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Lung Function in Congenital Heart Disease*

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INTRODUCTION

The importance of acquiring an understanding of the pulmonary circulation and lung function has become increasingly apparent in the diagnosis and treatment of children with congenital heart disease. Complicating heart failure and pneumonia are related to pulmonary blood flow in patients with left-to-right shunts. Outcome after surgical repair is dependent in many cases on the status of the pulmonary vascular bed.

This study was designed to determine whether alterations in ventilatory function reflect changes in the pulmonary vasculature and altered blood flow to the lungs.

Sixty-two children from six to 17 years of age were studied. All were admitted to the pediatric service at the UCLA Medical Center either for cardiac catheterization or surgical correction of congenital heart defects. None was in congestive failure at the time of study. There were 14 patients with ventricular septal defects (VSD), 16 with atrial septal defects (ASD), six with lesions causing right-to-left shunts (mainly tetralogy), 18 with various other congenital abnormalities (including patent ductus arteriosus, aortic stenosis, pulmonic stenosis, atrio-ventricular canal, and acyanotic tetralogy), and eight with innocent functional murmurs. Diagnoses were established at right heart catheterization and/or surgery.

METHODS

The various subdivisions of total lung volume were measured using a 9 liter Collins spirometer and the closed circuit helium dilution technique. Measurements included minute volume of respiration (MVR), oxygen consumption (VO₂), total and timed vital capacity or forced expired volume (FEV), 12 and 24 second maximal ventilatory volume (MVV), residual volume (RV), functional residual capacity (FRC) and total lung capacity (TLC). A series of "hospital normal" children consisting of those without evidence of cardiopulmonary disease or severe debilitating illness were initially studied and values for these children agreed well with the larger series of normals reported by Hellieson, et al.

MVV values were compared to figures obtained in normal children in our laboratory. In general, these control values averaged 15 per cent below levels found by Ferris. The values of Andrewes and Simmons were used for the ratios RV/TLC and FRC/TLC. Results were expressed as percentage of expected normal values so that children of varying heights could be compared on the same basis. A two-sided test with a significant level of 5 per cent was used to evaluate differences between the cardiac and control groups.

The data for patients with congenital heart disease were compared to expected normal values and to those for children with functional heart murmurs. Findings were compared for children with atrial and ventricular septal defects. Values for those with significant shunts (shunts greater than 50 per cent of total systemic flow) were compared to those with very small shunts (less than 50 per cent). Another comparison was made among children with systolic pulmonary artery pressures below 35 mm. Hg, between 35 and 50 mm. Hg and above 50 mm. Hg.

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RESULTS

In Table 1, data from children with functional murmurs are compared with data from patients with congenital heart disease (CHD). Ventilatory findings in patients with functional murmurs were not different from normal values in our laboratory. On the other hand, in the group of children with congenital heart disease, there were significant reductions in VC and MVV and significant increases in FRC and RV.

Although a trend was noted for greater reduction in VC, FRC, RV and TLC with increasing size of shunt, no statistical difference between the subdivisions was demonstrated.

In Table 2, findings in patients with atrial and ventricular septal defects are compared. Both groups show trends to decreased VC and increased FRC and RV. Only MVV is significantly lower than normal in both groups, and the FRC/TLC and RV/TLC ratios are elevated. No marked difference is found between the groups.

Table 3, which compares lung volumes on the basis of pulmonary artery pressure (PAP), shows the most striking findings. Patients with PAP over 50 mm. Hg showed reduction in all lung volumes with marked differences in FRC among all three groups. Progressive reduction in VC, FRC, RV and TLC is seen with increasing pulmonary hypertension.

Table 4—Summary of Significant Abnormalities in Ventilatory Studies

<table>
<thead>
<tr>
<th>Code: ↑=increase; ↓=decrease; — = no significant difference from normal (5 percent level).</th>
<th>PAP&lt;35 mm.Hg</th>
<th>PAP 35-50</th>
<th>PAP&gt;50</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. of Patients</td>
<td>28</td>
<td>13</td>
<td>8</td>
</tr>
<tr>
<td>VC</td>
<td>92± 3.5</td>
<td>86± 4.5</td>
<td>79± 3.6</td>
</tr>
<tr>
<td>MVV</td>
<td>91± 5.1</td>
<td>80± 5.8</td>
<td>96± 7.3</td>
</tr>
<tr>
<td>FRC</td>
<td>114± 6.5</td>
<td>106± 5.4</td>
<td>83± 8.8</td>
</tr>
<tr>
<td>RV</td>
<td>130±10.6</td>
<td>114±13.5</td>
<td>89±13.7</td>
</tr>
<tr>
<td>TLC</td>
<td>106± 4.9</td>
<td>95± 6.1</td>
<td>82± 5.0</td>
</tr>
<tr>
<td>FRC/TLC**</td>
<td>125± 4.2</td>
<td>121± 3.0</td>
<td>111±11.2</td>
</tr>
<tr>
<td>RV/TLC**</td>
<td>109±10.9</td>
<td>122± 8.7</td>
<td>117±14.8</td>
</tr>
</tbody>
</table>

All study figures are per cent of predicted normal.

---

**Table 1—Comparison of Ventilatory Studies in Children with Functional Murmurs and with Congenital Heart Disease (Means and Standard Errors)**

<table>
<thead>
<tr>
<th>Studies</th>
<th>Functional Murmur</th>
<th>All CHD</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. of Patients</td>
<td>8</td>
<td>54</td>
</tr>
<tr>
<td>VC</td>
<td>99± 5.0</td>
<td>89±2.4</td>
</tr>
<tr>
<td>MVV</td>
<td>100±11.2</td>
<td>89±2.9</td>
</tr>
<tr>
<td>FRC</td>
<td>101± 9.4</td>
<td>108±3.8</td>
</tr>
<tr>
<td>RV</td>
<td>117±14.4</td>
<td>125±6.2</td>
</tr>
<tr>
<td>TLC</td>
<td>103± 3.4</td>
<td>101±3.1</td>
</tr>
<tr>
<td>FRC/TLC**</td>
<td>112± 5.3</td>
<td>123±2.6</td>
</tr>
<tr>
<td>RV/TLC**</td>
<td>105± 7.0</td>
<td>126±4.8</td>
</tr>
</tbody>
</table>

*Expected=44
**Expected=24
Underlined figures indicate significant difference from normal (5% level).
All study figures are per cent of predicted normal.

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**Table 2—Comparison of Ventilatory Studies in Children with Interventricular and Interalial Septal Defects (Means and Standard Errors)**

<table>
<thead>
<tr>
<th>Studies</th>
<th>VSD</th>
<th>ASD</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. of Patients</td>
<td>14</td>
<td>16</td>
</tr>
<tr>
<td>VC</td>
<td>91± 5.0</td>
<td>90± 9.1</td>
</tr>
<tr>
<td>MVV</td>
<td>89± 4.6</td>
<td>90± 5.7</td>
</tr>
<tr>
<td>FRC</td>
<td>112± 6.9</td>
<td>103± 5.3</td>
</tr>
<tr>
<td>RV</td>
<td>120±14.2</td>
<td>122±12.5</td>
</tr>
<tr>
<td>TLC</td>
<td>105± 7.4</td>
<td>102± 5.1</td>
</tr>
<tr>
<td>FRC/TLC</td>
<td>121± 3.8</td>
<td>118± 3.7</td>
</tr>
<tr>
<td>RV/TLC</td>
<td>122± 5.6</td>
<td>122±10.4</td>
</tr>
</tbody>
</table>

All study figures are per cent of predicted normal. Underlined figures indicate significant difference from normal control (5 per cent level).
Table 4 summarizes the statistically significant findings in all children with congenital heart disease and those with functional murmurs, and further divides the groups according to pulmonary artery pressure.

Additional comparisons show reduction in VC and MVV in cardiac patients with left-to-right shunts, while these values for children with lesions without shunting were within the normal range.

None of the subjects in this study demonstrated evidence of obstructive ventilatory insufficiency, as gauged by reduction of one and three second forced expiration volumes or by air trapping during performance of the MVV.

**DISCUSSION**

In the last decade, there has been increased interest in ventilatory function and lung volumes in normal children and those with cardiopulmonary disease. The establishment of standards has greatly aided such study. Early work correlated a decrease in vital capacity with heart disease (mainly rheumatic) in children. Increased residual volumes were noted in other studies, particularly when patients suffered from cardiac decompensation. Gamalero and Segagni found that cyanotic patients with right-to-left shunts had the greatest reduction in VC and MVV with less marked reduction in those children with left-to-right shunts. They felt that impairment of ventilatory function in cyanotic patients was related to deficient tissue oxygenation with damage to the muscles of respiration while those with left-to-right shunts were thought to have decreased pulmonary elasticity. Larmi found decreased VC with normal RV and MVV in a study of 46 patients with cardiovascular disease. More recently, Cook and his group studied lung volumes and the mechanics of respiration in 100 patients from four to 42 years of age. They found that patients with left-to-right shunts had significant reduction in vital capacity and increase in residual volume and concluded that this was related to decreased effective respiratory muscle strength. Flow resistance in these patients was normal, ruling out air trapping and airway obstruction as a cause for the decreased VC and increased RV. They found 30 per cent of patients with left-to-right shunts had significant reductions of vital capacity compared with 29 per cent in our group. In our series, the incidence of significant reductions in VC was twice as high in the group with significant pulmonary hypertension as compared to those with normal pulmonary artery pressures.

It was hoped that our study might help define relationships between pulmonary air and blood volumes. Is increased pulmonary blood flow accompanied by increased thoracic blood volume with consequent displacement of gas volumes of the lung? Or does compensatory hyperinflation occur with increased pulmonary blood volume? Do changes in the viscoelastic properties of the lung also exert an effect? Is pulmonary function in children with heart disease affected by debilitation, associated disease or exercise limitations imposed by the physician or parent?

It has been demonstrated by Mills that when blood is accumulated in the legs by venous tourniquets, thoracic blood volume is decreased and vital capacity is increased. A change in body position from the erect to the supine results in an increased thoracic blood volume and causes a slight decrease in VC, RV and TLC. A trend toward progressive decrease in VC, FRC, RV and TLC with increasing left-to-right shunt suggests that these changes may be related to increased thoracic blood volume or cardiac volume.

Larmi showed that changes in the visco-elastic properties of the lung parenchyma altered lung volumes and ventilatory function. The "stiffened lung syndrome" with decreased TLC, VC and normal RV bears a resemblance to our findings in patients with high pulmonary artery pressure. Stiffening of lung parenchyma may result from vascular sclerosis.
which accompanies longstanding pulmonary hypertension.

Studies in our laboratory,8 and others,5,23,28 have shown that compliance is reduced in children with large left-to-right shunts and/or pulmonary hypertension. The correlation in our study between severity of pulmonary hypertension and reduction of vital capacity and total lung capacity suggests that progressive medial hypertrophy and intimal proliferation of pulmonary vessels might decrease lung distensibility and result in lower lung volume measurements. Increased pulmonary capillary blood volume and distention of large pulmonary blood vessels is probably not of sufficient magnitude per se to displace significantly lung gas volumes, even with large increases in pulmonary blood flow.4

In dogs with left-to-right shunts, Ellison and colleagues7 found that altered pulmonary function correlated with the degree of pulmonary hypertension. Those with medial hypertrophy and intimal thickening of the pulmonary vessels always had altered pulmonary function while those without pulmonary vascular pathology did not show a consistent change in lung function. Their study revealed an increasing venous admixture in systemic flow with increasing pulmonary hypertension, related to abnormal diffusion or pulmonary arteriovenous anastomoses. Previous data29 also demonstrated reduced arterial oxygen saturation in cases with increased cardiac output or pulmonary artery pressure. Bucci and Cook23 found increased diffusing capacity of the pulmonary membrane and over 50 per cent increase in pulmonary capillary blood volume in the preoperative patient with congenital heart disease and a large left-to-right shunt. These findings would fit with those of Ellison if certain areas of the lung were subject to increased flow and decreased ventilation.

Vital capacity decreases in disease and debilitation.30 Recurrent infection, poor general growth and development and enlarged hearts in small thoracic cages might reduce functional lung tissue in congenital heart disease. In addition, children with heart disease may have limited opportunity to develop ventilatory function through exercise. The lower TLC and VC in patients with the highest pulmonary artery pressures may be related to increased debilitation in these children.

Maximal ventilatory volume (MVV) was reduced in all heart patients, but not in those with functional murmurs. Since this latter group was subject to the same medical and psychologic management as the cardiacs, this suggests that this deficit was related to the cardiopulmonary defect rather than to decreased play activity, exercise and motivation. Normal values for all parameters in the children with functional murmurs suggest that our methods were reliable and that parental or iatrogenic restriction of physical activities in this group did not play a significant role in reduced pulmonary function.

SUMMARY

Pulmonary function and lung volume determinations in children with congenital heart disease demonstrated significant reduction in VC and MVV. FRC and RV were increased, this being reflected in increased RV/TLC and FRC/TLC ratios. Only the group with severe pulmonary hypertension had decreased FRC and RV, but even here the relatively greater reduction in TLC resulted in increased ratios. Changes appeared to correlate better with degree of pulmonary hypertension than with specific lesion or size of shunt.

ACKNOWLEDGMENT: The authors wish to express their appreciation for the technical assistance of Miss Martha Darling.

RESUMEN

Los estudios de la función pulmonar en los niños con enfermedades congénitas del corazón han demostrado una reducción significativa de CV y de MVV. CRF y VR están aumentados, lo que se refleja en aumento de las relaciones VR/CPT y de CFR/CPT. Sólo los grupos con grave hipertensión pulmonar tuvieron CRF y VR decrecidas pero aún aquí la relativamente mayor reducción de CPT dio por resultado un aumento de las relaciones. Los cambios parecen correlacionarse mejor con el grado de hipertensión pulmonar que con la lesión específica o con el grado de la intercomunicación.
ZUSAMMENFASSUNG


REFERENCES


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