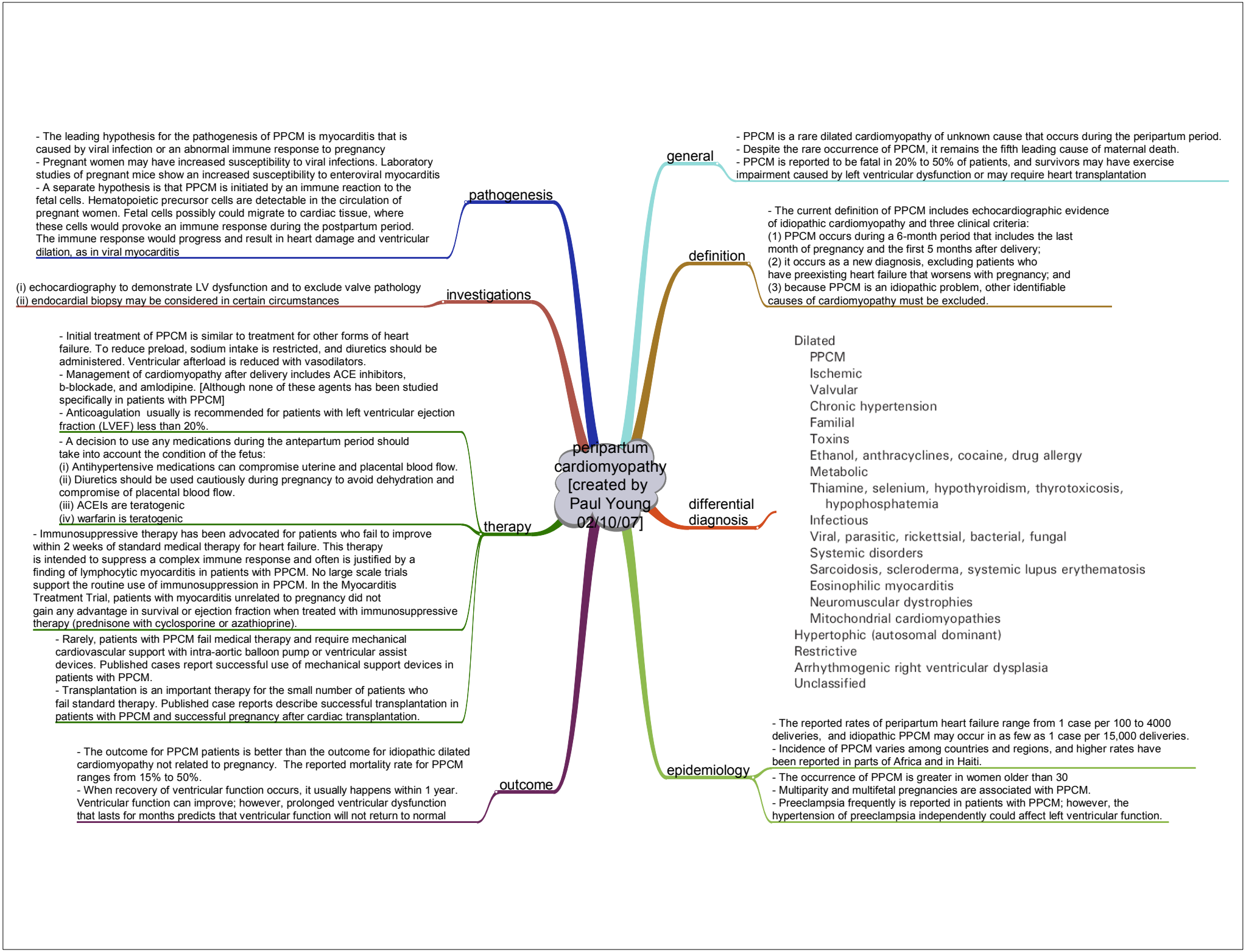
peripartum cardiomyopathy [created by Paul Young 02/10/07]
general
- PPCM is a rare dilated cardiomyopathy of unknown cause that occurs during the peripartum period.
- Despite the rare occurrence of PPCM, it remains the fifth leading cause of maternal death.
- PPCM is reported to be fatal in 20% to 50% of patients, and survivors may have exercise impairment caused by left ventricular dysfunction or may require heart transplantation
definition
- The current definition of PPCM includes echocardiographic evidence of idiopathic cardiomyopathy and three clinical criteria:
(1) PPCM occurs during a 6-month period that includes the last month of pregnancy and the first 5 months after delivery;
(2) it occurs as a new diagnosis, excluding patients who have preexisting heart failure that worsens with pregnancy; and
(3) because PPCM is an idiopathic problem, other identifiable causes of cardiomyopathy must be excluded.
differential diagnosis
Dilated
PPCM
Ischemic
Valvular
Chronic hypertension
Familial
Toxins
Ethanol, anthracyclines, cocaine, drug allergy
Metabolic
Thiamine, selenium, hypothyroidism, thyrotoxicosis, hypophosphatemia
Infectious
Viral, parasitic, rickettsial, bacterial, fungal
Systemic disorders
Sarcoidosis, scleroderma, systemic lupus erythematosis
Eosinophilic myocarditis
Neuromuscular dystrophies
Mitochondrial cardiomyopathies
Hypertophic (autosomal dominant)
Restrictive
Arrhythmogenic right ventricular dysplasia
Unclassified
epidemiology
- The reported rates of peripartum heart failure range from 1 case per 100 to 4000 deliveries, and idiopathic PPCM may occur in as few as 1 case per 15,000 deliveries.
- Incidence of PPCM varies among countries and regions, and higher rates have been reported in parts of Africa and in Haiti.
- The occurrence of PPCM is greater in women older than 30
- Multiparity and multifetal pregnancies are associated with PPCM.
- Preeclampsia frequently is reported in patients with PPCM; however, the hypertension of preeclampsia independently could affect left ventricular function.
outcome
- The outcome for PPCM patients is better than the outcome for idiopathic dilated cardiomyopathy not related to pregnancy. The reported mortality rate for PPCM ranges from 15% to 50%.
- When recovery of ventricular function occurs, it usually happens within 1 year. Ventricular function can improve; however, prolonged ventricular dysfunction that lasts for months predicts that ventricular function will not return to normal
therapy
- Initial treatment of PPCM is similar to treatment for other forms of heart failure. To reduce preload, sodium intake is restricted, and diuretics should be administered. Ventricular afterload is reduced with vasodilators.
- Management of cardiomyopathy after delivery includes ACE inhibitors, b-blockade, and amlodipine. [Although none of these agents has been studied specifically in patients with PPCM]
- Anticoagulation usually is recommended for patients with left ventricular ejection fraction (LVEF) less than 20%.
- A decision to use any medications during the antepartum period should take into account the condition of the fetus:
(i) Antihypertensive medications can compromise uterine and placental blood flow.
(ii) Diuretics should be used cautiously during pregnancy to avoid dehydration and compromise of placental blood flow.
(iii) ACEIs are teratogenic
(iv) warfarin is teratogenic
- Immunosuppressive therapy has been advocated for patients who fail to improve within 2 weeks of standard medical therapy for heart failure. This therapy is intended to suppress a complex immune response and often is justified by a finding of lymphocytic myocarditis in patients with PPCM. No large scale trials support the routine use of immunosuppression in PPCM. In the Myocarditis Treatment Trial, patients with myocarditis unrelated to pregnancy did not gain any advantage in survival or ejection fraction when treated with immunosuppressive therapy (prednisone with cyclosporine or azathioprine).
- Rarely, patients with PPCM fail medical therapy and require mechanical cardiovascular support with intra-aortic balloon pump or ventricular assist devices. Published cases report successful use of mechanical support devices in patients with PPCM.
- Transplantation is an important therapy for the small number of patients who fail standard therapy. Published case reports describe successful transplantation in patients with PPCM and successful pregnancy after cardiac transplantation.
investigations
(i) echocardiography to demonstrate LV dysfunction and to exclude valve pathology
(ii) endocardial biopsy may be considered in certain circumstances
pathogenesis
- The leading hypothesis for the pathogenesis of PPCM is myocarditis that is caused by viral infection or an abnormal immune response to pregnancy
- Pregnant women may have increased susceptibility to viral infections. Laboratory studies of pregnant mice show an increased susceptibility to enteroviral myocarditis
- A separate hypothesis is that PPCM is initiated by an immune reaction to the fetal cells. Hematopoietic precursor cells are detectable in the circulation of pregnant women. Fetal cells possibly could migrate to cardiac tissue, where these cells would provoke an immune response during the postpartum period. The immune response would progress and result in heart damage and ventricular dilation, as in viral myocarditis