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Short reports

Lung developmental abnormalities in severe scoliosis

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We report a case of severe scoliosis developing before the age of 1 year in which severe cardiopulmonary disorder led to death at the age of 36. Results of detailed physiological study and postmortem lung morphometry are presented.

Case history

The patient was first noted to have a thoracic scoliosis, convex to the left, soon after birth. She had no congenital abnormalities or neuromuscular disease but was always mildly dyspnoeic on exertion during childhood and early adult life. This began to worsen in 1979 at the age of 34. By 1981 she had ankle swelling and regular early morning headaches and nausea. At this time systemic blood pressure was 130/95 mm Hg and her jugular venous pressure was raised with a prominent "a" wave; her pulmonary second sound was loud and a 4th heart sound audible.

Radiographs showed that the scoliosis was at the level of the 5th thoracic vertebra. Her height was 142 cm and arm span 161 cm and the Cobb angle was 140°. Her electrocardiogram was normal. FEV_1 and forced vital capacity were both 0·6 l, the residual volume was 0·95 l, functional residual capacity 1·10 l, and total lung capacity 1·55 l. Physiological data at rest and during exercise on a bicycle ergometer at 25 watts are shown in table 1. A few hours afterwards her condition deteriorated and her respiration was monitored continuously with an impedance spirometer together with an electrocardiographic trace. Frequent periods of hypoventilation and apnoea associated with transient atrial fibrillation were noted before a prolonged apnoeic spell led to asystole. Attempted resuscitation was unsuccessful.

Pathology and morphometric analysis

Macroscopically the lungs were deformed and slightly oedematous. The left lung weighed 320 g and the right lung 440 g. The heart weighed 270 g and the right ventricle was grossly hypertrophied and up to 0·7 cm in thickness; the right ventricle weighed 89·8 g and the (LV + IVS)/RV ratio was 1·4, confirming right ventricular hypertrophy.[1] The lungs were inflated and fixed by continuous perfusion of formaldehyde through the airways.[2] The volume of each lung was measured by displacement of water and 1 cm square blocks were taken from slices, a random number sampling grid being used. The number of randomly obtained blocks taken from each lobe allowed the calculation of lobar volumes (table 2). With a Zeiss integrating micrometer disc Turret 1 at a magnification of 400 the radial alveolar count (RAC) (derived from 48 fields[3]) and the mean linear intercept (MLI) were estimated. The surface area of the alveolar-air interface (SAT) was calculated on the basis of the formula[4] SAT = (4 × volume of fixed parenchyma × 0·78)/MLI.

Point counting of 100 points was performed on sections from every block to calculate the absolute volume of air space, blood, and tissue in conjunction with the displacement lung volume. All the morphological estimates were performed blind (table 2).

The morphometric results showed that the lungs were abnormally small. The volume of air from point counting was 1·54 l, indicating a chronological age of 5–6 years[5]; and the SAT of 26·1 m^2 corresponded to a similar age.[5] The RAC, which gives a measure of the number of alveoli per acinus, was only 8·43, corresponding to a developmental age of less than 1 year. The left upper and lower lobes are normally of about the same size[6] but here the difference was very striking, the left upper lobe contributing only 28·2% of the volume of the left lung. The lobar proportions within the right lung were normal.

Discussion

This patient's scoliosis developed before the age of 1 year so it seems probable that it influenced the mechanical forces determining lung growth at this early age. This would account for the very low RAC value, corresponding to a child of less than 1 year old. Interestingly, her scoliosis was convex to the left and high thoracic. This might be expected to have had its most profound effect on the left upper lobe, which was indeed the lobe most diminished in volume.

Whether the alteration in lung growth was due to development of abnormally few alveoli or of alveoli that were too small, or both, cannot be definitely answered from our data. Alveolar multiplication is thought to be largely completed by the age of 2 years,[5] although it continues until 8 years.[7] The low RAC indicates that each acinus had fewer alveoli than normal but it is possible, although unlikely, that the total number of acini was increased. The similarity of the overall developmental age indicated by the lung volume and the SAT plus the lower age indicated by the RAC implies that the average alveolar size was larger than normal as it is in compensatory overinflation after, for example, pneumonectomy. This makes it likely that the primary abnormality in lung development in scoliosis of early onset is the failure to develop a normal number of alveoli.

Our patient's pulmonary artery pressure and cardiac output hardly changed during exercise. This has not been

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Table 1 *Arterial blood gas and right heart catheterisation data at rest and during exercise*

	pH	$PaCO_2$ (kPa)	PaO_2 (kPa)	SO_2 (%)	Pressures (mm Hg)			Cardiac output (l/min)
					RV	PA	PA (w)	
Rest, supine	7·37	7·8	4·2	61	102/12	102/43 mean 60	7·5 (mean)	3·7
Rest, sitting		7·5	4·7	68		140/40		
Exercise: 1 min						140/40		
2 min		7·2	5·0	70		140/40		
3 min						140/40		3·88

$PaCO_2$—arterial carbon dioxide tension; PaO_2—arterial oxygen tension; SO_2—oxygen saturation; RV—right ventricle; PA (w)—pulmonary artery (wedged).

Table 2 *Postmortem morphometric analysis*

	Volume after fixation (ml)	Volume of air (ml)	Volume of blood (ml)	Volume of structure (ml)	RAC	MLI (cm)	SAT (m^2)
Left lung (total)	820·0	644·4	38·7	136·9	8·78 (0·34)	0·0218	11·736
Left upper lobe	231·0	179·0	12·3	39·7	8·24 (0·34)	0·0222	3·254
Left lower lobe	589·0	478·2	18·1	89·0	9·50 (0·51)	0·0209	8·793
Right lung (total)	1120·0	897·5	62·9	159·6	7·47 (0·43)	0·0243	14·380
Right upper lobe	411·4	328·2	24·1	59·0	8·08 (0·81)	0·0234	5·491
Right middle lobe	228·6	185·8	12·4	30·3	7·40 (0·82)	0·0248	2·876
Right lower lobe	480·0	383·2	26·3	70·4	7·00 (0·78)	0·0250	5·995

RAC—radial alveolar count; MLI—mean linear intercept; SAT—surface area of the alveolar-air interface.

recorded before in scoliosis but is recognised in severe pulmonary hypertension from other causes.

Pulmonary hypertension in scoliosis has been attributed to hypoxaemia during the day[8] or night[9] and also to anatomical restriction of the pulmonary vascular bed.[10] In this patient hypoxaemia was probably important since her arterial oxygen tension was only 4·2 kPa during the day and prolonged apnoeic spells were recorded at night. The pulmonary capillary blood volume could not be estimated from our morphological techniques but the very low SAT and RAC indicate extreme alveolar underdevelopment. If the pulmonary microcirculation was correspondingly underdeveloped the increase in pulmonary vascular resistance would contribute appreciably to the pulmonary hypertension.

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