

Orthocyanosis: a sign of pulmonary arteriovenous malformation

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Normally the arterial oxygen tension (P_{aO_2}) remains the same or rises with the change from the supine to the upright position.¹ Several patients showing the opposite phenomenon (orthodeoxia) have been reported, some with a pulmonary arteriovenous malformation.²

While cyanosis is a recognised feature of pulmonary arteriovenous malformation,^{3,4} our patient is of interest because she had cyanosis when she was upright but not when she was supine (orthocyanosis).

Case report

A 17-year-old girl was referred for investigation of polycythaemia. She had had no dyspnoea, haemoptysis, chest pain, epistaxis, or melaena. When examined supine her blood pressure was 100/70 mm Hg, pulse 56 beats/min, respiratory rate 12/min, and temperature 37°C. She did not have cyanosis, clubbing, or telangiectasis. Examination of the lungs showed nothing abnormal. The heart size and sounds were normal. A systolic bruit was easily heard over the left anterolateral part of the chest. An inspiration greater than tidal volume increased the systolic intensity of the bruit. Examination showed cyanosis when she was upright. The bruit increased in intensity and became continuous with systolic accentuation.

The haemoglobin concentration was 16.7 g/100 ml with a haematocrit of 47%. The electrocardiogram was normal. The chest radiograph showed a 4.5 x 5.0 cm non-homogeneous mass in the left lower lobe obliterating part of the anterior hemidiaphragm. Blood gases were obtained with the patient in the supine and upright positions having 21% and 100% oxygen. The results are recorded in the table. The shunt fraction was obtained with Benatar's virtual isoshunt lines.⁵ Haemodynamic data from catheterisation of the left and right side of the heart showed normal pulmonary artery pressure, pulmonary resistance, and cardiac output, with pulmonary flow equal to systemic flow. A pulmonary angiogram showed an arteriovenous malformation in the left lower lobe (fig 1). An uneventful lobectomy was carried out. After surgery cyanosis was no longer observed and the blood gases showed improved gas exchange (table).

Discussion

In pulmonary disease the P_{aO_2} most commonly rises in the upright position above its level in the supine position. In contrast, orthodeoxia has been observed in congenital heart disease with right-to-left shunt. The explanation for both phenomena has been well documented.^{1,6} In these patients the cyanosis present in the

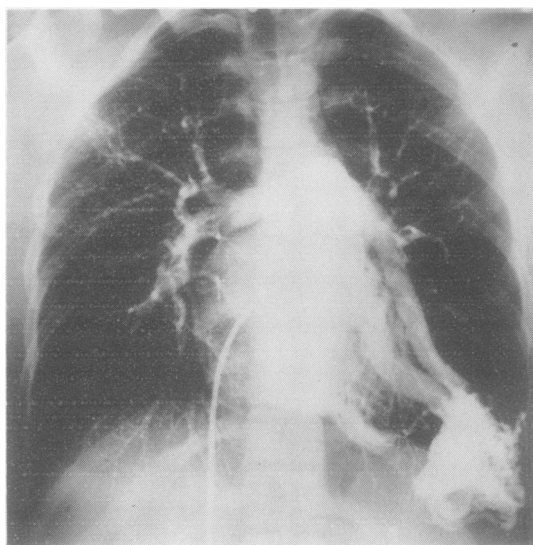


Fig 1 Pulmonary angiogram showing the malformation in the left lower lobe (patient in supine position).

Arterial blood gases before and after surgery

	Before surgery				After surgery			
	Upright		Supine		Upright		Supine	
F_{IO_2}	0.21	1.0	0.21	1.0	0.21	1.0	0.21	
pH	7.43	7.42	7.42	7.43	7.42	7.43	7.42	
P_{aCO_2} mm Hg	25	28	34	30	38	34	38	
P_{aO_2} mm Hg	45	52	59	136	82	500	83	
$S_{aO_2}\%$	83	88	91	99	96	100	96	
Q_s	0.45		0.23		0.08			

upright position has been diminished or reversed in the supine position. More recently orthodeoxia has been reported in true vascular lung shunts, several due to pulmonary arteriovenous malformation.²

The pathophysiology of orthodeoxia in lung vascular shunts is quite different from that in right-to-left intracardiac shunts. The lung has a two-compartment circulation, the alveolar or small vessel compartment and the extra-alveolar or large vessel compartment.^{7,8} With lung inflation, resistance rises in the alveolar vessels and falls in the extra-alveolar vessels. With deflation, the opposite occurs. This is due to the regional determinants of blood flow — namely, arterial, alveolar, and venous pressure.⁹

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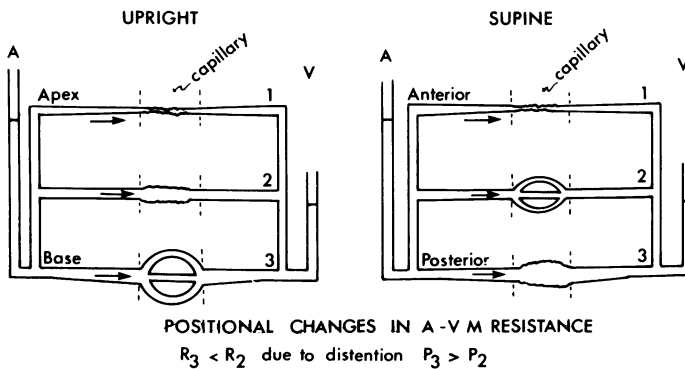


Fig 2 Hydrostatic pressure (P) relationships and blood flow through zones 1, 2 and 3 showing the change in resistance (R) and flow through the malformation with change in position: $R_3 < R_2$ because of distention; $P_3 > P_2$. A—arterial; V—venous.

In the arteriovenous malformation the vessels act as extra-alveolar vessels.^{3 10} Since they are not in series with alveolar vessels, the flow through them for a given pulmonary arteriovenous pressure difference is determined by the absolute hydrostatic distending pressure (P) at a given lung volume and the distending forces of the surrounding lung at varying lung volumes.^{7 10}

With our patient in the supine position the arteriovenous malformation was at the level of the heart, probably in zone 2 (fig 2) and the shunt was 23%. When she was breathing room air the P_{aO_2} was 59 mm Hg and no cyanosis was noted. When she was upright the arteriovenous malformation was below the level of the heart, probably in zone 3. It exerted less resistance to blood flow in the upright position because the distending P_3 was greater than P_2 (fig 2). Furthermore, the resistance in the alveolar vessels was probably higher in the upright position than in the supine position because of a predicted increase in functional residual capacity.¹⁰ With resistance probably increased in the alveolar vessels and diminished in the arteriovenous malformation when she was upright flow through it increased. Accordingly, the shunt increased to 45%, the P_{aO_2} fell to 45 mm Hg, and cyanosis became evident. With further increase in lung volume during inspiration, further increase in flow through the arteriovenous malformation would be expected. This is the probable explanation for the increase in the intensity of the bruit during inspiration.

Orthocyanosis differs from orthodeoxia in the degree of arterial oxygen desaturation. This case is of interest for this reason. The arteriovenous malformation, although congenital, generally does not become clinically manifest until the third decade.³ It results from malformation of arteries and veins from a common capillary plexus. It is thin walled and gradually distends with time owing to constant exposure to arterial pressures.³ It has been shown to increase further in size even after it becomes clinically evident.⁴ In the evolution of arteriovenous malformation orthocyanosis therefore may be a transient sign of a

long-term dynamic range of clinical manifestations. These include mild hypoxaemia with orthodeoxia, moderate hypoxaemia with orthocyanosis, and severe hypoxaemia with cyanosis in both the upright and the supine positions.

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