**WORKSHEET for Evidence-Based Review of Science for Emergency Cardiac Care**

**Worksheet author(s)**
Marino, Bradley S  

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**Clinical question.**

Ped-055B For infants and children with Fontan or hemi-Fontan/bidirectional Glenn (BDG) circulation who require resuscitation from cardiac arrest or pre-arrest states (pre-hospital [OHCA] or in-hospital [IHCA]) (P), does any specific modification to standard practice (I) compared with standard resuscitation practice (C) improve outcome (O) (e.g. ROSC, survival to discharge, survival with good neurologic outcome)?

**Is this question addressing an intervention/therapy, prognosis or diagnosis?** Intervention/therapy.

**State if this is a proposed new topic or revision of existing worksheet:** This is a proposed new topic.

**Conflict of interest specific to this question**

Do any of the authors listed above have conflict of interest disclosures relevant to this worksheet? No.

**Search strategy (including electronic databases searched).**

PubMed MeSH search using the following [MeSH] AND [Title/Abstract] criteria:

- "cardiopulmonary resuscitation"[MeSH] AND "univentricular heart"[Title/Abstract]
- "cardiopulmonary resuscitation"[MeSH] AND "hemi-fontan"[Title/Abstract]
- "cardiopulmonary resuscitation"[MeSH] AND "cavopulmonary anastomosis"[Title/Abstract]
- "cardiopulmonary resuscitation"[MeSH] AND "glenn"[Title/Abstract]
- "cardiopulmonary resuscitation"[MeSH] AND "bidirectional glenn"[Title/Abstract]
- "cardiopulmonary resuscitation"[MeSH] AND "cavopulmonary connection"[Title/Abstract]
- "heart arrest"[MeSH] AND "univentricular heart"[Title/Abstract]
- "heart arrest"[MeSH] AND "fontan"[Title/Abstract]
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- "heart arrest"[MeSH] AND "glenn"[Title/Abstract]
- "heart arrest"[MeSH] AND "bidirectional glenn"[Title/Abstract]
- "advanced cardiac life support"[MeSH] AND "univentricular heart"[Title/Abstract]
- "advanced cardiac life support"[MeSH] AND "fontan"[Title/Abstract]
- "advanced cardiac life support"[MeSH] AND "hemi-fontan"[Title/Abstract]
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- "advanced cardiac life support"[MeSH] AND "bidirectional glenn"[Title/Abstract]
- "advanced cardiac life support"[MeSH] AND "single ventricle"[Title/Abstract]
- "advanced cardiac life support"[MeSH] AND "superior cavopulmonary connection"[Title/Abstract]
- "advanced cardiac life support"[MeSH] AND "superior cavopulmonary anastomosis"[Title/Abstract]
- "advanced cardiac life support"[MeSH] AND "univentricular heart"[Title/Abstract]
- "advanced cardiac life support"[MeSH] AND "multiple ventricles"[Title/Abstract]
- "advanced cardiac life support"[MeSH] AND "single ventricle"[Title/Abstract]
- "advanced cardiac life support"[MeSH] AND "superior cavopulmonary connection"[Title/Abstract]
- "advanced cardiac life support"[MeSH] AND "superior cavopulmonary anastomosis"[Title/Abstract]
- "respiration, artificial"[MeSH] AND "univentricular heart"[Title/Abstract]
- "respiration, artificial"[MeSH] AND "fontan"[Title/Abstract]
- "respiration, artificial"[MeSH] AND "hemi-fontan"[Title/Abstract]
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- "respiration, artificial"[MeSH] AND "superior cavopulmonary anastomosis"[Title/Abstract]
- "respiration, artificial"[MeSH] AND "single ventricle"[Title/Abstract]
- "heart massage"[MeSH] AND "univentricular heart"[Title/Abstract]
- "heart massage"[MeSH] AND "fontan"[Title/Abstract]
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"heart massage"[MeSH] AND "superior cavopulmonary anastamosis"[Title/Abstract]
"heart bypass, right"[MeSH] AND "resuscitation"[Title/Abstract]
"heart bypass, right"[MeSH] AND "pediatric advanced cardiac life support"[Title/Abstract]
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-Cochrane Library, Ovid, Medline, Scopus, Google Scholar, ECC EndNote X Master Library

**State inclusion and exclusion criteria**

- Study must have some patients who have undergone Fontan or hemi-Fontan/BDG procedures
- Must include discussion of resuscitation techniques specific to Fontan or hemi-Fontan/BDG patient populations

**Number of articles/sources meeting criteria for further review:** 13 studies met criteria for further review. Of these studies, 2 were LOE 4 (no controls) and 11 were LOE 5 (not directly related).
## Summary of evidence

### Evidence Supporting Clinical Question

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### Evidence Neutral to Clinical question

<table>
<thead>
<tr>
<th>Level of evidence</th>
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### Level of evidence

- **A** = Return of spontaneous circulation
- **B** = Survival of event
- **C** = Survival to hospital discharge
- **D** = Intact neurological survival
- **E** = Other endpoint

*Italic* = Animal studies
Evidence Opposing Clinical Question

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REVIEWER'S FINAL COMMENTS AND ASSESSMENT OF BENEFIT / RISK:

Evidence from one case series and one case report indicated that ECMO and manual external abdominal compressions with closed chest cardiac compression may be useful in resuscitation of patients who have undergone single ventricle palliation. In the case series, Booth et al demonstrated that ECMO may be successfully utilized as a resuscitative strategy in single ventricle patients, although it was shown to be less successful in patients with BDG physiology than with Fontan physiology. BDG circulation was thought to be inadequately supported by ECMO secondary to cannulation strategy and specific issues relative to the cardio/cerebral/respiratory interaction in the SCPA patient. Elevated SVC pressure, from the presence of a venous cannula, combined with low systemic blood pressure in an SCPA patient during cardiac arrest puts patients at high risk for neurologic injury. Evidence from the case report by Tewari et al demonstrated that it may be beneficial to use manual external abdominal compressions with closed chest cardiac compression (CCCC) during cardiopulmonary resuscitation in patients with Fontan circulation. By administering periodic manual external abdominal compressions, flow into the lungs may increase as the chest relaxes. This may also decrease the backflow of blood during CCCC into the IVC secondary to the intrathoracic-extrathoracic pressure gradient created during CCCC.

Evidence from four physiological studies supported the use of increased CO\textsubscript{2} tensions and hypoventilation in pre-arrest situations. Hoskote et al found that hypoxemia may be reversed following the BCPA by utilizing higher PaCO\textsubscript{2}, while low PaCO\textsubscript{2} or alkalosis may be detrimental. Li et al demonstrated that oxygen transport may be improved through moderate hypercapnia in conjunction with respiratory acidosis. This effectively improved arterial oxygenation and reduced oxygen consumption and arterial lactate levels. Fogel et al showed that the cerebral CO\textsubscript{2} feedback loop dominates over the pulmonary feedback loop when in direct competition with one another. CO\textsubscript{2} plays a significant role in flow distribution, while O\textsubscript{2} has little impact, and increased CO\textsubscript{2} may improve cerebral oxygenation. Bradley et al demonstrated that hypoventilation may improve systemic oxygenation in patients after BCPA, as hypoventilation-induced hypercarbia increases cerebral, superior vena cava, and pulmonary blood flow by decreasing cerebral vascular resistance.

Evidence from two complimentary physiological studies demonstrated that hyperventilation may be harmful to single ventricle patients with Fontan or hemi-Fontan/BDG physiology. Bradley et al showed that hyperventilation significantly impairs oxygenation after BCPA, noting that this fall in oxygenation occurs despite a decrease in transpulmonary gradient. Bradley et al demonstrated this by showing that hyperventilation lowers arterial PCO\textsubscript{2}, thereby increasing cerebral vascular resistance and lowering cerebral, superior vena cava, and pulmonary blood flow. Mott et al found that hyperventilation caused a decrease in cerebral oxygenation, demonstrating that normo- or mild hypoventilation may be more successful at preserving cerebral oxygenation in BCPA patients than hyperventilation.

Evidence from two physiological studies supported the use of negative pressure ventilation (NPV) over intermittent positive pressure ventilation (IPPV) in single ventricle patients with Fontan or hemi-Fontan/BDG physiology. These studies demonstrated that NPV increases pulmonary blood flow by improving stroke volume and thereby enhancing cardiac output. In addition, Shekerdemian et al (1996) showed significant reduction in systemic and pulmonary vascular resistance with NPV.

Two physiological studies assessed the impact of high frequency ventilation on cardiovascular hemodynamics in the single ventricle population. Meliones et al supported the use of high frequency jet ventilation, showing that HFJV resulted in significant reduction in pulmonary vascular resistance and a 25% increase in cardiac index, at a reduced mean airway pressure. Kornecki et al found high frequency oscillation ventilation to have no effect on cardiac output or on pulmonary vascular resistance in children after Fontan operation.

In a case report, Tobias et al utilized MAST suit application on the lower extremities to provide venous compression ("autotransfusion") to improve pre-load to the single ventricle. MAST suit application increased RAP and augmented pulmonary
flow, which lessened the need for fluid administration. Tobias et al concluded that the phasic external compression of the lower extremities via the MAST suit may reduce morbidity associated with the Fontan procedure.

Acknowledgements:
Desmond Bohn, MB, BCh, BAO, FFARCS, FRCPC, LMCC
Emily Greenberg, BA

Citation List

Level 4. Supporting, Fair (no controls).
-Resuscitative techniques: extracorporeal membrane oxygenation (ECMO)
-Population: n=20; n=14 Fontan, n=6 BDG
-Outcome: successful decannulation, survival to discharge, alive at follow-up (median follow-up time=35 months)
-Results: 7 out of 14 Fontan patients survived to discharge and 5 were alive at follow-up. Out of the 6 BDG patients, only one patient survived and had adverse neurologic outcomes.

Level 5. Opposing, Good.
-Ventilatory techniques: sedated and paralyzed, hyperventilation by increasing ventilatory rate while keeping tidal volume constant
-Population: n=12 (all BCPA, median age 6.4 months, age range 6 to 32 months)
-Outcome: arterial blood gas, systemic oxygenation, transcranial Doppler
-Results: Hyperventilation resulted in decreases in systemic oxygen saturation, arterial PO$_2$, and transpulmonary gradient following bidirectional superior cavopulmonary connection procedures.

Level 5. Supporting, Good.
-Ventilatory techniques: sedated and paralyzed, baseline (normal ventilation) Æ sodium bicarbonate administration to create alkalosis with no change of ventilatory parameters Æ hypoventilation achieved by decreasing ventilatory rate while keeping tidal volume constant
-Population: n=15 (all BCPA, median age 8 months, age range 4.7-15.5 months)
-Outcome: transcranial Doppler, arterial blood gases, systemic oxygenation saturation, atrial and pulmonary artery pressures, transpulmonary gradient, pulmonary artery oxygen saturation, cerebral blood flow velocity, mean flow velocity
-Results: Hypoventilation induced hypercarbia, increased mean arterial PO$_2$, mean systemic oxygen saturation, and mean cerebral blood flow velocity, as well as causing a decrease in the arterial venous oxygen consistent with cerebral blood flow.

Level 5. Supporting, Good.
-Ventilatory techniques: 100% oxygen (n=8) Æ room air (n=12) Æ 3% CO$_2$ (n=12) for 10 to 15 minutes followed by MRI velocity mapping in the aorta and jugular veins; in 3 of the patients who did not receive 100% oxygen, velocity mapping of the SVC was performed under room air and CO$_2$ in order to assess the contribution of the subclavian and innominate veins to PBF; in the final patient, time constraints only allowed for study under room air and hypercarbic conditions
-Population: n=12 (all SCPC, age 2.2 ± 0.5 years)
-Outcome: arterial blood gases, pulse oxymetry, electrogardiogram, blood pressure cuff, direct visualization, Siemens 1.5-Tesla Vision MRI system
-Results: With the administration of CO$_2$ flow to the brain and lungs as well as cardiac index increased, and there was an improvement in PO$_2$. The increased cardiac index accounted for the increased cerebral and pulmonary blood flow and cerebral O$_2$ transport increased by 80% while preserving body O$_2$ delivery. Hyperoxia did not change cerebral and pulmonary blood flow and PO$_2$ increased by 94%.


samples)

Results: A short period of NPV resulted in a 42% mean increase in pulmonary blood flow. After an extended period of NPV pulmonary blood flow increased by 54%. These values fell back toward baseline when IPPV was reinstituted. Heart rate did not change and stroke volume increased from 25 to 37.


Level 5. Supporting, Good.

-Ventilatory techniques: intermittent positive pressure ventilation (IPPV) → negative pressure ventilation (NPV) for 15 minutes → IPPV

-Population: n=11; n=5 repair of TOF, n=6 TCPC with fenestration (median age 5.3 years)

-Outcomes: end-tidal carbon dioxide, mean pulmonary blood flow, heart rate, VO₂, arterial oxygen content, mixed venous oxygen, mixed venous saturation, arteriovenous oxygen content difference, mean systemic arterial or right arterial pressures, mean pulmonary arterial or left atrial pressures, systemic vascular resistance, pulmonary vascular resistance, right-to-left shunt fraction, pulmonary blood flow

-Results: Heart rate did not change and stroke volume increased by a mean of 48.5%. Mixed venous saturation increased by 4.6%, and arteriovenous oxygen content difference fell significantly as a consequence. The systemic and pulmonary vascular resistances were reduced significantly during NPV.


Level 4. Supporting, Poor (case report).

-Resuscitative techniques: manual external abdominal compressions with closed chest cardiac compression (CCCC), endotracheal intubation, artificial ventilation with oxygen, buffer correction, epinephrine infusion

-Population: n=1 (13 year-old female)

-Outcomes: survival of event, survival to discharge

-Results: Systolic blood pressure increased and the heart was restored to normal rhythm.


Level 5. Supporting, Poor (case report).

-Ventilatory techniques: mechanical ventilation, intubation, MAST suit, dopamine, dobutamine

-Population: n=2 (33-month-old female with modified Fontan, 45-month-old female with modified Fontan)

-Outcomes: mean arterial pressure, right atrial pressure, urine output

-Results: Both patients recovered fully and survived to discharge.