SCIENTIFIC LETTER

Ventilatory response to exercise in patients after correction of cyanotic congenital heart disease: relation with clinical outcome after surgery

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PATIENTS AND METHODS

One hundred and six patients were used for this analysis. At a routine outpatient visit, the patients were clinically assessed, and also underwent echocardiographic, electrocardiographic, and radiological examinations. Three patient categories with previous surgical repair for cyanotic congenital heart disease were studied: total repair for tetralogy of Fallot (n = 34), atrial switch operation for transposition of the great arteries (TGA) (n = 23), and Fontan operation for functionally univentricular hearts (Fontan) (n = 13). These patient groups were compared to a group of age matched patients who had also undergone sternotomy for surgical closure of a ventricular septal defect (VSD).

RESULTS

A significantly elevated value for total \( V_E \) (\( p = 0.001 \)) (range 23–48%) was found at different exercise levels, when compared to normal controls. This resulted from a significantly higher value of both alveolar (\( p = 0.03 \)) and physiological dead space ventilation (\( p = 0.01 \)) compared to normal controls. This larger difference between total and alveolar ventilation varied from 12–48% at different levels of exercise. The largest differences (\( VD = 9 \text{ l/min} \)) were found in patients with TGA and in patients with Fontan circulation at 8% inclination of the treadmill. Abnormal values for the physiological dead space ventilation (> 95% confidence limit of the normal mean value for at least four exercise levels) were found in a considerable number of patients in all three groups (table 1). The inefficient ventilatory response to exercise in the patients was also characterised by significantly higher than normal values (\( p < 0.05 \)) for the ventilation versus carbon dioxide slope.

Furthermore, to study the possible effect of thoracic surgery on the efficiency of gas exchange during exercise, we compared the results obtained in a group of patients with surgical closure of VSD via sternotomy, to the other groups. In patients with surgically closed VSD the slope of \( V_E \) versus \( V\text{CO}_2 \) averaged 3.2 (6.4) and was not significantly different from the normal controls (3.2 (8.2)) (\( p = 0.25 \)). The incidence of abnormal values for physiological dead space ventilation (\( VD \)) was associated with a significantly higher number of residual haemodynamic lesions (33–62%) (\( p < 0.05 \)), compared to patients with normal values (12–28%) (table 1).

DISCUSSION

Surgical results of congenital heart disease are excellent nowadays, with the majority of the patients leading a normal life. This is confirmed by the exercise results in the present study: 33–50% of the patients have a perfectly normal ventilatory response during exercise. The purpose of the present study was to evaluate whether the inefficient ventilatory response to exercise, as observed in a considerable number of patients, was associated with haemodynamic abnormalities after surgery. In 32–62% of the patients with an inefficient ventilatory response to exercise, residual haemodynamic lesions after surgery were observed. Typical lesions were right ventricular dilatation and pulmonary incompetence or stenosis in TF patients, severe right ventricular dilatation and tricuspid incompetence in patients.

Abbreviations: TGA, transposition of the great arteries; \( V_A \), alveolar ventilation; \( VD \), dead space ventilation; \( V_E \), total ventilation; VSD, ventricular septal defect.
with Senning operation, and atrioventricular insufficiency in patients with Fontan circulation.

The elevated value for physiological dead space ventilation and the increased ratio of ventilation over $V_{CO_2}$ as observed in about half of the patients, suggest a ventilation/perfusion mismatch. This can be attributed to an inability to appropriately increase the effective pulmonary blood flow in response to increasing metabolic needs. Concern could be raised about the effect of thoracic surgery on the ventilatory efficiency during exercise. Previous studies have shown restrictive lung function patterns in patients who underwent palliation or definitive repair of tetralogy of Fallot. Therefore, we compared the ventilatory efficiency of patients with repair of cyanotic congenital heart disease, to patients who underwent thoracic surgery for non-cyanotic congenital heart disease such as surgical closure of a VSD. In the group of patients after VSD closure, no significant difference was found for ventilatory efficiency with normal controls. Therefore, the increased values for physiological dead space ventilation and slope of $V_E$ versus $V_{CO_2}$ after surgery for cyanotic congenital heart disease cannot be ascribed to the effect of previous thoracic surgery.

Assessment of the functional status after surgery by anamnesis and evaluation of the medical history is unreliable and lacks sensitivity in patients with congenital heart disease. We would therefore recommend that exercise testing with direct measurements of gas exchange should be performed in these patients as this contributes to global functional evaluation.

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Table 1: Incidence of abnormal values for physiological dead space ventilation $V_D$ and $V_E$ versus $V_{CO_2}$ slope. Associated haemodynamic lesions in patients with surgical repair of cyanotic congenital heart disease are also shown.

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Incidence</th>
<th>Associated lesions</th>
</tr>
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<tbody>
<tr>
<td>Normal VD</td>
<td>Normal slope $V_E \div V_{CO_2}$</td>
<td>Abnormal VD</td>
</tr>
<tr>
<td>TP (n=34)</td>
<td>18 (53%)</td>
<td>16 (47%)</td>
</tr>
<tr>
<td>TGA (n=23)</td>
<td>8 (35%)</td>
<td>15 (65%)</td>
</tr>
<tr>
<td>Fontan (n=13)</td>
<td>5 (38%)</td>
<td>8 (62%)</td>
</tr>
</tbody>
</table>

*p<0.05

Abnormal values are defined as values which exceeded the 95% confidence limits of the normal mean value. The incidence of subnormal values was significantly higher in all patient groups ($\chi^2$, p<0.0001) compared to normal controls.

AV valve insuff, atrioventricular insufficiency; PR, pulmonary regurgitation; PS, pulmonary stenosis; RV dil, right ventricular dilatation; slope $V_E \div V_{CO_2}$, slope of ventilation versus carbon dioxide output; TI, tricuspid incompetence; VD, physiological dead space ventilation.

REFERENCES