Case reports

high triglyceride and cholesterol levels some months after the attacks in the absence of overt diabetes suggests a primary hyperlipoproteinaemia rather than a pancreatitis or hormone-induced abnormality.

The mechanism of development of pancreatitis in hyperlipaemic states is obscure, the most favoured explanation being vascular sludging due to the chylomicronaemia. It is possible that oral contraceptives, which have been claimed to increase the tendency to vascular thrombosis and also to elevate the serum levels of certain blood clotting factors, may set the foundation for enhanced chylomicron aggregation and clustering. Alternatively the hormonal effect of the Pill may aggravate one or more of the mechanisms suggested for the development of secondary pancreatitis-induced hyperlipaemia, viz., acute diabetes, the release of triglycerides from areas of fat necrosis, pancreatic a cell damage, increased release of glycerides from the liver into the plasma, defective intravascular clearing of glycerides due to lipoprotein lipase inhibition or indeed, the aggravation of the pre-existing defect in lipid metabolism (Marks, Bank & Louw, 1968). Fredrickson has found that oestrogens tend to raise and progesterone to reduce serum glyceride levels (Glueck et al., 1969).

The findings in the cases represented in the present paper suggest that pancreatitis be considered in patients on oral contraceptives who develop abdominal pain. While there is no evidence that oral contraceptives precipitate pancreatitis in patients with normal serum lipids, it is possible that pancreatitis may be a real hazard in patients with a pre-existing hyperlipaemia.

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References


Complete agenesis of the lung

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SCHNEIDER (1909–1913) divided pulmonary agenesis into three main degrees:

(a) True agenesis—a group in which there is complete absence of bronchi, alveolar tissue and their blood supply.

(b) A group in which a rudimentary bronchus arose from the trachea with no pulmonary tissue investing its tip.

(c) A group with a poorly developed main bronchus invested by a fleshy mass of ill-developed pulmonary tissue.

Agenesis may be unilateral or bilateral, involve the whole lung, or be lobar or segmental. The diagnosis is usually made accidentally in asymptomatic patients by X-ray or by physical signs of mediastinal shift. Recognition of the true nature of the lesion is important, so that unnecessary interference is avoided.

Cases of isolated pulmonary agenesis are relatively rare as the condition is more commonly associated with serious malformations of other organs that do not permit prolonged life. Oyamada, Gasul & Holinger (1953) found a high association between pulmonary agenesis and anomalies of musculo-skeletal, cardio-vascular, gastro-intestinal and urogenital systems. Bronchography and angiography may be required to establish the diagnosis.
Case reports

Case 1

A male patient aged 35 years with four healthy children. No history of congenital abnormalities could be obtained in his family.

When he was a schoolboy he suffered from occasional bronchitis and his family doctor mentioned to his parents that his heart was beating on the wrong side. Because of the finding of dextrocardia he was not accepted for military service.

In July 1958 when he was under treatment for aspirin poisoning, the cardiac impulse was observed to be on the right side of the chest. His only symptom was mild exertional dyspnoea. Because of this and the abnormal radiographic appearance he was referred for examination.

On examination: He was obese but otherwise healthy-looking, 196 pounds in weight, not cyanosed or dyspnoeic. Pulse 70 per minute, regular. Blood pressure 130/90 mmHg. The trachea was deviated to the right, but expansion of the chest was equal on both sides. FEV₁ was 1.8 litres and the FVC 2.85 litres. The heart sounds were normal and were best heard on the right fourth intercostal space one inch from the right sternal border. The electrocardiogram showed sinus rhythm. The left ventricular complexes from V₁ to V₆ indicated right sided displacement of the heart.

Radiographic examination showed that the right hemithorax was smaller than the left. The mediastinum was displaced to the right. The heart was posteriorly placed. Diaphragmatic movement was normal. The cause of dextrocardia was not apparent.

At bronchoscopy no right main bronchus was found and bronchography did not outline any of the right bronchial tree.

Angiography: A right ventricular angiogram did not outline a right pulmonary artery. There was no abnormality of the cardiac chambers. The excretion urogram showed that the left kidney was in the normal position and the right was displaced, lying vertically and lateral to the left border of the mid-lumbar spine.

No further investigation was undertaken and no treatment was advised.

Case 2

An asymptomatic girl aged 12 years. She had a normal birth but her mother suffered from hypermesis. The child was first examined when she was 9 months old because of episodes of bronchitis with tracheal rattles and an abnormal radiograph.

Radiographs of the chest showed that the heart occupied the posterior part of the right hemithorax. Bronchoscopy and bronchography confirmed the absence of a right bronchial tree.

At angiography, no right pulmonary artery was opacified.
When she was re-examined at the age of 12 she was free from respiratory complaints. She had a slight scoliosis of the spine and corrective exercises were advised.

Abnormalities of pulmonary vessels may be associated with pulmonary agenesis, (Ferencz, 1961).

Vascular compression of the trachea or bronchus between a posteriorly situated pulmonary artery and anteriorly situated aorta has been reported (Maier & Gauld, 1953).

Stenosis of the trachea caused either by compression or rarely by intrinsic congenital stenosis may lead to episodes of hypoxia and greatly raised pulmonary arterial blood pressure (Killingsworth & Hibbs, 1939; Nelson et al., 1967). This in turn may cause reversal of blood flow through a persistent ductus arteriosus necessitating urgent operative closure. Other vascular anomalies include patent ductus arteriosus, absent pulmonary arteries to both lungs and intra-cardiac septal defects.

It has been reported that in twins (Finkelstein, 1912, Yount, 1948) the agenesis may be on opposite sides.

Complete unilateral pulmonary agenesis was stated by Smart (1946) to occur once in 15000 autopsies. The majority of cases have been discovered at necropsy in infants.

In the absence of other serious defects, the condition is compatible with prolonged survival. Heerup (1927) described a patient who lived till the age of 72 and Lüdin & Werthemann (1930) another who lived to 58.

The single lung is much larger than normal. It fills the space that should be occupied by the absent lung and leads to displacement of the mediastinal contents away from itself.

The vital capacity may be within the normal range. Smart recorded as import, three clinical signs:
1. Symmetrical chest with almost equal expansion of both sides.
2. Gross displacement of trachea to one side.
3. Rotation of whole mediastinum.

Spirometric tests show that the enlargement of the single lung is due to hypertrophy and not to degenerative emphysematous changes.

Minor congenital abnormalities may be present in the other lung, in the chest wall and diaphragm.

The factors responsible for pulmonary agenesis are not fully understood.

The following have been recognized:
(a) Genetic (Neill, Ferencz, Sabiston & Sheldon, 1960) reported the case of a father and daughter with right pulmonary agenesis. Finkelstein (1912), Yount (1948) recorded agenesis in twins.

(b) Injury, threatened abortion and bacterial infection during first three months of pregnancy (Field, 1946).

(c) Viral infection following rubella in pregnancy (Campbell, 1961, Butler, et al., 1965; Forrester, Lees & Watson, 1966).

**Discussion**

Bilateral complete pulmonary agenesis was reported by Schmit (1893) in a foetus born after 8 months' gestation. Chaireaux & Ferreira (1958) described a full-term male infant whose trachea ended blindly. The pulmonary artery joined the aorta below the left subclavian artery (persistent left sixth aortic arch). The infant lived 15 min. No pulmonary veins or bronchial arteries were present and there was a left superior vena cava which entered the left atrium. Tuynman & Gardner (1952) described a similar case in which development had proceeded to the stage when primitive bronchial buds were present (26 day stage). The pulmonary artery joined the thoracic aorta at the point where the ductus arteriosus normally joins the pulmonary artery.

Morgagni (1762) was the first to describe pulmonary agenesis. Unilateral agenesis has been reviewed by Smart (1946), Burger (1947), Wexels (1951), Jimenez-Martinez et al. (1965) and Nelson, McMillan & Bharucha (1967).

Absence of the right lung is less common than absence of the left (70%). Males are more affected than females (Lucas, Dotter & Steinberg, 1953).
(d) Diet deficiency in Vitamin A during pregnancy in experimental rats (Wilson & Warkany, 1949).

The prognosis in congenital absence of one lung depends on the presence or absence of other associated congenital abnormalities, together with increased vulnerability to pulmonary infections.

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References

Smart, J. (1946) Complete congenital agenesis of lung. Quarterly Journal of Medicine, 15, 125.

Congenital abdominal coarctation with renal artery hypoplasia, hypertension and rheumatoid arthritis

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Stenosing lesions of the abdominal aorta are rare. They are of varied aetiology, and may be due to congenital anomalies, atherosclerosis, dissecting aneurysm and acquired lesions of the inflammatory type of specific and of non-specific origin. Abdominal coarctation can occur above or below the level of the renal arteries. If the renal arteries are involved a syndrome of Goldblatt hypertension is produced.

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This paper reports a case of congenital abdominal coarctation with renal artery hypoplasia and hypertension associated with rheumatoid arthritis in a girl of 13 years.

Case report

A girl, aged 13 years was admitted on 17 April 1967 with a history of swelling of her joints of 3 years' duration, headache and effort dyspnoea of 1 years' duration.
Complete agenesis of the lung.

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