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Brief Report

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Management of post-operative ventilator dependency in an operated case of tetralogy of Fallot with absent pulmonary valve: a team approach involving cardiac surgeon, cardiologist, intensivist, and radiologist

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Abstract

Tetralogy of Fallot with absent pulmonary valve syndrome is commonly associated with trachea-bronchial anomalies, possibly due to airway compression caused by massively dilated pulmonary arteries secondary to severe pulmonary regurgitation. This airway obstruction may persist post-operatively also. We report a case of an infant who required a series of management strategies including bronchial stenting to manage his obstructive symptoms.

Tetralogy of Fallot with absent pulmonary valve syndrome represents an extreme form of tetralogy of Fallot where pulmonary insufficiency and annular stenosis often results in massive pulmonary artery dilatation with consequent compression of adjacent trachea and bronchi, leading to **airway obstructive symptoms**.[1–3] Reduction pulmonary artery plasty is aimed at reducing pulmonary artery sizes with subsequent relief from airway obstruction, with outcomes essentially determined by airway status and its management.[3–6] In our case, **prolonged airway collapse and recurrent pneumothorax** required a series of imaging tests and novel management strategies like use of **biodegradable stent** to relieve airway obstruction and **digital suction devices** to manage air leaks.

Case description

A **5-month**-old boy baby with history of **recurrent pneumonia and feeding difficulty** was hospitalised in view of gradually worsening tachypnoea. Echocardiography showed tetralogy of Fallot with absent pulmonary valve syndrome with **hugely dilated pulmonary arteries (20 mm each)**. CT pulmonary angiography showed absent pulmonary valve with aneurysmal dilatation of pulmonary arteries and their lobar divisions, causing anteroposterior compression of the proximal lobar bronchi (Fig 1) along with large areas of air trapping in both upper lobes and superior segments of both lower lobes likely due to compression by aneurysmal lobar branches.

He underwent **total correction with valved conduit plus reduction pulmonary artery plasty**. However, he had a torrid post-operative course with **prolonged ventilator dependency and recurrent extubation** failures. Initial evaluation in post-operative course showed no residual defects with normal cardiac function. Ventilator dependency persisted despite treating the usual suspects including post-operative pneumonia with aggressive IV antibiotics, intermittent bilevel positive airway pressure and rigorous chest physiotherapy, and diaphragm palsy by **diaphragmatic plication**.

HRCT chest was done in view of serial chest X-rays showing collapsed left lung, which revealed narrowed left main bronchus which was subsequently confirmed by direct bronchoscopy. Confirmatory fluoroscopic bronchography showed **significant long segment left bronchial compression** with lack of dye uptake due to extrinsic compression by aneurysmal lobar pulmonary arteries. Patient continued to have recurrent pneumothorax secondary to air trapping

Intervention

He underwent endobronchial stenting of left main bronchus (Fig 2a and b) on post-operative day 6. A **biodegradable angioplasty stent (9 * 19 mm)** was put under direct bronchoscopy by both paediatric cardiology and interventional radiology teams. Post-procedure, his airway pressures dramatically reduced from 50 cmH2O to 23 cmH2O with subsequent rapid improvement in clinical status and chest X-ray. We also had to put multiple bilateral chest tubes to treat recurrent bilateral pneumothorax.

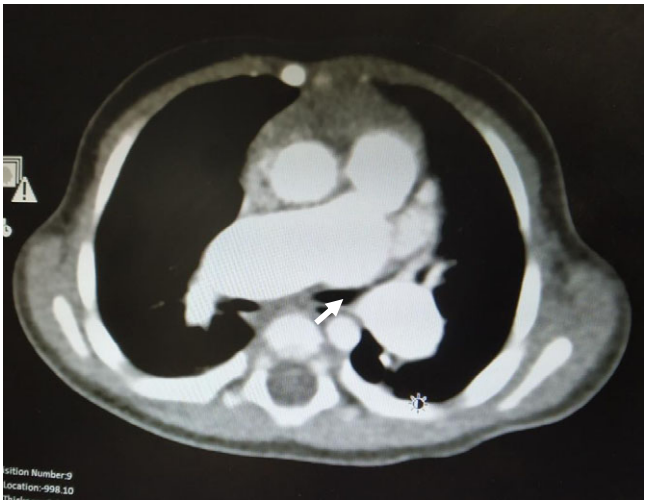

Figure 1. Axial CT pulmonary angiography image demonstrating enlarged right and left pulmonary arteries with aneurysmal left lobar artery causing left main bronchial compression (white arrow).

We used **digital suction devices with regulated negative pressure suction based on the patient's air leak** which resulted in suction being delivered only when pleural pressure reached the physiological limit. Patient responded to the above measures and was ventilator-free by 8th post-operative day and chest tube free by 21st post-operative day. Patient was discharged on room air and continued remaining symptom-free till 6 months of follow-up. Then, he was lost to follow-up.

Discussion

Our in-hospital data of last 10 years show that out of total 10 cases operated for tetralogy of Fallot with absent pulmonary valve syndrome, age ranging from 1 month to 46 years, airway complications, and overall complications were mainly seen in infancy age group, though most of which responded to usual measures.

In this case, we had to resort to novel measures in airway management in view of prolonged and complicated clinical course. In previous studies, metallic bronchial stenting in cases of tetralogy of Fallot with absent pulmonary valve syndrome has been reported. Metallic stents are found to be associated with excessive granulation tissue growth and airway perforation while silicon stents are fraught with risk of migration, impaired mucociliary clearance, and mucous plugging at the ends of the stent.[7–8]

We used bioabsorbable scaffold with sirolimus-eluting properties with subsequent lesser risk of granulation tissue formation and lower strut thickness which supports early endothelisation and minimises injury.[9] Endobronchial stenting for extrinsic compression is a low-risk procedure associated with good short-term outcome, yet long-term outcomes are still unknown. Also, the stenting does not relieve distal bronchial compression.

We also used Medela low-pressure suction devices which provide measurable air leak data and apply suction only when required; hence, patient is not harmed by continuous suction. They also hasten chest tube removal; thus, enabling early mobilisation and reducing the number of chest X-rays are ultimately cost effective to the patient. [10]

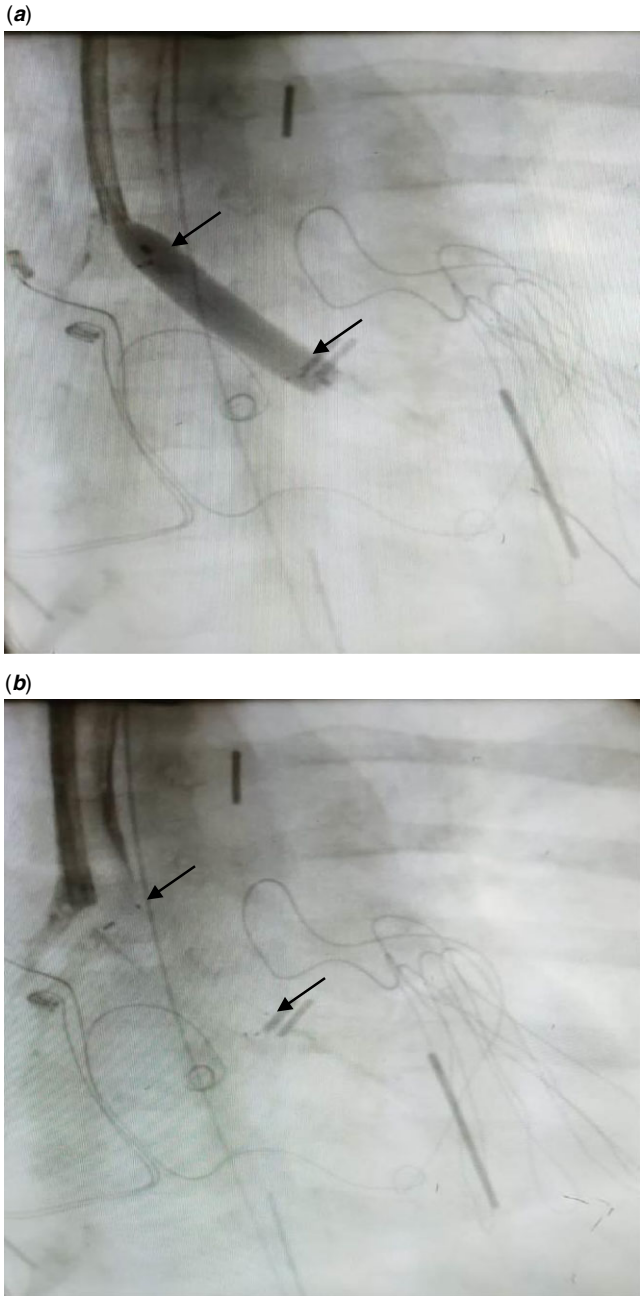

Figure 2. (***a*** and ***b***): Fluoroscopic bronchography done in AP view demonstrating successful endobronchial stenting of left main bronchus, shown by markings (black arrows) on the both ends of stent length with complete reopening of the collapsed left main bronchus.

Learning point

- Anticipate complicated airway management issues and prolonged post-operative course in a case of tetralogy of Fallot with absent pulmonary valve syndrome.
- State-of-the-art management strategies including use of bronchial stenting and low-pressure digital suction devices to be considered early to treat difficult scenarios.
- A close coordination amongst cardiac surgeon, cardiologist, intensivist, and radiologist is imperative in the real-time management of this complex anomaly.

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