Fontan Operation (Extra-Cardiac, Non-Fenestrated) Guideline

CHOC Children's

Overall Care Guideline: GRADE B

Admit to CVICU
POD #0

- **Sedation**
  - Intermittent fentanyl (1 mcg/kg IV q 1 hour PRN pain)
  - Dexmedetomidine infusion (0.5 mcg/kg/hour IV)
- **Respiratory**
  - PEEP 3
  - Ventilator mode
    - SIMV-PRVC
    - SIMV-PC
  - Consider iNO if evidence of pulmonary hypertension *(Refer to Patient Care Policy I-1004 Inhaled Nitric Oxide (iNO) Protocol)*
  - Early extubation
    - Utilize extubation readiness testing
- **Cardiac**
  - Vital sign goals
    - HR < 120
    - Oxygen saturation > 90%
    - Blood pressure per age normal
    - Temperature control/avoidance of fever
    - Fontan pressure < 10 mmHg
  - Monitor for low cardiac output syndrome
    - Consider milrinone infusion
    - Consider epinephrine infusion
  - Monitor Fontan pressure
  - Continuous atrial ECG monitoring for 6 hours and then PRN
- **GI**
  - Start clears 2 hours after successful extubation
- **Heme**
  - Monitor for bleeding
- **Monitor and trend Etiometry T3 data**

POD #1

- **Fluid management**
  - Negative fluid balance goal
    - Furosemide 1 mg/kg IV q 8 hours
    - Chlorothiazide 1 mg/kg po bid when taking po
    - Fluid restrict to 80% maintenance while chest tubes are in place
    - Spironolactone 1 mg/kg po bid when taking po
    - Consider enalapril in consultation with cardiologist when taking po
- **Wean milrinone infusion to off**
- **Advance diet**
  - Low fat
  - Consult Nutrition
- **Minimize positive pressure ventilation**
  - Wean HFNC
  - Keep NC 0.5 LPM minimum while CTs in place
- **Wean off iNO** *(Refer to Patient Care Policy I-1004 Inhaled Nitric Oxide (iNO) Protocol)*
  - Consider oral sildenafil
- **Discontinue arterial line**
- **Discontinue bladder catheter**
- **Discuss anticoagulation requirements and timing of initiation with surgeon and cardiologist**
- **Analgesia and anxiolysis**
  - Consider ketorolac in consultation with surgeon
  - Wean off dexmedetomidine
  - Ambulate
  - Establish normal sleep-wake cycle

POD #2 - 5

- Consider changing to oral diuretic regimen
- Consider removing CTs when total output is < 2 mL/kg/day
- Transition to oral pain medication PRN
- Complete discharge teaching

Discharge Criteria

- CTs out for > 24 hours with clear chest radiograph the day of discharge
- Discharge teaching complete
- Discharge echocardiogram complete
- Pain controlled on oral medication
- Ambulatory (per age normal)
- Normal sinus rhythm unless cleared by EP, cardiologist and surgeon

Common Complications

- **Tamponade**
  - Consider fluid bolus
  - Consider echocardiogram
  - Notify cardiologist
  - Notify surgeon
  - Consider bedside/catheterization lab drainage
- **Junctional ectopic tachycardia**
  - *Refer to Junctional Ectopic Tachycardia Guideline*
- **Systemic hypotension and low Fontan pressure**
  - Consider volume resuscitation
  - Consider PRBC transfusion if anemic
  - Consider vasopressin infusion
  - Consider assessing for adrenal insufficiency
- **Hypotension and high Fontan pressure**
  - Pulmonary hypertension
    - Consider iNO initiation and pCO_2 goal 35-40 mmHg *(Refer to Patient Care Policy I-1004 Inhaled Nitric Oxide (iNO) Protocol)*
    - Consider sedation
    - Consider paralysis
  - Fontan obstruction
    - Consult with cardiologist and surgeon
    - Consider urgent catheterization lab diagnosis and intervention
  - Ventricular failure
    - Consider echocardiogram
    - Consider epinephrine infusion
    - Consider milrinone infusion
- **Heart block**
  - Consider A-V temporary pacing
  - Minimize dexmedetomidine use
- **Bleeding**
  - Consider checking CBC
  - Consider PRBC transfusion
  - Measure coagulation panel and replace factors as indicated
    - Consider Factor 7 administration
  - Perform TEG
  - Call surgeon
- **Chylothorax**
  - *Refer to Chylothorax Guideline*

Patient Education

- Refer to CVICU unit specific education

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Approved Evidence Based Medicine Committee
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Reassess the appropriateness of Care Guidelines as condition changes and 24 hrs after admission. This guideline is a tool to aid clinical decision making. It is not a standard of care. The physician should deviate from the guideline when clinical judgment so indicates.

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