Intrauterine and Postnatal Atrial Fibrillation in the Wolff-Parkinson-White Syndrome

BERNARD BELHASSEN, M.D., DAVID PAUZNER, M.D., LEONARD BLEDEN, M.D., JACK SHEREZ, ALEXANDRA ZINGER, M.D., MENAHEM DAVID, M.D., BOAS MUHLBAUER, M.D., AND SHLOMO LANIAKO, M.D.

SUMMARY A fetal tachyarrhythmia was discovered at the thirty-second week of gestation of a 22-year-old woman. Fetal echocardiography revealed atrial fibrillation with rapid ventricular rate, without any other demonstrable cardiac abnormality. In spite of therapeutic maternal blood levels of digoxin, the fetal ventricular rate and cardiac size increased, which prompted us to perform cesarean section at the thirty-fourth week of gestation. A baby with a Wolff-Parkinson-White syndrome but no other cardiac anomaly was delivered. Recurrent episodes of nonsustained atrial fibrillation with conduction over the accessory pathway occurred in the first hours of life. The Wolff-Parkinson-White pattern was not present on subsequent ECG recordings. The use of echocardiography in the diagnosis and management of this rare fetal tachyarrhythmia is emphasized.

ATRIAL FIBRILLATION is a common complication of the Wolff-Parkinson-White syndrome (WPW) in adult patients.1-3 However, it has been rarely noted in children.2 4 5 We report a case of a newborn infant with WPW in whom atrial fibrillation was documented at birth and during intrauterine life.

Case Report

A 22-year-old was in her second pregnancy. The first pregnancy was voluntarily interrupted and was complicated by uterine perforation that required surgical repair. During routine examination in the thirty-second week of gestation, fetal cardiac tachyarrhythmia was noted by the family physician. The patient was referred to our Department of Obstetrics on December 6, 1980. On admission, the maternal examination was unremarkable. The blood pressure was 110/80 mm Hg and the pulse rate 90 beats/min. The maternal ECG and echocardiogram were normal. Routine blood and urine examinations, glucose tolerance test and thyroid function tests were all normal. Evaluation of the fetal status using a standard B-scan examination did not reveal any abnormality. The age of the fetus was assessed to be 32 weeks of gestation. However, the fetal pulse was rapid (more than 200 beats/min) and irregular. Several attempts to record a fetal ECG failed.

Fetal M-mode and two-dimensional echocardiographic examinations were performed. One-dimensional echocardiograms were performed using an Electronics for Medicine VR12 ultrasonoscope and a 2.25-MHz transducer, collimated to 7.5 cm and pulsed with repetition rate of 1000 pulses/sec. Two-dimensional studies were obtained using an Aloka Echocardiography SSD 1105 mechanical sector scanner. This imaging system uses a hand-held, single-element, 3.5-

MHz transducer with a variable scanning angle of 30–90° and variable scanning speed of 5–30 frames/sec. During the examination, the mother lay in a supine position. The fetal lie was determined using images of the fetal head, spine and liver. The total cardiac size was measured on the two-dimensional echocardiogram, as the distance from the right ventricular epicardial surface to the epicardial surface of the left ventricular posterior wall.6 Measurements of the cardiac size were made on each examination when the atrioventricular valves had just closed. The right and left ventricular inner diameters were measured from the M-mode recording.7

The first fetal M-mode echocardiogram (December 8) demonstrated an irregular ventricular tachyarrhythmia (cycle length 240–420 msec) as visualized on the tricuspid valve movements (fig. 1A). Irregular atrial tachyarrhythmia was also visualized on atrial wall movements. Whether these wall movements were those of the right or left atrium was undetermined. Later, the ventricular rhythm was regular (cycle length 240 msec) and the atrial rhythm was also regular at a rate twice the ventricular rate (fig. 1B). The right and left ventricular inner diameters were normal (table 1).7 Cross-sectional echocardiographic examination allowed analysis of the four chambers, and all had normal dimensions (table 1).6 No cardiac abnormality could be demonstrated (fig. 2).

In view of the findings of rapid atrial fibrillation without evident cardiac abnormality, we treated the fetus by administering digitalis to the mother. The initial dose was 0.25 mg twice daily. Fetal echocardiographic studies were performed on December 14, when the maternal digoxin blood level was 0.9 ng/ml. The echocardiogram revealed persistent atrial fibrillation with irregular ventricular rhythm (cycle length 240–420 msec), marked biventricular enlargement, and increase of the total cardiac size (fig. 3, table 1). The dose of digoxin was increased to 0.25 mg three times daily. Fetal echocardiographic examination was again performed on December 18, when the maternal digoxin level was 1.9 ng/ml. Atrial fibrillation was still present but the ventricular response was faster than

From the Departments of Cardiology, Gynecology and Obstetrics, and Neonatology, Tel-Aviv Medical Center, Tel-Aviv, and the Unit of Pediatric Cardiology, Beilinson Hospital, Petach Tikva, Israel.

Address for correspondence: Bernard Belhassen, M.D., Department of Cardiology, Ichilov Hospital, 64239 Tel-Aviv, Israel.

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during the previous studies (ventricular cycle length 220–340 msec) (fig. 4). A further slight increase in the cardiac size was suspected (table 1).

The acceleration of the fetal ventricular rate while on therapeutic maternal blood levels of digoxin prompted us to perform cesarean section on the same day, in the thirty-fourth week of gestation.

A male baby that weighed 2500 g was delivered with no apparent complication. The Apgar score was 7 at 1 minute (mild cyanosis) and 9 at 5 minutes. The heart rate at delivery was irregular and rapid (about 200 beats/min). The placenta was unremarkable on macroscopic examination. The following digoxin levels were measured at the time of delivery: maternal blood 1.6 ng/ml, umbilical cord 1.6 ng/ml, fetal blood 1.6 ng/ml and amniotic fluid 0.6 ng/ml.

In the neonatal intensive care unit, the baby’s general condition was satisfactory. At 10 minutes of age, slight peripheral cyanosis was present, the blood pressure was 40 mm Hg and the heart rate was regular at 170 beats/min. The liver was palpated 2 cm below the costal margin. A grade 2/6 systolic murmur was audible in the Erb area. The lungs were clear. The ECG revealed a normal sinus rhythm of 176 beats/min, with P waves of 3 mm amplitude, PR interval of 0.08 second, delta waves that were positive from V1 to V6 and diphasic in II, III, VF (fig. 5A). These findings were characteristic of a posterior-septal accessory pathway. Chest roentgenography showed mild-to-moderate cardiomegaly without signs of pulmonary congestion. The echocardiogram, which was performed 2 days later, was normal except for mild left ventricular enlargement.

During the first 36 hours of life, recurrent, nonsustained attacks of tachyarrhythmias were recorded. The ECG during the attacks revealed episodes of paroxysmal atrial fibrillation with an irregular ventricular response with antegrade conduction over the accessory pathway (rate 170–210 beats/min). The shortest RR interval during preexcited beats was 280 msec (fig. 6). Digoxin, 0.01 mg twice daily, was administered. No attacks of tachyarrhythmia occurred after the second day of life. On the thirtieth day, no signs of WPW were noted on the ECG (fig. 5B). Normal sinus rhythm was present at 167 beats/min, with normal P waves; the PR interval was of 0.10 second without a delta wave. Incomplete right bundle branch block was present. The echocardiogram was normal. At 9 months of follow-up, the infant was asymptomatic and in excellent general condition. The cardiac examination was normal. The ECG showed a normal sinus rhythm without evidence of preexcitation.

**Discussion**

We believe that this is the first report of an infant with WPW and atrial fibrillation demonstrated in utero and during the early hours after birth.

Although a fetal ECG was not obtained, we believe that the intratable atrial fibrillation occurred with conduction over the accessory pathway for three reasons: the high ventricular rate during the tachyarrhythmia; the increase of the ventricular rate after giving the mother digitalis; and the recording of paroxysmal atrial fibrillation with conduction over the accessory pathway at the birth. A high ventricular rate could have

### Table 1. Echocardiographic Measurements in the Fetus and the Newborn

<table>
<thead>
<tr>
<th>Date</th>
<th>Total Cardiac Size (mm)*</th>
<th>LV (mm)†</th>
<th>RV (mm)†</th>
</tr>
</thead>
<tbody>
<tr>
<td>December 8, 1980</td>
<td>39</td>
<td>12</td>
<td>12</td>
</tr>
<tr>
<td>December 14, 1980</td>
<td>47</td>
<td>17</td>
<td>17</td>
</tr>
<tr>
<td>December 18, 1980</td>
<td>50</td>
<td>18</td>
<td>18</td>
</tr>
<tr>
<td>January 1, 1981</td>
<td>50</td>
<td>18</td>
<td>10</td>
</tr>
</tbody>
</table>

*Measured by two-dimensional echocardiogram.
†Measured by M-mode echocardiogram.
Abbreviations: LV = left ventricle; RV = right ventricle.
resulted from enhanced atrioventricular nodal conduction due to enhanced sympathetic tone (secondary to congestive heart failure, for example) or to atrionodal bypass tracts. However, the fact that the tachyarrhythmia actually accelerated after maternal digoxin therapy makes this hypothesis unlikely.

Maternal digitalis administration is considered to be the therapy of choice in intrauterine tachyarrhythmias. Animal studies indicate that digoxin equilibrates across the placenta by passive diffusion. These studies have been confirmed in humans by the observation that fetal cord digoxin concentrations at term are similar to those in maternal venous blood. This was noted in our case. In addition, as previously reported, low digoxin levels were obtained in the mother during administration of digoxin at standard therapeutic dosages and high doses of digoxin were required to obtain therapeutic blood levels.

The use of digitalis has been shown to be dangerous during atrial fibrillation in patients with the WPW and
short antegrade refractory period of the accessory pathway.\textsuperscript{2, 14, 15} The latter is sometimes shortened by the administration of digitalis, resulting in the possibility of rapid ventricular response with the risk of ventricular fibrillation. We initially administered digoxin because we considered the probability of the association of atrial fibrillation and WPW very poor, since it had not been previously reported at this age. The use of digoxin at therapeutic maternal blood levels resulted in acceleration of the ventricular fetal rate and aggravated the fetal cardiac status, as shown by serial echocardiographic studies. This was probably because digoxin shortened the refractory period of the accessory pathway estimated by analysis of the shortest interval between the points of closure of the tricuspid valve during atrial fibrillation. Administration of digoxin at birth did not result in deleterious effects in our case. However, use of digitalis in this type of patient has not been shown to be safe and should be avoided. Differences between the values of the refractory periods of the accessory pathway during intra- and extrauterine life were found. These may be due to changes in environmental conditions, such as sympathetic tone, or to the natural lengthening of the refractory period of the accessory pathway.

Echocardiography has been introduced in the diagnosis of fetal cardiac arrhythmias.\textsuperscript{16} In our case, it allowed a precise characterization of the tachyarrhythmia as well as the evaluation of both medical therapy and tolerance of the arrhythmia. Although we cannot be certain that the repeated echocardiographic examinations were performed in exactly the same position, the fact that the same cardiac structures were visualized each time made it reasonable to compare the chamber sizes in the serial examinations. The dramatic increase in the fetal ventricular rate was the major factor in our decision to perform a cesarean section. The associated increase in cardiac size during serial echocardiographic examinations, although less precise, also played a part in this decision. The echocardiographic features in the normal fetus have been described.\textsuperscript{7, 17} The echocardiogram in our patient suggested to us that normal fetal cardiac anatomy was present. This was subsequently confirmed by postnatal examination.

Finally, the electrocardiographic evolution must be
pointed out. The WPW pattern was not present at the thirtieth day and was not observed again after a 9-month follow-up period. Such spontaneous disappearance of preexcitation has been noted in as many as 50% of the neonates with WPW in the first year of life.18-20

References

FIGURE 6. Electrocardiogram recorded a few hours after the birth. Shown are ECG leads I, III, V_L, and V_F. Repeated episodes of nonsustained atrial fibrillation with conduction over the accessory pathway are present.
Intrauterine and postnatal atrial fibrillation in the Wolff-Parkinson-White syndrome.
B Belhassen, D Pauzner, L Blieden, J Sherez, A Zinger, M David, B Muhlbauer and S Laniado

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