Wolff-Parkinson-White syndrome in infants and children

A long-term follow-up study

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Sixty-two infants and children with the Wolff-Parkinson-White syndrome (WPW) were under long-term follow-up at The New York Hospital over a 20-year period. Twenty had associated congenital heart disease. In 29 infants and 6 children in hospital for paroxysmal supraventricular tachycardia, digitalis relieved the episodes in all but one instance and prevented further recurrences in all but 4. One infant and 3 older children continued to experience brief episodes of paroxysmal tachycardia. Though three-quarters of the 42 babies were symptomatic, only 4 of the 46 followed as children and adolescents had tachyrhythmias. The prognosis into adult life for infants and children with WPW with or without episodes of tachycardia is good.

New developments in electrophysiology, anatomy, and cardiac therapy have brought new understanding and excitement to the fascinating syndrome that Wolff, Parkinson, and White first described in 1930 as an electrocardiographic combination of short PR interval and prolonged QRS duration in frequent association with attacks of paroxysmal tachycardia (Wolff, Parkinson, and White, 1930). Since that time, many case reports and several reviews of the syndrome occurring in infants and children have appeared (Engle, 1952; Schiebler, Adams, and Anderson, 1959; Swiderski, Lees, and Nadas, 1962), but there is little reported concerning long-term follow-up of these patients (Wolff and White, 1948; Malinow and Langendorf, 1950; Flensted-Jensen, 1969). It is the purpose of this paper to add to the knowledge of the natural history of the infant and child with Wolff-Parkinson-White syndrome based on our experience over the past 20 years.

Subjects and methods

This review includes 62 infants and children with WPW who were seen by the Pediatric Cardiology Division at The New York Hospital between 1950 and 1970. Each subject was 16 years of age or younger when an initial cardiogram showed the abnormality. A complete history, physical examination, and cardiac series of chest x-rays were obtained on each patient. Those with evidence of associated heart disease underwent additional cardiac diagnostic studies. Some continued under periodic cardiac supervision here. For the others, the subsequent course and present status were ascertained by recall for examination or by questionnaire filled out by the physician, the family, and/or the patient.

A diagnosis of this syndrome was made when a short PR interval together with a prolonged QRS complex and delta waves were present. The QRS measurements varied with age, being shorter in babies than in children, but in each instance the QRS duration was at least double the average for that age and twice that found in the same individual if periods of normal conduction were recorded. The tracings were classified into type A (false right bundle-branch block, Fig. 1) and type B (false left bundle-branch block, Fig. 2) conduction patterns (Rosenbaum et al., 1945). None with short PR and normal QRS as the only electrocardiographic pattern when not in tachycardia were included in this survey, but one child had these abnormalities transiently (Fig. 3).

Results

The manner of presentation to the hospital and the relation to associated congenital heart disease are shown in Table 1. Of the 62 cases, 65 per cent were male. None had a history of relatives with the syndrome. Table 2 illustrates the age at the time of the initial diagno-
TABLE I Relation of heart disease and tachycardia in Wolff-Parkinson-White syndrome

<table>
<thead>
<tr>
<th>No. of cases</th>
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<tbody>
<tr>
<td>With episodes of tachycardia</td>
</tr>
<tr>
<td>Without heart disease</td>
</tr>
<tr>
<td>With congenital heart disease</td>
</tr>
<tr>
<td>Without episodes of tachycardia</td>
</tr>
<tr>
<td>Without heart disease</td>
</tr>
<tr>
<td>With congenital heart disease</td>
</tr>
<tr>
<td>Total</td>
</tr>
</tbody>
</table>

sis and Table 3 at the time of the follow-up study. Two-thirds were younger than 2 years when first seen; almost half were older than 10 years when last seen, and 28 patients were adolescents. Table 4 shows the number of years from the first contact with the patient to the present follow-up study. Thirteen subjects were under observation for periods of 13 to 20 years.

Patients with paroxysmal tachycardia
Thirty-five individuals came because of paroxysmal tachycardia, documented in each by electrocardiography. Twenty-nine were infants aged 2 months or less when the first episode occurred, and two were less than 24 hours old. Fifteen had a history of one or multiple episodes of tachycardia, inadequately controlled, before referral. The others were seen in their first episode. In all but one, tachycardia responded to digitalization, and that one converted to normal rhythm with quinidine. In a few instances, brief repeated attacks occurred in the hospital while a suitable maintenance dose of digitalis was being determined.

Digitalis in the form of parenteral digoxin (12 patients) or digitoxin was administered over a 12- to 24-hour period as the initial therapy (Table 5). Of the patients who were treated with digitoxin, 22 per cent converted to normal sinus rhythm with less than 0.03 mg/kg (2 of these patients were under 1 year

TABLE 2 Age at initial examination

<table>
<thead>
<tr>
<th>Age (yr)</th>
<th>No. of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-2</td>
<td>42</td>
</tr>
<tr>
<td>3-6</td>
<td>7</td>
</tr>
<tr>
<td>7-10</td>
<td>7</td>
</tr>
<tr>
<td>11-16</td>
<td>6</td>
</tr>
<tr>
<td></td>
<td>62</td>
</tr>
</tbody>
</table>
of age and 3 were older than 2 years); 61 per cent needed 0.04 mg/kg (these patients were all under 1 year); and 11 per cent (2 patients who were aged 8 months and 2 months) required as much as 0.09 mg/kg in 24 hours for conversion.

Of the patients who were treated with digoxin, 36 per cent needed 0.025 mg/kg or less administered parenterally for conversion to normal sinus rhythm. These patients were all under 1 year of age. Another 36 per cent needed 0.06 mg/kg to 0.08 mg/kg intramuscularly for conversion (6 patients aged less than 1 year and 1 patient over 2 years). Thus these infants and children responded to digoxin or digoxin in the usual therapeutic range.

The average duration from hospital admission to conversion to normal sinus rhythm was approximately 10 hours with a range of 1 to 3 days. Eight infants who were admitted with tachyarrhythmia and congestive heart failure promptly converted with a digitalis preparation and were relieved of their cardiac decompensation. One infant presented in shock with rapid paroxysmal tachycardia but also did well with a digitalis preparation and maintenance therapy with digoxin. While most of the heart rates ranged from 220-260 a minute (Fig. 4), some were more rapid, and 5 had rates of 300 a minute or more (Fig. 5). Those infants with the fastest rates tended to be the sickest.

Among these 29 young infants was one who died with a recurrent episode of paroxysmal tachycardia. That event, early in this series, convinced us of the importance of maintenance digitalis in infants to prevent recurrences.

**TABLE 3** Age at follow-up study

<table>
<thead>
<tr>
<th>Age (yr)</th>
<th>No. of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-2</td>
<td>16</td>
</tr>
<tr>
<td>3-6</td>
<td>11</td>
</tr>
<tr>
<td>7-10</td>
<td>7</td>
</tr>
<tr>
<td>11-16</td>
<td>18</td>
</tr>
<tr>
<td>17-21</td>
<td>10</td>
</tr>
<tr>
<td></td>
<td>62</td>
</tr>
</tbody>
</table>

**Fig. 3** Spontaneous change from short PR and narrow QRS at a slower rate to short PR with wide QRS and delta waves in last 4 beats when rate accelerates. Baby boy, 2 months, with type B pre-excitation and paroxysmal tachycardia.

**Case report** The patient had complex cyanotic congenital heart disease with dextrocardia but functioned as a tetralogy of Fallot. His first episode of paroxysmal tachycardia occurred at 36 hours of age, before transfer on the fourth day of life. He was treated with digitoxin, 0.04 mg/kg, and normal sinus rhythm with type A WPW appeared. Maintenance digitoxin was given for the next 4 days and was discontinued. He was observed in the hospital for the next 2 weeks and was discharged on no medication. Two months later he was brought to the emergency room at 12:30 a.m. because of pallor and rapid respirations. The resident took an electrocardiogram which

**TABLE 4** Years of follow-up (interval from initial examination to follow-up study)

<table>
<thead>
<tr>
<th>Years</th>
<th>No. of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>1-4</td>
<td>28</td>
</tr>
<tr>
<td>5-8</td>
<td>10</td>
</tr>
<tr>
<td>9-12</td>
<td>11</td>
</tr>
<tr>
<td>13-20</td>
<td>13</td>
</tr>
<tr>
<td></td>
<td>62</td>
</tr>
</tbody>
</table>
showed paroxysmal tachycardia, but when the baby vomited and spontaneously reverted to sinus rhythm, he sent the infant home. The next recurrence was later that day, but the infant was moribund when brought back to the emergency room with a rate of 300 a minute. He died before any therapy could be given.

Since that time it has been our policy to maintain babies with WPW and paroxysmal tachycardia on digitalis in increasing doses commensurate with their weight gain, at least for the first 6 to 12 months, and to discontinue it, if they have remained free of recurrences, around the first birthday. After therapy was stopped, there was no recurrence during the period of follow-up in 85 per cent.

However, 4 subjects have continued to experience symptoms and electrocardiographic evidence of paroxysmal tachycardia, despite vigorous therapy with digitalis, quinidine and, in one case, propranolol (Fig. 1, 4, and 6) together with digitalis. One of these patients converted to normal sinus rhythm on one occasion immediately after intravenous injection of edrophonium. Three of the children continue on maintenance medication, and their attacks, though not completely eliminated, are less frequent, less prolonged, and better tolerated. The fourth, on no medication, has frequent momentary attacks that do not bother him in his normal activities. It is noteworthy that these three had their first episode of paroxysmal tachycardia after the age of 8 years and that one of them has Ebstein’s anomaly of the tricuspid valve, a condition in which episodes of tachycardia tend to occur even in the absence of the WPW syndrome. The fourth was treated successfully at 3 months for paroxysmal tachycardia at a rate of 280 a minute and was maintained on the drug, free of attacks, until the age of 2

### TABLE 5
**Digitalis therapy for conversion of paroxysmal tachycardia**

<table>
<thead>
<tr>
<th>Age of patient</th>
<th>Digitoxin</th>
<th>Digoxin</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No. of patients</td>
<td>Dose (mg/kg I.M.)</td>
</tr>
<tr>
<td>&lt; 1 year</td>
<td>2</td>
<td>0.03</td>
</tr>
<tr>
<td></td>
<td>11</td>
<td>0.04</td>
</tr>
<tr>
<td>&gt; 1 year</td>
<td>0</td>
<td>0.03</td>
</tr>
<tr>
<td></td>
<td>0</td>
<td>0.04</td>
</tr>
<tr>
<td>Total</td>
<td>18</td>
<td></td>
</tr>
</tbody>
</table>

### TABLE 6
**Associated congenital heart disease**

<table>
<thead>
<tr>
<th>Associated congenital heart disease</th>
<th>No. of cases</th>
<th>No. with paroxysmal tachycardia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ventricular septal defect, large*</td>
<td>4</td>
<td>1</td>
</tr>
<tr>
<td>Persistent ductus arteriosus</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>Ebstein’s anomaly</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Pulmonary stenosis</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Atrial septal defect, ventricular septal defect</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Cyanotic heart disease, complex</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Mitral atresia</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Ventricular septal defect, atrial stenosis, pulmonary stenosis, double outlet right ventricle</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Ventricular septal defect with pulmonary stenosis</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Transposition of great arteries</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>20</td>
<td>7</td>
</tr>
</tbody>
</table>

* With Down’s syndrome: 1.
determined, this situation seems to be an indication for DC conversion of the tachy-
rythmia rather than for digitalis therapy. Intravenous lignocaine, which is effective
against ventricular tachycardia, might also be
chosen to terminate such a tachycardia since
its use has been reported to be successful in
two adults with supraventricular tachycardia
and WPW (Dye, 1969).

The mechanism of tachycardia could not
be analysed in most, since the tracings were
often recorded during an attack or after sub-
sidence of the paroxysms but usually not dur-
ing onset or offset. In a few, however, one
could identify reciprocating tachycardia. Fig.
8 and 9 illustrated this in an infant with a his-
tory of repeated attacks of tachycardia. At a
normal heart rate, WPW (type B) was usually
present, but during paroxysms the QRS was
narrow (Fig. 8). When the PR interval was
prolonged, a secondary P wave, different in
form from the primary one, was clearly evi-
dent following the QRS and preceding the T
wave. When Wenckebach periods were pre-
sent, the secondary P was recorded after the
last and longest PR in the sequence. Some-
times the secondary P wave had a blocked
ventricular response; at other times it initiated
the onset of reciprocating tachycardia. These
variations were recorded in rapid succession
during the course of treatment with digitalis
to control the tachycardia that caused the ad-
mission to the hospital (Fig. 9).

This reciprocating rhythm represents circus
movement with antegrade conduction down
the normal pathway and reciprocal conduction
back up the accessory bundle (ventriculo-
atrial echo). The relation to delayed AV con-
duction is such that when the PR is long
enough, the atria are no longer refractory
when the retrograde impulse arrives and so
can respond. Reciprocal rhythm and reciproc-
ating tachycardia have recently been well

years. When he was about 10 years old,
attacks began again and occurred one to four
times monthly. His physician treated him by
inducing vomiting which terminated the
episode. He is the only patient in the series in
whom attacks recurred in late childhood after
several years of freedom from them.

A problem in differential diagnosis of the
dysrhythmia occurred when the electrocar-
diogram showed a wide QRS complex during an
episode of paroxysmal tachycardia. The
ventricular' tachycardia occurred in
4 subjects and was difficult to differentiate
from ventricular tachycardia (Fig. 5–7),
especially if no previous electrocardiogram
had been obtained or was available. Herr-
mann et al. (1957) suggested that the extreme-
ly high ventricular rate should suggest that
this dysrhythmia represents supraventricular
(ventricular) tachycardia in a patient
with accelerated AV conduction and false
bundle-branch block, rather than ventricular
tachycardia. If the proper diagnosis cannot be

FIG. 6 Response to propranolol of persisting
paroxysmal tachycardia that was unresponsive
to digitalis in an 8-year-old girl whose electro-
cardiograms are shown in Fig. 1 and 4. Note
the onsets and offsets of tachycardia and the
wide QRS form that suggests ventricular
tachycardia, especially at 42 seconds and 5
and 7 minutes after introduction of the drug.
As tachyrhythmia subsides, one sees fewer runs
of rapid rhythm and more periods of sinus
beats until, at 13 minutes, normal sinus
rhythm is restored.

FIG. 7 Pseudoventricular tachycardia with
offset and onset in an 8-month-old boy whose
sustained tachyrhythmia at 300/min is shown
in Fig. 5.

Our 62 cases were analysed in order to determine if there was an association between the conduction pattern of WPW and the incidence of dysrhythmias or ease of control. If the R wave was the sole or largest deflection in V1 and V2 (simulating complete RBBB), the electrocardiogram was classified as type A. If the sole or largest deflection in the right precordial leads was an S or QS (simulating complete LBBB), the electrocardiogram was classified as type B (Rosenbaum et al., 1945).

Thirty-two had type A; 29 had type B pattern; and one baby with paroxysmal tachycardia at 2 months later displayed both types, a rare occurrence (Matter and Hayes, 1964). Of the patients with type A, half have documented evidence of paroxysmal tachycardia, and of the patients with type B, 63 per cent had such evidence. Of the 4 patients in whom we have had difficulty in controlling recurrent bouts of tachycardia, 2 had type A pattern and 2 had type B.

Patients without tachycardia The Wolff-Parkinson-White electrocardiogram was detected in the other 27 patients during diagnostic workup for suspected heart disease in 22 patients and for other medical illnesses in 5 patients. Under observation, none has begun to experience episodic rapid heart action.

Associated congenital heart disease Table 6 gives the associated heart disease found in 20 children with Wolff-Parkinson-White syndrome. Of the patients with congenital heart disease, 7 (35%) had episodes of paroxysmal tachycardia. One baby with cyanotic heart disease died due to a recurrence of tachycardia (see above). Seven other deaths were related to the heart disease itself and not to the WPW syndrome.

Discussion

A paramount feature of the Wolff-Parkinson-White syndrome is an association with recurrent bouts of paroxysmal supraventricular tachyarrhythmia. It is well known that such an occurrence may have serious repercussions, since death may result from the altered haemodynamics of sustained tachyarrhythmias, especially in an individual with cyanotic heart disease. However, this series indicated that in the absence of primary cardiac disease, the
susceptibility to attacks of rapid heart action (early in infancy), and also the control of attacks with digitalis are concerned. Perhaps, as James (1970a) suggested, shaping and moulding postnatally of the AV node and His bundle are responsible for this vulnerable period in the first months of life. James (1970b) further proposed that the evolution into the final form of these structures might explain the disappearance after infancy of the electrocardiographic pattern of WPW, a change that occurred in 5 of the 29 infants in this series with documented tachycardia and WPW syndrome.

Recent developments in electrophysiology and in the functional and pathological anatomy of the cardiac conducting tissues have generated renewed interest in this unusual syndrome. Recording of His bundle electrocardiograms (Castellanos et al., 1970; Massumi, 1970) has confirmed the pathways of pre-excitation, as have the technique of epicardial excitation mapping combined with the introduction of intracardiac electrical stimulation (Durrer, Schuilenburg, and Wellens, 1970).

The historical background and evolving concepts of pathogenesis of the syndrome have been reviewed by James (1970b). He concluded that the syndrome could be explained by abnormalities in structure and function of the AV node, His bundle, and their environs in the septal AV junctional tissue. Ferrer (1970) has recently synthesized extensive electrocardiographic experience in a review of the condition and its management.

Our experience differs in one respect from her statement that during episodes of supraventricular tachycardia ‘the QRS is not abnormal because pre-excitation of the ventricles does not occur during tachycardias’. While generally this is so, it is not always the case, because 4 of our 35 subjects with paroxysmal supraventricular tachycardia had the abnormal and wide QRS during tachycardia just as during sinus rhythm (Fig. 6 and 7). One of these had both wide and narrow QRS on different occasions during repeated episodes of tachyarrhythmia (Fig. 4 and 6).

New pharmacological agents and the judicious use of DC conversion or cardiac pacing (Ryan et al., 1968) add to the physician’s armamentarium in treating the unusual patient who is resistant to more standard antiarrhythmia agents such as digitalis and quinidine.

The technique of surface-mapping of the heart has led to the successful surgical interruption of pathways of accelerated conduction in a patient with type B WPW who had been incapacitated because of repetitive and previously uncontrollable attacks of tachycardia (Cobb et al., 1968; Sealy et al., 1969). Surgery to modify the attacks was not considered to be indicated in the children in this report. None had disabling tachycardia. It is still too soon to evaluate the feasibility of surgery to interrupt accessory pathways of conduction. Though the first reported success in a type B Wolff-Parkinson-White was impressive, the operation requires the technique of surface mapping of conduction pathways, a detailed study not generally available. Even if that were carried out, surgical incision of the accessory tract might not be uniformly applicable or permanently successful (Burchell et al., 1967) because the accessory pathways might be inaccessible, multiple, or even located within the normal conduction system, or there might be a disparity between electrophysiological mapping and the accessory tract (Cole et al., 1970). More electrophysiological studies are needed, and more reports of long-term results of surgery are required before the role of this new form of treatment can be assessed.

Another surgical approach to the problem of recurrent, severe, medically uncontrollable tachycardia in adults with WPW was used by Dreifus et al. (1968) and Edmonds, Ellison, and Crews (1969). They proposed to abolish the attacks by interrupting the cirrus pathway on the antegrade limb of conduction. They induced atrioventricular block by surgically ligating the AV bundle (Dreifus et al., 1968) or by electrocoagulation (Edmonds et al., 1969) and then employed an electrical pacemaker.

It seems that these forms of surgery should be considered for the rare patient who is incapacitated by attacks that are unresponsive to medical management. In the paediatric years, however, our experience over a 20-year period is that about half had no episodes of tachycardia and that the others who did were usually infants who responded promptly to digitalis and remained free of tachyarrhythmia while maintained on this for the first year of life. The prognosis into adulthood is good.

References


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