Congenital Heart Disease and Pregnancy

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Pregnancy, labor, and delivery appear to be well tolerated in most patients with mild-to-moderate congenital heart disease (CHD). The management of these patients is uneventful. Patients with more severe problems require frequent assessment and reassurance by the obstetrician and cardiologist, who should work as a team. Therapy consists of curtailing physical activity, avoiding excessive weight gain, reducing salt intake, immediate treatment of infection, diuresis for fluid retention, and close observation for signs or symptoms of latent congestive heart failure. Induction of labor and cesarean section are indicated only for obstetrical reasons and not for cardiac reasons. Patients with uncorrected cyanotic CHD or Eisenmenger's syndrome constitute a high-risk group and should be advised against pregnancy. Sterilization is recommended in patients with severe cardiac disability that cannot be controlled with medical or surgical therapy and for whom pregnancy may be life threatening. Cardiovascular surgery during pregnancy is rarely indicated. Prophylactic antibiotics should be employed at the time of delivery if indicated by the underlying lesion. Hemodynamic and electrocardiographic monitoring should be used in all high-risk patients in order to identify and correct hemodynamic and electrical instability as soon as possible.

The likelihood of encountering pregnant females with congenital heart disease (CHD) in general medical practice has increased significantly over recent years, reflecting advances in medical and surgical care that have allowed patients with CHD to reach childbearing age. Most types of CHD have been reported in pregnant patients; however, the most commonly reported lesions are atrial septal defect (ASD), patent ductus arteriosus (PDA), ventricular septal defect (VSD), pulmonary stenosis, aortic stenosis, coarctation of the aorta, and Tetralogy of Fallot. In the largest reported group of 237 patients with CHD, who were followed up through 488 pregnancies, 36 patients had cyanotic diseases, 53 had obstruction of either left or right ventricular outflow, and 148 patients had volume overload caused by left-to-right shunt. The almost universal recommendation to either prevent conception or perform early interruption of pregnancy in women with CHD, a common practice until only the recent past, does not seem to be justified today. An understanding of the pathophysiology of the disease and of potential complications of pregnancy is essential, however, in order to minimize risks to both mother and fetus. The management of patients with CHD should begin before conception. Careful diagnostic and functional evaluation is needed in order to determine the accurate diagnosis, cardiac function, and functional capacity of the patient. Cardiologist and obstetrician should then counsel the patient and her family on the

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potential risks of pregnancy to both the mother and fetus, the expected physical limitations of the mother in years to come, and, when appropriate, the patient's limited life span. The statistical risk to the offspring of inheriting CHD should be assessed, based on the type of maternal defect and family history. In addition, guidance on the need for anticoagulant and prophylactic antibiotic therapy should be provided.

I. MATERNAL OUTCOME

Maternal outcome is determined by the nature of the CHD, the functional capacity of the patient, and the history of surgical repairs. Whittenmore et al. did not report maternal deaths in 237 women with CHD, who were followed through 488 pregnancies. These investigators rated a patient's functional status as "excellent to good" or "fair to poor." Cardiovascular morbidity was greater in patients with "fair to poor" function. Congestive heart failure was the most common complication, occurring in 25% of patients with abnormal functional capacity. Other complications, such as arrhythmias and hypertension, were also related to the functional status of the mother. Pregnancy-induced hypertension was more frequent in patients with CHD (15%) than in the total obstetric population (4.3%). Shime et al. reported the deterioration of functional capacity and development of heart failure in as many as 47% of patients with cyanotic and in only 13% of patients with acyanotic CHD. Surgical repair improves the outcome of pregnancy in women with cyanotic heart disease. Most often patients with repaired cyanotic CHD survived pregnancy, although functional deterioration, especially during the third trimester, is often seen. Since anticoagulation may be indicated with CHD during pregnancy, bleeding complications can be anticipated in some patients. Whittenmore and colleagues reported the development of angina in 2 of 60 patients with fair to poor functional capacity, and in none of the patients with good to excellent function. More recently, Presbitero et al. reported perinatal complications in 4 of 45 patients with cyanotic CHD who had 96 pregnancies. Two patients had infective endocarditis, which resulted in death in 1. One patient had pulmonary embolism, and 1 patient had central embolism.

In summary, most patients with acyanotic CHD, who are in New York Heart Association (NYHA) functional class I or II, will tolerate pregnancy without significant complications. In fact, the maternal mortality rate for these patients is essentially the same as in patients without cardiac disease. Pregnancy in patients with congestive heart failure (NYHA classes III and IV), despite drug therapy, unrelieved cyanotic CHD, and/or pulmonary hypertension, carries a markedly increased risk of complications and mortality for both the mother and fetus, and therefore, has to be considered contraindicated. If pregnancy occurs in patients with one of these conditions, an early abortion is recommended. If, however, the continuation of pregnancy is desired by the patient and her family, a multidisciplinary approach, involving experienced obstetricians, cardiologists, and anesthesiologists is required, in order to minimize risks to the mother and fetus.

II. FETAL OUTCOME

Only 45% to 55% of pregnancies in mothers with hypoxemia are expected to result in liveborn infants, an incidence significantly lower than that seen in mothers without cyanosis (80%). Fetal outcome seems to be related to maternal functional capacity and the presence of cyanosis. The number of interrupted pregnancies and therapeutic abortions in the group reported by Whittenmore and colleagues was significantly lower in patients with excellent to good, compared with fair to poor functional capacity (80% vs. 68% live-born infants, respectively). Women with cyanotic CHD, who had palliative surgery, had a higher incidence of live-born infants, than women with uncorrected cyanotic lesions (72% vs.42%). The majority of infants born to mothers with cyanotic CHD demonstrate low birthweight for gestational age and prematurity. The average birthweight for infants of women without cyanosis in Shime's group was 3300 ± 600 g, compared with 2400 ± 800 g in their cyanotic counterparts. Prematurity in infants born to
mothers with cyanotic CHD was reported to correlate with maternal hemoglobin and hematocrit levels. In summary, fetal loss, prematurity, and low birthweight for gestational age are common findings in women with cyanosis.

INCIDENCE OF CHD IN THE FETUS
The incidence of CHD in the general population is in the range of 5 to 8/1000. The reported incidence of CHD in the offspring of women with heart defects is 2 to 8/100. In addition, increased incidence of noricardiac anomalies, learning difficulties, and physical impairment occur in children born to mothers with CHD.

Dennis and Warren reported a 3.4% recurrence of CHD in a group of 308 patients with VSD, pulmonary valvular or infundibular stenosis, or any combination thereof, including patients with tetralogy of Fallot. Czeizel and coworkers documented the presence of CHD in 4.9% of children born to patients who had surgery for heart defects. Rose et al. reported substantial CRn in 8.8% of 385 children born to patients with one of four selected defects: ASD, coarctation of the aorta, aortic valve stenosis, or complex dextrocardia. Emanuel and associates found CHD in 14.3% of 36 offspring of mothers with atrioventricular septal defects. The highest incidence was reported by Whittenmore et al. who, in a prospective and nonselective evaluation of the offspring of all women with CHD, reported a 16.1% incidence of cardiac defects. The exclusion of cases with a dominant genetic syndrome (Noonan's syndrome and hypertrophic cardiomyopathy), familial clustering of CHD, and use of terato genetic medications, reduced the incidence rate to 13.5%. In addition, excluding children who had a VSD that closed spontaneously (within 3 years in 12 cases), the incidence of clinically important CHD was approximately 8%.

Evaluations over at least 3 years of 64 children born to mothers with CHD suggested a higher incidence of learning difficulties, especially in infants born to mothers with aortic stenosis. Children born prematurely to mothers with cyanotic CHD never exceeded the 50th percentile for height. The risk of recurrence of the mothers' anomalies in their offspring is between 2.5% and 4.0%. This risk increases by two- or threefold when the father is also affected.

PERIPARTUM MANAGEMENT
In the two large series of Whittenmore and colleagues and Shime et al., most women with CHD had vaginal deliveries. Although Shime's group reported a somewhat higher cesarean delivery rate, Whittenmore et al. thought that vaginal delivery may be better tolerated by many patients with CHD. Cesarean section is not indicated in most patients with CHD, and thus should be performed primarily for obstetric reasons. Oxygen should be given throughout labor and delivery in hypoxemic mothers. Hemodynamic evaluations with a balloon-tipped catheter is recommended for every patient with symptoms of heart failure during pregnancy, in patients with pulmonary hypertension, and in patients with cyanotic heart disease. Blood loss during delivery or postpartum should be replaced as rapidly as possible, especially in patients with right-to-left shunting, cyanosis, and pulmonary hypertension.

Maternal mortality may occur during anesthesia. The choice of anesthesia and anesthetics should be made jointly by the anesthesiologist, cardiologist, and obstetrician, and should be based on the form of maternal heart disease, the cardiovascular physiology of labor and delivery, and the expected hemodynamic consequences of anesthetic techniques and drugs.

PREVENTION OF INFECTIVE ENDOCARDITIS
Despite recent advances in antimicrobial therapy and cardiovascular surgery, infective endocarditis is still a serious complication with significant morbidity and mortality. Even though there is no documentation that prophylactic antibiotic therapy prevents infective endocarditis in humans, it is recommended in patients with CHD or acquired valvular defects in situations where bacteremia is highly predictable. Congenital cardiac anomalies, with the exception of the secundum-type ASD, can be complicated by endocarditis. Patients with CHD, who are at high risk for
Active endocarditis, should take measures to avoid or minimize potential sources of bacteremia. Poor dental hygiene and dental disease are associated with bacteremia, and therefore, the highest level of oral health should be maintained. The use of oral irrigational devices should be avoided, however, as the procedure can also cause bacteremia. Oral rather than nasotracheal intubation, should be utilized in patients with a high susceptibility to bacterial endocarditis. If bronchoscopy must be performed, the fiberoptic, rather than the rigid bronchoscope, should be used. As the incidence of bacteremia is often determined by the presence or absence of urinary infections, urine should be made sterile prior to any urologic procedures.20

The American Heart Association, in its most recent committee report,21 recommends antibiotic prophylaxis with all dental procedures that are likely to cause gingival bleeding, including professional cleaning. In addition, prophylactic treatment is recommended before certain procedures on the upper respiratory tract, including tonsillectomy or adenoidectomy, bronchoscopy, and surgical procedures involving the respiratory tract. Prophylactic therapy is also recommended before surgery or trulementation of the genitourinary tract, the gastrointestinal tract, or the gallbladder, in cases of obstetric infections, such as puerperal sepsis or peripartum infection. Infective endocarditis in an uncomplicated gynecologic delivery is extremely rare. Sugrue and coworkers22 reported an incidence of 0.09% in 35 women with rheumatic heart disease or iD, and Marquis23 did not observe a single case of endocarditis after normal delivery in over 1750 cardiac patients during a 25-year period. In view of the lower incidence of endocarditis and the fact that routine antibiotic prophylaxis may cause antibiotic resistance while in term, enhancing the risk of antibiotic-resistant endocarditis,24-25 the American Heart Association has recommended prophylactic antibiotic therapy before an uncomplicated vaginal delivery only in patients with a prosthetic heart valve or surgically constructed systemic-to-pulmonary shunt. These conditions are particularly prone to endocarditis. Despite the American Heart Association's recommendation, antibiotic prophylaxis in other forms of CHD at a time of normal, uncomplicated labor and delivery is not an uncommon practice in many hospitals, including our own.26 Prophylactic antibiotic therapy is advisable in every patient with CHD, except those with an isolated, secundum-type ASD, surgically uncorrected or corrected without a prosthetic patch, and those who underwent ligation and division of a PDA after a 6-month healing period postsurgery.21

An increased risk for endocarditis has been suggested after manual removal of the placenta, and antibiotic prophylaxis seems indicated in this situation.23,27

ATRIAL SEPTAL DEFECT

Three types of atrial septal defects (ASD) occur in adults: ostium primum, ostium secundum, and sinus venosus. Among ASDs, the secundum type is the most common. In fact, with the exception of the congenital bicuspid aortic valve, it is the most common form of CHD in the adult. The female to male ratio is approximately 2 to 1. Thus this defect is the most common defect seen in pregnancy.

Not infrequently, ASD is first diagnosed during pregnancy, when the murmur is detected. The left-to-right shunting places a significant load on the right ventricle and increases pulmonary blood flow. This is usually well tolerated during pregnancy, even when pulmonary blood flow is several times higher than aortic flow, and pregnancy, labor, and delivery are usually uneventful.28

Atrial septal defect associated with endocardial cushion defect can vary from a partial cushion defect with a cleft mitral valve to a complete endocardial cushion defect, in which the atrial and ventricular septal defects (VSDs) are one, and there is a large cleft in the mitral and tricuspid valve, essentially producing a common atrioventricular valve. Pregnancy in such cases may be associated with the development of congestive heart failure (CHF), hypertension, arrhythmias, and worsening of functional capacity. Recommendations concerning pregnancy in these patients must be considered on an individual basis.
Whittenmore's group \cite{9,10} reported CHD in 15% of the offspring born to women with left-to-right shunting. The exact incidence in women with ASD was not reported. Emanuel et al. \cite{16} reported CHD in 14.3% of the offspring of 36 mothers with atrioventricular defects, the majority of which had an ostium primum-type defect, with various degrees of cleft atrioventricular valves. Sixty percent of the affected children had concordant lesions, and 40% had discordant lesions. The recurrence risk of ASD in the offspring has been reported to be approximately 2.5% in all mothers with the same lesion.\cite{19}

If CHF occurs and persists, despite adequate medical therapy during pregnancy, surgical closure must be strongly considered. Normally, the shunting of blood in uncomplicated ASD is from left to right, but the direction may be reversed by sudden events, such as pulmonary embolization, leading to the development of pulmonary hypertension. If excessive hemorrhage occurs during delivery, systemic hypotension may result, causing a reversal of the shunting and producing cyanosis. This can be corrected by restoring volume, preferably with the administration of blood, and by increasing systemic vascular resistance with vasopressor agents.

As a result of the low risk of bacterial endocarditis in patients with an ostium secundum-type ASD, prophylactic antibiotic treatment before delivery is not indicated, unless the ASD has been repaired with a prosthetic patch.\cite{21} Pulmonary hypertension and the development of Eisenmenger's syndrome in patients with ASD are associated with a substantially higher risk of complications.\cite{29}

VENTRICULAR SEPTAL DEFECT

Women with VSD usually tolerate pregnancy very well. In one report, 56 women with VSD had 141 pregnancies, and only 2 maternal deaths occurred; both were patients with Eisenmenger's syndrome.\cite{6} The other pregnancies were essentially uncomplicated. Episodes of congestive heart failure or arrhythmias were rarely noted. Whittenmore and colleagues\cite{9} did not report any deaths in a group of 50 patients with VSD, having 98 pregnancies with 80% live-born infants. Pregnancy was complicated by heart failure and arrhythmias in patients with fair-to-poor functional capacity, but was uncomplicated in the patients with excellent-to-good function. The incidence of recurrence of a VSD in the offspring of mothers with the same cardiac anomaly is between 4% and 11%. Almost half of these cases demonstrated spontaneous closure of the defect by the age of 3 years.\cite{9,19}

Pregnancy after closure of an uncomplicated VSD is essentially unremarkable. Several cases of successful closure of VSDs during pregnancy have been reported.\cite{30} However, the need for closure of the VSD during pregnancy should be rare, and is only indicated when CHF is not controlled by adequate medical therapy or when complications of bacterial endocarditis supervene.

Shunt reversal may occur in the postpartum period in patients with elevated pulmonary pressures and systemic hypotension. Prompt replacement of volume and the use of vasopressors to restore blood pressure to normal will prevent catastrophic complications.

The development of pulmonary hypertension and Eisenmenger's syndrome in patients with VSD is associated with a significant risk to the mother and fetus during pregnancy.

PATENT DUCTUS ARTERIOSUS

Patent ductus arteriosus is one of the most common congenital cardiovascular anomalies,\cite{31} and is predominantly observed in women (2 to 1 ratio). It used to be the most common congenital lesion seen in pregnancy, with the exception of ASD. It has become, however, a rare finding in pregnancy, as most PDA cases are diagnosed and surgically corrected in early childhood.\cite{31}

Maternal outcome in patients with PDA is usually favorable. Most patients with PDA tolerate pregnancy without significant difficulties.\cite{21,28,32} However, some of the patients will demonstrate worsening functional capacity and symptomatic status, and occasionally patients may develop CHF during pregnancy.\cite{31} Maternal mortality of 5.5% in 204 patients with PDA had been reported previously.\cite{32} The mortality was mostly attributable to heart failure.
The risk of recurrence of a PDA in the offspring of a mother with the same anomaly was estimated to be 4%.\(^{19}\)

Surgical closure of PDA should ideally be performed before gestation. Most patients in whom the malformation is diagnosed during pregnancy will tolerate the hemodynamic burden of pregnancy without difficulty; the occasional individual who presents with heart failure would be treated medically with bed rest and diuretics. Digitalis should be added, if left ventricular systolic dysfunction is present. Since right ventricular afterload is reduced by blood lancing from the aorta into the low-resistance pulmonary circulation, the benefit of isodilator agents is questionable. Surgical intervention during pregnancy should be performed only when heart failure cannot be controlled by optimal medical therapy. Concern has been expressed regarding delivery during pregnancy in women with PDA because of hormone-mediated changes in the arterial wall.\(^{28}\) However, published data suggest that surgical closure, if indicated, can be performed during pregnancy without increased maternal or fetal risk.\(^{28}\)

In the early postpartum period, there is the potential risk for shunt reversal attributable to a fall in systemic blood pressure in patients with elevated pulmonary artery pressure. Sysmic hypotension caused by blood loss or asodilation should, therefore, be promptly reated by replacing blood volume and administering vasopressor agents.

If pulmonary hypertension is present and shunt reversal occurs (Eisenmenger's syndrome), then pregnancy carries a significantly higher complication rate, both for the mother and for the fetus.\(^{29}\) As in most other congenital heart defects (CHD), antibiotic prophylaxis before delivery is recommended in patients with PDA.

**ORTIC STENOSIS**

Congenital obstruction of left ventricular outflow can be valvular, supravalvular, or sub/valvular in origin.\(^{33}\) These defects comprise approximately 4% to 5% of all congenital heart malformations. Aortic stenosis is 3 to 4 times (more common in males than in females; hence, he incidence in pregnancy is very low.\(^{33}\)

There are only a few reports in the literature concerning pregnancy in women with congenital aortic stenosis. Although pregnancy can be occasionally uncomplicated, even in cases with severe stenosis (gradient over 100 mm Hg), maternal functional deterioration, CHF, sudden death, and fetal mortality have been repeatedly reported.\(^{1.2.9.33}\) Up to 20% of newborns delivered by mothers with left ventricular outflow obstruction have been documented to have had cardiac defects. Most of the cardiac defects occurred in the offspring of patients without surgical correction. Mother-child defect concordance was estimated to be about 4% in women with congenital aortic stenosis.\(^{19}\) Whittenmore and associates\(^{9.10}\) found obstructive defects in 12% of the offspring born to mothers with left and/or right ventricular obstruction.

In summary, aortic stenosis, especially if severe, can be associated with maternal morbidity and even mortality during pregnancy and the peripartum period. With early diagnosis and appropriate care—including hemodynamic monitoring during labor and delivery in patients with severe disease, and appropriate anesthesi-most patients should have a favorable outcome. A high incidence of cardiac defects in the newborn has been found in patients without surgical correction of the obstruction of left ventricular outflow.

**COARCTATION OF THE AORTA**

The incidence of aortic dissection or rupture in patients with coarctation appears to be higher during pregnancy.\(^{34}\) Nowadays, uncorrected coarctation of the aorta is rarely diagnosed during pregnancy, since surgical correction is usually completed before the childbearing years. In uncomplicated coarctation of the aorta, pregnancy is usually safe for the mother, but fetal development can be impaired because of decreased blood flow distal to the aortic narrowing. In complicated coarctation, there is an increased risk to the mother.\(^{35}\) Whittenmore's group\(^{9}\) did not report any fatality. They described complications, such as hypertension, CHF, and angina in patients with left ventricular obstruction. Although patients with
Pulmonary Stenosis

Pregnancy is usually well tolerated, although symptoms of fatigue, CHF, and right heart failure have been occasionally noted. The incidence of cardiac defects may be as high as 19% in newborns to women with pulmonary stenosis. The risk of inheriting pulmonary stenosis from a mother with this disorder is estimated to be 3.5%. The additional load imposed by pregnancy on the right ventricle of patients with pulmonary stenosis is tolerated fairly well, as evidenced by the low incidence of cardiac failure and other complications. Most of the pregnancies complicated by pulmonary stenosis described in the literature were reported 10 to 20 years ago. With modern improvements in medical therapy, the incidence of heart failure should be lower. When heart failure persists, despite adequate medical therapy, surgical valvotomy should be performed. Balloon valvuloplasty is not recommended during pregnancy; this procedure may result in a large amount of radiation and hemodynamic instability, both of which may be deleterious to the fetus.

Tetralogy of Fallot

Tetralogy of Fallot is the most common cyanotic CHD in children and adults today. It is also the most common cyanotic congenital lesion in pregnancy. As more children with this defect undergo palliative or definitive surgical repair, more will survive to childbearing age. The hemodynamic changes associated with pregnancy may result in clinical deterioration with both maternal and fetal complications. The fall in peripheral vascular resistance of the aorta were included in this report, the exact incidence of maternal complications in these patients was not disclosed. Pregnancy-induced hypertension was also reported by Shim et al in 4 of 9 pregnancies in 6 women with coarctation. In a recent review of the literature after 1958, Metcalfe and colleagues28 reported 13 maternal deaths in 565 pregnancies involving 230 women with coarctation of the aorta.

In early studies, fetal mortality ranged from 13% to 25%. Whittenmore and coworkers9 described live-born infants in 78% of patients with left ventricular obstruction. Cardiac defects were present in 20% of newborns who predominantly had ventricular obstruction. The incidence of CHD in newborns was higher in mothers who did not undergo surgical correction. The risk of coarctation in the offspring, if the mother is the only parent with the same lesion, has been estimated to be 2%. The incidence of fetal heart disease is higher in uncorrected coarctation. Accordingly, the coarctation should be corrected either surgically or by balloon dilatation before pregnancy. The mainstay of treatment during pregnancy are to limit physical activity and to control blood pressure. Rupture of the aorta and cerebral aneurysms are less likely to occur in the absence of significant hypertension. However, excessive reduction of blood pressure is not recommended, since it may result in reduction of placental blood flow, which could affect the fetus.

Surgical correction of coarctation during pregnancy has been successful9 but is not recommended, unless major complications such as aortic dissection, uncontrollable increased systolic blood pressure (> 200 mm Hg), or severe heart failure develop, despite optimal medical therapy.

Previously, the stress of labor and delivery was thought to increase maternal mortality caused by rupture of the aorta. Consequently, a cesarean section was recommended as the mode of delivery. Although aortic rupture and dissection do occur during pregnancy, they are not as common as previously suspect. Moreover, most aortic ruptures during pregnancy occur before labor and delivery, and death during or immediately after labor and delivery is rare.7 Because of that, vaginal delivery seems to be safe in patients with uncomplicated coarctation of the aorta.

Antibiotic prophylaxis before delivery is recommended because of the potential risk of endocarditis in patients with coarctation of the aorta.
resistance observed during pregnancy can lead to the development or worsening of right-to-left blood shunting and cyanosis. In addition, increased blood volume and venous return to the heart promote increased shunting and heart failure.

Labor and the period immediately after delivery are critical. Hypotension, attributable to blood loss or a fall in peripheral resistance, an increase right-to-left shunting, and thus result in more cyanosis and hypoxia, with possible death caused by circulatory collapse and arrhythmias.43 Pregnancy in women with tetralogy of Fallot, who have not been treated surgically, was associated, in early studies, with a maternal mortality greater than 4% and a rather high perinatal mortality. Improvement of cyanosis by palliative operation reduces the risk during pregnancy.9 Total surgical correction of tetralogy of Fallot has resulted in successful pregnancies in the majority of cases.1,2,7,9,43-48 Two recent reports by Whittenmore and colleagues9 and Metcalfe et al28 described pregnancies in 37 women with palliative or corrective surgery without maternal mortality. Eighteen pregnancies in patients with cyanosis during pregnancy, after palliative surgery, were described by Whittenmore and associates.9 Eight patients had tetralogy of Fallot and/or transposition of the great arteries, plus pulmonary stenosis and a RAS. Pregnancy was interrupted in 17% for unspecified reasons. Fifteen percent of the remaining pregnancies resulted in spontaneous abortions. Thus, 13 of 18 pregnancies (72%) resulted in live-born infants. A somewhat higher incidence of live-born infants (78%) was observed in 23 pregnancies in 13 women who were acyanotic following surgical repair. Eleven of these patients had tetralogy of Fallot. Metcalfe and coworkers28 recently reported 40 pregnancies without maternal morbidity or mortality in 18 women with surgically-corrected tetralogy of Fallot. Five of the pregnancies were interrupted, and 5 ended in spontaneous abortions. Although these data were collected in a relatively small number of patients, they are compatible with an improved fetal outcome in surgically repaired lesions of tetralogy of Fallot.

Cardiac defects were reported in 15% of infants born to women with cyanosis and in 17% of acyanotic women. In contrast, CHD was found in only 1 of 30 infants born to women with surgically-corrected tetralogy of Fallot. The expected incidence of tetralogy of Fallot in the newborn of a woman with the same lesion and a healthy husband has been estimated to be 4%.19

Such a procedure is desirable before pregnancy because of the obvious improvement in both maternal and fetal outcome after surgical repair of tetralogy of Fallot. Patients who have only palliative procedures, such as anastomosis of the subclavian artery to the pulmonary artery (Blalock-Taussig), descending aorta to left pulmonary artery (Potts), or ascending aorta to right pulmonary artery (Watson-Cooley), are still subjected to a significantly higher risk during pregnancy. The incidence of pregnancy in patients who have had a total correction is increasing because of the increased total surgical correction rate over recent years. Patients who have had surgical repair should tolerate pregnancy well without any excess risk.46

Pregnancy in women with uncorrected or partially corrected tetralogy of Fallot carries a high risk. A maternal hematocrit level of greater than 60%, peripheral arterial oxygen saturation less than 80%, right ventricular hypertension, and syncopal attacks are all poor prognostic signs. Although tetralogy of Fallot may be surgically corrected during pregnancy,30 surgical intervention should be performed ideally before the childbearing years.

In cyanotic or symptomatic patients, close hemodynamic monitoring and frequent arterial blood gas determinations during labor, delivery, and the early postpartum period are recommended for early recognition and correction of any circulatory deterioration. Oxygen administration is advisable during labor and delivery, and antibiotic prophylaxis should be given to prevent bacterial endocarditis. Hypotension, resulting from blood loss or systemic arterial vasodilatation, should be promptly corrected with blood products and vasoconstrictive drugs.
**EBSTEIN'S ANOMALY**
Connolly and Warnes have recently reported the outcome of 49 pregnancies in 40 women with Ebstein's anomaly; cyanosis was present in 15 of the 40 women. Serious maternal cardiovascular complications were not observed. However, there was increased fetal loss, prematurity, and cardiac anomalies (7%) in the offspring.

**TRICUSPID ATRESIA**
Several cases of tricuspid atresia have been reported in association with pregnancy. A few cases of pregnancy with delivery of infants, who subsequently developed normally, have been described. The most recent case of a successful pregnancy in a patient with tricuspid atresia was reported by Hatjis and coworkers. They described a 30-year-old patient with tricuspid atresia who had undergone a Glenn procedure (anastomosis of the superior vena cava to the right pulmonary artery) at the age of 13 years and had a VSD with left-to-right shunting, cyanosis, clubbing, and arterial P02 of 38 mm Hg. At 32 to 33 weeks gestation, the patient delivered by cesarean section an infant with fetal growth retardation, who subsequently showed normal development.

Despite these reports, pregnancy in patients with tricuspid atresia carries a high risk of complications to both the mother and fetus. Heart failure, hemoptysis, stroke, and pulmonary embolism frequently occur. Fetal outcome is also poor in the majority of cases, with a high incidence of spontaneous abortions, premature deliveries, growth retardation, and neonatal deaths.

**TRANSPOSITION OF THE GREAT VESSELS**
Wilson et al have recently reported a favorable outcome in 14 pregnancies in 9 women with transposition of the great arteries after the Mustard operation. However, none of these women had cardiac symptoms before pregnancy. In contrast, a patient post-Mustard operation was recently reported to have developed atrial fibrillation and pulmonary edema at the 37th week of gestation, despite the absence of cardiac symptoms before pregnancy.

**TRUNCUS ARTERIOSUS**
Simon and Lustberg described a patient with truncus arteriosus, who underwent a successful pregnancy and full-term delivery of a healthy infant. However, the mother died suddenly on the third day postpartum. The cause of death was thought to be pulmonary infarction. We reported a 20-year-old patient with truncus arteriosus, who had a successful pregnancy and delivered a healthy infant without any complications.

**SINGLE VENTRICLE**
Successful pregnancies have been reported in mothers with a single ventricle. Yuzpe and colleagues reported on a 17-year-old patient with a single left ventricle, absent right ventricular sinus, levo-transposition of the great vessels with sinus solitus of viscera and atria, pulmonary valvular stenosis, PDA, bicuspid aortic valve, hypoplasia of the aortic arch, and origin of all coronary arteries from the posterior aortic sinus. The patient complained of limitation of exercise tolerance and exertional dyspnea, but otherwise tolerated pregnancy well. She delivered under general anesthesia a small, healthy baby by cesarean section.

Ahmed and associates reported a successful pregnancy in a 19-year-old primigravida with a single ventricle. The patient had a vaginal forceps delivery at 36 weeks gestation. Intrathecal morphine was administered for analgesia and resulted in hemodynamic stability. The baby was small for gestational age and weighed only 1700 g. Successful pregnancies were also described in patients with a single ventricle with transposition of the great vessels. In contrast, Mandel and Hirsch reported on a 29-year-old patient who had to undergo a therapeutic termination of pregnancy at 20 weeks' gestation because of severe
~eart failure. Eighteen hours after the procedure, the patient died.

In summary, although successful pregnancies have been reported in women with complex cyanotic CHD, pregnancy is associated with deterioration of functional capacity and worsening of symptoms in many patients and can even lead to maternal death. In addition, the incidence of spontaneous abortions, premature deliveries, small-for-gestational age infants, and cardiac as well as noncardiac congenital defects is high. Pregnancy should be prevented when a maternal hazard is believed to exist. In such patients, an early therapeutic abortion is indicated should a pregnancy occur. If termination of pregnancy is declined by the patient, care should consist of advice to the patient to reduce all strenuous activity during pregnancy, early detection and management of CHF, and treatment of cardiac arrhythmias. Antibiotic prophylaxis is recommended before delivery. Oxygen therapy should be administered from the onset of labor through 24-48 hours postpartum, with frequent assessment of arterial blood gases, in order to assure hemodynamic stability. Although vaginal delivery seems to be well tolerated in most cases, attempts should be made to shorten the second stage by forceps or vacuum delivery.

**Eisenmenger's Syndrome**

Review of the literature published until 1990 reveals that 70 women with Eisenmenger's syndrome became pregnant, at least once. Forty-four percent of these patients died in connection with one of their pregnancies. No statistically significant difference in maternal mortality among the first, second, and third pregnancies could be identified. Thus, a successful pregnancy in patients with Eisenmenger's syndrome cannot be taken as a positive prediction for further pregnancies. An especially high incidence of maternal death is associated with hypovolemia, thromboembolic phenomena, and preeclampsia/eclampsia. Cesarean sections and other surgical procedures are associated with extremely high maternal mortality. However, it is possible that patients requiring cesarean section represented a biased study group, indicating a severely compromised situation. Since maternal death was significantly greater when any kind of delivery was compared with elective termination of pregnancy, termination of pregnancy appears indicated in patients with Eisenmenger's syndrome.

Among the underlying malformations that may cause Eisenmenger's syndrome, VSD is the most frequent shunt. Maternal mortality in cases with VSD is 60%. Maternal mortality with ASD is 44%, followed by 42% mortality in association with PDA. The vast majority of all maternal deaths occurs during or within the first week after delivery.

In addition to the unfavorable maternal effect, pregnancy in women with Eisenmenger's syndrome is also associated with extremely poor fetal outcome. Only 25% of all pregnancies were reported by Gleicher's group to have reached term. At least 55% of all deliveries were clearly premature, and at least 30% of all delivered infants showed evidence of intrauterine growth retardation. This number represented almost half of all newborns for whom available information allowed valid clinical evaluation. The total perinatal mortality for this group reached 28% and was found to be strongly associated with prematurity. Based on these data, we recommend the following:

1. Cooperation of all involved medical services to obtain the best possible results.
2. Hospitalization for any sign of symptomatic or hemodynamic instability at any time during pregnancy.
3. Anticoagulation, starting at approximately 20 weeks' gestational age and continuing at least 1 month postpartum, even though the value of anticoagulation during pregnancy is based on theoretical grounds and remains to be proved.
4. With an increase in the number of episodes of exertional dyspnea—With advancing gestational age, the beneficial effect of oxygen on pulmonary vascular resistance and shunt flow 74,75 should be utilized.

5. Congestive heart failure, if it develops, should be treated with digitalis and diuretics.

6. Fetal well being should be followed throughout pregnancy with routine antepartum testing, such as the measurement of fetal growth by ultrasound, nonstress tests, and oxtocin challenge tests.

7. Premature delivery should be anticipated, and the patient should be hospitalized for any sign of premature uterine activity.

8. Spontaneous labor is preferable to induction. Such a policy should decrease the chance for prematurity and cesarean section delivery. It should be recognized, however, that in some institutions, the risk of a night delivery with an incomplete or a diminished medical staff might outweigh the risk of an induction. In cases of elective induction, evaluation of fetal lung maturity is recommended before delivery.

9. Labor and delivery should occur in an intensive care setting, next to an immediately available operating room.

10. Hemodynamic and electrocardiographic monitoring is suggested during labor, delivery, and early postpartum period.

11. Anticoagulation with heparin should be stopped with the onset of labor and reinstituted as soon as feasible after delivery.

12. Administration of high concentrations of oxygen is recommended during labor and delivery.

13. Epidural anesthesia represents the best method of anesthesia for these patients.

14. The repeated determination of blood gases during labor, delivery, and the puerperium is essential to detect changes in shunt flow.

15. Elective low forceps delivery seems preferable to normal spontaneous vaginal delivery, in order to shorten the second stage of labor. Vaginal delivery seems preferable to cesarean section delivery; however, cesarean section does not seem contraindicated.

16. A postpartum hospitalization of at least 2 weeks is suggested.

REFERENCES


62. Spinnato JA, Imarynck BJ, Cooper MW. Eisenmenger's syndrome in pregnancy: Epidural anesthesia for...