Single Ventricle: Recommendations for the Treatment of Postoperative Stage II (Bidirectional Glenn/Hemi-Fontan) and Stage III (Fontan) Palliation Patients

2020 (New): For patients in a prearrest state with superior cavopulmonary anastomosis physiology and severe hypoxemia due to inadequate pulmonary blood flow (Qp), ventilatory strategies that target a mild respiratory acidosis and a minimum mean airway pressure without atelectasis can be useful to increase cerebral and systemic arterial oxygenation.

2020 (New): ECLS in patients with superior cavopulmonary anastomosis or Fontan circulation may be considered to treat low DO_2 from reversible causes or as a bridge to a ventricular assist device or surgical revision.

Why: Approximately 1 in 600 infants and children are born with critical congenital heart disease. Staged surgery for children born with single ventricle physiology, such as hypoplastic left heart syndrome, spans the first several years of life.[15] Resuscitation of these infants and children is complex and differs in important ways from standard PALS care. Previous PALS guidelines did not contain recommendations for this specialized patient population. These recommendations are consistent with the 2018 AHA scientific statement on CPR in infants and children with cardiac disease.[14]

Pulmonary Hypertension

2020 (Updated): Inhaled nitric oxide or prostacyclin should be used as the initial therapy to treat pulmonary hypertensive crises or acute right-sided heart failure secondary to increased pulmonary vascular resistance.

2020 (New): Provide careful respiratory management and monitoring to avoid hypoxia and acidosis in the postoperative care of the child with pulmonary hypertension.

2020 (New): For pediatric patients who are at high risk for pulmonary hypertensive crises, provide adequate analgesics, sedatives, and neuromuscular blocking agents.

2020 (New): For the initial treatment of pulmonary hypertensive crises, oxygen administration and induction of alkalosis through hyperventilation or alkali administration can be useful while pulmonary-specific vasodilators are administered.

2020 (New): For children who develop refractory pulmonary hypertension, including signs of low cardiac output or profound respiratory failure despite optimal medical therapy, ECLS may be considered.

2010 (Old): Consider administering inhaled nitric oxide or aerosolized prostacyclin or analogue to reduce pulmonary vascular resistance.

Why: Pulmonary hypertension, a rare disease in infants and children, is associated with significant morbidity and mortality and requires specialized management. Previous PALS guidelines did not provide recommendations for managing pulmonary hypertension in infants and children. These recommendations are consistent with guidelines on pediatric pulmonary hypertension published by the AHA and the American Thoracic Society in 2015,[16] and with recommendations contained in a 2020 AHA scientific statement on CPR in infants and children with cardiac disease.[14]

Neonatal Life Support

There are over 4 million births every year in the United States and Canada. Up to 1 of every 10 of these newborns will need help to transition from the fluid-filled environment of the womb to the air-filled room. It is essential that every newborn have a caregiver dedicated to facilitating that transition and for that caregiver to be trained and equipped for the role. Also, a significant proportion of newborns who need facilitated transition are at risk for complications that require additional trained personnel. All perinatal settings should be ready for this scenario.

The process of facilitating transition is described in the Neonatal Resuscitation Algorithm that starts with the needs of every newborn and proceeds to steps that address the needs of at-risk newborns. In the 2020 Guidelines, we provide recommendations on how to follow the algorithm, including anticipation and preparation, umbilical cord management at delivery, initial actions, heart rate monitoring, respiratory support, chest compressions, intravascular access and therapies, withholding and discontinuing resuscitation, postresuscitation care, and human factors and performance. Here, we highlight new and updated recommendations that we believe will have a significant impact on outcomes from cardiac arrest.

Summary of Key Issues and Major Changes

- Newborn resuscitation requires anticipation and preparation by providers who train individually and as teams.
- Most newly born infants do not require immediate cord clamping or resuscitation and can be evaluated and monitored during skin-to-skin contact with their mothers after birth.
- Prevention of hypothermia is an important focus for neonatal resuscitation. The importance of skin-to-skin care in healthy babies is reinforced as a means of promoting parental bonding, breastfeeding, and normothermia.

- Inflation and ventilation of the lungs are the priority in newly born infants who need support after birth.
- A rise in heart rate is the most important indicator of effective ventilation and response to resuscitative interventions.
- Pulse oximetry is used to guide oxygen therapy and meet oxygen saturation goals.
- Routine endotracheal suctioning for both vigorous and nonvigorous infants born with meconium-stained amniotic fluid (MSAF) is not recommended. Endotracheal suctioning is indicated only if airway obstruction is suspected after providing positive-pressure ventilation (PPV).
- Chest compressions are provided if there is a poor heart rate response to ventilation after appropriate ventilation-corrective steps, which preferably include endotracheal intubation.
- The heart rate response to chest compressions and medications should be monitored electrocardiographically.
- When vascular access is required in newly born infants, the umbilical venous route is preferred. When IV access is not feasible, the IO route may be considered.
- If the response to chest compressions is poor, it may be reasonable to provide epinephrine, preferably via the intravascular route.
- Newborns who fail to respond to epinephrine and have a history or an exam consistent with blood loss may require volume expansion.
- If all these steps of resuscitation are effectively completed and there is no heart rate response by 20 minutes, redirection of care should be discussed with the team and family.

Major New and Updated Recommendations

Anticipation of Resuscitation Need

2020 (New): Every birth should be attended by at least 1 person who can perform the initial steps of newborn resuscitation and initiate PPV and whose only responsibility is the care of the newborn.

Why: To support a smooth and safe newborn transition from being in the womb to breathing air, every birth should be attended by at least 1 person whose primary responsibility is to the newly born and who is trained and equipped to begin PPV without delay. Observational and quality-improvement studies indicate that this approach enables identification of at-risk newborns, promotes use of checklists to prepare equipment, and facilitates team briefing. A systematic review of neonatal resuscitation training in low-resourced settings showed a reduction in both stillbirth and 7-day mortality.

Temperature Management for Newly Born Infants

2020 (New): Placing healthy newborn infants who do not require resuscitation skin-to-skin after birth can be effective in improving breastfeeding, temperature control, and blood glucose stability.

Why: Evidence from a Cochrane systematic review showed that early skin-to-skin contact promotes normothermia in healthy newborns. In addition, 2 meta-analyses of RCTs and observational studies of extended skin-to-skin care after initial resuscitation and/or stabilization showed reduced mortality, improved breastfeeding, shortened length of stay, and improved weight gain in preterm and low-birth-weight babies.

Clearing the Airway When Meconium Is Present

2020 (Updated): For nonvigorous newborns (presenting with apnea or ineffective breathing effort) delivered through MSAF, routine laryngoscopy with or without tracheal suctioning is not recommended.

2020 (Updated): For nonvigorous newborns delivered through MSAF who have evidence of airway obstruction during PPV, intubation and tracheal suction can be beneficial.

2015 (Old): When meconium is present, routine intubation for tracheal suction in this setting is not suggested because there is insufficient evidence to continue recommending this practice.

Why: In newly born infants with MSAF who are not vigorous at birth, initial steps and PPV may be provided. Endotracheal suctioning is indicated only if airway obstruction is suspected after providing PPV. Evidence from RCTs suggests that nonvigorous newborns delivered through MSAF have the same outcomes (survival, need for respiratory support) whether they are suctioned before or after the initiation of PPV. Direct laryngoscopy and endotracheal suctioning are not routinely required for newborns delivered through MSAF, but they can be beneficial in newborns who have evidence of airway obstruction while receiving PPV.

Vascular Access

2020 (New): For babies requiring vascular access at the time of delivery, the umbilical vein is the recommended route. If IV access is not feasible, it may be reasonable to use the IO route.

Why: Newborns who have failed to respond to PPV and chest compressions require vascular access to infuse epinephrine and/or volume expanders. Umbilical venous catheterization is the preferred technique in the delivery room. IO access is an alternative if umbilical venous access is not feasible or care is being provided outside of the delivery room. Several case reports have described local complications associated with IO needle placement.