Question 44
During pregnancy, many cardiovascular conditions are associated with an increased risk of maternal mortality.
Which one of the following cardiovascular conditions is associated with the highest rate of maternal mortality?

Moderate to severe:
A. Pulmonary hypertension
B. Hypertrophic cardiomyopathy
C. Coarctation
D. Aortic regurgitation
E. Peripartum cardiomyopathy

Answer A

FROM HARRISONS MEDICAL CONDITIONS IN PREGNANCY CHAPTER 6
CARDIAC DISEASE
Valvular Heart Disease
Mitral Stenosis
This is the valvular disease most likely to cause death during pregnancy. The pregnancy-induced increase in blood volume, cardiac output, and tachycardia can cause pulmonary edema in women with mitral stenosis. Pregnancy associated with long-standing mitral stenosis may result in pulmonary hypertension. Sudden death has been reported when hypovolemia has been allowed to occur in this condition. Careful control of heart rate, especially during labor and delivery, minimizes the impact of tachycardia and reduced ventricular filling times on cardiac function. Pregnant women with mitral stenosis are at increased risk for the development of atrial fibrillation and other tachyarrhythmias. Medical management of severe mitral stenosis and atrial fibrillation with digoxin and beta blockers is recommended. Balloon valvulotomy can be carried out during pregnancy.

Mitral Regurgitation and Aortic Regurgitation
These are both generally well tolerated during pregnancy. The pregnancy-induced decrease in systemic vascular resistance reduces the risk of cardiac failure with these conditions. As a rule, mitral valve prolapse does not present problems for the pregnant patient, and aortic stenosis, unless very severe, is well tolerated. In the most severe cases of aortic stenosis, limitation of activity or balloon valvuloplasty may be indicated.

For women with artificial valves contemplating pregnancy, it is important that warfarin be stopped and heparin initiated prior to conception. Warfarin therapy during the first trimester of pregnancy has been associated with fetal chondrodysplasia punctata. In the second and third trimester of pregnancy, warfarin may cause fetal optic atrophy and mental retardation. For women with prosthetic heart valves, prophylaxis against thrombosis with low-molecular-weight heparin (LMWH) is not recommended due to reports of valvular thrombosis despite adequate anticoagulation. Prophylaxis with unfractionated heparin is recommended for this group of women.

Congenital Heart Disease
The presence of a congenital cardiac lesion in the mother increases the risk of congenital cardiac disease in the newborn. Prenatal screening of the fetus for congenital cardiac disease with ultrasound is recommended. Atrial or ventricular septal defect is usually well tolerated during pregnancy in the absence of pulmonary hypertension, provided that the woman’s prepregnancy cardiac status is favorable. Use of air filters on intravenous sets during labor and delivery in patients with intracardiac shunts is generally recommended.

Other Cardiac Disorders
Supraventricular tachycardia is a common cardiac complication of pregnancy. Treatment is the same as in the nonpregnant patient, and fetal tolerance of medications such as adenosine and calcium channel blockers is
acceptable. When necessary, electrocardioversion may be performed and is generally well tolerated by mother
and fetus.

Peripartum cardiomyopathy is a rare disorder of pregnancy associated with myocarditis, and its etiology remains
unknown. Treatment is directed toward symptomatic relief and improvement of cardiac function. Many patients
recover completely; others are left with a progressive dilated cardiomyopathy. Recurrence in a subsequent
pregnancy has been reported, and women should be counseled to avoid pregnancy after a diagnosis of
peripartum cardiomyopathy.

**Specific High-Risk Cardiac Lesions**

**Marfan Syndrome**

is an autosomal dominant disease, associated with a high risk of maternal morbidity. Approximately
15% of pregnant women with Marfan syndrome develop a major cardiovascular manifestation during pregnancy,
with almost all women surviving. An aortic root diameter <40 mm is considered to be associated with a favorable
outcome of pregnancy. Prophylactic therapy with beta blockers has been advocated, although large-scale clinical
trials in pregnancy have not been performed.

**Pulmonary Hypertension**

Maternal mortality in the setting of severe pulmonary hypertension is high, and primary pulmonary hypertension
is a contraindication to pregnancy. Termination of pregnancy may be advisable in these circumstances to
preserve the life of the mother. In the Eisenmenger syndrome, i.e., the combination of pulmonary hypertension
with right-to-left shunting due to congenital abnormalities maternal and fetal death occur frequently. Systemic
hypotension may occur after blood loss, prolonged Valsalva maneuver, or regional anesthesia; sudden death
secondary to hypotension is a dreaded complication. Management of these patients is challenging, and invasive
hemodynamic monitoring during labor and delivery is generally recommended.

In patients with pulmonary hypertension, vaginal delivery is less stressful hemodynamically than Cesarean
section, which should be reserved for accepted obstetric indications.

From
Valvular Heart Disease in Pregnancy  Sharon C. Reimold, M.D., and John D. Rutherford, M.B., Ch.B.

primary pulmonary hypertension is associated with high maternal mortality (33 to 40 percent), as
well as with an increased rate of adverse neonatal events. Secondary pulmonary hypertension due
to valvular disease is associated with an increased rate of adverse maternal events, but the absolute
risk of such events is unclear. A systolic pulmonary-artery pressure that is more than 75 percent as
high as the systemic pressure places the woman at high risk.

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Heart disease and pregnancy Samuel C Siu, Jack M Colman

Marternal cardiac status and risk of cardiac complications during pregnancy
Low risk

- Small left to right shunts
- Repaired lesions without residual cardiac dysfunction’
- Isolated mitral valve prolapse without significant regurgitation
- Bicuspid aortic valve without stenosis
- Mild to moderate pulmonic stenosis
- Valvar regurgitation with normal ventricular systolic function
Intermediate risk
- Unrepaired or palliated cyanotic congenital heart disease
- Large left to right shunt
- Uncorrected coarctation of the aorta
- Mitral or aortic stenosis
- Mechanical prosthetic valves
- Severe pulmonic stenosis
- Moderate to severe systemic ventricular dysfunction
- History of peripartum cardiomyopathy with no residual ventricular dysfunction

High Risk
- New York Heart Association (NYHA) class III of IV symptoms
- Severe pulmonary hypertension
- Marfan syndrome with aortic root or major valvar involvement
- Severe aortic stenosis

The New York Heart Association (NYHA) Functional Classification:

I  No symptoms and no limitation in ordinary physical activity.
II  Mild symptoms and slight limitation during ordinary activity. Comfortable at rest.
III  Marked limitation in activity due to symptoms, even during less-than-ordinary activity. Comfortable only at rest.
IV  Severe limitations. Experiences symptoms even while at rest.