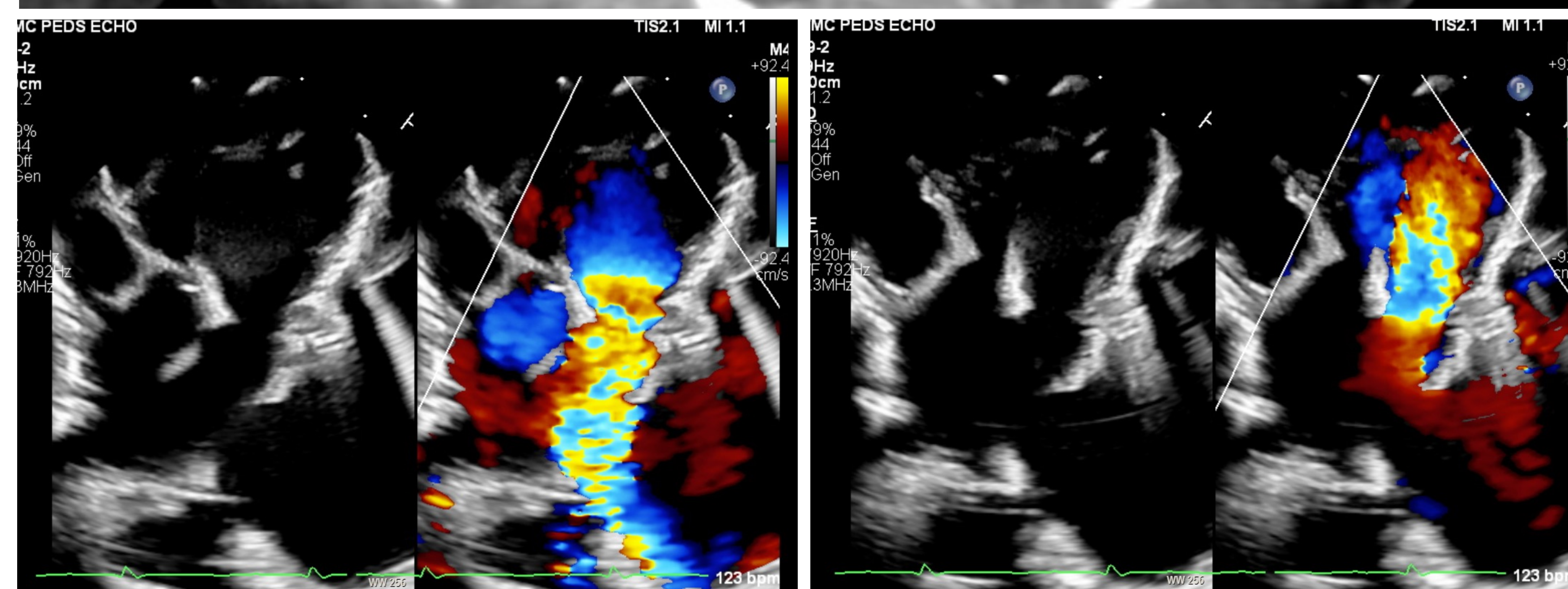
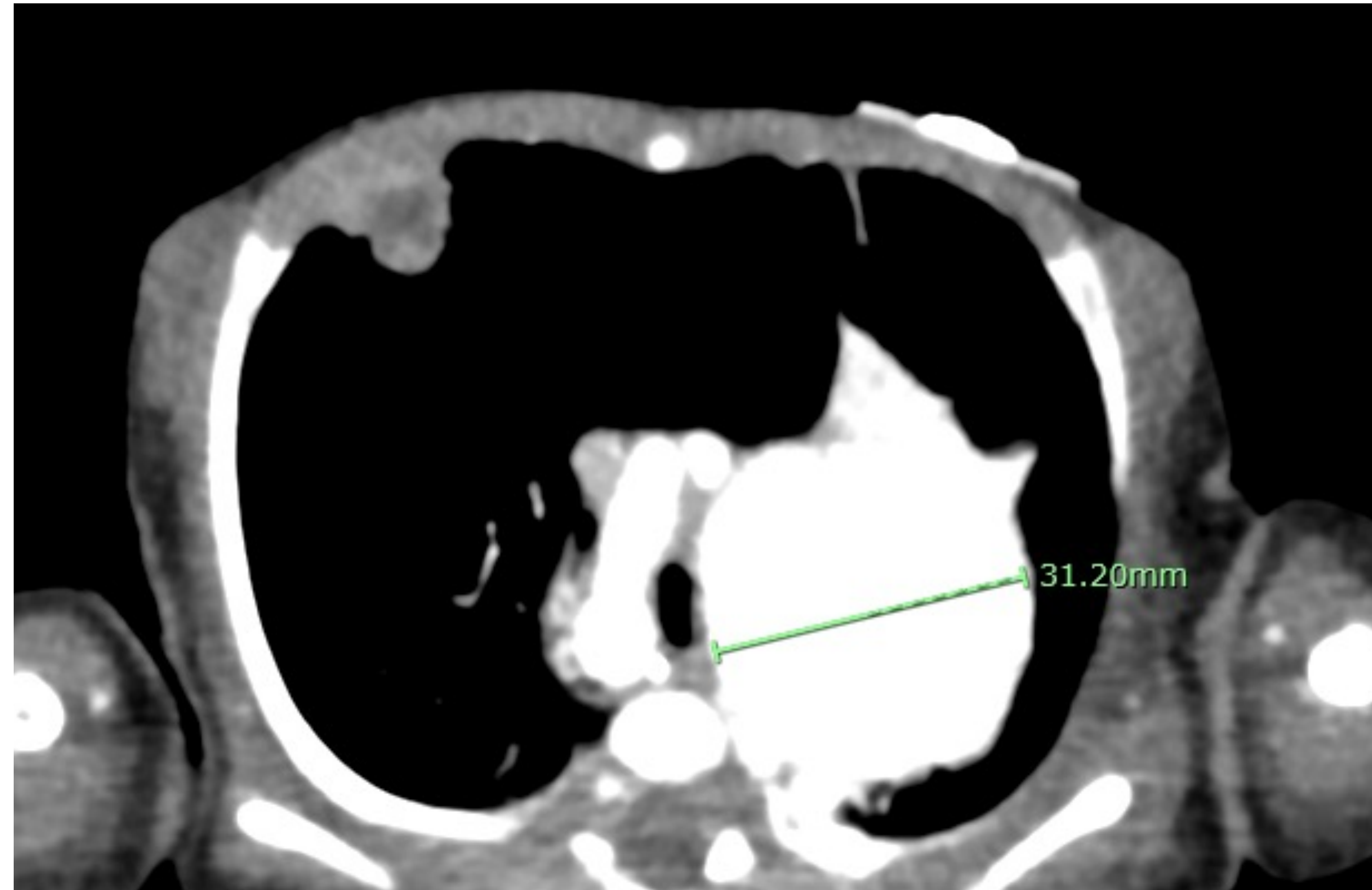


BACKGROUND

- Tetralogy of Fallot with absent pulmonary valve (TOF/APV) is a rare congenital defect associated with high morbidity and mortality.
- Lack of pulmonary valve cusps leads to extreme pulmonary regurgitation as well as aneurysmal dilation of the main and central branch pulmonary arteries, and eventually airway compression.
- We describe an extreme case of Tetralogy of Fallot (TOF) with absent pulmonary valve syndrome (APVS) and its management.

CASE DESCRIPTION

- 8-week-old male infant presented as a transfer from an overseas institution with TOF/APV.
- He required high PEEP with modest pressure control settings, however, weaning off respiratory support not achieved
- CT chest and TTE showed severely dilated branch pulmonary arteries, left > right, and severe compression of the left lung parenchyma and airways.
- Bronchoscopy was limited as there was severe obstruction to both main bronchi. Surgical repair of TOF took place.
- Patient arrived to the OR intubated and was kept spontaneously breathing on sevoflurane throughout bronchoscopy. Intraoperatively, special attention was applied to prevent ↑PVR and to avoid reversing L → R shunting (no ↑ in PEEP and avoidance of hypercarbia and acidosis).
- On post-operative bronchoscopy, much more patent main bronchi but still with abnormal flattened shape.
- Patient had delayed chest closure and was discharged with a tracheostomy.



Pulmonic Valve in Systole

Pulmonic Valve in Diastole

DISCUSSION

- Tetralogy of Fallot with absent pulmonary valve leads to extremely severe pulmonary regurgitation and aneurysmal dilation of the main and central branch pulmonary arteries and thus can result in tracheobronchial compression.
- Right ventricular dysfunction independently predicts overall mortality.
- TOF/APV can present as a spectrum of symptoms, from mild respiratory distress during infancy to severe postnatal cardiopulmonary symptoms, or even fetal hydrops.
- Management of mild symptoms include oxygen therapy, non-invasive PPV, and medical management.
- Moderate symptoms may require prone position to alleviate airway compression, or intubation and mechanical ventilation
- Severe obstructive symptoms may require ECMO until surgical repair.
- Special anesthesia considerations during surgical repair include airway management due to bronchial compression and careful monitoring and management of PVR

CONCLUSIONS

Although it is rare, Tetralogy of Fallot with absent pulmonary valves is a significant cause of cardio-respiratory failure in neonates and infants. The severity of the symptoms dictates early treatment modalities with medical and surgical approaches.

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