The Wolff-Parkinson-White Syndrome and Ventricular Septal Defect*
A Case Report of Successful Surgery in an 18-Year-Old Man

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INTRODUCTION

The occurrence of the Wolff-Parkinson-White (W-P-W) syndrome in patients who have ventricular septal defect has been stated to be common. A review of the largest surveys in the literature in which the W-P-W syndrome has been correlated with cases of congenital heart disease reveals, however, that this combination has been rare in cases documented at surgery or necropsy.

The patient described in this report had successful closure of his ventricular septal defect, though his electrocardiogram showed an intermittent W-P-W pattern.

CASE REPORT

This 18-year-old man entered the Diagnostic Clinic at the University of Florida Hospital in 1961 complaining of fatigue. Heart murmurs had been noted since infancy, and these were heard at subsequent physical examinations. He developed normally, participated in competitive sports without experiencing difficulty, but had noted more fatigue than usual following exercise during the year prior to admission. He denied, however, awareness of an irregular heart beat, and there were no documented occurrences of paroxysmal tachycardia.

A review of systems and past history yielded only that he had an asymptomatic congenital deformity of his cervical vertebrae, and probable congenital deafness of his left ear. There was no family history of congenital abnormalities, and no history of maternal rubella during pregnancy.

Physical Examination: He was a muscular young man whose salient physical findings were: pulse was regularly irregular with the usual rhythm being two beats followed by normal diastolic intervals and then three more rapid beats; peripheral arterial pulses were normal in intensity and contour; blood pressure was 120/80 mm.Hg. His hearing was normal in the right ear, but both air conduction and bone conduction were absent in the left ear. No abnormal venous or arterial pulsations were present in the neck. The chest moved symmetrically, and the lungs were clear to auscultation and percussion. The heart was enlarged with a forceful apical impulse at the sixth left intercostal space, 15 cm. to the left of the midsternal line. There was a mild left parasternal heave and a systolic thrill along the lower left sternal border. The heart sounds were of normal quality, but the components of the second sound were widely split and did not vary with respiration. A protodiastolic gallop was audible at the apex. Holo-systolic regurgitant murmur was present maximally in the fourth left intercostal space along the sternum and was widely transmitted over the entire chest. Abdominal findings were normal, except that the tip of the spleen was palpable. Leftward deviation of the cervical vertebrae at the level of C5 and C6 was noted on musculoskeletal examination.

Accessory Clinical Data: Routine blood and urine studies were normal. The electrocardiogram and vectorcardiogram revealed a basic pattern of mild biventricular hypertrophy with the intermittent occurrence of the W-P-W syndrome (Fig. 1). The phonocardiogram confirmed the auscultatory findings. Biventricular hypertrophy, slight enlargement of the main pulmonary artery and normal pulmonary vasculature were noted at cardiac fluoroscopy and on routine thoracic roentgenograms.

Cardiac Catheterization: A routine right heart catheterization revealed a significant left-to-right shunt at the ventricular level (Table 1).

Cardiac Surgery: A 1.5 cm. membranous septum type of septal defect, located under the
medial leaf of the tricuspid valve, was closed with interrupted and continuous sutures employing cardiopulmonary bypass and hypothermia. (Operation performed by Dr. Thomas D. Bartley, Assistant Professor of Surgery.) There was no postoperative complication.

Follow-up: This man, now two years after surgery, has been asymptomatic. The intermittent occurrence of the W-P-W syndrome persists on the electrocardiograms, but less frequently than prior to surgery.

**COMMENT**

Each lead of the electrocardiogram in this case showed the intermittent occurrence of the W-P-W syndrome. The electrocardiogram and vectorcardiogram (Fig. 1) show two basic patterns; normal sinus rhythm without W-P-W syndrome, and normal sinus rhythm with the W-P-W syndrome. The latter meets the criteria of W-P-W syndrome as put forth by Katz and Pick: a short PR interval occurring with normal sinus rhythm; an abnormal QRS complex revealing a “delta wave;” and an ST-T vector which is usually 180° from the initial QRS vector.

A review of the world’s literature correlating the W-P-W syndrome with congenital heart disease noted that only one case of ventricular septal defect documented at surgery or necropsy was associated with the W-P-W syndrome even though many clinical reports of this combination were said to exist. Though Tamm had stated that the W-P-W syndrome occurred

**FIGURE 1:** The electrocardiogram in an 18-year-old man with ventricular septal defect. Each lead shows two distinct patterns, a normal sinus rhythm without the Wolff-Parkinson-White syndrome, and a normal sinus rhythm with the Wolff-Parkinson-White syndrome. The rs'r' pattern in V1 when a normal sinus rhythm exists, suggests greater anterior QRS forces than normal and right ventricular hypertrophy. The tall and narrow contour of the T waves in the left precordial leads suggests diastolic (volume) overload of the left ventricle.
Table 1—Summary of a Conventional Right Heart Catheterization Carried Out by Inserting a No. 18 Thin Wall Arterial Needle in the Left Femoral Artery and Passing a No. 7 Catheter Through the Left Basilic Vein. This Catheterization Data Shows a Left-to-Right Shunt at the Ventricular Level

<table>
<thead>
<tr>
<th>Vol. Per Cent</th>
<th>Pressure</th>
<th>Heart Rate</th>
</tr>
</thead>
<tbody>
<tr>
<td>Femoral artery</td>
<td>16.6 (96%)</td>
<td>158/86</td>
</tr>
<tr>
<td>Superior vena cava</td>
<td>12.8</td>
<td></td>
</tr>
<tr>
<td>Superior vena cava</td>
<td>12.9</td>
<td></td>
</tr>
<tr>
<td>Hepatic vein</td>
<td>13.2</td>
<td></td>
</tr>
<tr>
<td>Right atrium (high)</td>
<td>12.7</td>
<td></td>
</tr>
<tr>
<td>Right atrium (mid)</td>
<td>12.8</td>
<td>0</td>
</tr>
<tr>
<td>Right atrium (low)</td>
<td>11.7</td>
<td></td>
</tr>
<tr>
<td>Right ventricle (in)</td>
<td>14.6</td>
<td></td>
</tr>
<tr>
<td>Right ventricle (mid)</td>
<td>14.9</td>
<td></td>
</tr>
<tr>
<td>Right ventricle (out)</td>
<td>14.7</td>
<td>26/0 (11)</td>
</tr>
<tr>
<td>Right pulmonary artery</td>
<td>14.1</td>
<td></td>
</tr>
<tr>
<td>Right pulmonary artery</td>
<td>14.1</td>
<td>21/7 (13)</td>
</tr>
<tr>
<td>Pulmonary wedge</td>
<td>ac 7</td>
<td>v 8</td>
</tr>
</tbody>
</table>

O₂ consumed=274 ml./min.=145 ml./min./M²; BMR=+22; Peripheral cardiac output=7.49 L./min.=3.96 L./min./M²; Pulmonary blood flow=10.75 L./min.=5.69 L./min./M²; Left to right shunt=3.3 L./min.=1.73 L./min./M²; Pulmonary arterial resistance=61 dynes sec. cm⁻²; Total pulmonary resistance 97 dynes sec. cm⁻²; Total peripheral resistance=1154 dynes sec. cm⁻².

frequently in patients having a ventricular septal defect, the University of Minnesota review noted that this combination had occurred only once in 1000 cases of ventricular septal defect.4

Later, Bernreiter5 described two cases of infants with the W-P-W syndrome and severe bouts of arrhythmias. These arrhythmias, plus the presence of large ventricular septal defects, led to congestive heart failure and early death. Necropsy revealed a large defect in the membranous portion of the ventricular septum in one case, and multiple defects in the muscular septum in the second case. Thus, this becomes only the fourth reported incidence of the W-P-W syndrome occurring in a documented case of isolated ventricular septal defect, although ventricular septal defects in association with other anomalies and the W-P-W syndrome have been noted.6

Approximately 50 per cent of patients who have W-P-W syndrome have bouts of paroxysmal atrial tachycardia, though the great majority of such attacks appear to occur under the age of 18 months.4 True ventricular tachycardia apparently do not occur in this syndrome.4 Trigger mechanisms such as fever, fatigue, and emotional distress should be determined when possible in cases who have persistent bouts of paroxysmal tachycardia. These episodes are usually treated with digitalis, quinidine or both. Although the increased incidence of arrhythmias in patients having the W-P-W syndrome may have frequently been a hindrance to the performance of surgical

Figure 1C: The initial portion of the QRS forces of the vectorcardiogram in the horizontal plane (H) is markedly slowed where the Wolff-Parkinson-White syndrome exists. This initial slowing corresponds to the "delta wave" on the electrocardiogram.
procedures, this patient underwent cardiac catheterization and corrective surgery without incident. The available evidence does not suggest that the presence of the W-P-W syndrome should be a deterrent to diagnostic or surgical procedures, particularly in an individual who has no history of tachycardia.

References
1 TAMM, R. H.: "Das Wolff-Parkinson-White Syndrom Eine Häufige Erkrankung im Kinde-
2 SCHIEBLER, G. L., ADAMS, P., JR. AND ANDER-
son, R. C.: "The Wolff-Parkinson-White Syn-
drome in Infants and Children. A Review and
a Report of 28 Cases," Pediatrics, 24:585,
1959.
3 SWIDERSKI, J., LEES, M. H. AND NADAS, A. S.: 
"The Wolff-Parkinson-White Syndrome in In-
fancy and Childhood," Brit. Heart J., 24:561,
1962.
4 KATZ, L. N. AND PICK, A.: Clinical Electro-
cardiography, Part I: The Arrhythmias, Lea
5 BERNREITER, M.: "The Association of W-P-W
Syndrome and Paroxysmal Tachycardia in Two
Cases of Ventricular Septal Defect," Missouri

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CHRONIC OBSTRUCTIONS PSEUDOEMPHYSEMA

An asymptomatic young man in whom routine
x-ray study showed a left hyperlucent lung, is de-
scribed. Air trapping and diminished perfusion were
demonstrated by laminography and angiography.
Bronchograms had the typical "pruned-tree" ap-
ppearance. Lung function studies showed a minor
obstructive impairment, whereas bronchospirometry
displayed no ventilation, marked trapping and little
blood flow on the affected side. The condition mimics
chronic obstructive emphysema, but lacks alveolar
disruption by definition associated with this disease.

Therefore, we have preferred to use the term pseu-
doemphysema. The increased tendency of diffuse expir-
atory airway collapse may well be the result of a
severe respiratory infection in childhood. Evidence
is presented to contradict the view that this entity
is due to primarily diminished perfusion. Surgical
excision is not indicated unless the disease is com-
plicated by respiratory infections.

PROWS, C. M., FUCHS, J. F., KAUFMAN, S. A. AND
GANNISLER, E. A.: "Chronic Obstructive Pseu-

DIAGNOSIS AND SURGICAL TREATMENT OF AORTIC COARCTATION
WITH PATENT DUCTUS

The article touches upon problems pertaining to
the diagnosis and surgical treatment of patients pre-
senting aortic coarctation associated with patent ductus
arteriosus. A study of 12 cases furnished the back-
ground for describing a characteristic clinical picture
and roentgenologic signs of this abnormality. The
x-ray investigation included fluoroscopy, roentgeno-
graphy in three projections, roentgenokymography
and, in a number of cases, catheterization of right
division of the heart and angiography. In the au-
thors' opinion, the main symptoms which may point
to the presence of patent ductus arteriosus in aortic
coarctation are systolic-diastolic murmur in the sec-
don intercostal space to the left of the sternum,
lowered diastolic pressure and an x-ray picture show-
ing congestion of the pulmonary circulation. The
authors outline the surgical technique in dealing
with aortic coarctation combined with patent ductus
arteriosus.

Berezov, Yu. E., POKOVSKY, A. V., GOLONKO, R. R.,
RUSHANOV, I. I., AND MELNIIK, I. Z.: "Diagnosis and Sur-
gical Treatment of Aortic Coarction in Conjuction with Patent