TOTAL ANOMALOUS DRAINAGE OF PULMONARY VEINS INTO THE LEFT GASTRIC VEIN

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Total anomalous pulmonary venous drainage is uncommon and probably accounts for about 2 per cent of all congenital cardiac malformations (Keith et al., 1958). It may be found as an isolated anomaly or in association with gross malformations of the heart. These are usually referred to as the isolated form and the complicated form respectively, and the former is the more common variety. Further subdivision into different types is made on the basis of the site of drainage of the anomalous pulmonary veins. This is of practical importance because it leads to wide differences in the clinical picture, each with its own particular diagnostic, prognostic, and therapeutic problems. The subject has been well reviewed by Gott et al. (1956), Darling et al. (1957), and Keith et al. (1958).

The case to be described, one of pulmonary venous drainage via the left gastric vein, represents an exceedingly rare type of the isolated form. Indeed, we have been unable to trace any previous report of this type of drainage except for a brief reference to it in the complicated form (Keith et al., 1958). A remarkable feature of the case is the occurrence of a fatal hæmatemesis from oesophageal varices at the age of three days. This in itself must be almost unique, though the nature of the malformation provides an apt explanation for it.

CASE HISTORY

P.W.H. was a full-term male infant born by a difficult forceps extraction after a lengthy second stage. During the night the baby had a prolonged apneic attack and became deeply cyanosed. He was revived by oxygen given by nasal catheter, but the cyanosis persisted and remained intense. The infant was seen by one of us (R.J.K.B.) when just 24 hours old. He was a well developed newborn with no detectable abnormality apart from the intense cyanosis and a grade I systolic murmur in the third left interspace. There was no cardiac enlargement and no sign of heart failure. Femoral pulses were easily palpable. Respiration were normal and the cry was lusty though productive of some increase in cyanosis. Cyanotic congenital heart disease was diagnosed. A chest radiograph showing a normal cardiac outline but some oligemia of the lungs (Fig. 1), and an electrocardiogram showing right ventricular preponderance, normal for the age (Fig. 2), afforded no help in making an accurate anatomical diagnosis.

On the second day he vomited a small quantity of bright red blood and was immediately given 10 mg. of synkavit (vitamin K) by intra-muscular injection. A few hours later there followed a severe hæmatemesis with considerable deterioration in the infant’s condition and he was transferred to the children’s ward of another hospital. That night there was no further bleeding and with oxygen his condition remained quite good. However, the following morning, after several small hæmatemeses and some melena, his pallor increased so that cyanosis was no longer detectable and his haemoglobin level was 40 per cent (5·9 g.). Before a blood transfusion could be started, there was a profuse vomit of bright red blood leading to severe circulatory collapse. This blood looked so much like arterialized blood that it was at first thought to be coming from the lungs, but the full significance of this observation was not appreciated at the time. In

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spite of repeated attempts to transfuse blood through small collapsed veins, it was impossible to keep pace with the blood loss, and a catheter was inserted into the umbilical vein and 60 ml. fresh whole blood injected by syringe. During this procedure the infant died.

*Necropsy.* The body was pale and weighed 3.24 Kg.

The heart showed some petechial haemorrhages on the pericardial surface and was moderately enlarged, with a dilated and hypertrophied right atrium and ventricle. A foramen ovale, though probe patent, appeared functionally closed. The left atrium was small, almost vestigial, with no vessels draining into it. The left ventricle was hypoplastic, all the valves were normal, and the interventricular septum was intact.

The pulmonary trunk and aorta were anatomically normal, and a patent ductus arteriosus, 7 mm. in length, and 6 mm. in diameter (after fixation), joined these two structures, which is not unusual for an infant of this age.

Both pleural cavities were dry; the lungs, which were conspicuously pale, were well aerated and showed a few pulmonary haemorrhages. No other abnormalities on the surface or in the parenchyma were noted. The pulmonary arteries were normal. The pulmonary veins, however, were very unusual, consisting of a
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comparatively slender vessel from the upper and middle lobes and another very much larger one from the lower lobes on each side. These vessels joined to form a single wide, thick-walled trunk 7 mm. in diameter, which traversed the diaphragm. As the heart was not anchored by pulmonary veins entering the left atrium, it was of unusual mobility and could be easily lifted forwards and upwards (Fig. 3).

A length of fine polythene tubing was lying in the umbilical vein. The areolar tissue surrounding the vessel, which was torn at one point, was distended with blood as far as the liver, and at one point the peritoneal covering was broken. The peritoneal cavity itself contained about 60 ml. of fresh unclotted blood.

The whole of the oesophagus and stomach and the first 4 cm. of duodenum were filled with clotted blood which formed a complete cast of these structures (Fig. 4). Large anastomosing varices, partially distended with blood, ran the length of the oesophagus immediately beneath the mucosa. One of these, at a point 8 mm. from the gastro-oesophageal junction, had ruptured and this could be demonstrated by injection techniques. These varices were fed by the common pulmonary trunk which terminated in the very large left gastric vein.

The lower end of the small and most of the large intestine were distended with melàena. No noteworthy features were found elsewhere in the body.
FIG. 4—The oesophagus, stomach, and duodenum after fixation. Very large submucous anastomosing veins run the length of the oesophagus. The bleeding point could just be distinguished near the cardiac sphincter. The inset shows the venous trunk from the lungs, anatomically the left gastric vein, entering the stomach. The complete blood cast of the oesophagus, stomach and duodenum is shown on the right.

DISCUSSION

Keith et al. (1958) have combined their own series with those reported by Darling et al. (1957) and Gott et al. (1956) to provide information on 103 cases of the isolated form of total anomalous pulmonary venous drainage proved by autopsy. They have divided them into four groups as suggested by Darling et al. The first consists of 55 cases in which the drainage was supracardiac, mostly into a left superior vena cava; the second of 30 cases in which the drainage was directly into the right atrium, though in half it was via a coronary sinus. In their fourth group, of only 3 cases, mixed multiple drainage was found. It is their third group of 12 cases where drainage was entirely infracardiac, that concerns this discussion. Of these twelve, nine had drainage into the portal vein, two into the ductus venosus, and one into the inferior vena cava: there was no case in which the veins drained into the left gastric vein.

Although the embryology of this anomaly is controversial, there is no doubt that in the very young embryo the splanchnic plexus of the foregut communicates with all the systemic and visceral
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veins in its vicinity. Since the lung develops as an outpouching from the ventral wall of the fore-gut, there is a stage in the normal development of the embryo at which the venous plexuses of the lung bud communicate with the pre- and post-cardinal veins, which are the precursors of the superior vena cava, innominate veins, azygos vein, and coronary sinus. There are also communications with umbilical and vitelline veins which in due course give rise to the portal vein, ductus venosus, and inferior vena cava. When the pulmonary stem of the sinus venosus fails to connect with the pulmonary plexus, some of these other venous channels may persist. Their multiplicity accounts for the several possible varieties of anomalous pulmonary venous drainage.

Diagnosis of some of the more common varieties is made possible because of certain features such as a characteristic radiographic appearance, but those in Group III of Darling et al. exhibit no typical diagnostic features. In this group cyanosis is intense from birth and life is seldom prolonged beyond the newborn period. In the present case the vomiting of arterialized blood in a cyanosed infant indicated that it had come from the lungs via a communication between pulmonary and portal venous systems, and this might have suggested the correct diagnosis, had we been alive to the possibility. However, it is perhaps not surprising that such an anomaly was never suspected until it was revealed as a result of the post-mortem examination. Indeed, in cases such as this nothing remarkable may be noticed at autopsy unless a methodical routine is adopted, although Keith et al. (1958) stress the important practical point that the heart can be lifted upward and forward on its pedicle because the left atrium is not anchored by the pulmonary veins. This point was well illustrated in our case.

The occurrences of such gross oesophageal varices at birth is arresting and, had the infant’s condition permitted investigation with a radiopaque swallow, these could undoubtedly have been demonstrated during life. Though some of the pulmonary venous blood would have drained into the portal vein, a great deal must have coursed along these varices to link up with the azygos and left hemi-azygous veins from the upper end of the oesophagus and to drain into the superior vena cava.

SUMMARY

Fatal hæmatemesis in a 3-day-old male infant with congenital cyanotic heart disease proved at autopsy to be due to rupture of oesophageal varices. The latter were produced as a result of total anomalous pulmonary venous drainage via a common pulmonary trunk into the left gastric vein. It is believed that this is the first time this type of drainage of the pulmonary veins has been described in association with a heart that was otherwise normal.

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Darling, R. C., Rothney, W. B., and Craig, J. M. (1957). Lab. Invest., 6, 44.