Response to Exercise in Congenital Complete Atrioventricular Block

By Danae Ikkos, M.D., and John S. Hanson, M.D.

The clinical and hemodynamic findings in patients with complete heart block have been reported by many authors. There is general agreement that irrespective of the etiology of the block, cardiac enlargement and increased stroke volume are the compensatory mechanisms for the slow rate. In a few studies ventricular rate and other hemodynamic changes have been followed during exercise. The prevailing opinion is that in patients with the congenital form of complete heart block the ventricular rate increases more readily than in patients with the acquired form of the arrhythmia. This opinion is based principally on observations in isolated cases. Therefore, the effect of exercise and exercise tolerance were studied in patients with the congenital form of complete atrioventricular dissociation.

Materials and Methods

The present studies were conducted in 11 such patients ranging from 7 to 23 years of age. Diagnosis of congenital A-V block had been made in infancy or early childhood in all patients. Exhaustive histories had failed to disclose any infective, toxic or other bases for the supposition that the blocks might be acquired rather than congenital. None of the patients had subjective cardiac symptoms, including Stokes-Adams attacks.

Exercise tolerance tests were performed on an electrically braked bicycle ergometer for 2 6-minute periods. The work load during the first period was approximately half the submaximal working capacity as predicted on the basis of the patient's physical characteristics (sex, age, and body weight). During the second period the full load was applied. In the usual calculation of the submaximal working capacity the load causing a pulse frequency of 170 per minute at a relatively steady state is taken as the desired value. In the present series such a reference value could not be obtained because of the patients' cardiac arrhythmia. Therefore, in duplicate determinations each subject was asked to perform exercise at increasing loads up to his limit of tolerance. The highest work load tolerated was arbitrarily taken as the submaximal working capacity. During exercise additional correlative signs proposed by others such as atrial heart rate and electrocardiographic changes were followed.

A standard 12-lead electrocardiogram was recorded at rest. During exercise the electrocardiogram was obtained by placing the indifferent electrode on the forehead. Heart volume was determined radiologically by means of biplane, right angle radiographs. Predicted normal values in standing position were calculated according to Maurea. Total amount of hemoglobin was determined by the alveolar carbon monoxide method with certain modifications. Total blood volume was calculated from total hemoglobin and hemoglobin concentration. Right heart catheterization was performed according to methods previously described from this laboratory. Oxygen content and capacity of blood samples were determined by Van Slyke manometric apparatus, while oxygen saturation of cardiac catheter samples was determined in a Kipp Haemoreflector according to Brinkman.

Results

The primary data concerning heart size, working capacity, electrocardiographic findings, and total amount of hemoglobin as well as atrial and ventricular rates before, during, and after exercise are presented in table 1. The heart size, relative to body size, is presented graphically in figure 1. Since the age and body size of the patients varied widely, the relative heart size is presented as the difference between the found heart volume (in standing position) and the one predicted on the basis of the data of Maurea, the difference
### Clinical and Hemodynamic Data

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<th>Weight (Kg)</th>
<th>Body surface area (M²)</th>
<th>Heart volume (ml)</th>
<th>Total amount (Hb g)</th>
<th>Working capacity (Kg/min)</th>
<th>Rest</th>
<th>Exercise 1st load</th>
<th>Exercise 2nd load</th>
<th>Exercise 1 min.</th>
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Fig. 1: Heart volume, ml. (in standing position) in relation to body size, expressed as the difference between observed and predicted values from Menege et al.

Fig. 2: Heart volume in cm³, difference between observed and predicted values.

Fig. 3: Difference between heart volume in cm³ (g = standard deviation)
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![Figure 2](image)

Heart volume, ml., (in prone position) in relation to total amount of hemoglobin. Normal values and their ± 2σ limits from the data of Holmgren et al.29

![Figure 3](image)

Working capacity in per cent of the normal value (normals from Bengtsson21) related to heart size expressed in standard deviations.

cise, disregarding periods with extrasystoles, varied between 68 and 130 per minute. No correlation between the ventricular rate at rest and that during exercise was established (fig. 5). Nor was there any correlation between heart size and ventricular rate (fig. 6). When the exercise was stopped, extrasystoles disappeared immediately in 4 patients and within 1 minute in another. In the sixth instance the bigeminy, which appeared during the test, persisted for 5 minutes after exercise. This particular patient had at the same time a sinoatrial block and demonstrated the largest degree of heart volume increase observed in the present group. Because of the sinoatrial block this patient has been excluded from figures 5, 6, 7, and 10.

The relative increase in the atrial and ventricular rates during exercise are shown in figure 7. The former increased almost stepwise at increasing work loads, but a stepwise increase in ventricular rate was found in only a few cases. In several instances the maximal ventricular increase occurred between rest and the first load, while in others it was observed during the second load.

Although it was previously assumed14 that in congenital heart block the relation between the atrial and ventricular rates remained unchanged by exercise, this was not, however, the case in our material. Changes in the ratio of atrial-ventricular rates were observed in every case, being pronounced in 3, both during (fig. 8) and after exercise (fig. 9).

The relation between the gain in the atrial and ventricular pulse rate during exercise (in per cent of the rate at rest) is presented in figure 10. The present findings demonstrate a clear positive correlation between these 2 variables, the gain in the ventricular rate in-
The relation between the ventricular rates at rest and exercise.

Relation between ventricular rate at rest and exercise and relative heart size (expressed in S.D. from normal).

creasing parallel with the gain in the atrial rate.

Upon cessation of exercise atrial and ventricular rates decreased and returned almost to previous values within 10 minutes (fig. 4). The rapidity with which this decrease occurred was, however, not always the same for the 2 rates. In most instances the ventricular rate decreased more rapidly than the atrial as is shown by the increase in the atrial-ventricular ratio 1 minute after exercise (fig. 9).

Cardiac catheterization was carried out in 4 of the 11 subjects, evidence of an atrial septal defect being found twice (table 2). All right atrial, right ventricular, and pulmonary artery pressures were normal in the 2 patients without atrial septal defect, and response to exercise during catheterization was determined in these instances (fig. 11). Presence of increased cardiac output, stroke volume, and a widened A-V oxygen difference was confirmed during exercise.

Discussion

The calculation of the working capacity of patients with atrioventricular block with the exercise tolerance test utilized presents some difficulties. In patients without arrhythmias the working capacity corresponds to the submaximal work load, i.e., the load which at a steady state causes a pulse frequency of 170 per minute.\textsuperscript{16, 17} It is evident that such rates
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Figure 8
Changes in the ratio of atrial:ventricular rates during exercise.

Figure 9
The change in ratio of atrial:ventricular rates during recovery following exercise.

could not be obtained in patients with congenital atrioventricular block. Therefore in the present study the work load that could be tolerated by the patients was taken as the submaximal work load and compared to the one predicted for each case. Holmgren et al. have suggested that the atrial rate instead of the ventricular rate should be used in cases with atrioventricular block. From the results in table 1 it is obvious that the submaximal working capacity as determined above meets the criteria of Holmgren et al. In all patients but 1 (case 2) the atrial rate during the second work load was around 170 per minute (range 160 to 185).

That patients with congenital atrioventricular block have a normal or nearly normal working capacity in a standardized exercise tolerance test demonstrates that this cardiac arrhythmia is not in itself an incapacitating disease, at least in the age range studied. Not only are such patients able to cope with the muscular activity of everyday life without symptoms, but they can also tolerate strenuous muscular exercise. For example, patients 3, 6, 7, and 9 were engaged in usual athletics and patient 4 was an ardent ice-hockey player.

Similar isolated examples have been previously reported in the literature.

The ventricular acceleration during exercise observed in these patients with atrioventricular block shows that this circulatory adjustment, which is the most important one during physical effort in normal people, is also of importance in such patients. The maximal ventricular rate obtained during exercise at submaximal working load showed large individual variations (table 1, fig. 4). Gilechrist studied the effect of a standard exercise tolerance test on the atrial and ventricular rates in patients with acquired complete heart block and found a negative correlation between the atrial and ventricular gains during exercise. He therefore suggested that the Bainbridge reflex was of importance in the large atrial gain with small ventricular acceleration during exercise. Our observations of a positive correlation (fig. 10) are, however, opposite to his. The much larger ventricular accelerations in our patients gave us observations in a different range from Gilchrist's that are not comparable. The difference in myocardial competence between his elderly patients and our young ones may well
account in large part for the amount of exercise tolerated, in ventricular acceleration, and in the ratios of atrial-ventricular gains.

The factor that determined the maximal ventricular rate was not the rate at rest nor the heart size, since no correlation could be demonstrated between these variables and the maximum ventricular rate (figs. 5 and 6). Furthermore, although the ventricular and atrial rates increased, by and large, in parallel (fig. 4), the agreement between these increases was not complete. The atrial-ventricular ratio showed changes both during transit from rest to exercise and from exercise to rest (fig. 9). These changes were very pronounced in a few cases (fig. 8). The reason for these changes in atrial-ventricular ratio is demonstrated in figure 7. The atrial rate increased in a normal manner, i.e., stepwise, while the increase in the ventricular rate was irregular in the sense that it occurred in many cases either between the rest and the first load or between the first and second load.

Although it might be postulated that the occurrence of ventricular extrasystoles, either isolated or in the form of bigeminy or trigeminy, could be a factor in increasing cardiac output, this does not seem to be the case. Holmgren\textsuperscript{14} has demonstrated decreases in mean arterial pressure during periods of bigeminal rhythm. As the increase in ventricular rate produced by exercise is limited in these cases, however, other hemodynamic changes must take place to effect maintenance of a normal exercise tolerance, despite the relative bradycardia. Measurement of the heart size in the present group confirmed previous findings of others that cardiac enlargement is present in atrioventricular block. This increase in heart size permits normal minute volumes by increasing stroke volume\textsuperscript{11, 12} and thus aids in supporting a normal or nearly normal working capacity (fig. 1). The importance of cardiac enlargement as a compensatory mechanism in atrioventricular block has even been experimentally demonstrated. Starzl et al.\textsuperscript{27, 28} found that dogs with experimentally produced atrioventricular blocks re-
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Findings at Right Heart Catheterization

<table>
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<th>Case no.</th>
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<th>O_{2} Difference</th>
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<th>Stroke Volume</th>
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<th>Ventricular Rate</th>
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<td>R 11.0</td>
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gained their preoperative working capacity a few months later when cardiac enlargement and increased stroke volume were obvious.

In 2 patients of our group the hemodynamic adjustments to exercise were observed during cardiac catheterization (fig. 11). The results clearly show the interplay between heart volume, ventricular rate, and stroke volume in such cases. The first case was a 12-year-old girl with normal heart size. The cardiac output during work increased from 4.8 to 8.8 liters per minute by virtue of an increase in ventricular rate from 53 to 110 per minute, the stroke volume remaining practically unchanged. Even the A-V oxygen difference was within the normal range for the changes in heart rate. An increase in cardiac output was also seen in the second patient, a girl of 15 years. But here the increase of minute volume from 4.8 to 10.5 liters per minute was achieved principally through a 50 per cent increase in stroke volume, the ventricular rate increasing only slightly from 48 to 68 per minute. In this case another hemodynamic adjustment was recognized, the A-V oxygen difference increasing above the normal limit to be expected from the normal rate.

Summary

The electrocardiographic and hemodynamic responses to graded exercise in 11 patients with congenital complete atrioventricular block are described. Ventricular rate increases during exercise were unrelated to the resting rate, atrial rate, or heart size. Changes in the ratio of atrial:ventricular rates were observed in all cases during physical work. Physical working capacity was normal or only slightly reduced in all instances. This is accomplished through maintenance of normal minute volumes, which in turn reflects an interplay of several factors: acceleration of ventricular rate, increased heart volume and stroke volume, and a normal or supernormal oxygen-transporting capacity of the blood as reflected in increased amount of total hemoglobin.

Summario in Interlingua

Es desribite le responsas electrocardiographic e hemodynamic constatate post graduate exercitio in 11 patientes con congenitate completo bloco atrioventricular. Le augmentos del proratas ventricular durante le exercitio revelava nulle correlasion con le proratas in reposo, le proratas atrial, o le dimensiones del corde. Alterationes in le proportion inter le proratas atrial e ventricular esseva observate in omne le casos durante le effortio physic. Le capacitate de effortio physic esseva normal o levemente reduce in omne le casos. Isto es le resultato del mantenentia de un normal volumine per minuta, un phenomeno que, de su parte, reflete un concerto de plure factores, i.e. le acceleration del prorata ventricular, le augmento del volumine cardiade e del volumine per pulso, e un capacitate normal o supernormal de transporto de oxigeno in le sanguine con le resultato de un augmento del quantitate total de hemoglobin.

References

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