

# CHOA Delivery Room Protocol

## Tetralogy of Fallot-Absent Pulmonary Valve (TOF-APV)

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Children's  
Healthcare of Atlanta

Tetralogy of Fallot-Absent pulmonary valve (TOF-APV) is a rare subtype of TOF. In TOF-APV, the pulmonary valve leaflets are absent or dysplastic, leading to severe regurgitation across the valve and severe dilation of the main and branch pulmonary arteries. Additional unique features of TOF-APV include right ventricular dilation—a result of pulmonary regurgitation—and absence of the ductus arteriosus in most cases. Right ventricular outflow tract obstruction (RVOTO) can also still occur at the valvar level. Importantly, pulmonary artery dilation—which can be massive—can externally compress the trachea/ bronchi, which can produce mechanical airway obstruction. In utero, this can lead to lung hypoplasia or air /fluid trapping in the lungs due to a ball-valve effect. Postnatally, this can lead to airway obstruction, bronchomalacia, and air-trapping in the lungs. Airway obstruction is often the most critical clinical issue facing these newborns.

### Prenatal clinical checklist:

1. Is there ventricular dysfunction or hydrops noted on fetal echocardiography?
2. Is there evidence of airway obstruction/fluid trapping or lung hypoplasia on fetal MRI?

### Goals for delivery room and transport:

Assessment of degree of hypoxia and respiratory distress. Hypoxia can occur due to airway obstruction, but also due to the effects of elevated pulmonary vascular resistance (PVR), which can worsen pulmonary regurgitation and reduce right ventricular output/pulmonary blood flow.

Clinical goals in the DR/during transport include:

- Securing airway for ventilation
- Rapidly dropping PVR with 100% FiO<sub>2</sub> and iNO if clinically necessary
- Minimize oxygen consumption with sedation/paralysis
- Support systemic perfusion

### Access plan:

1. Low-lying single lumen 5 French UVC—XR confirmation of position unnecessary if blood return documented
2. Peripheral IV if able to place in timely fashion.

### Medication plan:

Have the following available in the delivery room:

1. D10W at 60 ml/kg/day (80 ml/kg/day: maternal DM, IUGR, preterm, or low DS)
2. Medications to aid intubation OR for sedation and reduction in metabolic demands:
  - a. Fentanyl IV 1 mcg/kg/dose (fentanyl may be given rapidly if immediately followed by vecuronium)
  - b. Vecuronium IV 0.1 mg/kg/dose or Rocuronium 1 mg/kg dose for neuromuscular blockade
3. Rescue medications to have available, not necessarily drawn up:
  - a. IV atropine (0.02 mg/kg/dose) for bradycardia
  - b. IV epinephrine (0.02 mg/kg/dose) for circulatory collapse
4. Inotropic support (if concern for fetal hydrops or severe ventricular dysfunction):
  - a. Epinephrine IV infusion (0.02 mcg/kg/min)
  - b. Dopamine IV infusion (start at 5 mcg/kg/min, may escalate to 10 mcg/kg/min) when epinephrine unavailable

### Equipment/supplies:

1. Recommend cuffed endotracheal tube for term infants
2. Blood pressure cuff
3. Pulse-oximetry monitoring

### Delivery room management plan:

1. Delayed cord clamping per institutional protocol. May not be feasible in setting of fetal distress.
2. Follow NRP guidelines for initial resuscitation. Supplemental oxygen may be used
3. Saturations should be >75% by 10 min. Desaturation more likely due to airway/pulmonary than cardiac. If initiating respiratory support, would escalate FiO<sub>2</sub> to 100% to augment pulmonary blood flow.
4. If infant in respiratory distress, place infant in prone position, which can relieve the compression on the bronchi from the pulmonary arteries
5. If hypoxemia is not improved or respiratory distress is not stabilized, proceed to intubation
  - a. If intubating, recommend providing sedation and paralysis to both facilitate intubation and decrease metabolic demand post intubation
6. Given the risk of air trapping with bagging via mask or ETT, use low pressures (PIP) and a short inspiratory to expiratory ratio to facilitate exhalation. Recommend a PEEP of 10 mmHg.
7. If poor systemic perfusion or hemodynamic compromise, the infant may need to be started on inotropy
  - a. Recommend epinephrine as first line, 0.02 mcg/kg/min
  - b. Dopamine infusion (per medication plan) also appropriate if epinephrine unavailable

### Post delivery room strategies:

Ongoing ventilatory strategies:

- High PEEP low TV to avoid air trapping
- Insure full exhalation of every ventilator breath
- Prone positioning
- Consider HFOV if ongoing issues with oxygenation and ventilation
- FiO<sub>2</sub> of 1.0 and iNO to promote pulmonary blood flow
- Sedation with neuromuscular blockade while mechanically ventilated