demonstrating longer ICTs (60.9 ms vs. 13.6 ms for the blocked PACs).[346] Blocked atrial bigeminy typically results in ventricular rates between 75 and 90 bpm.[347] Rates below 65 should increase suspicion for AVB.[347] Fetuses with normal hearts are usually asymptomatic, thus blocked atrial bigeminy does not require antiarrhythmic therapy. Expectant mothers should be advised to avoid stimulants such as caffeine. While atrial ectopy persists, the fetal heart rate should be monitored weekly, as blocked atrial bigeminy increases the risk for SVT more than random isolated PACs.[336,343]

Bradycardia due to Atrioventricular Block

Between 90% and 95% of fetal AVB is related to either an isoimmune process (40%) or a structural abnormality of the conduction system in complex CHD (50% to 55%).[30] Management in either situation includes close serial assessment of the fetal heart rate and observation for the development of heart failure. If the fetal heart rate drops below 55 bpm or if there are signs of fetal heart failure, hydrops or underlying severe CHD, maternal treatment with beta-sympathomimetics may be used to augment the escape ventricular rate.[348] Terbutaline is a commonly used, generally well-tolerated agent.[348]

Structural defects associated with AVB include CCTGA, heterotaxy with LAI, DORV, and AVSD.[30] The prognosis for fetuses with congenital AVB and structural heart disease is generally poor. In one study from two tertiary referral centers, only 9 of 16 such pregnancies intended to carry to term resulted in live births, and only 3 were still alive at 1 year of age.[349] In a more recent and optimistic study at a single institution, 5 of 7 fetuses (71%) with AVB and LAI survived to live birth, and remained alive at a year of age.[344]

The true prevalence of anti-SSA anti-SSB maternal autoantibodies in the general population is not known, but fetal AVB may develop regardless of the presence or absence of overt maternal symptoms of lupus or Sjögren syndrome. High SSA levels (≥50 U/mL) appear to correlate with increased fetal risk.[315] In pregnancies of known serology positive mothers with no previously affected children, between 1% and 5% of fetuses develop AVB.[106,350–352] If a previous child had AVB or neonatal lupus, the risk increased strikingly to between 11% and 19%. The presence of maternal hypothyroidism or vitamin D deficiency further increases the risk.[353]

AV conduction can be assessed by measuring the mechanical PR interval in at-risk fetuses. However, it has not been possible to demonstrate a progression from first-degree AV block to second-degree AV block and then complete AV block in all fetuses. Serial fetal echocardiographic screening is recommended in expectant mothers with

known +SSA/SSB autoantibodies beginning at 16 weeks, then weekly or every other week to 28 weeks in a first child.[30] For mothers with a prior child with AVB or neonatal lupus, screening at least weekly from 16 to 28 weeks is recommended.[30]

In addition to screening for AVB, serial fetal echocardiograms should include observation for tachyarrhythmia, including atrial flutter (AFL), VT, or junctional ectopic tachycardia, all of which have been reported in fetuses with isoimmunization.[354,355] Mitral or tricuspid valvitis, pericarditis, myocardial dysfunction, or endocardial fibroelastosis (EFE) may coexist.[110,356] Concern for myocardial involvement may justify additional fetal echocardiographic assessment in the third trimester even if the rhythm has remained sinus throughout gestation.[30] Untreated, fetal mortality ranges from 9% to 34%, with risk of death increased in those diagnosed before 20 weeks, or with a ventricular rate <50 bpm, fetal hydrops, or impaired left ventricular function at diagnosis.[357–359]

Treatment with dexamethasone (4 to 8 mg/day) may be considered for the fetus with second-degree AV block or first-degree block with other evidence of cardiac involvement as reversal or stabilization of incomplete block and improvement in hydrops, ventricular function, and EFE has been reported.[30,358,360] Treatment may also be considered in fetuses manifesting with complete block to prevent dilated cardiomyopathy.[30,357,359] However, the potential, and not universally established, benefits of dexamethasone must also be weighed against the potential for significant side effects such as growth restriction, ductal constriction, maternal diabetes, oligohydramnios, and possible CNS side effects.[357,361,362] A recent study evaluated a small cohort of 16 patients treated with dexamethasone as fetuses for AVB and found normal neurodevelopment, including overall intelligence and the subdomains of cognitive function at 6 years of age.[363] In fetuses with systolic dysfunction or EFE, IVIG may also be considered to potentially improve survival.[30]

Channelopathies such as long QT and Brugada syndromes are far less frequent etiologies of AVB in the fetus.[364–367] Prenatal management entails maternal avoidance of QT prolonging agents and serial surveillance for the development of VT, progressive conduction disease, or dilated cardiomyopathy.[30]