Clinical vignette
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Aortic atresia, interrupted aortic arch type C perfused by bilateral arterial duct

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A non-syndromatic, 4.2 kg male newborn was admitted to the hospital at 24 h of age because of grunting and pale colour. Suspecting cardiogenic shock, prostaglandin infusion was initiated with prompt improvement of the clinical condition.

Echocardiography revealed a complex congenital heart disease. There was an unbalanced atrioventricular septal defect with a small left ventricle. There were bilateral superior caval veins, the left draining to the coronary sinus. The pulmonary veins drained to a collecting vessel which connected to the right atrium near its junction with the right superior caval vein. The aortic valve is atretic with a very small ascending aorta (AoA).

Catheterization showed a complex form of interrupted aortic arch with a peculiar relationship to the pulmonary arteries. The aortic arch was interrupted between the brachiocephalic trunk (BCT) and the left common carotid artery (LCCA). Blood supply to the very hypoplastic AoA and coronary arteries was provided by a right arterial duct (RAD), connecting the right pulmonary artery (RPA) and the BCT. This was nicely shown by retrograde injection of the right radial artery (Panel A). Through a left arterial duct (LAD), the descending aorta (AoD) was perfused as well as the distal part of the aortic arch (Panel B). Panels C and D show all great arteries and their relationship. Of particular interest is a sharp angle at the origin of the BCT and the LCCA, suggesting fibrous continuity of the arch rather than complete interruption.

A bilateral duct is a rare find. In this case of interrupted aortic arch and aortic atresia, blood supply to the coronary arteries depended on the patency of the RAD. Constriction of ductal tissue in the first hours after birth resulted in cardiac ischaemia and an early presentation with cardiogenic shock. Considering the complexity of the malformation, no further therapy was offered.

Panel A. Retrograde injection in the right radial artery. Blood supply to the AoA from the RPA through the RAD and BCT.
Panel B. Injection in the AoD. Blood supply to the AoD and distal aortic arch, with LCCA and left subclavian artery, was provided by an LAD.
Panel C. Injection in the LAD. LAD supplying the AoD and the distal arch. Right aortic arch supplying AoA. Fibrous continuity of arch (arrow) suggested by sharp angle at the origin of the BCT and the LCCA (LPA, left pulmonary artery).
Panel D. Schematic representation of great arteries. Interrupted line, presumed fibrous continuity of aortic arch.