PARTIAL AND TOTAL ANOMALOUS PULMONARY VENOUS DRAINAGE

An Essay

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By

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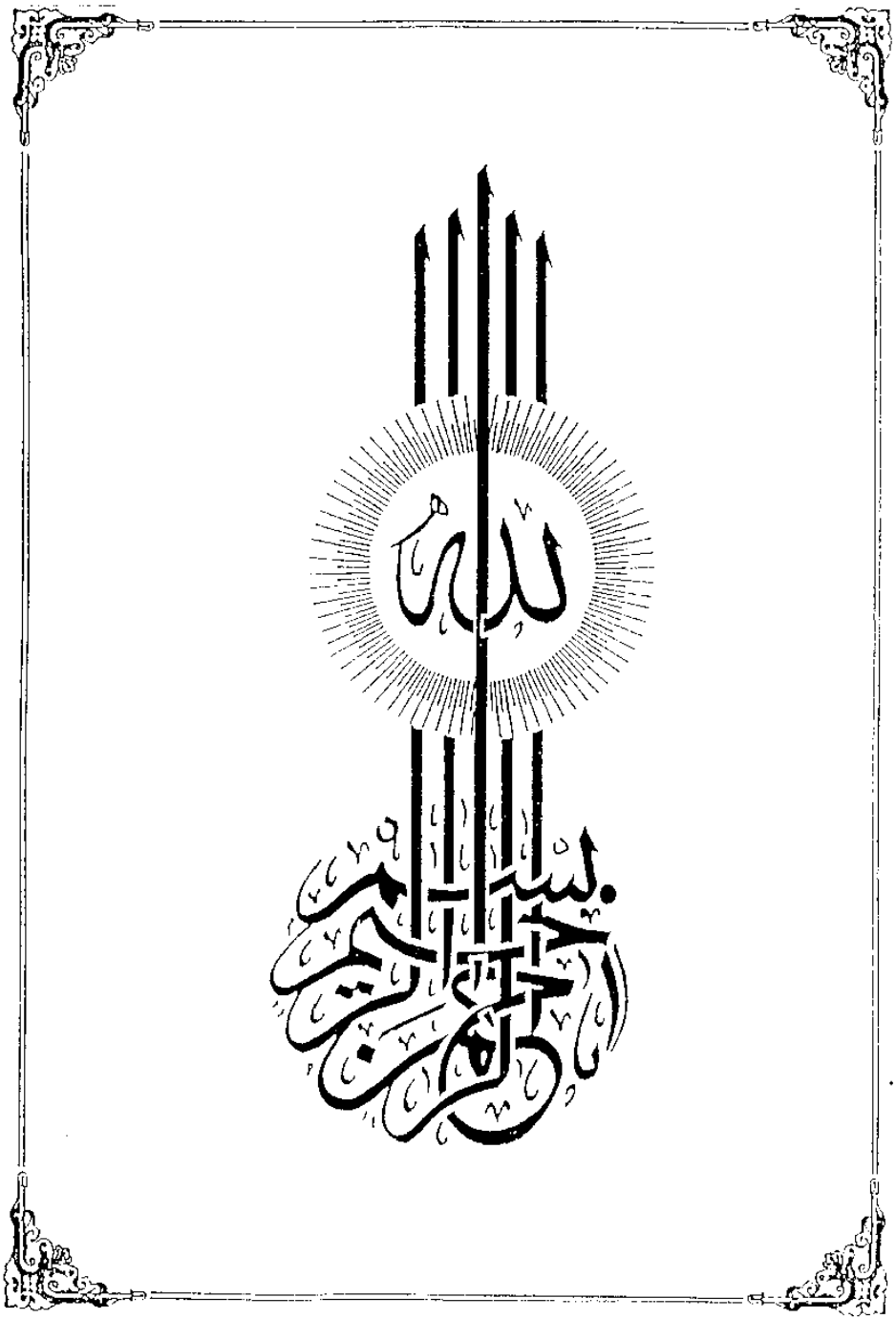

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INTRODUCTION

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Anomalous pulmonary venous drainage is either total or partial. Total anomalous pulmonary venous drainage is considered a rare congenital anomaly constituting 1.5-2% of congenital heart diseases. The condition was originally described by *James Wilson, 1978* and was usually fatal in infancy until cardiopulmonary by-pass with or without deep hypothermia became established.

Compensation is achieved in utero because the blood is partly diverted to the left side via a patent foramen ovale and the aorta and systemic circulation by a right to left shunt via a patent ductus arteriosus, so a baby at birth can cope with it's abnormal circulation until the ductus arteriosus starts to close when he will die.

Early paliation by *Rashkind* ballon or atrial septostomy were performed.

Few patients will survive with right sided failure and failure to thrive.

Very rare cases may be asymptomatic until school age. The left side is underdeveloped due to the fact that most of the blood has been avoiding it, this makes corrective surgery more hazardous.

The three main dominant factors which determine the clinical evolution are:

1. The degree of obstruction of the common pulmonary vein.
2. The relative size of the P.D.A.
3. The relative size of the A.S.D.

and also the ability of the right ventricle to cope with a large volume overload.

Partial anomalous pulmonary venous drainage ispresent in 10-15% of cases of atrial septal defect of the ostium secondum type.

This anomaly is almost always present in presence of persistant sinous venosus.

In the later type the right superior pulmonary vein opens at the base of the superior vena caval orifice in the right atrium.

Aim of the Work

The aim of the work is to write an essay on the total and partial anomalous pulmonary venous drainage.

EMBRYOLOGY

EMBRYOLOGY

The pulmonary veins start to develop in the 5 mm human embryo (Skidmore[1], 1975).

The lung buds arise from the primitive foregut, then, they become surrounded by a plexus of veins called the pulmonary venous plexus. (Fig. 1.A.).

As differentiation proceeds this venous system drains into the heart via the splanchnic plexus of veins. Normally this pulmonary venous plexus becomes connected to a vessel called the common pulmonary vein. (Fig. 1.B).

This vessel is formed by the sinus venous as follows:

Haemocytoblasts develop immediately adjacent to, and in the substance of, the left atrial wall. At the 6 mm & 7 mm stages these cells accumulate in the inferior tracheo bronchial angle. This chain of cells becomes converted to a primitive vascular channel. by the 8 mm stage, a left sided diverticulum is formed. The latter is the precursor of the pulmonary vein. The vascular outgrowth grows posteriorly and medially, passing backwards behind the left horn of Curvier, to join the angioblast in the inferior tracheo-bronchial angle (Skidmore[2], 1975).

The common pulmonary vein is a transient structure that arises from the undivided sinus venous, just to the left of

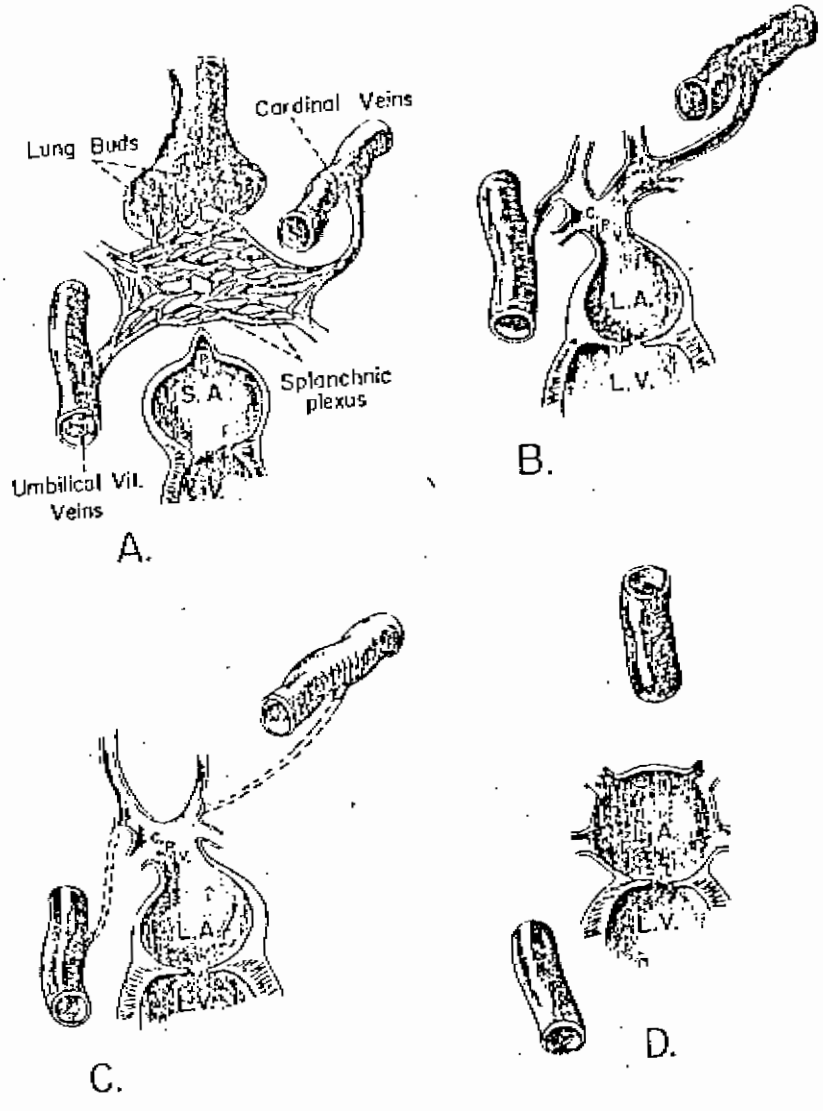

Fig. 1: Development of the pulmonary venous system. From Gibbon's surgery of the chest, 1983.

the area in which the septum primum will develop (Hammon and Bender[1], 1983).

Later in development, the common vein itself and part of its four pulmonary veins expands tremendously to become incorporated into the embryonic left atrium. The intrapulmonary part of the splanchnic venous plexus loses its connections with the systemic veins, and drains exclusively by the pulmonary veins (Van Mierop, 1986) (Fig. 1. B,C,D).

Abnormal development of the common pulmonary vein provides the embryological bases for most of the congenital anomalies or the pulmonary veins (Lucas et al., 1982).

The total form of the anomalous pulmonary venous connection presumably is due to either lack of the development of the embryonic common pulmonary vein, or the early involution and disappearance of this vein (Van Mierop, 1986).

If atresia of the common pulmonary vein occurs when systemic communications are still present, any one or several of the collateral channels can enlarge to form total anomalous pulmonary venous connection to the systemic venous system (Hammon and Bender[1], 1983) (Fig. 2B).

Partial anomalous pulmonary venous drainage is due to residual connection between part of the pulmonary venous system and the systemic circulation (Van Meirop, 1986).

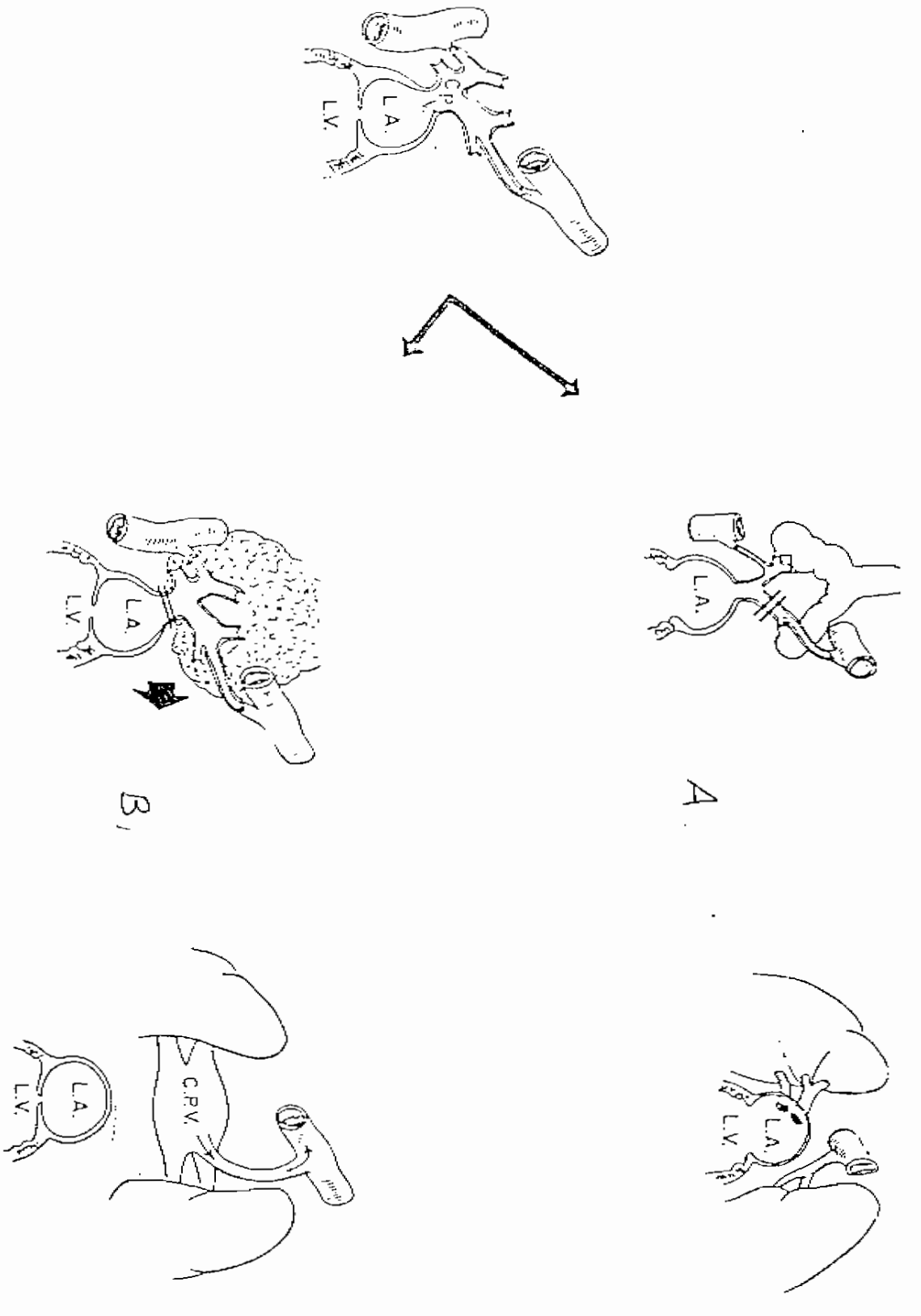

Fig. 2: Development of anomalous pulmonary venous drainage. From Gibbon's surgery of the chest, 1983.

Also, if only the right or left portion of the common pulmonary vein is atretic, partial anomalous pulmonary venous connection occurs (Fig. 2A). The individual anatomy of the anomalous pulmonary venous connection is directly related to the embryological connection between pulmonary and splanchnic connection that persists. (Hammon Bender[1], 1983).

Direct anomalous connection to the right atrium are probably best attributed to an abnormal septation of the two atria. As the septum may be formed abnormally to the left of its original site (Shaner, 1961).

Also as a result of close relation ship between the splanchnic and vitelline networks some of the vitelline veins may serve as a channel for the splanchnic drainage which when persist account for the direct connection between the pulmonary veins and the sinus venous. If such connection is localized in an area which will be absorbed by the right atrium, the definite anomaly is connection of the pulmonary vein to the right atrium. If the embryonic persisting drainage is localized into a part of the sinus venous that will not be absorbed into the right atrium, an anomalous connection to the coronary sinus may develop. This also the case if the connection is to the horn duct of Cuvier (Goor[1] and Lillehei, 1975).

TOTAL ANOMALOUS PULMONARY VENOUS CONNECTION

ANATOMY OF TOTAL ANOMALOUS PULMONARY VENOUS CONNECTION

Total anomalous pulmonary venous drainage results from failure of development of the common pulmonary vein and persistent pulmonary venous connection to the right side of the heart (Paster et al., 1977).

Anomalous pulmonary venous connection may appear as an isolated lesion, or may be associated with numerous other malformations. The most common are atrial septal defect of various types, persistent left superior vena cava, VSD, F4 (Vouhé, 1986).

Patent ductus arteriosus is persistent in 20-25% of cases (Bourroughs and Associates, 1960).

The individual right and left pulmonary veins nearly always converge to form a common pulmonary venous sinus that lies posterior to the left atrium and retro pericardial. Which in turn connects to the systemic venous system. But that sinus may be absent in some cases with cardiac or mixed connections (Small Horn, 1981).

Darling and associates in 1957 classified total anomalous pulmonary venous connection into four types based on the anatomical site of connection of the pulmonary veins to the systemic venous circulation (Darling and associates, 1957).