cardiological re-intervention.

Tracheobronchial decompression by suspension of the pulmonary artery to the retrosternal fascia in addition to an aneurysmorrhaphy in a patient with tetralogy of Fallot with APV was described earlier. But pulmonary arteriopexy with aortopexy based on intraoperative bronchoscopy findings as was done in this current child was not reported to the best of the authors’ knowledge.

In conclusion, in patients with tetralogy of Fallot with APV, real time intraoperative bronchoscopy can be a valuable tool in gaining an optimal airway patency during the primary repair of this complex cardiac lesion.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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