transposition, Ebstein's anomaly, or left atrial isomerism. Detailed use of doppler velocimetry and motion-mode analysis may differentiate second-degree heart block from atrial bigeminy resulting from frequent blocked premature atrial beats. Echocardiography may also reveal cardiomyopathy and heart failure that may evolve in the setting of autoimmune-mediated heart block.[395]

4. The presence of fetal bradycardia may suggest an underlying diagnosis of IAS.[413] Options for prenatal diagnosis are limited, but magnetocardiography, if available, can provide diagnostic confirmation.[414] Prenatal evaluation also includes monitoring fetuses for ventricular arrhythmias. Postnatal evaluation with assessment of neonatal QT interval and mutational ion panels can confirm the diagnosis of suspected IAS. Recommendations for the management of IAS during pregnancy are provided in Section 12.
5. Since the fetal cardiovascular system has an immature Starling capability, fetal cardiac output is rate dependent.[415] Nonstress testing may be unreliable for assessing fetal status in the setting of fetal bradycardia because of blunted autonomic response, which impairs the normal variability of fetal heart patterns generally observed in otherwise healthy fetuses.[416] A normal biophysical profile correlates with reassuring fetal oxygenation and acid/base status.
6. Autoimmune heart block and cardiomyopathy can evolve over a variable period of time and have been reported to occur between weekly fetal assessments.[417] Periodic monitoring to detect evolving disease enables shared decision-making between the patient and her cardio-obstetrics team regarding interventions such as maternal steroid initiation. Additionally, depending upon the gestational age, steroids for fetal lung maturity can be given if further fetal deterioration prompts consideration of preterm birth.
7. The literature demonstrates variables results in the efficacy of antenatal fluorinated steroids for the treatment of fetal heart block. Current literature reporting on steroid treatment for isoimmune heart block is nonrandomized and underpowered. While third-degree heart block has not been demonstrated to be reversible, at least 1 study demonstrates that mild forms of fetal heart block may be reversible.[402] Chronic use of high-dose fluorinated steroids antenatally can lead to potential fetal complications including growth restriction and oligohydramnios. Detailed discussions between the patient and the cardio-obstetrics team are required to weigh potential risks and benefits.
8. Treatment of maternal autoantibody-mediated fetal cardiomyopathy/endocardial fibroelastosis with dual therapy including fluorinated steroids and IVIG may improve outcomes.[404] However, data are limited regarding timing, dosage, and frequency of treatment, and IVIG can cause maternal complications, including infection and hypersensitivity reactions, leading to premature delivery.
9. While beta-adrenergic agents can increase the heart rate in fetuses with fetal bradycardia, overall fetal outcome improvement has not been documented in the literature.[418] Chronic maternal administration of beta-adrenergic agents can lead to severe maternal complications, including pulmonary edema, electrolyte abnormalities, and maternal cardiac dysfunction.[419,420]

Section 12 Inherited arrhythmia syndromes

12.1. Management and risk stratification of inherited arrhythmia syndromes during pregnancy

Inherited arrhythmias syndromes (IAS) encompass a number of conditions, namely LQTS, Brugada syndrome (BrS), CPVT, early repolarization syndrome, Andersen-Tawil syndrome, and short QT syndrome (SQTS). Whether or not a genotype is identified, the vast majority are inherited in an autosomal dominant manner. To date, although genetic testing is exceptionally useful in confirming a clinical diagnosis and providing the opportunity for testing at-risk relatives, it is not universally diagnostic. As such, some individuals will have this diagnosis made clinically with no accompanying pathogenic variant identified. This is particularly the case for BrS, for which there is currently interest in whether the monogenic approach has limited the understanding of this condition, and advice to treat exists regardless of the results of genotyping availability.[15] All of these conditions carry an increased risk for ventricular arrhythmias and sudden cardiac death. They often share the unusual features of being sensitive to precipitants, such as medications or particular environmental situations. Patient education is exceptionally important in these conditions, as affected individuals must learn to modulate their risk. Though many data center on genotype- and phenotype-positive individuals, in some conditions the advice to treat exists regardless of genotyping availability or results.

Management of the pregnant woman with an inherited arrhythmia has a limited evidence base. In the absence of pregnancy-specific data, management is guided by evidence-based treatment established in nonpregnant patients. For the management of the fetus in pregnancies complicated by IAS, specifically LQTS, see Section 12.2.