THE DIAGNOSIS AND TREATMENT OF COMPLETE ANOMALOUS PULMONARY VENOUS DRAINAGE

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Anomalous pulmonary venous drainage, expressed in the simplest of terms, means that pulmonary veins fail to enter the left auricle. If one or two pulmonary veins fail to enter the left side of the heart there may be no outward signs of cardiac disability and the patient leads a relatively normal life. Even if one half of the oxygenated blood enters directly or by venous channels into the right auricle there may be very few symptoms. However, more than half the venous drainage from the lungs entering the right side of the heart will constitute some disability, and total anomalous pulmonary venous drainage in which no pulmonary veins enter the left atrium is usually accompanied by a poor prognosis and death in infancy. In order to survive there must be an intracardiac communication, usually intra-atrial, in the form of a patent foramen ovale or intra-auricular septal defect. The intracardiac shunt is obviously from right to left, and survival beyond infancy will depend largely on the size of this communication between the two auricles.

An embryological study of this lesion is interesting since it concerns not only the rudimentary systemic venous return but also the venous return of the lungs. In early developmental stages, the lungs, which are considered to develop by division of the foregut, are drained by umbilico-vitelline veins which in turn enter the systemic pre-cardinal and post-cardinal veins. The post-cardinal veins unite to form the inferior vena cava and the pre-cardinal veins the right superior vena cava. While these systemic channels are becoming delineated into the normal situation, a channel from the sinal atrial region of the primitive heart tube outpouches to become the left atrium which grows towards the lungs and develops a communication which eventually forms the normal pulmonary venous drainage.

If one were to postulate that, at the stage at which the pre-cardinal veins are busy disappearing and the outgrowth of the left atrium is equally busy trying to reach the lungs, growth in these venous channels were to stop, then one is left with the left atrium trying in vain to reach the lungs and the lungs quite happy with their drainage into the systemic system of veins. If such a situation persisted it would result in total anomalous pulmonary venous drainage in which the pulmonary veins entered the embryological cardinal venous system. Partial interruption would result in one or more veins making an anastomosis with the outpouching of the left atrium. The venous channels connecting the lungs to the systemic venous system would become elongated and develop into recognizable channels. Due to the disappearance of certain parts of the systemic venous channels there persists a common vein which drains the pulmonary veins and enters the right auricle by way of established systemic venous channels (Fig. 1).

![Fig. 1](imageurl)

**Fig. 1.**—Schematic illustration of the pulmonary veins entering the persistent left superior vena cava rather than the left auricle.

Fortunately, Snellen and Albers (1952), closely followed by Gardner and Oram (1953), suspected that this condition could be diagnosed clinically if the presence of a common venous channel was recognized both by clinical investigation and by radiography. Brown (1950) and Whitaker (1954) have contributed in drawing attention to this
anomaly. Darling and Rothney (1956) have defined accurately the clinical pathology. If complicated cardiac anomalies can be excluded, an isolated form of anomalous pulmonary venous drainage, total in nature, which can be classified either supracardiac, cardiac, infracardiac and mixed, can be recognized.

Fortunately for the surgeon, almost 50% of cases of total anomalous pulmonary venous drainage are supracardiac and drain into a common venous channel, which, on a clinical basis, can be described as a persistent left superior vena cava or a precardial vein. Unfortunately, to be a true precardial vein, it should drain into the sinus venosus, but this is not necessarily true. Drainage into the sinus venosus or coronary sinus is at a cardiac level and drainage into the ductus venosus and the portal system is infracardiac. The mixed type, which is exceedingly rare, can be a combination of these forms.

As this anomaly is recognized by pathologists, the incidence may increase, since at necropsy, if the heart is removed from its pericardial sac and it lifts free, an anomalous pulmonary venous drainage is certainly there because of the loss of the tethering effect of the pulmonary veins entering the left atrium. At necropsy, both right ventricle and right auricle are enlarged, the extreme dilatation of the right ventricle masking, to some effect, hypertrophy.

The intra-atrial communication is usually a patent foramen ovale (75% of cases) but may be a large secondary intra-auricular septal defect. The ductus arteriosus is patent in approximately 25% of patients. The pulmonary artery is larger than normal, usually one and a half times the diameter of the aorta. The tricuspid valve ring is larger than the right mitral valve ring and the diameter of the common pulmonary vein, which drains into the right auricle, may be twice the diameter of the waist of the left auricle—of some consideration when one entertains the possibility of surgical approach. One-third of cases show intimal thickening and evidence of pulmonary hypertension. This anomaly probably represents, to the best of our knowledge, 2% of all cardiac malformations in infancy and childhood.

The haemodynamics of this condition are interesting. Pulmonary venous blood of high oxygen saturation enters the right auricle through the common venous channel and mixes with systemic blood of lower saturation, and a variable proportion of both streams passes from the right auricle through a defect in the auricular septum (either a patent foramen ovale or an auricular septal defect) to the left heart and aorta. Also a proportion passes through the tricuspid valve to the right ventricle and out through the pulmonary artery. For this reason the oxygen saturation of the blood in the heart and the systemic circulation is approximately the same.

When one considers the clinical features of this isolated form of total anomalous pulmonary venous drainage, there are certain signs or symptoms during the first two years of life. The first symptom is usually one of tachypnoea. Cyanosis is usually absent in the next few weeks or a month. The infant fails to gain weight satisfactorily and becomes intermittently cyanosed. Distinct cyanosis, gross tachypnoea or failure demands further investigation in the older infant. A typical case is dyspnoeic and dyspnoeic. Equivocal cyanosis produces a muddy discoloration. Clubbing is rare. The liver is usually enlarged and the praecordium bulging. One-third of cases show no murmurs. A systolic murmur along the sternal border may be present and occasionally a soft continuous venous hum, due to turbulence in the common venous channel, can be discovered.

Radiographically, increased lung vascularity, progressive cardiac enlargement, a left-sided aorta and a 'snowman' appearance, due to the persistence of a left superior vena cava, develop around 4 months of age (Fig. 2). This appearance is described in the English literature as a 'cottage loaf' appearance. The difference in nomenclature is possibly climatic in origin. Most Englishmen have never made a snowman and most Canadians think a cottage loaf is a summer vacation.

Fig. 2.—'Cottage loaf' appearance due to persistent superior vena cava.
Electrocardiographic tracings show a uniform pattern: right axis deviation, high p waves, right ventricular hypertrophy and q wave in the right praecordial lead.

Cardiac catheterization demonstrates oxygen saturation high in the right auricle and in the superior vena cava. An increase in right auricular pressure, double the average mean pressure, is usually present, and pressure is increased in the pulmonary artery to, or nearly to, systemic level. Occasionally, exploration by catheter may reveal anomalous pulmonary venous drainage entering the left innominate vein (Fig. 3).

**Fig. 3.—Oxygen saturation in total anomalous pulmonary venous drainage.**

Indication dilution techniques, after injection of Evans blue into a peripheral vein, may demonstrate a short appearance time with a double tip characteristic of right-to-left shunt.

With the physician cognizant of the condition and the diagnosis confirmed, exploration by thoracotomy is indicated. In our early cases, an anterior approach was used, but more recently we have favoured a lateral approach, somewhat posterior in the fourth interspace on the left side.

Our first three cases in 1952 were operated on at normothermic temperatures. One case was operated on at the extracorporeal circuit using a pump and biological oxygenator. Since that time we have used hypothermia, cooling down to $31^\circ$ C. and allowing a drift to $28^\circ$ C. Since these are infants they are cooled in a water bath; over the age of 3 years we use a cooling blanket.

It has been our experience that a common venous channel is immediately seen and dissection is carried down both extra- and intrapericardially to expose this common venous channel with the pulmonary veins entering it, and the waist of the left atrium is brought up (Fig. 4). We feel that it is unwise to anastomose the atrial appendage to the common venous channel because of the size of the waist of the left auricle, which in most cases is only one-half the diameter of the common venous channel. In order to obtain a large anastomosis, the waist of the left atrial appendage is divided longitudinally, which, when it becomes open after anastomosis, will be roughly one and one-half times its normal size. The occlusion of the left atrium in its entirety, including its appendage, and of the venous return from the lungs required, in our opinion, the added protection of hypothermia. Side-to-side anastomosis is carried out, and the surgeon is always tempted and has indeed, on many occasions, tied off the communication to the right auricle via the persistent left superior vena cava (Fig. 5).

**Fig. 4.—Appearance of this anomaly as seen at operation with a posterior-lateral incision in left chest.**

**Fig. 5.—Anastomosis between the common venous pool and the waist of the auricle.**
ARCHIVES OF DISEASE IN CHILDHOOD

Of our first case surviving operation, in 1954, we have no follow-up study as the patient died two months post-operatively with an ear infection. In this patient the superior vena cava had been ligated. On discharge from hospital the patient was in excellent condition. Our subsequent experience with cardiac irregularity and death after ligation of the superior vena cava led us to believe that the persistent superior vena cava should not be ligated, and in our seventh case in 1954 this was not done and the baby, operated upon at 3 months, is thriving and well two years post-operatively. This experience led us, in our eighth case, to leave the vena cava open and re-catheterization of this patient five months post-operatively showed no evidence of right-to-left shunt. The oxygen saturation in both the superior and inferior vena cava was normal (superior vena cava 63%; inferior vena cava 75%). A large volume left-to-right shunt was present between the auricles and indicated the presence of an auricular septal defect. A gain of 10 lb. in weight, lack of symptoms and reduction in right heart strain in the E.C.G. indicated that the new haemodynamic situation of an isolated septal defect was much better tolerated than the combination of total anomalous pulmonary venous drainage and auricular septal defect.

In our next three cases, we became foolishly brave and ligated the superior vena cava. These patients died and demonstrated a relatively small intra-atrial septal defect and a patent foramen ovale. We then felt that it would be unwise to tie the superior vena cava in any case because of the size of the left heart and its inability to withstand the additional load of all the pulmonary venous return to it.

In our last case in February of this year we were fortunate in that it was a mixed lesion in which the left pulmonary vein entered the persistent left superior vena cava and all other veins entered the coronary sinus. We anastomosed the auricular appendage to the persistent left superior vena cava and then ligated it. The waist of the left auricle was then anastomosed to the coronary sinus. This child is alive and well and has not been re-catheterized.

In conclusion I should like to comment upon this interesting condition of total anomalous pulmonary venous drainage and its possible surgical correction. With experience of 12 patients operated upon, four of whom survived operation, one dying two months post-operatively, and three alive and well, we feel justified in making certain recommendations.

First, the diagnosis, since total anomalous pulmonary venous drainage represents 2% of cardiac anomalies, should be made more frequently both on the clinical and radiological findings. Secondly, surgery should be undertaken in all cases and an anastomosis between the common venous pool and the waist of the left auricle should be attempted. This should be done under the protection of hypothermia or an extracorporeal circulation. Finally, the common venous drainage to the right auricle should not be ligated at the time of operation because of the additional load thrust on the left heart and the subsequent failure. Whether this common venous channel to the left auricle should be ligated subsequently or not is a decision I would leave to wiser surgeons.

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REFERENCES
Darling and Rothney (1956). Personal communication.
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