Peripartum Cardiomyopathy

Background

1. Definition
   - Heart failure without identifiable cause, with onset in late pregnancy or early postpartum

Pathophysiology

1. Pathology of disease
   - Unclear
   - Suspected factors include myocarditis (viral), genetic, increased pregnancy-related hemodynamic changes
2. Incidence, prevalence
   - Varies worldwide
   - 1:15,000 in US
   - Higher in other areas, especially Haiti & some African nations (as high as 1:100 in some regions)
3. Risk factors
   - Age
     - Higher risk with age >30 years
   - Genetic
     - African-Americans at higher risk
     - Some increased risk within families with history of peripartum cardiomyopathy - genetic vs. exposure?
   - Pregnancy-related factors
     - Multiparity
     - Multiple gestation
     - History of pregnancy-induced hypertension, preeclampsia, or eclampsia
     - Long term use of beta-blockers
   - Environmental
     - Cocaine use during pregnancy
4. Morbidity / mortality
   - Mortality
     - 6-23% in US based case series
   - Cardiac transplant rate
     - 4-7% eventually require heart transplant
   - One-half of patients reach EF >50% by 2 years
     - Most of this recovery occurs in first 6 mos
   - Potential risk in future pregnancies
     - Further pregnancies may worsen EF - risk may exist even if cardiac function had normalized

Diagnostics

1. History
   - Dyspnea, cough, orthopnea, PND, hemoptysis, fatigue, occasionally abdominal pain, palpitations
2. Physical exam
   - Displaced PMI, gallop, edema

3. Diagnostic testing
    - Echo and EKG most important
    - Laboratory evaluation
        - Enzymes- usually normal
        - Consider viral cultures/titers (including coxsackie B)
    - Diagnostic imaging
        - Echocardiogram
        - LV enlargement
        - Contractility impaired
        - Measurements of ventricle size, EF important
    - Consider CXR if post-delivery
    - Other studies
        - EKG
            - May show LVH, sinus tachycardia, nonspecific ST changes
            - Occasionally other arrhythmia, Q waves
        - Less commonly used, may be needed for severe case:
            - Heart Catheterization
            - Endomyocardial biopsy
        - Tests needed to exclude other diagnostic concerns:
            - Spiral CT or VQ scan to rule-out PE
4. Criteria for diagnosis
    - Time of onset
        - After 36 weeks of pregnancy or first 5 mos. postpartum
        - Those with idiopathic cardiomyopathy before 36 wks excluded from this definition, but may have similar prognosis & course
    - No identifiable cause
    - No signs/symptoms of heart disease prior to last month of pregnancy
    - Signs of left ventricular systolic dysfunction (LSVD) on echo (EF<45%)

Differential Diagnosis

1. Key DDx
    - Pulmonary embolus
    - Pneumonia
    - CHF from other causes
    - Preexisting cardiac disease
2. Extensive DDx
    - Pulmonary edema related to preeclampsia or eclampsia or treatment for these conditions

Therapeutics

1. Acute treatment
    - Similar to other heart failure
        - Oxygen
        - Diuretics
        - Na restriction
        - Digoxin
        - Beta blockers
        - ACE-I or ARB if post-delivery

        - Anticoagulation (esp. for severe dilation)
        - Avoid warfarin prior to delivery
    2. Further management (24 hrs)
        - Cardiac monitoring
        - Treat arrhythmias
        - Pulmonary emboli more common
    3. Long-term care
        - Monitor with follow-up echos (after 2-3 wks, again q 6 mos. until complete recovery or plateau in improvement)
        - Severely decreased EF without improvement may require heart transplant
        - Document normalized LV function prior to attempting future pregnancy & counsel about potential risks
        - Counsel those with persistent LVSD to avoid future pregnancy

Follow-Up

1. Admit to hospital
    - All patients with new onset heart failure or decompensation
2. Refer to specialist

Prognosis

1. Most who regain EF >50%, do so within 6 mos
2. Best prognosis for regaining EF is in those with initial EF >30%

References

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