New coronary congenital disease classification aids identification of secondary defects

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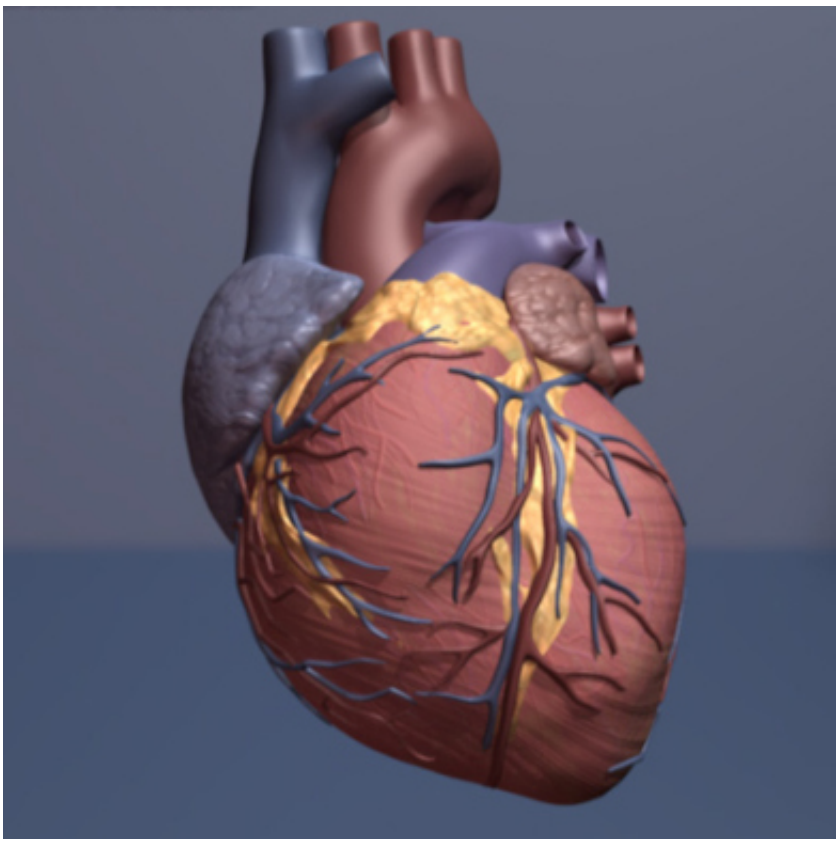

Human heart. Credit: copyright American Heart Association

A new classification of coronary congenital diseases is set to help surgeons identify secondary defects in the operating theatre. The scheme is outlined in a novel European Society of Cardiology (ESC) position

paper published today in *Cardiovascular Research*. Clinical cardiologists will also know what to look for on cardiovascular images.

"It can be difficult to spot further defects in the stressful environment of the operating theatre."

"Coronary congenital diseases affect less than 1% of newborns but are an important cause of myocardial infarction and sudden death particularly in children and competitive athletes," said Professor Cristina Basso, chairperson of the ESC Development, Anatomy and Pathology Working Group. "These conditions are often forgotten in the clinical setting since atherosclerotic coronary artery disease is far more common."

Coronary arteries are the blood vessels that nourish the heart muscle. Disruption of coronary development during embryogenesis results in coronary congenital defects that change blood flow. These defects can severely affect cardiovascular health.

The paper launches a new classification of coronary artery anomalies that explains common points of origin between different coronary defects. By identifying the origin of the primary defect, doctors can evaluate the probability of finding specific secondary defects with an origin mechanistically related to the main anomaly.

First author Dr José María Pérez-Pomares said: "We have established links between coronary congenital diseases sharing a common mechanism. When operating a diagnosed coronary anomaly, it can be difficult for surgeons to spot further defects in the stressful environment of the operating theatre and having an idea of the anomalies you might find can be extremely helpful. The new classification will also help clinicians using imaging to diagnose coronary artery anomalies and prevent future complications."

He added: "We have been able to produce this classification because we understand more about how coronary arteries develop in the embryo and how they relate to major diseases. The paper provides basic cardiovascular research scientists with new, updated information on the complex embryonic development of coronary arteries to throw light on the aetiology of coronary congenital anomalies."

The authors give their expert opinion on the embryonic origin of the coronary endothelium, which is a controversial topic in cardiovascular developmental biology. Dr Pérez-Pomares said: "For a long time scientists have wanted to identify a single and unique source for coronary endothelial cells but we now know that they come from different sources that merge together. This appears to have an impact on what happens to coronary vessels during embryonic development but perhaps also in the adult."

Although not explicitly stated in the paper, the authors think that this heterogeneity of cell sources contributing to the endothelium may be important in the development of adult coronary disease, as endothelial cells with different origins might respond differentially to pathological stimuli.

The diverse origin of coronary smooth muscle cells and fibroblasts are also described. Remarkably, adventitial fibroblasts that cover the arteries share a common embryonic origin with adult interstitial fibroblasts located between the myocardial fibres which are responsible for fibrotic scar formation after myocardial infarction. This finding also suggests that embryonic cues related to coronary development could be relevant for understanding cardiac fibrosis in the adult ischaemic heart.

The authors say that this "mosaic-like embryonic development of the coronary vascular system" is key to understanding the complex spectrum of coronary artery anomalies. Dr Pérez-Pomares said: "The more we dig

in, the more we have the impression that important events happening in the embryo, including the activation of regulatory gene networks, signalling molecular pathways and specific cellular mechanisms also have a clear function in adult responses to pathological stimuli."

Professor Basso concluded: "This is a translational paper written by basic scientists, including developmental biologists, anatomists and pathologists. It contains practical information to help clinicians diagnose coronary artery anomalies and prevent devastating complications including sudden death."

More information: Pérez-Pomares JM, de la Pompa JL, Franco D, Henderson D, Ho SY, Houyel L, Kelly RG,Sedmera D, Sheppard M, Sperling S, Thiene G, van den Hoff M, Basso C. Congenital coronary artery anomalies: a bridge from embryology to anatomy and pathophysiology—a position statement of the development, anatomy, and pathology ESC Working Group. *Cardiovascular Research*. 2016: DOI: 10.1093/cvr/cvv251

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