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Pulmonary Atresia With Ventricular Septal Defect: Preoperative and Postoperative Responses to Exercise

GERALD BARBER, MD, GORDON K. DANIELSON, MD, FACC, FRANCISCO J. PUGA, MD, FACC, CHARLES T. HEISE, CRTT, DAVID J. DRISCOLL, MD, FACC

Rochester, Minnesota

Between April 1982 and June 1984, maximal exercise testing was performed 35 times in 34 consecutive patients with pulmonary atresia and ventricular septal defect (14 studies in patients without repair, 11 studies in patients with partial repair [insertion of a right ventricle to pulmonary artery conduit without ventricular septal defect closure] and 10 studies in patients with complete repair [insertion of a conduit with septal defect closure]). Total work performed, maximal power achieved, exercise time and maximal oxygen uptake were significantly greater in patients after partial or complete repair than in patients without repair. Systemic arterial blood oxygen saturations at rest and during exercise were directly related to the degree of repair. Although heart rate at rest in the three study groups was similar to that in a separate group of normal control subjects, patients in all three study groups had a blunted heart rate response to exercise. The ventilatory equivalent for oxygen was increased both at rest and during exercise for patients without conduit repair and those with a right ventricle to pulmonary artery conduit without ventricular septal defect closure but was similar to that of control subjects in the group with conduit insertion and septal defect closure.

This study indicates that patients with pulmonary atresia and ventricular septal defect have decreased exercise tolerance both before and after corrective surgery. Exercise tolerance improves significantly after placement of a conduit from the right ventricle to the pulmonary artery with or without ventricular septal defect closure. Although no further improvement in exercise tolerance occurs with closure of the septal defect, ventilatory function and systemic arterial blood oxygen saturation are improved.

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Subjective exercise intolerance in patients with unrepaired pulmonary atresia and ventricular septal defect (tetralogy of Fallot with pulmonary atresia) has been noted by several investigators (1–4). Surgical correction of this condition is now possible either primarily (5) or using a staged procedure (2,3,6). Primary repair consists of interposing a conduit between the right ventricular outflow tract and the pulmonary artery and closing the ventricular septal defect. It is used only if the size and distribution of the pulmonary artery are reasonably normal. If the pulmonary arteries are hypoplastic, a staged procedure can be used. In this procedure, right ventricle to pulmonary artery continuity is established but the ventricular septal defect is not closed. The defect can be closed subsequently if sufficient enlargement of the pulmonary artery occurs. Subjective improvement in exercise tolerance has been reported (5) after right ventricle to pulmonary artery conduit placement with or without closure of the ventricular septal defect. To our knowledge, the precise level of exercise tolerance before operation and the degree of improvement after operation have not been documented using formal exercise testing. Therefore, the purpose of this study was to measure the effect of operation on exercise tolerance and the cardiorespiratory responses to exercise in patients with pulmonary atresia and ventricular septal defect.

Methods

Study groups. Thirty-four consecutive patients with pulmonary atresia and ventricular septal defect who completed a maximal exercise test at the Mayo Clinic between April 1982 and June 1984 were included in the study. There were 16 male and 18 female patients (Table 1). Two additional patients were unable to complete a maximal exercise test because of arrhythmia.

The study patients were classified into three groups on the basis of prior operative procedure: group 1 (no repair) included patients without surgical correction or patients with a prior systemic to pulmonary artery anastomosis only; group

From the Mayo Clinic and Mayo Foundation, Rochester, Minnesota.

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Address for reprints. David J. Driscoll, MD, Mayo Clinic, 200 First Street SW, Rochester, Minnesota 55905.

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Table 1. Demographic Data on Patients* and Control Subjects

	Control Subjects (n = 22)	Group 1 (n = 14)	Group 2 (n = 11)	Group 3 (n = 10)
Sex (M/F)	16/6	7/7	5/6	5/5
Age (yr)†	12 ± 4	12 ± 6	12 ± 4	16 ± 4
Range	(6 to 18)	(4 to 20)	(5 to 21)	(13 to 27)
Height (cm)†	152 ± 62	135 ± 24	141 ± 17	162 ± 7
Range	(108 to 183)	(101 to 172)	(115 to 173)	(150 to 176)
Body surface area (m^2)†	1.4 ± 0.4	1.1 ± 0 4	1 1 ± 0.3	1 6 ± 0.2
Range	(0.75 to 2.04)	(0.7 to 1.6)	(0.6 to 1.6)	(1 3 to 1 9)

*See text and figures for definition of groups. †Mean ± SD F = female; M = male.

2 (partial repair) included patients with a right ventricle to pulmonary conduit who did not have the ventricular septal defect closed because of abnormal distribution or diminutive size of the pulmonary artery; group 3 (complete repair) included patients with a right ventricle to pulmonary conduit and ventricular septal defect closure. An additional group of 22 normal subjects from our laboratory served as a control group (see *Data analysis*).

Of the 14 patients in group 1, 8 had no prior cardiac operative procedures and 6 had a systemic to pulmonary artery anastomosis (Blalock-Taussig anastomosis in 4 and Waterston anastomosis in 2) at least 1 year before the exercise test.

The 11 patients in group 2 underwent conduit insertion 4.2 ± 1.8 years before the exercise test; 1 patient in this group had an exercise test both before and after operation.

The 10 patients in group 3 had exercise testing 6.1 ± 3.2 years after conduit insertion and septal defect repair. Intraoperative right and left ventricular pressures were measured for 9 of the 10 patients in group 3. The ratio of right to left ventricular pressure ranged from 0.5 to 0.9 (mean 0.69), and the right ventricle to pulmonary artery pressure gradient (six patients) ranged from 1 to 40 mm Hg (mean 24). Systolic pulmonary artery pressure ranged from 35 to 64 mm Hg (mean 49).

Study protocol. Spirometry was performed before 32 of the 35 exercise tests. The forced vital capacity, forced expiratory volume in 1 second and maximal voluntary ventilation were obtained.

The patients' heart rate, respiratory rate, blood pressure, electrocardiogram, oxygen consumption, carbon dioxide production and minute ventilation were measured at rest and at each work load during the exercise test. Three electrocardiographic leads were monitored throughout the test; a 12 lead electrocardiogram was obtained before the exercise test and at each work load. Heart rate was computed as the average of five consecutive RR cycles on the electrocardiogram recorded at a paper speed of 50 mm/s. Blood pressure was measured with an appropriately sized cuff (7) connected to a programmable air compression cuff system (Narco Systems, PE-300). Carbon dioxide and mixed expiratory oxygen were measured by sampling from a port at the distal end of a 6 liter mixing box for patients studied before February 15, 1983. After that date, computer-assisted breath by breath measurements were made from a sampling port placed at the mouthpiece. The gases were analyzed utilizing a mass spectrometer (Centronic 200 MGA). The mass spectrometer was calibrated before each exercise test using gases analyzed by the Haldane techniques. Flow was measured as expired gas passed through a pneumotachygraph (Fleisch no. 3) connected to a pressure transducer (Validyne DP 45). The flow signal was integrated electronically to obtain volume. An ear oximeter (Hewlett-Packard no. 47201 A) measured systemic arterial blood oxygen saturation at rest and throughout the exercise test. An acetylene-helium rebreathing technique was utilized to measure pulmonary blood flow (cardiac output) (8) at rest while the patient was seated on the cycle ergometer and at each work load for 6 of the 10 patients in group 3.

Exercise was performed on a previously calibrated cycle ergometer (Siemens-Elema 300 B) using the protocol described by James et al. (9). This is a 3 minute incremental cycle exercise protocol. The work loads are based on the patient's sex, height and body surface area. The patients were encouraged to exercise to the point of exhaustion, and only the results that the examiner considered to represent maximal cardiorespiratory effort were included in the study.

Data analysis. The results of the exercise tests were analyzed by comparing them with previously published normal values (9,10) and the values obtained in 22 normal control subjects in our laboratory. The demographic data for the control subjects and study patients are summarized in Table 1.

Statistical analysis. Two-tailed unpaired Student's *t* test and linear regression were used in the statistical analysis with $p < 0.05$ considered significant. All data were expressed as mean ± SD.

Results

Spirometry. In group 1 (no repair), there was no significant difference in forced vital capacity (percent pre-

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Table 2. Exercise Test Results in Control Subjects and Patients With Pulmonary Atresia and Ventricular Septal Defect*

Result	Control Subjects (n = 22)		Group 1 (n = 14)		Group 2 (n = 11)		Group 3 (n = 10)	
	Range	Mean	Range	Mean	Range	Mean	Range	Mean
Total work performed (%)	63 to 199	102	6 to 45	21	25 to 69	45	4 to 73	38
Maximal power achieved (%)	68 to 130	108	17 to 69	45	29 to 82	61	13 to 73	58
Total exercise time (%)	83 to 149	103	15 to 60	37	48 to 83	62	19 to 86	58
Maximal oxygen uptake (%)	70 to 245	98	21 to 70	39	31 to 71	53	24 to 75	51

*See text and figures for definition of groups. All values are expressed as percent of predicted normal.

dicted) between patients with a prior thoracotomy (72.7 ± 13.2%) and those without one (77.3 ± 22.4%). The ratio of forced expiratory volume in 1 second to forced vital capacity was less than 80% in two patients from group 1, one from group 2 (partial repair) and one from group 3 (complete repair).

Exercise capacity. All but two patients exercised to exhaustion, as indicated by audible ventilation and the inability to maintain a pedaling frequency of 60 rpm. No patient had syncope, chest pain or other adverse effect from exercise testing. Two patients were unable to complete a maximal exercise test because of arrhythmia. Because their tests were submaximal, their data are not included with the data from the other 34 patients in this study. The results of the exercise test for the three groups of patients as well as the 22 control subjects are summarized in Table 2.

All four indexes of exercise tolerance were significantly ($p < 0.001$) lower than values for normal control subjects in each of the three study groups (Fig. 1). The four indexes mean total work performed ($p < 0.001$), maximal power attained ($p < 0.05$), mean total exercise time ($p < 0.001$) and mean maximal oxygen uptake ($p < 0.05$), were significantly greater for patients with partial repair (group 2) than for patients without repair (group 1). There was no significant difference in any of the four indexes between patients with partial and complete repair (group 3), and there were no statistical differences in exercise performance between the six patients in group 1 who had had prior systemic to pulmonary anastomosis and the eight patients who had not.

In the 10 patients in group 3, there was no apparent relation between the ratio of right and left ventricular pressures, the right ventricular to pulmonary artery pressure gradient or pulmonary artery pressure at the end of complete repair and the work performed, exercise time or maximal oxygen uptake.

Effect of age on exercise tolerance. Only in patients in group 1 was there a significant negative correlation between age and exercise tolerance ($r = -0.53$, $p = 0.05$).

Degree of hypoxemia and exercise tolerance. Systemic arterial blood oxygen saturation values at rest and during exercise were not correlated with exercise tolerance

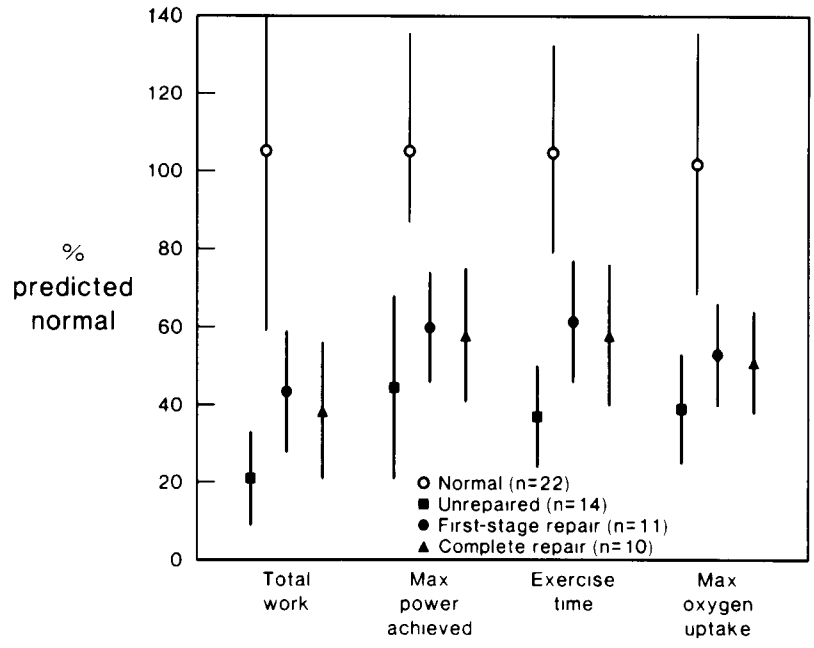

Figure 1. Comparison of total work performed, maximal (Max) power achieved, total exercise time and maximal oxygen uptake among the control group and patients with unrepaired pulmonary atresia and ventricular septal defect (group 1, unrepaired), patients after right ventricle to pulmonary artery conduit insertion without ventricular septal defect closure (group 2, first-stage repair) and patients after right ventricle to pulmonary artery conduit insertion and ventricular septal defect (group 3, complete repair). **Open circles** = control group; **closed squares** = group 1; **closed circles** = group 2; **closed triangles** = group 3. Each **bar** represents 1 SD above and below the mean.

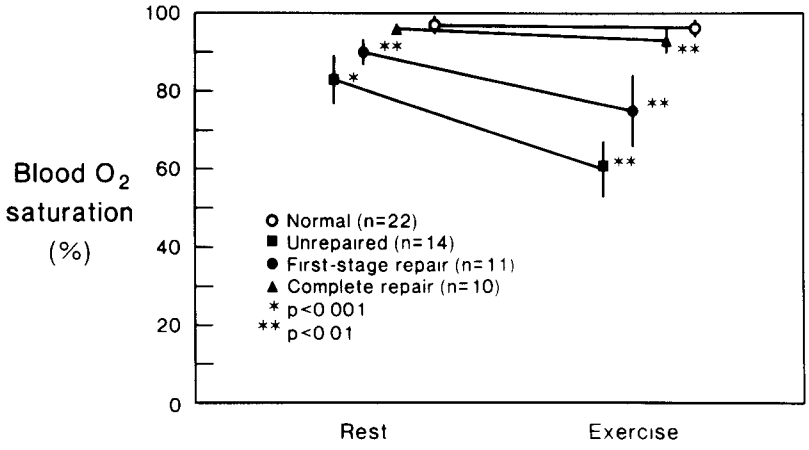

Figure 2. Comparison of exercise systemic arterial blood oxygen saturation at rest and during exercise among patients in the control group and groups 1, 2 and 3. **Symbols** as in Figure 1.

in any of the three groups (Fig. 2). Arterial blood oxygen saturations both at rest and during exercise increased with increasing degree of repair.

Pulmonary blood flow at rest and exercise tolerance. Pulmonary blood flow, measured at rest in 6 of the 10 patients in group 3, ranged from 2.4 to 3.5 liters/min per m^2 (mean 2.9). There was no correlation between exercise tolerance and pulmonary blood flow at rest.

Heart rate and blood pressure response to exercise. The heart rate and blood pressure responses to exercise of the three patient groups are summarized in Table 3. Heart rate at maximal exercise was abnormally decreased for all three groups.

Exercise ventilation. In groups 1 and 2, the ventilatory equivalent for oxygen ($\dot{V}_E/\dot{V}_{O_2}$) was significantly greater than values in normal control subjects both at rest and during exercise (Fig. 3). In group 1, however, the ventilatory equivalent for oxygen increased even further with exercise, but in group 2 it did not. The ventilatory equivalent for oxygen in group 3 was similar to the value in the control group both at rest and during exercise.

Minute ventilation, expressed as a percent of maximal voluntary ventilation ($\dot{V}_E$/MVV), was greater ($p < 0.01$) for groups 1 and 2 at rest than for the normal control group, but was similar to the control value during maximal exercise. In group 3, $\dot{V}_E$/MVV was similar to the control value at rest but lower ($p < 0.05$) than the control value during maximal exercise. For any given level of oxygen uptake, the ratio of minute ventilation to maximal voluntary ventilation was related to the degree of repair; that is, it was greater in group 1 than in group 2, greater in group 2 than in group 3 and greater in group 3 than in the control group (Fig. 4). Regardless of the degree of repair, patients would have had to exceed their maximal voluntary ventilation to achieve the same level of maximal oxygen uptake achieved by control subjects.

Rest and exercise electrocardiography. Thirteen patients in group 1, 10 in group 2 and 2 in group 3 had evidence

Table 3. Heart Rate and Blood Pressure Responses to Exercise in Control Subjects and in Patients With Pulmonary Atresia and Ventricular Septal Defect*

	Control Subjects (n = 22)		Group 1 (n = 14)		Group 2 (n = 11)		Group 3 (n = 10)	
Characteristics	Range	Mean	Range	Mean	Range	Mean	Range	Mean
Heart rate (%)†								
Rest	70 to 130	95	95 to 127	104	64 to 122	100	71 to 108	88
Exercise	80 to 105	95	55 to 88	73‡	68 to 81	74‡	39 to 94	78‡
Systolic blood pressure (%)†								
Rest	80 to 120	97	85 to 119	97	75 to 118	100	71 to 112	94
Exercise	75 to 105	93	76 to 105	90	78 to 109	91	65 to 103	76‡
Diastolic blood pressure (%)†								
Rest	80 to 120	95	64 to 108	90	75 to 161	98	72 to 103	86‡
Exercise	75 to 105	91	76 to 100	94	71 to 116	88	73 to 99	86

*See text and figures for definition of groups. †Percent of predicted normal. ‡Significantly lower than control value ($p < 0.05$).

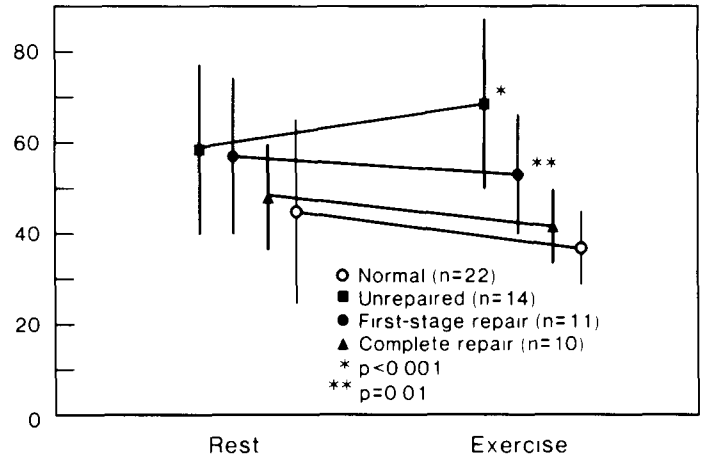

Figure 3. Comparison of the ventilatory equivalent for oxygen ($\dot{V}_E/\dot{V}_{O_2}$) at rest and during exercise among patients in the control group and groups 1, 2 and 3. **Symbols** as in Figure 1.

of right ventricular hypertrophy on the electrocardiogram at rest. The remaining patient in group 2 and seven of the remaining eight patients in group 3 had evidence of complete right bundle branch block. Only one patient from group 1 and one from group 3 had a normal electrocardiogram at rest.

In two patients the exercise tests were terminated because of ventricular arrhythmia. One of these patients had unrepaired congenital heart disease and the other had complete repair.

One patient in group 1 had ventricular bigeminy at rest. This disappeared during exercise and returned after exercise. There were no other arrhythmias in group 1, and no patient in this group had ST segment changes either at rest or during exercise.

One patient in group 2 had multiform premature ventricular complexes with variable coupling at rest. These decreased in frequency during exercise but returned to the preexercise frequency after the test. One patient had two premature ventricular complexes during exercise and two patients had ventricular bigeminy after exercise; in both, bigeminy resolved within 5 minutes. Of the nine patients without evidence of right bundle branch block whose ST segments could be evaluated, two had 1.5 mm ST depression at rest. During exercise one of the patients developed 2.5 mm depression and the other 3.5 mm depression. Three other patients developed ST segment depression during exercise (1.5, 3.0 and 3.5 mm). Only four patients in group 2 had no ST segment changes.

One patient in group 3 had multiple premature ventricular complexes during exercise, one had sinus arrest after exercise and one had three premature ventricular complexes after exercise. There were no other arrhythmias during exercise in group 3. Because 7 of the 10 patients had evidence of right bundle branch block, ST segment changes could be evaluated in only 3 patients. One of these had ST segment depression of 1.5 mm at rest and 3.0 mm during exercise. The other two patients had no ST depression at rest but had ST depression with exercise (1.0 and 2.0 mm).

Hemodynamic responses to exercise (group 3). Stroke volume, systemic vascular resistance and cardiac output at rest were similar to normal values for the six patients in

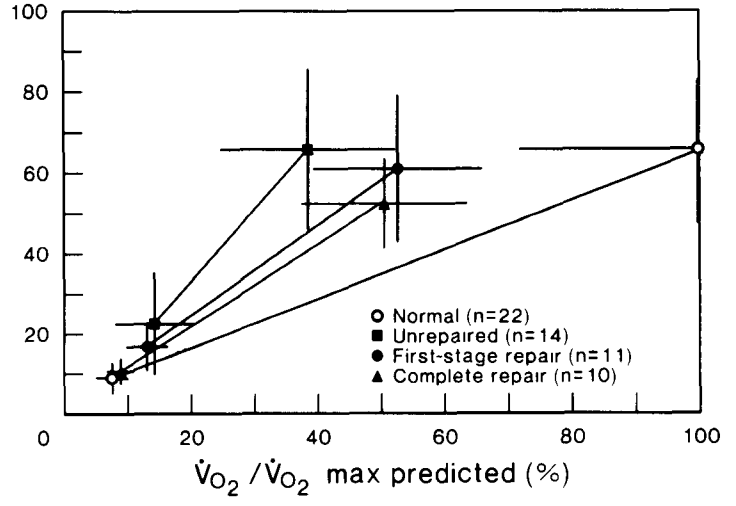

Figure 4. Relation between minute ventilation expressed as percent of maximal voluntary ventilation ($\dot{V}_E$/MVV) and oxygen uptake expressed as percent of maximal oxygen uptake ($\dot{V}_{O_2}/\dot{V}_{O_2}$ max) for patients in the control group and in groups 1, 2 and 3. **Symbols** as in Figure 1.

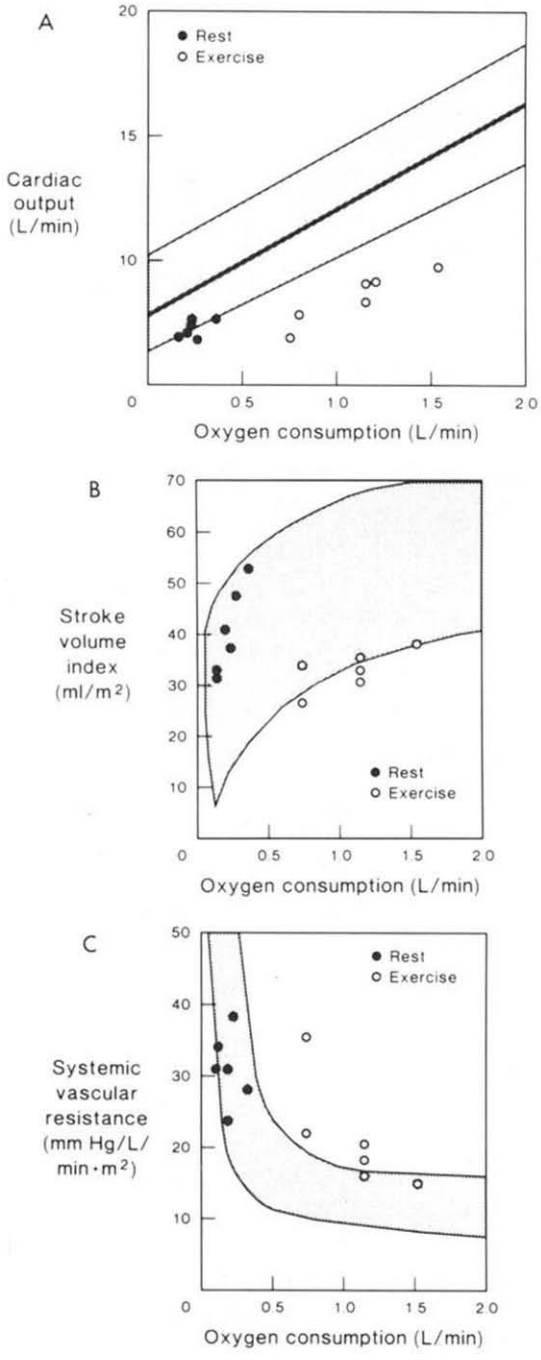

Figure 5. Cardiac output (**A**), stroke volume index (**B**) and total systemic vascular resistance (**C**) at rest and during exercise for six patients in group 3. **Hatched area** represents normal distributions for our laboratory.

group 3 in whom these measurements were made. With exercise, however, systemic vascular resistance was greater than normal and stroke volume and cardiac output were less than normal for our laboratory (Fig. 5).

Age at complete repair and exercise tolerance. There was no relation ($r = -0.33$, $p = 0.35$) between age at the time of complete repair and exercise tolerance for patients in group 3.

Discussion

Subjective assessments of exercise tolerance overestimate measured exercise capacity in children (11–13). Formal exercise testing is essential to document precisely the cardiorespiratory response to exercise of patients with complex cyanotic congenital heart disease and the effect of therapeutic interventions on patients with these lesions.

Although exercise tolerance and the presence of exercise-associated arrhythmia can be determined with a simpler exercise protocol than we utilized, a detailed exercise protocol in which multiple indexes of cardiac and respiratory functions are assessed is essential to define fully the cardiorespiratory responses to exercise.

Exercise tolerance. This study demonstrates that patients with pulmonary atresia and ventricular septal defect have a reduction in total work performed, power achieved, exercise time and maximal oxygen uptake both before and after the insertion of a right ventricle to pulmonary artery conduit.

Group 1: no operation versus systemic-pulmonary shunt. Although the total number of patients in our series is small, there was no difference in exercise tolerance, as measured by the four indexes outlined previously, between patients who had never been operated on and those who had either a Blalock-Taussig or a Waterston anastomosis. Because the shunt procedures were performed at an average age of 7.8 ± 3.8 years, this lack of difference should not be the result of a small shunt performed in a critically ill child during the neonatal period. Also, if this lack of difference is only the result of the small number of patients, one would expect that patients who had undergone a shunt procedure would have a small increase in the four indexes of exercise capacity, as compared with patients who did not undergo repair. The mean total work (as percent of predicted normal) was slightly higher in patients who were never operated on (21.4 ± 11.1%) than in those who had undergone a shunt procedure (21.3 ± 14.9%).

Right ventricle to pulmonary artery conduit without (group 2) and with (group 3) ventricular septal defect closure. Unlike patients with pulmonary atresia and ventricular septal defect who have undergone a Blalock-Taussig or Waterston anastomosis, patients with partial repair (insertion of a right ventricle to pulmonary artery conduit without closure of the septal defect) have significantly greater exercise tolerance than do patients without repair. Exercise tolerance was assessed before and after conduit insertion in only one patient in our study, and it increased even more strikingly for this patient than for the groups as a whole. This difference suggests that longitudinal measurements of exercise tolerance of individual patients may reveal even greater changes of exercise tolerance than are noted when groups are compared. In addition, there is no further increase in exercise tolerance after complete repair (right ventricle

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to pulmonary artery conduit insertion with closure of the ventricular septal defect). Although exercise intolerance persists after complete repair, it is improved sufficiently postoperatively to allow reasonably normal activity.

Age at time of complete repair. We found no relation between exercise tolerance and age at the time of complete repair. Similarly in a study of 24 patients with tetralogy of Fallot in our laboratory (unpublished data), we found no relation between age at time of complete repair and exercise tolerance. When Wessel et al. (14) eliminated the data of patients who had had intracardiac repair of tetralogy of Fallot during their late teens, there was no relation between exercise tolerance and age at intracardiac repair. To our knowledge, there are no data comparing postoperative exercise tolerance in patients undergoing repair very early (within 2 years) and in those undergoing repair in their preteen years.

Comparison with tetralogy of Fallot. Exercise tolerance is greater after repair of tetralogy of Fallot than after complete repair of pulmonary atresia with ventricular septal defect. In our laboratory, the mean maximal oxygen consumption (percent predicted) was 70% for 24 patients after repair of tetralogy of Fallot, which is similar to that of 135 patients (72%) reported by Wessel et al. (14). This compares with 51% in patients after complete repair of pulmonary atresia and ventricular septal defect.

Residual right ventricular hypertension. We could document no apparent relation between exercise tolerance and residual right ventricular hypertension after complete repair. However, the exercise studies were performed an average of 6.1 years after the pressure measurements were made, and the level of right ventricular hypertension may have changed during the interim. Also, the number of patients studied after complete repair (10, but only 9 with prior pressure measurements) may be too small to allow a meaningful statistical analysis. Indeed, Wessel et al. (14) found that patients with a right ventricular pressure of less than 50 mm Hg had significantly better exercise tolerance than did those with residual right ventricular hypertension (>50 mm Hg) after repair of tetralogy of Fallot. In our series, right ventricular pressure was greater than 50 mm Hg in eight of nine patients who were exercised after complete repair.

The decrease in exercise tolerance in our patients without repair is similar to that reported by Driscoll et al. (15) for "functional single ventricle" and by Barber et al. (16) for Ebstein's anomaly. The present study demonstrates that exercise tolerance correlates negatively with age in unrepaired pulmonary atresia and ventricular septal defect. It also was found to correlate negatively with age in "functional single ventricle" (15).

Systemic arterial blood oxygen saturation. A relation between exercise tolerance and systemic arterial blood oxygen saturation has been shown for tetralogy of Fallot (11), "functional single ventricle" (15) and Ebstein's anomaly (17). No such relation was apparent in our present study. This is probably the result of the narrow range of systemic arterial blood oxygen saturation at rest (9 of 14 patients in group 1 had a blood oxygen saturation at rest of 82 to 84%).

Heart rate and blood pressure responses during exercise. Similar to patients with "functional single ventricle" (15), but in contrast to those with tetralogy of Fallot (12) or Ebstein's anomaly (16), patients with pulmonary atresia and ventricular septal defect have a normal heart rate at rest. However, in our series the maximal heart rate during exercise was significantly lower than control values. A blunted heart rate response to exercise has been shown for patients with tetralogy of Fallot (11,13), "functional single ventricle" (15) and Ebstein's anomaly (16). Hypoxemia may decrease the heart rate response to exercise (18,19). This, however, does not explain the blunted heart rate response after complete repair. As noted with Ebstein's anomaly (16), there was no relation in our patients between heart rate and blood oxygen saturation during exercise. Patients with heart disease have abnormal sympathetic and parasympathetic control of heart rate (20,21) and this, along with possible sinus node dysfunction after surgery utilizing cardiopulmonary bypass, might explain the blunted heart rate response to exercise.

Blood pressure at rest and during exercise was normal for our patients without repair (group 1) or partial repair (group 2). During exercise, systolic blood pressure was decreased after complete repair (group 3). Patients in this group have slightly elevated systemic vascular resistance and have a decreased stroke volume and cardiac output response to exercise. Because patients after insertion of a right ventricle to pulmonary artery conduit without ventricular septal defect repair have blunted heart rate responses but normal blood pressure responses to exercise, their stroke volume responses are probably normal. Patients with unrepaired pulmonary atresia and ventricular septal defect and patients after insertion of a conduit without septal defect repair may maintain their stroke volume response to exercise by increasing the volume of right to left interventricular shunt, but when the septal defect is closed these patients no longer can maintain a normal stroke volume response to exercise. This may explain why, during exercise, normal systolic blood pressure was found in patients with a "functional single ventricle" (15), whereas a decreased systolic blood pressure was found in patients with Ebstein's anomaly (16). After repair of tetralogy of Fallot, patients have abnormal right ventricular function (22). Similarly, right ventricular dysfunction and obstruction to right ventricular outflow could explain the decreased stroke volume response to exercise seen in our patients.

Exercise ventilation. Similar to patients with tetralogy of Fallot (12,13), "functional single ventricle" (15) or Ebstein's anomaly (16), patients with unrepaired pulmonary atresia and ventricular septal defect ventilate excessively at

rest and during exercise. In contrast to normal subjects, the ventilatory equivalent for oxygen increases with exercise. After insertion of a right ventricle to pulmonary artery conduit without ventricular septal defect closure, the ventilatory equivalent for oxygen is still increased at rest. In this group, however, it decreases with exercise. After ventricular septal defect repair, the ventilatory equivalent for oxygen is normal at rest and during exercise. Hyperventilation associated with cyanotic congenital heart disease has been attributed to true or relative hypercapnia (23), hypoxemia (23) or increased dead space ventilation (24). The present study demonstrates that this hyperventilation can be reversed by corrective surgery.

All three groups of patients had decreased forced vital capacity. This has been attributed to small lungs from congenital underdevelopment secondary to hypoperfusion and low pulmonary artery pressure, lack of physical activity during periods of lung growth, prior thoracotomy, or pulmonary congestion (16,25). Whether exercise training either before or after operation could enhance lung growth and improve respiratory function in these patients is unclear.

Exercise electrocardiography. In two patients the exercise test was stopped because of arrhythmia. One patient had unrepaired pulmonary atresia and ventricular septal defect (group 1) and the other had complete repair (group 3). There was no significant difference in the incidence of exercise-induced arrhythmia among the three groups in this study.

Exercise-induced ventricular ectopic activity is associated with increased risk of sudden death in patients with repaired tetralogy of Fallot (17,26–28). The presence or absence of ventricular ectopic rhythm may be related to the degree of residual right ventricular outflow tract obstruction. It is reasonable to suspect that a similar relation exists among exercise-associated ventricular arrhythmia, sudden death and suboptimal hemodynamic surgical results for patients with repaired pulmonary atresia and ventricular septal defect.

There was a significant increase in the prevalence of ST segment depression at rest and during exercise, referable to the degree of repair in those patients in whom it could be evaluated. None of the 14 patients in group 1, 2 of 9 patients in group 2 and 1 of 3 patients in group 3 had ST segment depression at rest. During exercise there was still no ST depression in patients in group 1, whereas five of nine patients in group 2 and three of three patients in group 3 had ST depression.

Conclusions. This study demonstrates that patients with pulmonary atresia and ventricular septal defect have a significant decrease in exercise tolerance that is significantly alleviated by insertion of a right ventricle to pulmonary artery conduit with or without septal defect closure. Patients without repair have hyperventilation at rest and during exercise. This hyperventilation is partially decreased by insertion of a right ventricle to pulmonary artery conduit without ventricular septal defect closure and eliminated by insertion of a conduit with septal defect closure. Detailed exercise testing provides an objective measure of operative intervention on cardiorespiratory function.

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