Pseudo-Wolff-Parkinson-White Syndrome Observed in a Patient with Lutembacher’s Syndrome

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SUMMARY

Pre- and post-operative vectorcardiograms (Frank system) and angiocardiograms in a patient with Lutembacher’s syndrome, were correlated. Preoperative vectorcardiogram showed the initial conduction delay, complete RBBB, increased QRS voltage, and open QRS loop, while left ventriculography and left atrio-graphy demonstrated the markedly posteriorly dislocated left ventricular major axis surrounded by the huge right ventricle and right atrium. Two months after operation, when the left ventricular major axis shifted from abnormally posteriorly directed to left and inferiorly, with the decreased right-sided heart chambers, the initial conduction delay disappeared with decreased QRS voltage. As these vectorcardiographic and angiocardiographic changes at the postoperative stage were parallel, we concluded that the initial conduction delay was a representation of “the pseudo-W-P-W syndrome” and due to the abnormally posteriorly dislocated left ventricular apex and the delayed excitation of the right ventricular free wall caused by the huge dilatation.

Additional Indexing Words:

Initial conduction delay Angiocardiogram Huge dilatation of right-sided heart chambers Markedly posteriorly dislocated left ventricular apex

The combination of an atrial septal defect with mitral stenosis is known as Lutembacher’s syndrome.1) In a patient with this syndrome who is presented here, the heart was very huge. The enlargement particularly involved the right ventricle and right atrium. The left ventricular apex dislocated markedly posteriorly. The vectorcardiogram showed the initial conduction delay in addition to complete RBBB, increased QRS voltage and open QRS loop. The purposes of this paper are to demonstrate the pre- and post-operative vectorcardiograms and angiocardiograms, and to propose
the possible mechanisms regarding the postoperative disappearance of the initial conduction delay which we suspected to be the WPW syndrome complicated with Lutembacher's syndrome.

**Case Report**

A 37-year-old male was admitted in May, 1973, because of exertional dyspnea and palpitation. The severity of the heart failure was class III of New York Heart Association. First at age 12, he was told after auscultation of the heart that he had an abnormal heart murmur, and heart disease was suggested. Since about that time, there had been general malaise, exertional dyspnea, and palpitation, particularly increasing in severity in winter. In addition, since at age 31, he had taken the attack of “paroxysmal tachycardia” approximately a time per year after over work. Treatment with digitalis was started at the local hospital. But the patient's symptoms remained unchanged. On admission, the patient was a slender person. Weight was 49.6 Kg; height, 172.5 cm. Blood pressure was 92/60 in the upper extremities. The appearance was sick with the cyanosis. Auscultation revealed a harsh-blowing, medium-pitched, pan-systolic murmur best heard at apex (Grade 4/6), and a rumbling, low-pitched, mid-diastolic murmur (Grade 2/6) at apex. The splitting of the second heart sounds was wide and fixed in respiration. Although there was no liver enlargement, the prominences of jugular veins were noticed. The electrocardiogram showed notching or slurring in the first portion of the QRS complex, complete RBBB, atrial fibrillation and ventricular premature contractions. The vectorcardiogram showed the initial conduction delay in addition to complete RBBB, QRS high voltage and open QRS loop (Fig. 1). Echocardiogram revealed typical pattern of mitral stenosis, in addition to paradoxical septal movement and increased right ventricular dimension. At a right heart catheterization, pressures in right side slightly increased. The cardiac index was 4.65 L per minute per M² BS by Fick method. Left to right shunt was 18.28 L per minute (76.3% of pulmonary blood flow) and right to left shunt 1.44 L per minute (20.2%) at atrial level. Left ventriculography and left atriography showed the markedly, posteriorly dislocated left ventricle, the huge right ventricle and right atrium (Figs. 3a and 4). Based on the preoperative diagnoses (Ostium secundum atrial septal defect, mitral stenosis and tricuspid insufficiency), direct closure of intraatrial septal defect, mitral commissurotomy and annuloplasty of the tricuspid valve were performed successfully on September 10, 1973. All procedures were approached through a right atriotomy with the patient on cardiopulmonary bypass. The patient improved progressively. About 2 months after operation, right and left heart catheterizations were done. At a right heart catheterization, pressures in right side normalized. Left ventriculography demonstrated that the major axis of the left ventricle shifted from markedly posteriorly directed to left and inferiorly with markedly decreased right-sided heart chambers (Fig. 3b). The vectorcardiogram showed the disappearance of the initial conduction delay and decreased QRS voltage (Fig. 2). But the main feature, complete RBBB, remained unchanged. The QRS duration slightly delayed after operation (preoperative: 140 msec, postoperative: 150 msec). The patient was discharged on November 16, 1973.
Fig. 1. Preoperative vectorcardiogram (Frank system). The initial conduction delay, in addition to the complete RBBB, QRS high voltage and open QRS loop, were observed. The initial conduction delay consisted of 2 parts; the former directed left and anteriorly (septal Q vector) and the latter directed posteriorly, mainly depending on the electrical forces of the left ventricle (a dash time: 2.5 msec).
Fig. 2. Postoperative vectorcardiogram. Two months after operation, the initial conduction delay disappeared. But main feature, complete RBBB, remained unchanged. QRS voltage definitely decreased. QRS duration slightly delayed from preoperative 140 msec to postoperative 150 msec. The direction of septal Q vector shifted from left and anteriorly to straight anteriorly, after operation (a dash time: 2.5 msec).
Fig. 3a. Preoperative left ventriculogram. (A-P and lateral films at end diastole). Markedly, posteriorly dislocated left ventricular apex is demonstrated (preceding RR interval: 460 msec).

Fig. 3b. Postoperative left ventriculogram (A-P and lateral films at end-diastole). Left ventricular major axis shifted from markedly, posteriorly directed to left and inferiorly with decreased cardiac size (preceding RR interval: 740 msec).
Fig. 4. Preoperative left atriogram (A-P film). The left ventricle surrounded by the huge right ventricle and right atrium, is demonstrated (preceding RR interval: 1,400 msec). The left ventricular apex directed posteriorly, as shown in Fig. 3a.

DISCUSSION

The most prominent feature of the W-P-W syndrome in the vectorcardiogram is the initial conduction delay. In this study, this conduction delay (duration: 22 msec) disappeared after operation, accompanied by the decreased right-sided heart chambers and the anatomical positional change of the left ventricle. As these changes in the vectorcardiogram and the angiocardiogram were parallel, we considered the initial conduction delay to be due to "pseudo-W-P-W syndrome". This delay consisted of 2 parts in the horizontal plane: the former directed left and anteriorly (septal Q vector) and the latter directed posteriorly, mainly depending on the electrical forces of the left ventricular free wall. Usually, in the vectorcardiogram, most parts of instantaneous QRS vectors after initial Q vector are the summation vectors of the electrical forces of the left and right ventricular free walls as well as the septum. However, in this patient, there was the markedly posterior dislocation of the left ventricle caused by the huge dilatation of the right-sided heart chambers. Therefore, the second part of the conduction delay which directed posteriorly in the horizontal plane, was considered to be due to the abnormally posteriorly dislocated left ventricular apex and the
delayed excitation of the right ventricular free wall caused by the huge dilatation. In addition, it may play a role as another factor that the electrical forces of the left ventricular free wall are relatively much smaller than those of the right ventricular free wall under such circumstances in the marked right ventricular hypertrophy. In this meaning, the conduction delay in the vectorcardiogram does not indicate necessarily the slow conduction velocity of the excitation front in the ventricular depolarization. It also supports our hypothesis that the QRS duration did not decreased after operation, because the disappearance of the initial conduction delay in the true W-P-W syndrome was usually accompanied by the shortened QRS duration. The large spatial maximal leftward vector decreased from preoperative 3.58 mV to postoperative 2.88 mV. Here, the magnitude of this vector at preoperative stage was considered to be due to the electrical forces of the right ventricular free wall mainly, since the left ventricle was surrounded by the huge right ventricle and right atrium, and the left ventricular apex abnormally posteriorly dislocated. In the previous paper, the author concluded that the left ventricle should not be neglected when assessing vectorcardiograms. Also, in this patient with huge right ventricular dilatation and hypertrophy, we could not neglect the anatomical position of the left ventricle, especially to evaluate the initial conduction delay.

References