1. WHO:
   a. PATIENT:
      i. Risk Stratify: Identify maternal cardiac lesion and severity of disease
         1. Modified WHO scale (mWHO II-III, III, IV consider transfer to referral hospital)\(^1\)
         2. CARPREG II Risk score (CARPREG >1 consider transfer to referral hospital)\(^2\)
      ii. Early Assessment and Management
         1. Evaluate maternal function and pregnancy-related changes NYHA classes I-IV, symptoms: ask about climbing stairs and for examples of usual physical activity.
            a. Identify baseline function: Pre-pregnancy function
            b. Identify changes in function: Function in each trimester (particularly at 26-30 weeks)
            c. Assess current symptoms
         2. Gather additional (baseline) data at antepartum visit
            a. BNP, ECG, TTE
            b. Elevated BNP levels are associated with cardiac events, NT-proBNP >128 pg/mL at 20 weeks pregnancy is predictive of cardiac events later in the pregnancy.\(^1\)
            c. Exercise test
            d. ECG:
               i. Leftward heart rotation 15–20° with leftward axis deviation.
               ii. Transient ST/T wave changes
               iii. Lead III: Q wave and inverted T waves
               iv. aVF: attenuated Q wave
               v. V1-3: inverted T wave
               vi. LV hypertrophy pattern may be seen
               vii. Holter monitoring for patients with paroxysmal or persistent arrhythmias such as ventricular tachycardia, atrial fibrillation or atrial flutter or in patients who experience palpitations.
            e. TTE: some changes in size and function parameters may be normal but all changes from baseline should be assessed with a cardiologist
            f. If aortic pathology exists consider complete aortic imaging by computed tomography (CT) scanning or magnetic resonance imaging (MRI)
   3. Optimize early anticoagulation for:
      a. Mechanical valves
b. Low ejection fraction

c. Pulmonary hypertension

d. Arrhythmias

4. Optimize cardiac function medically or surgically depending on specific lesions, consult cardiologists1,3,4

5. Assess and address non-cardiac medical issues
   a. Medical history
   b. Surgical history
   c. Obstetric history (fetal concerns)
   d. Current medications
   e. Allergies
   f. Social history/support (services needed to help patient to follow regimen)

b. TEAM: (severity of lesion will determine need for additional team members – italicized specialties are optional)
   i. Obstetrician/Maternal Fetal Medicine
   ii. Anesthesiologist (Obstetric and Cardiothoracic)
   iii. Cardiologist
   iv. Neonatologist
   v. Hematologist
   vi. Cardiothoracic surgeon
   vii. ECMO surgeon
   viii. Perfusionist
   ix. Intensivist
   x. Critical Care Obstetric Nurse
   xi. Critical Care Nurse

2. WHAT:
   a. Route of delivery per obstetrician:
      i. Vaginal delivery: preferred mode of delivery for majority of women with heart disease unless maternal or fetal instability is present or highly likely
         1. Active phase of the second stage may be delayed for 1-2h to allow maximal descent of the fetal head, as to shorten the active phase of the second stage: While a prolonged second state followed by instrumented delivery can increase bleeding risk, some obstetricians may choose to prolong the passive second stage to promote a shortened active stage during which the patient does bare down, but only a few effective pushes are required to deliver the fetus.1
         2. Assisted delivery: forceps or a vacuum may be performed further reduce maternal effort, as indicated by the underlying cardiac lesion.1
      ii. Potential cesarean delivery indications:1
         1. Maternal or fetal instability:
Acute decompensated heart failure (severe right heart failure or LVEF<25%, failing Fontan, oxygen saturation <85%, severe peripartum cardiomyopathy)

Pulmonary hypertensive crisis (controversial in severe pulmonary hypertension or Eisenmenger’s in absence of “crisis”)

Severe, symptomatic aortic stenosis

Maternal intolerance to labor

Fetal intolerance to labor

Current anticoagulation precluding neuraxial anesthesia or vaginal delivery (fetal coagulopathy, maternal warfarin, and risk of intracerebral hemorrhage)

Aortopathy: significant aortic dilation (should not perform repeated valsalva maneuver)

Marfan’s Syndrome: >40mm

Bicuspid aortic valve: >45mm

Turner: Aortic size index >20mm/m²

Elective termination (considered in women with mWHO Class IV disease)

Contraception plan:

- Tubal ligation
- Intrauterine device
- Oral
- Partner sterilization

WHEN: The goal is to deliver the fetus as close to term as possible but pre-term deliveries may be indicated for fetal or maternal instability or for obstetric indications.

TARGET DELIVERY DATE (gestational weeks/days):

WHERE:

- Type of medical center:
  - Local
  - Referral hospital (maternal or fetal indication (neonatal ICU))

- Location within medical center: consider severity of maternal illness, need and availability of monitoring, need for cardiothoracic surgeon/surgery and/or ECMO
  - Labor and delivery suite labor room (may need ICU nursing support or training)
  - Labor and delivery suite operating room (elective CS)
  - Cardiothoracic operating room
  - Intensive care unit (may need OB nursing and OB anesthesia support)

- Transfer plan:
  - Identify accepting center and physicians
  - Identify transport team
  - Communicate current patient status to transport team and accepting medical center
  - Seek and provide follow up and debriefing
5. HOW: Peripartum plan
   a. Hemodynamic goals: Identify the lesion-specific hemodynamic goals and create a plan for maintaining hemodynamic stability
      i. Potential medications needed: phenylephrine, norepinephrine, vasopressin, dobutamine, dopamine, milrinone, epinephrine, anti-pulmonary hypertensives, oxygen
   b. Peripartum risks: Identify the peripartum risks associated with this patient’s disease(s) and create a plan for mitigating risks, and (if possible) how (and who) to respond if the “risk event” occurs.
   c. Delivery plan:
      i. Anesthesia:
         1. Neuraxial anesthesia:
            a. relative contraindications: current anticoagulation, respiratory failure
         2. General anesthesia
      ii. Monitoring:
        1. Blood pressure: non-invasive or arterial line
        2. Telemetry in labor: Yes/No
        3. Central venous pressure: Yes/No
        4. Pulmonary artery catheter: Yes/No
        5. Transthoracic echocardiogram: Yes/No
        6. Transesophageal echocardiogram (requires sedation or general anesthesia): Yes/No
      iii. Access:
        1. Venous: peripheral or central (consider central venous access if central venous pressure monitoring is desired, or use of inotropes or vasopressors is anticipated)
        2. ECMO: back-up, place wires, place sheaths, place cannulas
           a. Veno-venous ECMO: respiratory failure
           b. Veno-arterial ECMO: cardiac failure with or without respiratory failure
      iv. Antibiotic prophylaxis
      v. Management of anticoagulation/thromboprophylaxis:
         1. When to stop
         2. When to re-start
      vi. Emergency plan: Who is on the care team if an urgent (unscheduled) delivery is necessary?
   d. Hemorrhage prevention/management:
      i. Uterotonics:
         1. Oxytocin: intravenous infusion 15-30 units/hour preferred over bolus, treat accompanying vasodilation with vasopressor, coronary vasospasm is rare
         2. Methergine: relative contraindication in hypertensive disorders
         3. Hemabate: relative contraindication in pulmonary disease
4. Misoprostol: no contraindications

ii. Procedures:
   1. B-lynch suture
   2. Bakri balloon
   3. Uterine artery embolization
   4. Hysterectomy

iii. Blood products:
   1. pRBC
   2. Fresh frozen plasma
   3. Cryoprecipitate
   4. Platelets (caution in pulmonary hypertensive crisis)

e. Post care:
   i. Recovery location:
      1. Intensive care unit
      2. High risk maternal unit
      3. Post-partum unit
   ii. Monitoring and plan for continuation, removal or addition
      1. Blood pressure: non-invasive or arterial line
      2. Telemetry: Yes/No
      3. Central venous pressure: Yes/No
      4. Pulmonary artery catheter: Yes/No
      5. Transthoracic echocardiogram: Yes/No
      6. Transesophageal echocardiogram: Yes/No
   iii. Treatment goals: most common post-partum issues are arrhythmia and heart failure
      1. Diuresis
      2. Venous thromboembolism prophylaxis
      3. Stool softening for prevention of Valsalva
      4. Diet: Sodium restriction as needed
   iv. Subspecialty follow up

References:

<table>
<thead>
<tr>
<th>Diagnosis (otherwise well and uncomplicated)</th>
<th>mWHO I</th>
<th>mWHO II</th>
<th>mWHO II-III</th>
<th>mWHO III</th>
<th>mWHO IV</th>
</tr>
</thead>
<tbody>
<tr>
<td>Small or mild</td>
<td>Unoperated atrial or ventricular septal defect</td>
<td>Mild left ventricular impairment (EF &gt;45%)</td>
<td>Moderate left ventricular impairment (EF 30–45%)</td>
<td>Pulmonary arterial hypertension</td>
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<tr>
<td>– pulmonary stenosis</td>
<td></td>
<td>Hypertrophic cardiomyopathy</td>
<td>Previous peripartum cardiomyopathy without any residual left ventricular impairment</td>
<td>Severe systemic ventricular dysfunction (EF &lt;30% or NYHA class III–IV)</td>
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<tr>
<td>– patent ductus arteriosus</td>
<td></td>
<td>Native or tissue valve disease not considered WHO I or IV (mild mitral stenosis, moderate aortic stenosis)</td>
<td>Mechanical valve</td>
<td>Previous peripartum cardiomyopathy with any residual left ventricular impairment</td>
<td></td>
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<tr>
<td>– mitral valve prolapse</td>
<td></td>
<td>Marfan or other HTAD syndrome without aortic dilation</td>
<td>Systemic right ventricle with good or mildly decreased ventricular function</td>
<td>Severe mitral stenosis</td>
<td></td>
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<tr>
<td>Successfully repaired simple lesions (atrial or ventricular septal defect, patent ductus arteriosus, anomalous pulmonary venous drainage)</td>
<td></td>
<td>Aorta &lt;45 mm in bicuspid aortic valve pathology</td>
<td>Fontan circulation. If otherwise the patient is well and the cardiac condition uncomplicated</td>
<td>Severe symptomatic aortic stenosis</td>
<td></td>
</tr>
<tr>
<td>Atrial or ventricular ectopic beats, isolated</td>
<td></td>
<td>Repaired coarctation</td>
<td>Unrepaired cyanotic heart disease</td>
<td>Systemic right ventricle with moderate or severely decreased ventricular function</td>
<td></td>
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<tr>
<td></td>
<td></td>
<td>Atrioventricular septal defect</td>
<td>Other complex heart disease</td>
<td>Severe aortic dilatation (&gt;45 mm in Marfan syndrome or other HTAD, &gt;50 mm in bicuspid aortic valve, Turner syndrome ASI &gt;25 mm²/m², size of Fallot &gt;50 mm)</td>
<td></td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>Moderate mitral stenosis</td>
<td>Vascular Ehlers-Danlos</td>
<td></td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>Severe asymptomatic aortic stenosis</td>
<td>Severe (re)coarctation</td>
<td></td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>Moderate aortic dilatation (40–45 mm in Marfan syndrome or other HTAD; 45–50 mm in bicuspid aortic valve, Turner syndrome ASI 20–25 mm²/m², size of Fallot &lt;50 mm)</td>
<td>Fontan with any complication</td>
<td></td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>Ventricular tachycardia</td>
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</table>

| Risk                                                                 | No detectable increased risk of maternal mortality and no/mild increased risk in morbidity | Small increased risk of maternal mortality or moderate increase in morbidity | Intermediate increased risk of maternal mortality or moderate to severe increase in morbidity | Significantly increased risk of maternal mortality or severe morbidity | Extremely high risk of maternal mortality or severe morbidity |
| Maternal cardiac event rate                                             | 2.5–5% | 5.7–10.5% | 10–19% | 19–27% | 40–100% |
| Counselling                                                            | Yes | Yes | Yes | Yes: expert counselling required | Yes: pregnancy contraindicated; if pregnancy occurs, termination should be discussed |
| Care during pregnancy                                                   | Local hospital | Local hospital | Referral hospital | Expert centre for pregnancy and cardiac disease | Expert centre for pregnancy and cardiac disease |
| Minimal follow-up visits during pregnancy                              | Once or twice | Once per trimester | Bimonthly | Monthly or bimonthly | Monthly |
| Location of delivery                                                    | Local hospital | Local hospital | Referral hospital | Expert centre for pregnancy and cardiac disease | Expert centre for pregnancy and cardiac disease |

ASI = aortic size index; EF = ejection fraction; HTAD = heritable thoracic aortic disease; mWHO = modified World Health Organization classification; NYHA = New York Heart Association; WHO = World Health Organization.
## NYHA Class

<table>
<thead>
<tr>
<th>NYHA Class</th>
<th>Severity</th>
<th>Symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Mild</td>
<td>No limitation of physical activity.</td>
</tr>
<tr>
<td>II</td>
<td>Mild</td>
<td>Slight limitation of physical activity. Patient is comfortable at rest. Ordinary physical activity results in fatigue, palpitation (rapid or pounding heartbeat, dyspnea, (shortness of breath) or anginal pain (chest pain).</td>
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<tr>
<td>III</td>
<td>Moderate</td>
<td>Marked limitation of physical activity. Patient is comfortable at rest. Less than ordinary activity causes fatigue, palpitation, dyspnea, or anginal pain.</td>
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<tr>
<td>IV</td>
<td>Severe</td>
<td>Patient is unable to do any physical activity without discomfort. Symptoms of heart failure or the anginal syndrome may be present even at rest. If any physical activity is undertaken, discomfort is increased.</td>
</tr>
</tbody>
</table>

The CARPREG (Cardiac Disease in Pregnancy Study) II risk score is based on 10 predictors, shown in the box. Each predictor is assigned a weighted point score. The sum of points represents the risk score. Risk scores are categorized into the 5 groups (x-axis). The predicted (light blue) and the observed frequency of primary cardiac events in the derivation (medium blue) and validation (dark blue) groups are shown on the y-axis. NYHA = New York Heart Association.