Oximetry in Congenital Heart Disease with Special Reference to the Effects of Voluntary Hyperventilation

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An oximeter has been used to measure the arterial oxygen saturation of patients with a variety of congenital heart anomalies. The effects of exercise and oxygen administration have been of value in ascertaining the presence of a venous-arterial shunt. The rise in arterial oxygen saturation during a vigorously performed voluntary hyperventilation appears to offer a means of gauging the extent to which the pulmonary circulation may be increased; a less than normal increase has only been found in the presence of pulmonic stenosis or in some instances of pulmonary hypertension and a relatively fixed pulmonary resistance. The results of hyperventilation are sufficiently distinctive to be of diagnostic value in cases of Fallot’s tetralogy, Eisenmenger’s complex, isolated pulmonic stenosis, pulmonic stenosis with intact ventricular septum and patent foramen ovale, and Ebstein’s anomaly of the tricuspid valve with patent foramen ovale.

The changes in arterial oxygen saturation during exercise and oxygen administration have recently become an integral part of the study of congenital heart disease. A low arterial saturation at rest or a fall during exercise has been associated with venous-arterial shunts. A failure to reach full arterial saturation and an unduly prolonged time to reach maximal saturation during oxygen administration have been accepted as evidence of large venous-arterial shunts. We wish to present the effects of voluntary hyperventilation on arterial oxygen saturation and to discuss the possibility of using this procedure as a diagnostic test.

Methods and Material

Arterial oxygen saturation was measured by an ear oximeter designed by Wood and Geraci (Waters-Conley Absolute Reading Oximeter). Readings were made after a period of rest and a sufficient time to heat the ear. These were repeated at intervals until consecutive readings were constant. Oxygen (100 per cent) was administered through a mask for at least five minutes and continued for a longer time in many of the cases with cyanosis, the patient being in a recumbent position. Voluntary hyperventilation was carried out for one minute in the sitting position. The patient was shown how to breathe as rapidly and as deeply as possible and during the test was exhorted to make a maximum and constant effort. The exercise consisted of stepping up and down a platform 9 inches high 30 times in one minute. The changes in arterial oxygen saturation were called out by one observer and noted by another according to time recorded by a stop-watch, and were followed after the completion of the test until the saturation had returned to its resting level. The oximeter was reset with each change of posture and during recovery from the test. A sufficient time was allowed between each procedure to enable the patient to recover. Movement of the oximeter ear-piece during the exercise test was prevented by fixing the cables to the head of the patient by means of a bandage.

Twenty-five normal subjects (10 males and 15 females) between the ages of 18 and 35 years were subjected to the above procedure and their results used to establish normal limits.

Cooperation and the ability to undertake a certain degree of effort were necessary for the performance of the exercise and voluntary hyperventilation. The results from the latter test were dependent upon the vigor with which it was performed and failure to do so was the most frequent cause for rejection from this series.

The congenital heart anomalies studied consisted of the following types with the number in each group in parenthesis: Atrial septal defect (5); high ventricular septal defect (2); Eisenmenger’s complex (4); isolated pulmonic stenosis without septal defects (2); Fallot’s tetralogy and variants (8); pulmonic stenosis with intact ventricular septum...
and patent foramen ovale (4); Ebstein’s anomaly of the tricuspid valve with patent foramen ovale (2); patent ductus arteriosus with partial reversal of shunt (2). (See table 1.) It has been considered advisable to distinguish between an atrial septal defect and a patent foramen ovale in the conditions studied, although this cannot be decided by clinical and cardiac catheterization findings. An atrial septal defect allows a free communication between the left and right atrium whereas a patent foramen ovale has a valve-like action which will be partial or complete depending upon the size of the foramen. Both pulmonic stenosis with intact ventricular septum14, 15 and Ebstein’s anomaly11 are so commonly (if not invariably) associated with a patent foramen ovale when a defect between the atra exists that it has been described as a patent foramen ovale in preference to an atrial septal defect. The possible significance of the valve-like action of the patent foramen ovale in the results obtained will be discussed.

The diagnosis of atrial septal defect had been confirmed by venous catheterization of the heart. The cases of high ventricular septal defect and Eisenmenger’s complex were considered together because of the similarity of the findings obtained by cardiac catheterization. Systolic and diastolic hypertension were present in the pulmonary artery, and pulmonic stenosis was excluded by the absence of a systolic pressure increase as the catheter was withdrawn from the pulmonary artery to the right ventricle. A ventricular septal defect was suggested by the finding of an oxygen content of blood from the right ventricle exceeding that from the right auricle by more than 1 volume per cent, by catheterization of the aorta from the right ventricle, or by early opacification of the aorta during angiography. The criteria for classifying a case as one of Eisenmenger’s complex as opposed to a high ventricular septal defect was a resting or effort induced arterial unsaturation in the former. Four of the six cases of high ventricular septal defect and Eisenmenger’s complex had undergone cardiac catheterization. An angiogram had demonstrated a high ventricular septal defect by early opacification of the aorta in one case in which this had not been shown by blood studies. The aorta had been catheterized from the right ventricle in two cases. Typical clinical, radiologic and electrocardiographic evidence of Eisenmenger’s complex was present in the two remaining cases (M. J. and C. G.).

Isolated pulmonic stenosis had been demonstrated by cardiac catheterization in two cases. Blood oxygen studies excluded the presence of arteriovenous and veno-arterial shunts. Six cases were considered to have Fallot’s tetralogy. Pulmonic valvular or infundibular stenosis had been demonstrated by catheterization in four cases (S. C., C. W., N. F. and D. G.). An over-riding aorta had been catheterized in one of those cases (N. F.), while an angiogram had revealed a similar defect in another (C. S.). The diagnosis of Fallot’s tetralogy was confirmed at postmortem in case C. W., who died postoperatively. Two cases were diagnosed on clinical, electrocardiographic and radiologic findings of this condition (G. T. and N. N.). One of these cases of Fallot’s tetralogy (G. T.) had a murmur of patent ductus arteriosus. Two cases were considered to have pulmonary atresia, an over-riding aorta and well developed (clinical and radiologic) findings of a bronchial artery collateral circulation to the lung (V. H. and J. S.). This variant of Fallot’s tetralogy is generally referred to as “pseudo-truncus arteriosus.” A high ventricular septal defect was shown by catheterizing the aorta in case J. S., and pulmonary arteries could not be found at operation.

The diagnosis in four cases was pulmonic stenosis with intact ventricular septum and patent foramen ovale. Case V. A. showed pulmonic stenosis on cardiac catheterization. A blood sample taken from the left auricle showed an oxygen saturation similar to that of the femoral artery blood, indicating that a venous to arterial shunt took place entirely through an atrial septal defect. The diagnosis of this condition was confirmed at autopsy in one case (T. R.) in which infundibular stenosis of the right ventricle and a widely patent foramen ovale were present. Clinical evidence of pulmonic valvular stenosis was present in two cases (P. B. and B. M.) and an atrial septal defect was demonstrated during cardiac catheterization.

Ebstein’s anomaly of the tricuspid valve with patent foramen ovale was considered to be present in two cases.11 The details of these cases have been published separately.12 One case was cyanotic at rest while both showed a marked reduction in arterial saturation during effort. Cardiac catheterization showed normal pressures in the pulmonary artery, absence of pulmonic stenosis, and the division of the right ventricle into two distinct pressure zones. The pressures in the proximal (or inflow) part of the right ventricle and right auricle were similar. That the pressure was actually recorded from the proximal part of the right ventricle was indicated by the position of the catheter tip well to the left of the vertebral column during fluoroscopy. The catheter had been withdrawn from the pulmonary artery to this site and could not have entered any other chamber than the right ventricle. The catheter was withdrawn from this site well to the right of the vertebral column and under these circumstances was considered to be in the right auricle. These changes were demonstrated by a continuous pressure tracing from pulmonary artery to right auricle in case M. H. It was concluded that the right ventricle was divided by a displaced tricuspid valve. The only possible shunt capable of explaining the cyanosis on the basis of findings at catheterization was a venous-arterial shunt through a patent foramen ovale.
Two cases were diagnosed as patent ductus arteriosus with partial reversal of the shunt. The classical
artery shunt. The pressures in the systemic and pulmonary artery circulations were such that a

Table 1.—The Effects of Voluntary Hyperventilation, Exercise and Oxygen Administration on the Arterial Oxygen Saturation (Per Cent)

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Initials</th>
<th>Age yrs.</th>
<th>Sex</th>
<th>Voluntary Hyperventilation</th>
<th>Exercise</th>
<th>Oxygen</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>0 20 40 60</td>
<td>0 20 40 60</td>
<td>0 1 2 3 4 5</td>
</tr>
<tr>
<td>Atrial septal defect</td>
<td>H. C.</td>
<td>13</td>
<td>F</td>
<td>91 92 92 93</td>
<td>91 91 91 92</td>
<td>93 93 94 94 94</td>
</tr>
<tr>
<td>G. H.</td>
<td>27</td>
<td></td>
<td>F</td>
<td>94 95 96 96</td>
<td>92 94 94 94</td>
<td>94 97 97 97 97</td>
</tr>
<tr>
<td>A. S. P.</td>
<td>56</td>
<td></td>
<td>F</td>
<td>93 94 96 96</td>
<td>92 92 92 88</td>
<td>93 101 100 100 100</td>
</tr>
<tr>
<td>E. R.</td>
<td>18</td>
<td></td>
<td>F</td>
<td>93 94 95 95</td>
<td>90 92 92 88</td>
<td>93 98 98 98 98</td>
</tr>
<tr>
<td>P. W.</td>
<td>13</td>
<td>M</td>
<td></td>
<td>93 96 98 98</td>
<td>93 94 92 94</td>
<td>93 100 100 100</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>High ventricular septal</td>
<td>F. A.</td>
<td>12</td>
<td>F</td>
<td>93 95 95 95</td>
<td>93 93 92 92</td>
<td>93 97 97 97 97</td>
</tr>
<tr>
<td>Defect &amp; Eisenmenger's complex</td>
<td>C. J.</td>
<td>11</td>
<td>F</td>
<td>94 95 96 96</td>
<td>93 94 93 93</td>
<td>94 97 97 97 97</td>
</tr>
<tr>
<td>C. F.</td>
<td>11</td>
<td>M</td>
<td></td>
<td>92 93 93 94</td>
<td>93 91 88 84</td>
<td>95 99 100 100 101</td>
</tr>
<tr>
<td>M. J.</td>
<td>20</td>
<td>M</td>
<td></td>
<td>90 89 83 90</td>
<td>93 92 86 86</td>
<td>92 97 98 99 99</td>
</tr>
<tr>
<td>C. G.</td>
<td>28</td>
<td>F</td>
<td></td>
<td>83 80 80 79</td>
<td>83 79 73 66</td>
<td>82 88 88 88 88</td>
</tr>
<tr>
<td>A. S.</td>
<td>21</td>
<td>F</td>
<td></td>
<td>79 80 76 75</td>
<td>80 71 58 46</td>
<td>83 87 91 89 89</td>
</tr>
<tr>
<td>Isolated pulmonic stenosis</td>
<td>B. M.</td>
<td>10</td>
<td>F</td>
<td>94 95 96 96</td>
<td>94 95 95 94</td>
<td>94 97 98 98 98</td>
</tr>
<tr>
<td>I. R.</td>
<td>7</td>
<td>M</td>
<td></td>
<td>91 92 93 93</td>
<td>— — — — —</td>
<td>92 96 96 96 96</td>
</tr>
<tr>
<td>Fallot's tetralogy</td>
<td>S. C.</td>
<td>18</td>
<td>F</td>
<td>66 61 60 62</td>
<td>66 58 45 36</td>
<td>61 68 68 70 72</td>
</tr>
<tr>
<td>(Postop.)</td>
<td>89</td>
<td>90 89 89</td>
<td></td>
<td>89 91 91 90</td>
<td>91 97 98 98</td>
<td>98 98 98</td>
</tr>
<tr>
<td>C. W.</td>
<td>33</td>
<td>F</td>
<td></td>
<td>66 64 63 60</td>
<td>50 46 40 36</td>
<td>68 70 70 72 73</td>
</tr>
<tr>
<td>N. F.</td>
<td>7</td>
<td>F</td>
<td></td>
<td>69 70 68 68</td>
<td>69 70 66 64</td>
<td>70 74 76 77 77</td>
</tr>
<tr>
<td>R. N.</td>
<td>7</td>
<td>M</td>
<td></td>
<td>90 91 87 87</td>
<td>90 89 87 84</td>
<td>90 97 97 97 97</td>
</tr>
<tr>
<td>T. G.</td>
<td>20</td>
<td>F</td>
<td></td>
<td>90 90 88 87</td>
<td>89 86 76 69</td>
<td>91 101 102 102 102</td>
</tr>
<tr>
<td>D. G.</td>
<td>13</td>
<td>M</td>
<td></td>
<td>75 76 75 73</td>
<td>70 69 67 66</td>
<td>62 73 73 73 76</td>
</tr>
<tr>
<td>T. V.</td>
<td>7</td>
<td>M</td>
<td></td>
<td>80 81 78 79</td>
<td>80 78 74 72</td>
<td>80 87 88 88 88</td>
</tr>
<tr>
<td>T. J. S.</td>
<td>7</td>
<td>M</td>
<td></td>
<td>84 85 84 84</td>
<td>80 78 72 69</td>
<td>85 89 91 91 91</td>
</tr>
<tr>
<td>Pulmonic stenosis</td>
<td>P. B.</td>
<td>13</td>
<td>F</td>
<td>81 82 82 82</td>
<td>79 73 65 61</td>
<td>81 87 88 90 90</td>
</tr>
<tr>
<td>with intact ventricular septum &amp; patent foramen ovale</td>
<td>T. R.</td>
<td>55</td>
<td>M</td>
<td>78 78 78 78</td>
<td>— — — — —</td>
<td>— 79 81 82 82</td>
</tr>
<tr>
<td>V. A.</td>
<td>24</td>
<td>F</td>
<td></td>
<td>84 87 85 84</td>
<td>85 80 70 63</td>
<td>86 93 96 97 97</td>
</tr>
<tr>
<td>B. M.</td>
<td>22</td>
<td>M</td>
<td></td>
<td>81 81 83 81</td>
<td>81 78 75 73</td>
<td>80 86 88 88 88</td>
</tr>
<tr>
<td>Ebstein's anomaly with patent foramen ovale</td>
<td>M. A.</td>
<td>21</td>
<td>F</td>
<td>87 91 92 93</td>
<td>87 85 80 77</td>
<td>91 96 97 97 97</td>
</tr>
<tr>
<td>I. N.</td>
<td>20</td>
<td>F</td>
<td></td>
<td>67 70 72 76</td>
<td>72 66 68 59</td>
<td>72 84 86 86 86</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>H. G.</td>
<td>16</td>
<td>F</td>
<td>93 96 96 96</td>
<td>93 91 90 89</td>
<td>93 98 99 98 98</td>
</tr>
<tr>
<td>with partial reversal of the shunt</td>
<td>D. H.</td>
<td>34</td>
<td>F</td>
<td>87 88 89 88</td>
<td>87 86 86 86</td>
<td>87 96 96 — — —</td>
</tr>
</tbody>
</table>

* This reading was preceded by a period of coughing.
† Fallot's tetralogy and patent ductus arteriosus.
‡ Classified as pulmonic atresia with well developed bronchial artery collateral supply to the lungs: "pseudotruncus arteriosus."

The diastolic component of the murmur of patent ductus arteriosus was absent, yet cardiac catheterization demonstrated the presence of an aortic to pulmonary pulmonary artery to aortic shunt was possible. The reversal of the shunt was demonstrated in case D. H. by the finding of a higher saturation in the right
brachial artery than in the femoral artery, and a patent ductus arteriosus was demonstrated and ligated in the other (H. G.).

**Results**

The change in arterial oxygen saturation was calculated as an increase or decrease above or below the resting level. Normal subjects, 25 in number, showed a progressive rise in arterial oxygen saturation during maximal voluntary ventilation. The average increase was 3.5 per cent at the end of one minute, with a range of +2 to +7 per cent. The standard exercise produced very little change. An average decrease of 1 per cent was present at the end of one minute with a maximal increase of 1 per cent and a maximal decrease of 3 per cent. A further slight fall took place when exercise was stopped. The majority returned to the resting level one minute after completing the test. Oxygen administration caused an average increase of 4.5 per cent at the end of one minute. A further slight increase took place so that at the end of five minutes the average increase was 5 per cent, with a range of +3 to +8 per cent. A return to the resting level was generally achieved within five minutes of discontinuing oxygen. The normal limits are indicated by broken lines in figures 1 to 4.

Voluntary hyperventilation, exercise and oxygen administration resulted in changes within the normal limits in cases of uncomplicated atrial septal defect. (See table 1.) Exer-

**Fig. 1.** Cases F. A. and C. J. had high ventricular septal defects while the remainder were those of Eisenmenger's complex. The deviation from the resting arterial oxygen saturation is shown in this and subsequent figures during voluntary hyperventilation, exercise and oxygen administration. The normal limits are shown by broken lines while the average is indicated by a faint solid line. The broken vertical line indicates the end of the test procedure. The graph was drawn from data presented in table 1 and the results are discussed in the text.

**Fig. 2.** The results of five cases of Fallot's tetralogy are shown. Number 1 was case R. N., number 2 case G. T., number 3 case C. W., number 4 case N. N., number 5 case S. C., table 1.
cise and oxygen administration gave a normal response in two cases of uncomplicated pulmonic stenosis. The increase in arterial saturation during hyperventilation in these cases reached the lower limits of normal at the end of one minute. (See table 1.)

Two cases (C. J. and F. A.) in the group diagnosed as high ventricular septal defect or Eisenmenger's complex showed an essentially normal response and were classified as high ventricular septal defects. The remaining cases were diagnosed as Eisenmenger's complex because of a resting or effort induced arterial unsaturation (fig. 1). One case (C. F.) had a normal resting arterial saturation and the results of voluntary hyperventilation and oxygen administration were normal. Exercise, however, produced an abnormal fall. The remaining three cases (M. J., C. G. and A. S.) showed a marked drop during hyperventilation and exercise. In case M. J. the resting saturation was within normal limits, while in the other two cases they were reduced (C. G. 83 per cent, and A. S. 79 per cent). A greater than normal average increase took place in all cases during oxygen administration and there was a tendency for the maximal saturation to be reached later than occurred normally.

Voluntary hyperventilation and exercise caused an abnormal decrease in arterial saturation in all six cases diagnosed as Fallot's tetralogy, while only two cases showed a greater than normal increase in arterial saturation during oxygen administration (fig. 2). The postexercise drop in saturation was greater than normal in all cases. The two cases (V. H. and J. S.) with pulmonary atresia and a well-developed collateral circulation to the lungs (pseudo-truncus arteriosus), had resting arterial unsaturation and an abnormal decrease during exercise (table 1). Case V. H. showed an abnormal decrease during hyperventilation which was not found in case J. S. Both showed a greater than average increase on oxygen administration.

Voluntary hyperventilation produced a slight rise (case P. B.), no change (cases T. R. and B. M.), and an initial fall and then a rise (case V. A.) in the four cases diagnosed as pulmonary
stenosis with intact ventricular septum and patent foramen ovale (fig. 3). Exercise produced a marked drop in saturation in the three cases in which this test was performed. Two cases showed a greater than normal decrease in saturation immediately after exercise. Oxygen administration produced a greater than normal rise in three cases at the end of five minutes, and in all four cases the maximal saturation took longer to achieve than in normal subjects.

The cases of Ebstein's anomaly of the tricuspid valve associated with patent foramen ovale showed a marked increase in arterial saturation during hyperventilation (fig. 4). Exercise produced a greater than normal decrease in both. Oxygen administration produced a normal increase in one case (M. H.) and a greater than normal increase in the other (I. N.).

Partial reversal of a shunt through a patent ductus arteriosus caused resting arterial unsaturation in case D. H. and a greater than normal decrease during exercise in case H. G. (table 1). The arterial oxygen saturation of the right ear in case H. G. exceeded that in the left ear by 4 per cent, while no difference between the two ears was detected in case D. H. Whereas the fall in arterial oxygen saturation during exercise was of equal magnitude in both ears of case H. G., that in the left ear was greater than the right ear in case D. H.

**Discussion**

The results confirm the well documented effects of exercise and oxygen administration on the arterial blood oxygen saturation in congenital cardiac anomalies. The presence of a venous-arterial shunt was associated with one or more of the following findings: a resting arterial oxygen unsaturation; an abnormal decrease during exercise and a further decrease immediately after exercise; an abnormally prolonged time to attain maximal saturation and failure to attain normal saturation on oxygen administration. The results of exercise, oxygen administration, and voluntary hyperventilation were within the normal limits in cases with arteriovenous shunts. This has been shown in atrial septal defects in the present study and in cases of patent ductus arteriosus and coarctation not included here. The differentiation between an arteriovenous and venous-arterial shunt can be made in the majority of cases by the changes in arterial saturation that result from exercise and oxygen administration. These tests, however, do not indicate the site of the shunt. The changes in arterial oxygen saturation during voluntary hyperventilation appeared to be of more definitive value.

A less than normal rise or an actual fall in arterial oxygen saturation during hyperventilation was found in cases with pulmonic stenosis such as Fallot's tetralogy, pulmonic stenosis with intact ventricular septum and patent foramen ovale, and isolated pulmonic stenosis. A similar result was obtained in some cases of pulmonary hypertension. With two exceptions, this occurred in the cases of Eisenmenger's complex and in one of the patients with a partial reversal of a shunt through a patent ductus arteriosus. It is likely that this result will only be obtained in pulmonary hypertension when the pulmonary resistance is relatively fixed. Under such circumstances it will limit the increase in arterial saturation in the same way as pulmonic stenosis. A less than normal increase in arterial oxygen saturation during hyperventilation was only found in the presence of pulmonic stenosis and in some cases of pulmonary hypertension, and did not occur when these conditions were absent.

In normal subjects the rise in arterial oxygen saturation during hyperventilation is due to the net effects of factors favoring the increased uptake of oxygen and the increased utilization of oxygen brought about by the effort involved in the procedure. Hyperventilation will increase the partial pressure of oxygen in the alveoli and capillary blood in the absence of lung disease. It will also increase the blood flow through the lungs by the "pumping" action of breathing. The less than normal increase in arterial oxygen saturation in pulmonic stenosis and some cases of pulmonary hypertension can be ascribed to the limitations these conditions imposed on any increase in the blood flow through the lungs.

The presence of a resting arterial unsaturation in some and a greater than normal fall during exercise in all the cases diagnosed as Eisenmenger's complex, Fallot's tetralogy, pul-
monic stenosis with intact ventricular septum and patent foramen ovale, and Ebstein's anomaly with patent foramen ovale established the presence of a potential or actual venous-arterial shunt. Voluntary hyperventilation, however, produced distinctive changes. The cases with pulmonic stenosis with intact ventricular septum and patent foramen ovale showed little change from the resting arterial saturation while those with Fallot's tetralogy and Eisenmenger's complex showed a fall. This procedure appears to be of clinical value in the differentiation of pulmonic stenosis with intact ventricular septum and patent foramen ovale and Fallot's tetralogy. Eisenmenger's complex is readily distinguished from these anomalies on clinical and radiologic grounds. The well marked increase in arterial oxygen saturation during hyperventilation and a fall during exercise is a distinctive finding in Ebstein's anomaly of the tricuspid valve associated with a patent foramen ovale and has not been found in any other type of cyanotic congenital heart disease.

Voluntary hyperventilation produced a fall in arterial saturation in the conditions associated with a high ventricular septal defect (Fallot's tetralogy and some cases of Eisenmenger's complex), whereas those associated with a patent foramen ovale (pulmonic stenosis with intact ventricular septum and Ebstein's anomaly) showed little change or a rise, respectively. Preliminary observations in normal subjects indicate that voluntary hyperventilation lowers the total systemic peripheral resistance since it increases the cardiac output proportionately more than it increases the mean systemic blood pressure. The resistance of a stenosed pulmonic valve or lungs with marked pulmonary hypertension is likely to remain fixed. Hyperventilation in the presence of a high ventricular septal defect and pulmonic stenosis or marked pulmonary hypertension would thus favor an increase in right ventricle to aorta shunt and cause a decrease in arterial saturation. That hyperventilation does not alter the gradient of the shunt in pulmonic stenosis with intact ventricular septum and patent foramen ovale was indicated by the negligible changes that occurred in arterial saturation. The well marked increase in arterial saturation in the two cases of Ebstein's anomaly during hyperventilation was most probably indicative of a reversal of the shunt between the atria caused by a marked increase in pulmonary blood flow.

A patent foramen ovale opens from the right to the left atrium and will remain open only when the pressure in the right atrium remains higher than that in the left. Hyperventilation may increase pulmonary blood flow in Ebstein's anomaly sufficiently to raise the pressure in the left atrium and cause a partial or complete closure of the foramen ovale. A similar mechanism may operate in pulmonic stenosis with intact ventricular septum and patent foramen ovale, although any increase in blood flow will be limited by the pulmonic stenosis.

The diagnosis of Eisenmenger's complex is justified when a high ventricular septal defect and pulmonary hypertension occur in a patient with cyanosis or arterial unsaturation. The cyanosis is generally late in onset and before this occurs it is extremely difficult to distinguish these cases from those with atrial septal defect by clinical means. The presence of cyanosis at rest in two of the six cases classified as Eisenmenger's complex or high ventricular septal defect made the differentiation from atrial septal defect possible. The remaining four cases showed a normal resting arterial saturation. In two of these it was possible to cause an abnormal decrease in saturation during exercise and on this basis they were separated from cases of atrial septal defect in whom the resting saturation and the change during oxygen administration, exercise and voluntary hyperventilation were normal. It is justified to consider the two cases with effort-induced unsaturation as cases of Eisenmenger's complex and to classify those in whom it did not occur as cases of high ventricular septal defect. Eisenmenger's complex is associated with dextroposition of the aorta by definition. This feature may be absent in cases of high ventricular septal defect and on these grounds it is advisable to differentiate between these conditions.

An increase in pulmonary artery pressure may occur in some cases of patent ductus arteriosus and be so great that a partial or complete reversal of the shunt results. The classic murmur of patent ductus arteriosus dis-
appears under such circumstances. In one such case Cournand and colleagues have shown that the oxygen content of blood from the right brachial artery exceeded that from the femoral artery. The nature of the blood flow from pulmonary artery to aorta causes venous admixture of blood going to the femoral artery but not (or very little) to that going to the right brachial artery. Our cases demonstrate that venous blood may flow into the left carotid artery and cause the arterial saturation in the left ear to be lower than that in the right. When this difference is not present at rest it may become apparent during exercise.

The oximeter provides a means of obtaining reasonably accurate measures of the arterial oxygen saturation during a variety of tests which appear to be of value in the differentiation of a number of congenital heart anomalies. Measurement of the arterial oxygen saturation at rest and during oxygen administration provides no practical difficulties. The performance of a sufficiently vigorous exercise and voluntary hyperventilation, however, necessitates cooperation and a certain degree of physical development and fitness. These latter tests can generally be performed by patients over the age of six years. The interpretation of the results from voluntary hyperventilation are only justified when the test has been performed strenuously. A better assessment of the results of this test would be to compare the changes in arterial saturation with the amount of ventilation achieved and measured as currently done for the maximum breathing capacity.

Conclusions

1. An oximeter has been used to measure the changes in arterial oxygen saturation during voluntary hyperventilation, exercise and oxygen administration in 25 normal subjects and 30 cases of congenital heart disease.

2. The nature of the congenital heart anomaly was established by clinical study and venous catheterization of the heart in the majority of cases. The congenital heart anomalies studied consisted of the following types with the number in each group in parentheses: atrial septal defect (5); high ventricular septal defect (2); Eisenmenger's complex (4); isolated pulmonic stenosis without septal defects (2); Fallot's tetralogy and variants (8); pulmonic stenosis with intact ventricular septum and patent foramen ovale (4); Ebstein's anomaly of the tricuspid valve with patent foramen ovale (2); Patent ductus arteriosus with partial reversal of shunt (2).

3. Congenital heart disease associated with an arteriovenous shunt was characterized by an essentially normal response. This was found to be true for cases of atrial septal defect described in the present study and cases of uncomplicated patent ductus arteriosus and coarctation not included in this report.

4. Congenital heart disease associated with a venous-arterial shunt showed one or more of the following results: A resting arterial oxygen unsaturation; an abnormal reduction in saturation during exercise; an abnormally prolonged time to attain maximal saturation, and a failure to attain full saturation on oxygen administration. Although these findings indicate the presence of a venous-arterial shunt, they do not indicate the site of the shunt.

5. The effect of voluntary hyperventilation depended upon the presence or absence of pulmonic stenosis or pulmonary hypertension and the site of the venous-arterial shunt. A less than normal increase in arterial oxygen saturation was found in cases of pulmonic stenosis and in some cases with marked pulmonary hypertension irrespective of the associated abnormalities. This was attributed to a failure to increase pulmonary blood flow during this maneuver. Pulmonic stenosis with intact ventricular septum and a patent foramen ovale was found to produce little change in saturation during hyperventilation, whereas high ventricular septal defects in association with pulmonic stenosis (Fallot's tetralogy) or pulmonary hypertension (Eisenmenger's complex) resulted in a distinct fall. The response to voluntary hyperventilation would appear to offer a means of distinguishing a patent foramen ovale and a high ventricular septal defect when these conditions are associated with pulmonic stenosis or pulmonary hypertension.

6. A marked fall in arterial saturation during exercise was found in two cases of Ebstein's anomaly of the tricuspid valve and patent
foramen ovale, but unlike other types of cyanotic congenital heart disease, there was a normal response during hyperventilation.

7. The possible mechanisms of these different responses during voluntary hyperventilation have been discussed, and it is suggested that this procedure may be of differential diagnostic value in the study of congenital heart disease.

Acknowledgements

We wish to express our appreciation of the help received from Prof. G. A. Elliott and Drs. M. McGregor, J. Kaye, J. H. Gear and T. H. Bothwell. Dr. H. B. Taussig kindly reviewed the manuscript.

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Oximetry in Congenital Heart Disease with Special Reference to the Effects of Voluntary Hyperventilation

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_Circulation_. 1952;6:740-748
doi: 10.1161/01.CIR.6.5.740

_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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