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Committee on Adolescent Health Care

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Gynecologic Considerations for Adolescents and Young Women With Cardiac Conditions

ABSTRACT: Congenital heart conditions are the most common type of congenital anomaly, affecting nearly 1% of U.S. births, or approximately 40,000 neonates, each year. As more female patients with congenital heart disease enter adolescence and adulthood, there is a growing need to address reproductive health in this population. Addressing contraceptive needs is particularly important for adolescents and young women with congenital heart disease, many of whom may have limited knowledge about how their condition or medications may affect their long-term health, including reproductive health. Decisions regarding the most appropriate contraceptive method require discussion of future pregnancy desires and personal preferences, as well as critical assessment of the patient's underlying disease and the relative risks and benefits of the contraceptive option. Because of the morbidity associated with pregnancy in individuals with cardiac conditions, the initiation of contraception should not be delayed due to concerns about potential contraindication. For those patients with valvular heart disease, cardiomyopathy, or hypertension, it is reasonable to initiate a progestin-only method until clarification of the safety of an estrogen-containing method is determined in conjunction with the patient's cardiologist. Contraceptive counseling should be patient-centered, free of coercion, and should address the most common misperceptions about contraceptive methods in a way that is age-appropriate and compatible with the patient's health literacy. To optimize maternal and infant health outcomes, planning for future pregnancies in these patients should be done in collaboration with maternal–fetal medicine subspecialists and cardiology specialists. Patients who continue their pregnancy should be referred to a pregnancy heart team.

Recommendations and Conclusions

The American College of Obstetricians and Gynecologists (ACOG) makes the following recommendations and conclusions on gynecologic considerations for adolescents and young women with cardiac conditions:

- Gynecologic care of adolescent girls and young women with cardiac conditions should occur in collaboration with the patient's cardiologist.
- Adolescent patients and young women with congenital heart disease may have a high unmet need for contraception compared with other adolescents.
- Because of the morbidity associated with pregnancy in individuals with cardiac conditions, the initiation of contraception should not be delayed due to concerns about potential contraindication. For those patients with valvular heart disease, cardiomyopathy, or hypertension, it is reasonable to initiate a progestin-only method until clarification of the safety of an estrogen-containing method is determined in conjunction with the patient's cardiologist.
- All progestin-only-containing contraceptives are categorized by the Centers for Disease Control and Prevention's (CDC) U.S. Medical Eligibility Criteria for Contraceptive Use (USMEC) as category 1 methods (no restriction) for those with uncomplicated or complicated valvular heart disease.
- Intrauterine devices (IUDs) are the recommended reversible option for those with high-risk cardiac

conditions. The levonorgestrel-containing (LNG) IUD provides excellent contraception while decreasing menstrual bleeding, which may be of substantial benefit to patients requiring anticoagulation or for whom anemia exacerbates their underlying cardiovascular status.

- Given the low risk of pelvic infection associated with IUD insertion, the risk of infectious endocarditis in IUD insertion is low, and antibiotic prophylaxis is not indicated for insertion.
- Potential risks posed by contraceptive use should be weighed against the increased risks of morbidity associated with pregnancy.
- Menstrual suppression may be helpful in patients with congenital heart disease to reduce anemia, and in patients with postural orthostatic tachycardia syndrome to reduce orthostatic symptoms in the premenstrual or menstrual period. Reduced menstrual bleeding, a noncontraceptive benefit of progestin-only contraceptives, also may improve anemia, regardless of whether menstrual suppression is achieved.

Introduction

Patients with congenital heart disease are living longer and healthier lives and may seek a range of gynecologic and obstetric care. Adolescents and young women with cardiac conditions may present to an obstetrician-gynecologist for management of menstrual symptoms, including those that exacerbate cardiac symptoms, and may want to discuss options for contraception. Adolescent patients and young women with congenital heart disease may have a high unmet need for contraception compared with other adolescents (1). The American Heart Association recommends contraceptive counseling for women of reproductive potential who have congenital heart disease (2). Cardiovascular changes in pregnancy generally result in more morbidity and mortality than they do from changes in contraceptive use.

This document is not comprehensive and will focus on the most common major cardiac conditions in adolescents and young women: congenital heart disease and postural tachycardia syndrome. Gynecologic care of adolescent girls and young women with cardiac conditions should occur in collaboration with the patient's cardiologist. For more information, see the USMEC (3); ACOG Practice Bulletin No. 206, *Use of Hormonal Contraception in Women With Coexisting Medical Conditions* (4); and the American Heart Association's guidance on management of adults with congenital heart disease (5, 6). For guidance on the care of pregnant and postpartum individuals with preexisting or new-onset acquired heart disease and interpregnancy care for women with heart disease, see ACOG Practice Bulletin No. 212, *Pregnancy and Heart Disease* (7).

Congenital Heart Disease

Congenital heart conditions are the most common type of congenital anomaly, affecting nearly 1% of U.S. births, or approximately 40,000 neonates, each year (8, 9). With advances in medical care, 1-year survival of infants born with congenital heart disease has risen to 83%, and many more individuals with congenital heart disease are surviving to adulthood (10). As of 2010, there were an estimated 225,000 female adolescents and young women (aged 13–24 years) living with congenital heart disease in the United States, and this number is expected to increase (11). As more female patients with congenital heart disease enter adolescence and adulthood, there is a growing need to address reproductive health in this population. Congenital heart disease may occur in the setting of other anomalies or genetic conditions, including Down syndrome, Marfan syndrome, DiGeorge syndrome, Noonan syndrome, Turner syndrome, CHARGE (coloboma, heart defects, atresia choanae, growth retardation, genital abnormalities, and ear abnormalities) syndrome, and Williams syndrome, all of which have reproductive health considerations (2, 6, 12). See Box 1 for a glossary of congenital heart conditions and the associated anatomic variants.

The American Heart Association's *Adult Congenital Heart Disease Anatomic and Physiologic* classification system incorporates anatomic variables and physiologic functioning and acknowledges that individuals with the same anomaly may have different levels of function (6). Patients are scored as I (simple), II (moderate), and III (great) based on complexity of the anomaly, and A to D based on physiologic stages. In general, patients with isolated small atrial septal defects, ventricular septal defects, mild isolated pulmonic stenosis, a previously ligated ductus arteriosus, and repaired atrial septal defects and ventricular septal defects are considered to have simple anatomy (or anatomic level 1). Those with no hemodynamic or anatomic sequelae, no arrhythmias, normal exercise capacity, and normal renal, pulmonary, and hepatic function are at physiologic stage A. These patients comprise the majority of adolescents and young women presenting to obstetrician-gynecologists for care and they are candidates for all forms of contraception.

In contrast, in adolescents and young adults with more complex cardiac conditions, such as Fontan circulation procedure or single ventricle (anatomic level III) or severe pulmonary hypertension (physiologic stage D), the heart often is unable to tolerate abrupt changes in hemodynamic status. These patients cannot tolerate vasovagal events that result in reduction in preload. In general, estrogen-containing contraceptives are contraindicated in these patients who also are at risk of thromboembolic events.

Pregnancy in individuals with congenital heart disease is associated with increased risk of maternal and infant morbidity and mortality (7, 13). Rates of

Box 1. Common Diagnoses in Patients With Congenital Heart Disease

Atrioventricular Septal Defect (atrioventricular canal): A defect in the atrioventricular septum involving the atria and ventricles. A common valve replaces the mitral and tricuspid valves. Atrioventricular septal defects are often found in children with trisomy 21.

Ebstein's Anomaly: Apical displacement of the tricuspid valve creates a large right atrium and small right ventricle leading to right-sided heart failure.

Eisenmenger Syndrome: Because of severe pulmonary hypertension from left-to-right shunting through a congenital cardiac defect, a reversal of the original shunt occurs (now right-to-left), leading to cyanosis.

Fontan Circulation: A result of a series of three surgeries done for hypoplastic left heart. The superior and inferior vena cava connect to the pulmonary artery, and the single ventricle is used to pump oxygenated blood to the body.

Paradoxical Embolism: Occurs when a thrombus in a systemic vein dislodges and crosses across an atrial communication into the systemic arterial circulation. This may be associated with a thromboembolic stroke (cerebrovascular accident).

Patent Foramen Ovale: Present in approximately 20% of the general population. It is not a true atrial septal defect (eg, no atrial septal tissue is missing), but represents persistence of normal fetal anatomy. Patent foramen ovale is usually clinically silent.

Tetralogy of Fallot: The most common form of congenital cyanotic heart disease. The 4 components include: ventricular septal defect; valvular or subvalvular pulmonary stenosis, or both; an overriding aorta; and right ventricular hypertrophy. There also may be a right-sided aortic arch and atrial septal defect. This is usually repaired at age 6–12 months.

D-transposition of the Great Vessels: Occurs when the pulmonary artery arises from the left ventricle, and the aortic from the right ventricle, causing two separate parallel circulations and leading to cyanosis. Presents with cyanosis on the first day of life when the ductus arteriosus closes. Historically, d-transposition of the great vessels was palliated by the atrial switch (Mustard or Senning) procedure, but in the past 3 decades, it has been surgically corrected by the arterial switch operation (Jatene procedure) which carries less thrombotic risk.

Data from Cunningham JW, Brown DW. Congenital heart disease. In: Lilly LS, editor. Pathophysiology of heart disease: an introduction to cardiovascular medicine. 7th ed. Philadelphia, PA: Wolters Kluwer; 2021. p. 398–426 and Webb GD, Smallhorn JF, Therrien J, Redington AN. Congenital heart disease in the adult and pediatric patient. In: Zipes DP, Libby P, Bonow RO, Mann DL and Tomaselli GF, editors. Braunwald's heart disease: a textbook of cardiovascular medicine. 11th ed. Philadelphia, PA: Elsevier; 2019. p. 1519–73.

unintended pregnancy in adult women with congenital heart disease are reported to be approximately 54% (14), yet studies suggest many women with congenital heart disease do not receive contraceptive counseling that addresses their condition from any health care professional (14). A study of prepregnancy health care among adolescents and women with congenital heart defects reported that of 2,003 female patients aged 15–44 years who became pregnant, 9% were prescribed potentially teratogenic medication for their condition in the year before pregnancy (15). Additionally, less than 1% of adolescents and women received all American Heart Association-recommended tests and assessments before pregnancy. For patients with severe heart disease at risk of morbidity and mortality, working with a pregnancy heart team (including obstetric care professionals, maternal–fetal medicine subspecialists, cardiologists, and an anesthesiologist, at a minimum) increases positive outcomes. Current guidelines for pregnancy and heart disease recommend counseling patients to avoid pregnancy and to discuss options, including induced abortion, if they have severe heart disease (eg, an ejection fraction less than 30% or class III or IV heart failure, severe valvular stenosis, Marfan syndrome with aortic diameter more than 45 mm, bicuspid aortic valve with aortic diameter more than 50 mm, or pulmonary arterial hypertension) (7). Patients who continue their pregnancy should be referred to a pregnancy heart team (7).

Addressing contraceptive needs is particularly important for adolescents and young women with congenital heart disease, many of whom may have limited knowledge about how their condition or medications may affect their long-term health, including reproductive health. In a small survey of adolescents and young adults with congenital heart disease, only one half of those not trying to get pregnant and not currently pregnant reported using a moderately or highly effective contraceptive method (pill, contraceptive patch, ring, or implant) during their last sexual intercourse, suggesting an unmet need for contraceptive counseling and services (16, 17). Additionally, a survey of adult and pediatric cardiology fellows suggested that many do not feel prepared to counsel their patients on contraception and instead refer patients to obstetrician–gynecologists (18).

Contraceptive Considerations in Adolescents and Young Women With Congenital Heart Disease

See Box 2 for general considerations regarding contraception. Decisions regarding the most appropriate contraceptive method require discussion of future pregnancy desires and personal preferences, as well as critical assessment of the patient's underlying disease and the relative risks and benefits of the contraceptive option (7). Because of the morbidity associated with pregnancy in individuals with cardiac conditions, the initiation of

contraception should not be delayed due to concerns about potential contraindication. For those with valvular heart disease, cardiomyopathy, or hypertension, it is reasonable to initiate a progestin-only method (eg, quick start depot medroxyprogesterone acetate [DMPA] or progestin-only pill) until clarification of the safety of an estrogen-containing method is determined in conjunction with the patient's cardiologist. Depot medroxyprogesterone acetate is categorized as USMEC category 3 (the theoretical or proven risks usually outweigh the advantages of using the method) for women with ischemic heart disease or stroke (3). Short-acting methods have a higher rate of failure when compared with long-acting methods. Dual methods are encouraged to decrease the risk of sexually transmitted infections and unintended pregnancy. See Table 1 for considerations by contraceptive method. Obstetrician-gynecologists should collaborate with a cardiologist knowledgeable in congenital heart disease for additional considerations regarding contraceptive choices for patients with these conditions.

Intrauterine Devices

Intrauterine devices are the recommended reversible option for those with high-risk cardiac conditions (7). The LNG-IUD provides excellent contraception while decreasing menstrual bleeding, which may be of substantial benefit to patients requiring anticoagulation or for whom anemia exacerbates their underlying cardiovascular status. Obstetrician-gynecologists should be aware of the association between cardiac conditions and Müllerian anomalies and the potential for difficulty when inserting an LNG-IUD (19). Data show no increased thrombotic risk with use of an IUD among healthy women compared with nonusers (20). Patients with Fontan circulation, single ventricle, or pulmonary hypertension should have IUDs inserted in a facility with cardiac monitoring during placement to observe for hypotension or arrhythmia (12) and with health care professionals able to manage vasovagal events in patients with complex cardiac physiology. Health care professionals should have the ability to monitor the patient and provide intravenous hydration, if needed. Given the low risk of pelvic infection associated with IUD insertion, the risk of infectious endocarditis in IUD insertion is low, and antibiotic prophylaxis is not indicated for insertion (21). The CDC's USMEC notes that the insertion of an LNG-IUD does not pose major bleeding risks in individuals receiving long-term anticoagulant therapy (3). In adolescents with multiple complex medical conditions who may benefit from IUD insertion under anesthesia, coordinating insertion during other scheduled surgical procedures can minimize anesthesia exposure. However, because of the potential harm of delaying provision of contraception, initiating contraception should not be dependent on the presence of another scheduled procedure. A short-term contraceptive method may be offered as a bridge if the balance of risks and

Box 2. General Contraceptive Considerations in Adolescents and Young Women With Cardiac Conditions

- Cardiovascular changes in pregnancy generally are more dangerous than changes with contraceptive use.
- In all women, progestins are associated with lower thrombotic risks than estrogen-containing methods.
- Risk of estrogen use is stratified depending on cardiac defect and physiology.
- Although package inserts for progestin-containing medications suggest that progestins may cause fluid retention, current evidence does not support this effect.

benefits supports waiting until an IUD can be inserted during another scheduled procedure.

In patients who are receiving anticoagulant therapy, or in whom anemia would worsen their cardiac status, copper IUDs may not be the optimal first choice because they can increase menstrual blood loss for some individuals. Also, prompt attention to heavy or persistent breakthrough bleeding with the use of any hormonal method should be given to patients with complex cardiac conditions because anemia may compromise their cardiac physiological status.

Progestin-only Methods

All progestin-only containing contraceptives are categorized by the CDC's USMEC as category 1 methods (no restriction) for those with uncomplicated or complicated valvular heart disease (3). A systematic review of progestin-only contraception noted there are some data suggesting increased odds of venous thromboembolism (VTE) with the use of high-dose methods; however, the authors acknowledge confounders, including that the population who choose to use DMPA may differ from the general population of all hormonal contraceptive users. Depot medroxyprogesterone acetate often is used for menstrual suppression in patients with underlying medical conditions for whom estrogen is contraindicated (20). Reduced menstrual bleeding, a noncontraceptive benefit of progestin-only contraceptives, also may improve anemia, regardless of whether menstrual suppression is achieved. A retrospective study of 237 reproductive-aged female patients (aged 14 to 52 years) with congenital heart disease showed no worsening of cardiovascular status in those who used oral desogestrel (without ethinyl estradiol), LNG-IUDs, or contraceptive implants (22). In addition to the norethindrone .35-mg progesterone-only pill, another progestin-only method (drospirenone 4 mg) was approved by the U.S. Food and Drug Administration; with a regimen of 24 active tablets with 4 inactive tablets. Based

Table 1. Considerations for the Use of Contraceptive Methods in Patients With Cardiac Conditions*

Hormonal Contraceptive Method	Considerations for Use
Intrauterine device	Potential for vagal reaction with insertion; those patients with complex cardiac defects may need cardiac monitoring during placement to observe for arrhythmia. Health care professionals should be prepared to handle hypotension, arrhythmia, and syncopal events in these patients. Intrauterine device insertion and removal is safe for patients receiving anticoagulation therapy. In patients who are anticoagulated, or in those whom anemia would worsen their cardiac status, copper IUDs may not be the optimal first choice because they can increase menstrual blood loss for some individuals. The insertion of a levonorgestrel IUD does not pose major bleeding risks in individuals receiving chronic anticoagulant therapy.
Implants	Health care professionals should be prepared to handle syncopal episodes in these patients. Anticoagulation status should be considered with insertion and removal.
Depot medroxyprogesterone acetate	Depot medroxyprogesterone may exacerbate bone loss in patients on long-term warfarin therapy.
Combined hormonal contraception (eg, oral contraceptives, vaginal rings, contraceptive patch)	Contraindicated in patients with a history of thrombotic events or with these conditions: Fontan physiology, cyanotic disease, pulmonary arterial hypertension, mechanical valves, and D-transposition of the great vessels after atrial switch surgery (Mustard or Senning procedure). There is an increased risk of thrombosis.
Norethindrone progestin-only pill	Bosentan, used for pulmonary hypertension, may lower efficacy of this method.
Emergency contraception	The presence of a cardiac condition is not a contraindication to the use of emergency contraception, including emergency contraceptives or insertion of the copper IUD.

Abbreviation: IUD, intrauterine device.

*For additional information, see Curtis KM, Tepper NK, Jatlaoui TC, Berry-Bibee E, Horton LG, Zapata LB, et al. U.S. medical eligibility criteria for contraceptive use, 2016. *MMWR Recomm Rep* 2016;65(RR-3):1–103. <https://www.cdc.gov/reproductivehealth/contraception/mmwr/mec/summary.html>.

on a single-arm study of 953 females, the label reports a Pearl Index (the number of contraceptive failures per 100 women-years of exposure) (95% CI) of 4.0 (23).

In general, progestin-only methods have a lower risk of thrombosis and are safer than combined hormonal contraception. The effect of progestins on risk of thrombosis depends on the progestin used, the dose, and the route of delivery. Initiation of most progestin-only-containing contraceptives are categorized as US-MEC category 2 (the advantages of using the method generally outweigh the theoretical or proven risks) for women with VTE, ischemic heart disease, and stroke (3). Initiation of DMPA is safe for those with valvular heart disease, cardiomyopathy, and hypertension, yet is categorized as USMEC category 3 (the theoretical or proven risks usually outweigh the advantages of using the method) for women with ischemic heart disease or stroke. Depot medroxyprogesterone acetate is associated with a hypoestrogenic effect and reduced high-density lipoprotein cholesterol levels. In patients with a personal history of ischemic heart disease, the risks of DMPA may outweigh the benefits (4).

Depot medroxyprogesterone acetate use has been associated with weight gain and increased body fat composition (24, 25). Studies on other progestin-only methods are limited and do not report consistent results (25, 26). In patients with substantial impairment in cardiac function and inability to tolerate weight gain

because of cardiac status, other forms of contraception should be considered.

The primary source of the elevation of blood pressure for users of combined hormonal contraception is the estrogen component; however, synthetic progestins, with the exception of drospirenone, lack the natriuretic and blood pressure-lowering effects of natural progesterone (27). One physiology study has shown an increase in plasma volume and total extracellular volume with progesterone administration (28). A systematic review of the data on contraceptive safety for women with congenital heart disease concluded that the effects of fluid balance are likely of minimal clinical meaning (21). The use of drospirenone 4 mg is contraindicated in those at high risk of hyperkalemia (23). Potential risks posed by contraceptive use should be weighed against the increased risks of morbidity associated with pregnancy.

Combination (Estrogen and Progestin) Methods

The American College of Cardiology and American Heart Association's 2018 *Guidelines for Adults with Congenital Heart Disease* state that when compared with estrogen-containing contraceptives in individuals without congenital heart disease, estrogen-containing contraceptives potentially are harmful for women with congenital heart disease who are at increased risk of thromboembolic events, specifically cyanotic disease,

Fontan physiology, mechanical valves, prior thrombotic events, and pulmonary arterial hypertension (6). Additionally, although high-quality data on contraceptive use in patients with congenital heart disease are lacking, estrogen-containing contraceptives also may be harmful for individuals who have D-transposition of the great vessels after atrial switch surgery (Mustard or Senning procedure) (21). See Table 2 for cardiac conditions associated with an increased risk of VTE that preclude the use of combined (containing estrogen and progestin) hormonal contraception. For the general population, use of combined hormonal contraception independently is associated with a small increase in risk of myocardial infarction and ischemic stroke. Whereas the overall risks are extremely low in healthy adolescents and young women, these risks may be elevated for those with underlying complex heart disease. Ethinyl estradiol increases hepatic production of clotting factors that increase VTE risk. A 15-year cohort study of nonpregnant female individuals of reproductive age with no history of cardiovascular disease or cancer reported a baseline rate of VTE risk of 2.1 per 10,000 exposure years for nonusers of hormonal contraception. This increases to 6.2 per 10,000 exposure

years with users of 30–40-microgram-containing combined oral contraceptives (OCs) (29). For women with complicated valvular disease (pulmonary hypertension, risk for atrial fibrillation, or history of subacute bacterial endocarditis), combined hormonal contraception is classified as an USMEC category 4 method (condition that represents an unacceptable health risk if the contraceptive method is used) (3).

In healthy adolescents, combined oral contraceptive use does not appear to increase blood pressure (30). In susceptible individuals, ethinyl estradiol may increase blood pressure due to an increased hepatic production of angiotensin II or direct effect on the brain and the central sympathetic nervous system (27). Estradiol administration also increases intravascular volume caused by decreased transcapillary escape of albumin (28). Relative risk of myocardial infarction and ischemic stroke is ethinyl estradiol-dose-dependent (31), yet these risks still are lower than the risk associated with pregnancy. Given the anomalous circulation in patients with certain underlying congenital heart disease, expert opinion suggests that the risk of myocardial infarction and ischemic stroke would be magnified in these individuals.

Table 2. General Guide for the Use of Estrogen-Containing Contraception by Cardiac Condition in the Setting of Good Cardiac Function

Cardiac Condition	Can This Patient Use Estrogen-Containing Contraceptives? (Yes/No)
Simple	
Atrial septal defect (secundum), patent foramen ovale, and ventricular septal defects (small)	Yes, if no reversal of flow or pulmonary hypertension. A right-to-left shunt through a septum increases the risk of thromboembolic stroke (paradoxical embolism).
Repaired atrial septal defect with no residual or ventricular septal defect spontaneously closed	Yes
Mitral valve prolapse or mild pulmonic stenosis	Yes
Moderate	
Mechanical valve	No
Aortic stenosis	Yes
Coarctation of the aorta	Yes
Tetralogy of Fallot (repaired)	Yes
Moderate pulmonary stenosis	Yes, if not cyanotic
Repaired atrioventricular septal defect (atrioventricular canal) without pulmonary hypertension	Yes
Complex	
Cyanotic heart disease	No
Fontan circulation	No
D-transposition of the great vessels	<ul style="list-style-type: none"> ● Status post-atrial switch (Mustard and Senning) procedure: progestin methods are preferred. ● Status post-arterial switch (Jatene) procedure: estrogen methods are not contraindicated.
Single ventricle	No
Pulmonary hypertension	No

Emergency Contraception

The presence of a cardiac condition is not a contraindication to the use of emergency contraception (3, 7). Progestin-only and ulipristal acetate emergency contraceptive methods are generally better tolerated and are more efficacious than combined hormonal regimens and may be preferred in the setting of cardiac conditions. Insertion of a copper IUD is the most effective method of emergency contraception when inserted within 5 days after unprotected intercourse, provides ongoing contraception, and should be made available to patients at high risk of pregnancy morbidity and mortality (32). However, copper IUDs may not be the optimal first choice for individuals who have received anticoagulation therapy or in whom anemia would worsen their cardiac status because they can increase menstrual blood loss for some individuals. See Table 1 for more information on IUD use.

Cardiac Medications With Gynecologic Considerations

Patients with mechanical heart valves, Fontan circulation, or at substantial thromboembolic risk may be placed on warfarin therapy. Warfarin is a known teratogen. Adequate contraception that minimizes bleeding is paramount in these patients. About 10% of children with long-term warfarin therapy will have decreased bone mineral density ($Z\text{-score} \leq -2.0$). Children at the highest risk are those with complex medical conditions and low body mass index (33). Although bone loss in patients using DMPA for contraception is reversible, patients with known decreased bone density on warfarin therapy theoretically may not be able to recover bone density loss. Because of the potential long-term consequences of warfarin use on bone density, use of DMPA should be individualized in patients who were treated with warfarin as children.

Bosentan is used for the treatment of pulmonary hypertension. Pharmacokinetic studies show a decrease in ethinyl estradiol and norethisterone (norethindrone) concentrations through induction of CYP3A4 activity with the use of this medication, which may reduce efficacy of oral contraceptives (34, 35).

Patients with cardiac conditions may be prescribed angiotensin-converting enzyme inhibitors, aldosterone antagonists, coumadin, or other medications with potential fetal adverse effects (7). Adolescents and their parents or guardians often are unaware of the potential risks of some cardiac medications to offspring. This information can provide an opportunity for patients to engage in reproductive life planning with their cardiologist and their obstetrician-gynecologist and to discuss contraceptive options as well as the best treatment while trying to become pregnant.

Postural Tachycardia Syndrome

Postural tachycardia syndrome, also called postural orthostatic tachycardia syndrome (known as POTS), is

defined by daily symptoms of chronic orthostatic intolerance associated with tachycardia but without hypotension when standing. Adolescents may seek gynecologic care because symptoms of postural tachycardia syndrome can worsen in the premenstrual period or during menses. Common symptoms, including fatigue, nausea, and dizziness, can limit school and sports participation. Ninety percent of cases occur in female patients, with symptoms presenting usually within 1–2 years after puberty and often improving in late adolescence (36, 37). The reported prevalence of postural orthostatic tachycardia syndrome is 0.2% (38). The etiology of postural orthostatic tachycardia syndrome is varied, but often is most associated with autonomic dysfunction. Other causes include excessive sympathetic discharge (hyperadrenergic form), Ehlers Danlos syndrome, low intravascular volume, and mast cell activation syndrome (39, 40). Prolonged bed rest (greater than 23 hours) can lead to gravitational deconditioning and induce symptoms (39). The diagnosis often is clinical but can be confirmed by tilt table testing. Management is through a combination of hydration and exercise combined with pharmacologic methods to support volume (eg, fludrocortisone) and mitigate autonomic dysfunction (eg, beta blockers, selective serotonin reuptake inhibitors) (39).

Contraceptive Considerations for Patients With Postural Orthostatic Tachycardia Syndrome

The estrogen in hormonal contraception can increase angiotensin II and cause fluid retention, which may relieve symptoms in patients with postural orthostatic tachycardia syndrome. Use of continuous or extended-cycle combined hormonal contraception may alleviate symptoms triggered by menses. Inappropriate low aldosterone and low blood volume have been identified in some patients with postural orthostatic tachycardia syndrome, so drospirenone-containing oral contraceptives are not recommended for use in this population (36). Because pain and anxiety associated with the insertion of IUDs and contraceptive implants may trigger postural tachycardia orthostatic syndrome symptoms, health care professionals should be prepared to handle syncopal episodes in these patients.

Menstrual Suppression in Patients With Congenital Heart Disease and Postural Orthostatic Tachycardia Syndrome

If choosing contraceptives for menstrual suppression, the same relative rates of amenorrhea apply, and risk stratification based on cardiac defect is required. Menstrual suppression may be helpful in patients with congenital heart disease to reduce anemia, and in patients with postural orthostatic tachycardia syndrome to reduce orthostatic symptoms in the premenstrual or menstrual period. Norethindrone in 5-mg tablets is not indicated for contraceptive use but can be titrated in

dosages of 5–15 mg daily for menstrual suppression in patients who have underlying cardiac conditions (41). A small proportion of norethindrone is metabolized to ethinyl estradiol (approximately equivalent to 4 micrograms of ethinyl estradiol per 5 mg of norethindrone) (42). Norethindrone is still classified as a progestin in terms of thrombotic risk. For information on management of acute bleeding, see ACOG Committee Opinion No. 785, *Screening and Management of Bleeding Disorders in Adolescents With Heavy Menstrual Bleeding* (41).

Other Cardiac Conditions

See the USMEC for recommendations for patients who have peripartum cardiomyopathy or who have undergone cardiac transplant (3). Conditions such as hyperlipidemia, VTE, and cardiomyopathy are covered in ACOG's Practice Bulletin No. 206, *Use of Hormonal Contraception in Women With Coexisting Medical Conditions* (4).

Shared Decision Making and Patient-Centered Counseling

As with all adolescents, the obstetrician–gynecologist routinely should address the contraceptive needs, expectations, and concerns of adolescents and young women with cardiac conditions regardless of age or previous sexual activity (43). Special consideration should be given to the risks related to contraceptive method use and the risks of pregnancy related to the specific cardiac condition. Obstetrician–gynecologists should engage in shared decision making with the patient and engage in conversations with the patient's cardiologist. Contraceptive counseling should be patient-centered, free of coercion, and should address the most common misperceptions about contraceptive methods in a way that is age-appropriate and compatible with the patient's health literacy (43). To optimize maternal and infant health outcomes, planning for future pregnancies in these patients should be done in collaboration with maternal–fetal medicine subspecialists and cardiology specialists. Individuals with congenital heart conditions should be aware of the potential long-term sequelae of pregnancy on their cardiac condition and recurrence risk of congenital heart disease in their offspring. Individuals with complex congenital or noncongenital heart disease should be treated by a pregnancy heart team during pregnancy (7). The composition of the team will vary depending on the patient's cardiac condition and the resources of the individual institution; however, the team should include obstetric care professionals, maternal–fetal medicine subspecialists, cardiologists, and an anesthesiologist, at a minimum (7).

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