

## CASE REPORT

# Left common carotid artery arising from the pulmonary artery in a patient with DiGeorge syndrome

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## Abstract

**A female infant, born at 33 weeks' gestation with tetralogy of Fallot, died of severe perinatal asphyxia 6 hours after birth. Necropsy disclosed two associated vascular anomalies: a right aortic arch with a left common carotid artery arising from the pulmonary artery (isolated left common carotid artery) and an aberrant left subclavian artery arising from the descending aorta. Agenesis of the thymus and parathyroid gland was also found, suggesting that the child also had DiGeorge syndrome. Origin of the left common carotid artery from the pulmonary artery is exceedingly rare. When planning surgical treatment it is important to be aware of the possibility of this anomaly occurring in association with congenital heart disease, particularly in the presence of tetralogy of Fallot, right sided aortic arch, or DiGeorge syndrome.**

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**Keywords:** aortic arch anomaly; congenital vascular anomaly; DiGeorge syndrome; tetralogy of Fallot

Patients with a right aortic arch can be classified into several types according to the nature of the origin of the branches of the arch. The most common type is mirror image branches, the second is aberrant left subclavian artery, and the third is isolation of the left subclavian artery. Isolation of the left innominate artery is very rare, because its occurrence requires partial regression of the aortic sac and the left fourth aortic arch in early fetal life, which is very uncommon. Only six cases have been reported.<sup>1-6</sup> Isolation of the left common carotid artery—that is, the left common carotid artery arising from the pulmonary artery, is even rarer, with only two cases having been reported.<sup>5,7</sup> One of them was associated with tetralogy of Fallot.<sup>7</sup> The other one was associated with only an atrial septal defect and persistent left superior vena cava.<sup>5</sup> We report a patient with DiGeorge syndrome with tetralogy of Fallot and an isolated left common carotid artery. To the best of our knowledge, this association has not been reported before. The clinical implications are discussed.

## Case report

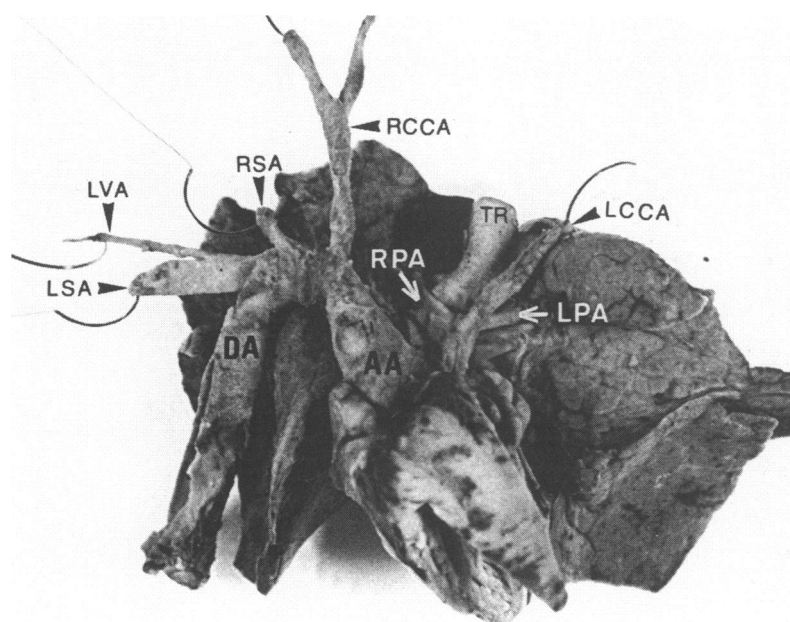
A 2116 g, premature (gestational age 33 weeks) female infant of a 33-year-old mother was delivered by emergency caesarean section because of progressive fetal bradycardia. After birth, the infant had low Apgar scores (1 and 1 at one minute and five minutes, respectively) and required resuscitation. A single umbilical artery was noted at birth. Physical examination revealed extreme pallor and no measurable blood pressure. Laboratory investigations showed severe anaemia (haemoglobin 2.3 g/dl, haematocrit 7.3%), thrombocytopenia (platelets: 69 000 per cm<sup>3</sup>), and severe metabolic and respiratory acidosis. The serum electrolyte concentrations after resuscitation and calcium gluconate supplementation were all within normal limits. Brain and abdominal ultrasonography revealed no evidence of haemorrhage. Echocardiography showed pulmonary atresia, overriding of the aorta, right sided aortic arch, and patent ductus arteriosus. The haemoglobin concentration was rapidly corrected to 10.3 g/dl after transfusion. Fetomaternal transfusion was suspected.

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**Figure 1** Heart and great vessels viewed from front. Arising from right aortic arch are the right common carotid artery (RCCA), right subclavian artery (RSA), and aberrant left subclavian artery (LSA). An isolated left common carotid artery (LCCA) arises from pulmonary artery (PA) via left patent ductus arteriosus. AA, ascending aorta; DA, descending aorta; LPA, left pulmonary artery; RPA, right pulmonary artery; LVA, left vertebral artery; TR, trachea.

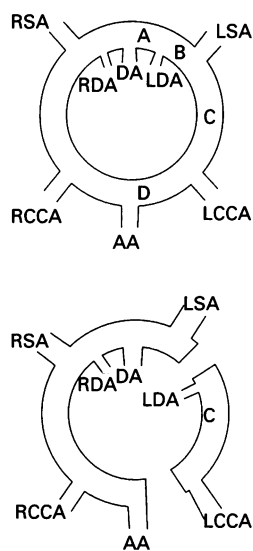


Figure 2 (upper) The hypothetical double aortic arch of Edwards. (lower) Concurrent interruption of the left posterior aortic arch and migration of the left subclavian artery (LSA) distal to left ductus arteriosus (LDA) resulted in aberrant left subclavian artery. Another interruption which occurred at zone D resulted in isolation of left common carotid artery (LCCA). AA, ascending aorta; DA, descending aorta; RSA, right subclavian artery; RDA, right ductus arteriosus; RCCA, right common carotid artery.

Prostaglandin E1 was given to maintain the patency of ductus arteriosus. The oxygen saturation improved gradually. However, metabolic acidosis and carbon dioxide retention persisted. The patient died when she was 6 hours old.

At necropsy 40 ml serosanguinous peritoneal effusion and moderate hepatomegaly were noted. No thymus or parathyroid gland tissue was found. A right aortic arch with right descending aorta was noted (fig 1). The aortic arch had only two branches: the right common carotid artery and the right subclavian artery. An aberrant left subclavian artery, arising from the descending aorta and distal to the right subclavian artery, was also found. This traversed behind the oesophagus to the left side of neck and gave branch to a large vertebral artery. The left common carotid artery arose from the junction of the distal main pulmonary artery and the proximal left pulmonary artery, via a left patent ductus arteriosus. The main pulmonary artery was atretic and cord-like. The right and left pulmonary arteries measured 4 mm and 3 mm in external diameter respectively. The heart showed overriding of the aorta with pulmonary atresia and a perimembranous type ventricular septal defect (0.5 cm in diameter).

### Discussion

The Edwards' hypothetical double aortic arch explains aortic arch abnormalities by selective regression of various parts of either arch (fig 2A).<sup>8</sup> In cases of isolation of the left innominate artery, the regression occurs at two sites: in the left posterior arch distal to the ductus arteriosus and in the left anterior arch proximal to the left common carotid artery.<sup>4,5</sup> In our case, the interruption should have occurred similarly in these two sites, but was also accompanied by concurrent migration of the left subclavian artery distal to the left ductus arteriosus (fig 2B). This resulted in an aberrant left subclavian artery as well as isolation of the left common carotid artery. Fong and Venables, who reported one case with the same aortic arch anomaly, considered that the regression occurred at zone B.<sup>5</sup> The case reported by Tozzi *et al* is similar to our case in also having tetralogy of Fallot.<sup>7</sup> However, the pulmonary blood flow in that case was not ductus dependent. In our case, the association of pulmonary atresia resulted in common carotid steal syndrome. The existence of a

patent ductus arteriosus, therefore, became vital for survival after birth. Nevertheless, early elective surgery is needed to prevent excessive steal syndrome. Preoperative recognition of such rare vascular anomalies in ductus-dependent cardiac lesions may be aided by the identification of weak left radial pulse on physical examination and by echocardiography and angiography.

The coexistence of agenesis of the thymus and parathyroid gland in our case is diagnostic of DiGeorge syndrome. The patient's normal serum calcium concentration was probably the result of the calcium supplement given during resuscitation. Absence or hypoplasia of the thymus is commonly associated with aortic arch anomalies or tetralogy of Fallot. Experimental studies have shown that normal aorticopulmonary septation and thymic morphogenesis depend on derivatives of the cephalic neural crest. Failure of cephalic neural crest derivatives to interact with pharyngeal pouch endoderm in sufficient quantity at the right time will result in partial or complete absence of the derivatives of the third and fourth pharyngeal pouches (the thymus and parathyroid gland) and conotruncal cardiac defects.<sup>9,10</sup> The data presented in this report support this idea.

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