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Total Anomalous Pulmonary Venous Drainage Associated with Tetralogy of Fallot: Report of a Case

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SUMMARY. **The association of total anomalous pulmonary venous drainage and Tetralogy of Fallot is a rare occurrence; only six cases have been reported and only in one of these was the pulmonary drainage by the infracardiac route. A further such case is reported in a girl twin, with cleft palate and umbilical hernia and a normal spleen, who died at the age of three weeks and the clinical and post mortem features are described. The masking effect of the pulmonary outflow stenosis on pulmonary venous obstruction is discussed together with the relevance to clinical diagnosis and surgical intervention as "palliative" surgery may be dangerous.**

KEY WORDS: **Anomalous, pulmonary veins — Pulmonary veins, infracardiac drainage — Pulmonary venous obstruction, masked — Tetralogy of Fallot**

The association of total anomalous pulmonary venous drainage (TAPVD) and tetralogy of Fallot (TF) is a rare occurrence; only six cases have been reported and only in one of these was the pulmonary drainage by the infracardiac route. A further such case is reported here in a girl twin, with cleft palate and umbilical hernia and a normal spleen, who died at the age of 3 weeks.

The six reported cases of combined TF and TAPVD are listed in Table 1 [1, 2, 5–7]. In only two of these was the pulmonary venous anomaly demonstrated angiographically; in a further two it was diagnosed during surgical intervention for the TF and in one case it was found at autopsy.

Case Report

M.H. was a female caucasian child from the first pregnancy of a normal 22-year-old mother and 26-year-old father. She was the second of twins and was delivered by forceps at 35 weeks' gestational age.

Conception followed administration of clomiphene and human menopausal gonadotrophin (Pergonal) and pregnancy was normal. The first twin, a male, was normal at birth and has subsequently progressed well with no evidence of heart disease. M.H. weighed 1.92 kg at birth; she was noted to have a cleft lip and palate and a small, easily reducible hernia into the umbilical cord and she required 70% oxygen to remain pink. At 7 h there was mild respiratory distress and a chest X-ray showed a boot-shaped heart with normal lung fields. Tetralogy of Fallot was suspected and she was transferred to a paediatric cardiology unit on ventilation, but no active investigations were done because of her serious clinical condition. She developed hypoxaemia, hypocalcaemia, hypokalaemia and right middle lobe lung collapse. Her condition steadily deteriorated; it was not possible to wean her from the ventilator and she died on the 20th day.

Post-mortem Examination

There was a bilateral hare-lip and cleft palate with a free premaxilla and an easily reducible umbilical hernia. There were no other abnormalities apart from those of the cardiovascular system. The spleen was normal and the lungs were normally lobulated. The heart was small and, on raising the apex, no anchoring pulmonary veins were seen to enter the left atrium and the heart lifted completely forwards—the so-called positive

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Table 1. Reported cases of total anomalous pulmonary venous drainage with tetralogy of Fallot

No.	Authors	Year	Sex	Age and outcome	Type of pulmonary venous drainage	Additional anomalies	Clinical notes
1	Muster et al. [7]	1973	F	Died at 4 months	Mixed supracardiac, into coronary sinus and l. innominate vien	Ostium secundum atrial septal defect	Angiographic diagnosis at 8 weeks Palliative surgery at 10 weeks Late postoperative death at 16 weeks
2	Muster et al. [7]	1973	M	Died at 3 months	Infracardiac, into portal vien	Right-sided aortic arch	Potts anastomosis at 8 weeks Died 24 days postoperatively TAPVD found at autopsy
3	Delisle et al. [1]	1976	?	Died at 19 years	Supracardiac, into left superior vena cava into coronary sinus	?	?
4	Field et al. [2]	1978	F	Died at 19 months	Supracardiac, into innominate vein into superior vena cava	Atrial septal defect (unspecified); right-sided aortic arch; aberrant l. subclavian artery	Angiographic diagnosis at 11 months Corrective surgery at 19 months Died on 4th postoperative day
5	Kobayashi et al. [5]	1978	M	Alive 7 years when reported	Cardiac, into right atrium	Total conus defect; intact atrial septum	Diagnosis at operation Successful surgical correction
6	Miyamoto et al. [6]	1979	F	Alive 2 years when reported	Cardiac, into right atrium	Situs inversus totalis	Diagnosis at operation Successful surgical correction
7	Present case	1980	F	Died at 3 weeks	Infracardiac, into ? hepatic vein	Ostium secundum atrial septal defect; patent ductus arteriosus; bilateral hare- lip and cleft palate; umbilical hernia	Diagnosed at autopsy

Taussig manoeuvre. The pulmonary veins from each lung were found to drain posteriorly into a transverse common pulmonary vein (Fig. 1) from which a large vessel passed downwards through the diaphragm into the liver to join the hepatic veins (not shown in the Figure). The systemic veins were normal. The atria were normal except that no veins entered the left atrium; there was a widely patent foramen ovale (ostium secundum) 12 mm in diameter. The atrioventricular valves were normal. The ventricles were normal and of equal size and wall thickness; they communicated through a high 4-mm-diameter subcristal septal defect situated behind the medial leaf of the tricuspid valve, immediately anterior to the membranous septum and immediately below the aortic valve. The aorta was moderately dextraposed and its orifice straddled the ventricular septal defect. The aortic valve was normal, the arch was to the left and the branches were normal. The right ventricular outflow tract was narrowed to 2 mm diameter by a moderate degree of muscular infundibular stenosis. The pulmonary valve was bicuspid but was not significantly stenosed and the pulmonary artery trunk and main branches were not unduly narrowed. There was a long thin persistent ductus arteriosus.

Histological examination showed poor expansion and perfusion of the lungs; the other organs were normal. Skin fibroblast culture showed a normal female chromosome complement.

Discussion

Although combination of TF and TAPVD is rare, it presents important diagnostic and surgical difficulties which require special consideration.

In the two cases described by Muster et al. [7], palliative surgery for the TF to increase the pulmonary blood flow by establishment of aortic–pulmonary artery anastomoses resulted in worsening of the unsuspected pulmonary venous obstruction associated with the TAPVD. It may be difficult to diagnose anomalous pulmonary venous return in the presence of pulmonary outflow stenosis as with decreased pulmonary flow pulmonary venous obstruction may not be manifest and the pulmonary venous pressure not unduly elevated [4]. A surgical

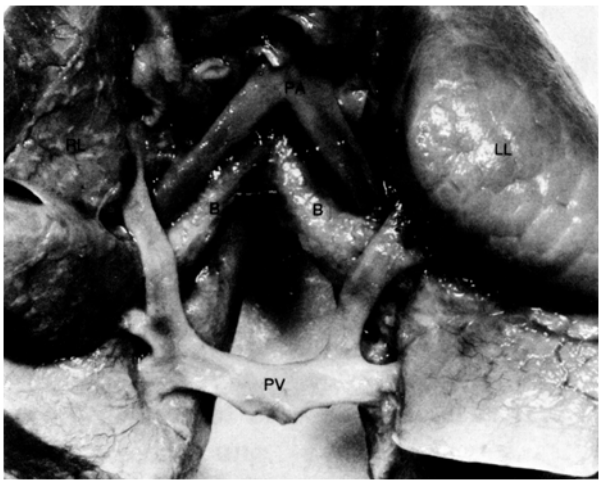

Fig. 1. The heart has been lifted forwards to show the anomalous pulmonary veins (PV). The descending vessel has been cut short above the diaphragm. *H*, heart; *PA*, pulmonary artery; *B*, bronchi; *RL*, right lung; *LL*, left lung.

aortic–pulmonary artery shunt in such a case may cause a precipitous rise in pulmonary venous pressure and acute pulmonary oedema. Muster et al. [7] stress that where both conditions are found in combination a palliative pulmonary artery shunt must be combined with correction of the anomalous pulmonary venous drainage. The policy of total correction of both anomalies had successful results in the two reported Japanese cases [5, 6].

Freedom et al. [3] suggested that where there is a possibility that critical pulmonary outflow obstruction and anomalous pulmonary venous drainage are part of the constellation of complex cardiac malformations, particularly those associated with the congenital asplenia syndrome, an otherwise silent pulmonary venous obstruction may be unmasked by injection of prostaglandin with resultant development of severe pulmonary oedema. They pointed out that neonates with pulmonary outflow tract obstruction would be most unlikely to develop pulmonary oedema as a result of increased ductal flow alone. This procedure would be contraindicated if pulmonary venous obstruction is known to exist.

According to Gersony [4], the possibility of an additional left-sided obstructive (venous) lesion indicates that patients who develop congestive heart failure after operations should have prompt repeat cardiac catheterization, especially if medical response is marginal.

The combination of these two lesions is uncommon, but it may not be as rare as it appears and should be considered in those cases of TF in which the pulmonary veins are not visualized on angiography. This is especially desirable when surgery is contemplated, because the condition is amenable to total correction, whereas palliative procedures directed against the decreased pulmonary flow alone would probably make matters worse.

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