The Problem Patient

Mother Without Cardiac History Bears Second Child with Heart Block

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Case Presentation

A 23-year-old black woman (gravida V, para IV) was admitted to the labor floor of the medical center at 39 weeks with regular contractions every five minutes. She had had two children (with normal pregnancies and deliveries) by a previous partner. Her next three pregnancies (which included the current one) were by a different man. Her youngest child, a girl, had been in congestive failure at birth. Congenital complete heart block was diagnosed, and a pacemaker was placed immediately. The child is now two years old and doing well. The mother was sickle cell trait positive. Her history was otherwise unremarkable. Her physical examination was normal. Blood pressure was 110/70. Other vital signs were stable. The fundal height was 34 cm, and the fetal presentation was vertex. The patient was placed on an external monitor, which recorded a normal baseline fetal heart rate of 130 to 140.

After an hour of apparently normal external monitoring, the baseline heart rate converted to a bradycardia of approximately 50 to 60. The maternal pulse at this time was 80, and the fetal heart rate was checked by fetoscopy. Artificial rupture of membranes was performed; the amniotic fluid was clear. An internal scalp electrode was placed; it revealed a heart rate of 47 or 48, with no variability. A fetal ECG showed complete heart block with an atrial rate of approximately 125 and a ventricular rate of 47. Real-time sonography and echocardiography, performed to rule out congestive heart failure or a congenital cardiac abnormality, were negative. Fetal scalp 

The infant was immediately transferred to the neonatal intensive care unit for observation. Congenital heart block was confirmed. Because of cardiac enlargement, a rapid atrial rate of 166, and a slow ventricular rate of 47, a permanent pacemaker was placed on the second day of life.

The mother was transferred to the recovery room, where an irregular heart rate was noted about 30 minutes after delivery. However, she was asymptomatic. Her irregular cardiac rhythm was intermittent, lasting for only about two to five minutes before converting to normal. An ECG initially revealed a normal sinus rhythm and nonspecific T-wave changes. A second ECG taken during a period of pulse irregularity revealed a second-degree atrioventricular block of the Mobitz type II variety. The block lasted only two hours. During her hospital stay the patient showed no further cardiac irregularity. On the third day postpartum, 24-hour Holter monitoring recorded normal sinus rhythm with a heart rate between 60 and 100.

At that time the patient was evaluated for connective tissue disease. A VDRL was reactive (it had been nonreactive two months before admission), microhemagglutination titer was nonreactive (ruling out syphilis), and ESR was 78. Antinuclear antibody was normal at 1:20; C3 (continued on page IODE)
Heart block (from page lock)

and C4 were normal. Rheumatoid factor was negative. A repeat ANA was less than 1: 20. and ESR was 12. SS-A antibody assay was positive: SS-B and antiribonucleoprotein was negative.

The Case in Context

Congenital complete heart block (CCHB) is a common enough condition to be encountered in even small obstetric services. Familial appearance of heart block, however, is distinctly rare. With the advent of routine fetal heart rate monitoring, more and more cases of congenital heart block are being diagnosed prenatally.

The incidence of congenital complete heart block is one per 20,000. About 40% to 50% of cases are secondary to a cardiac malformation and carry a very poor prognosis: the recurrence rate is 2.5% to 5%. In about 33% to 40% of cases, the condition is related to a maternal connective tissue disease. Most often systemic lupus erythematosus. A small but undefined number of cases are genetically transmitted. and for those patients the recurrence rate is up to 50%. Sporadic cases were thought to be associated with intrauterine myocarditis. An association between CCHB and the presence of antiribonucleoproteins (SS-A and SS-B antibodies) in the maternal and neonatal circulation was recently reported.

Occurrence of CCHB in siblings can be related to maternal connective tissue disease or can be hereditary. In most cases. the exact mechanism is not clear. since the maternal disease may not become overt until years after the birth of an affected child. In this case. it is impossible to determine the exact etiology. Conversely, the undiagnosed intermittent Mobitz type II cardiac block in the mother is related to the CCHB of her offspring and reflects an incomplete expression of the same gene. Mobitz type II cardiac block is an uncommon and serious arrhythmia. which may progress to complete heart block. In a large series (599 patients) of CCHB patients, there was one case of concomitant CCHB in the father and child. C. M. McCue reported one infant with CCHB whose mother was also affected.

The initial finding in CCHB is frequently a baseline fetal bradycardia. Baseline bradycardia during labor can reflect either fetal distress or cardiac dysrhythmia. Most cases that involve a heart rate of approximately 110 with an associated good variability and acceleration pattern represent uncomplicated bradycardia. which may be indicative of postmature infant. If the heart rate is below 100. one should consider the possibility of fetal dysrhythmia. With only intermittent conduction. External fetal heart rate monitoring can be misleading in those cases. since it may reflect only the atrial heart rate. which is usually within the normal range.

That happened in the initial stages of this case. It can be avoided by using direct fetal ECG or real-time ultrasonography, which allows visualization of atrial and ventricular rates separately. Real-time ultrasound (intrauterine echocardiography) can also help diagnose major congenital malformations of the heart, which represent the most common association with complete atrioventricular block. Fetal scalp sampling was performed to ensure that the fetal heart block had not led to acidosis. Therefore, spontaneous vaginal delivery was allowed to proceed.

Selected Reading

Shenker L: Fetal cardiac arrhythmias. Obstet Gynecol Surv 34:561. 1979
McCue CM et al: Congenital complete heart block In newborns of mothers with connective tissue disease. Circulation 56:82. 1977
Gleicher N. Elkayam U: Intrauterine dysrhythmias. Ibid. pp 535-564