Letter to the Editor

Hypoplastic left coronary artery with large collateral vessels from an ectatic right coronary artery
Multimodality imaging-based diagnostic work-up

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Hypoplastic coronary artery (HCA) is a rare form of coronary artery anomaly involving only 0.02% of the general population and 2.2% of all congenital coronary artery anomalies [1,2]. HCA is diagnosed by the presence of congenital underdevelopment or significant narrowing of one or more major coronary arteries, and it has only been rarely diagnosed in live individuals because even though most patients are asymptomatic, a high proportion of them can experience sudden death or myocardial infarction [2,3]. Hypoplasia of the entire left coronary artery is one of the rarest variations of HCA [2,3].

A 16-year-old high-school football player experienced multiple episodes of syncope during exercise. Resting electrocardiogram showed only incomplete right bundle branch block (Fig. 1; Panel A) and echocardiogram raised the suspicion for a hypoplastic left main coronary artery and an ectatic right coronary artery (Panel B). Cardiac computed tomography angiography was performed to further assess the coronary anatomy which revealed a diffusely hypoplastic left coronary system and an ectatic right coronary artery (Panel C) giving rise to large collateral vessels to the hypoplastic left anterior descending artery and obtuse marginal artery (Panels C–E). Functional assessment of this coronary anomaly with exercise stress N13 positron emission tomography revealed no evidence of stress-inducible ischemia (Panel F). During the stress test, the patient did not experience syncope or chest pain. He was finally discharged home without restrictions of athletic activities.

While the majority of HCA cases are sporadic, HCA can be associated with other congenital cardiac anomalies and the limited coronary blood flow may be further exacerbated by coexisting conditions, such as coronary artery spasm, thrombosis, myocardial bridge or increasing demand during exercise [2–4]. The correlation among ischemia, stress testing and presence or absence of collateral circulation has not been clarified yet.

Unexplained recurrent episodes of syncope in otherwise healthy and asymptomatic young individuals should always raise the suspicion of ischemia related to coronary anomaly and should be thoroughly investigated with functional and anatomical imaging. With regard to treatment, bypass surgery is not effective in treating HCA due to the diffuse and extensive coronary arterial narrowing [2]. Implantable cardioverter-defibrillator may be beneficial by preventing life-threatening arrhythmias, whereas heart transplantation may represent a reasonable alternative in cases of end-stage ischemic cardiomyopathy [2,3].

Hypoplastic left coronary artery is a rare and potentially life threatening coronary anomaly, usually detected by echocardiography. Coronary CTA and stress perfusion studies should always follow for further anatomic and functional assessment.

References


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Fig. 1. A. Resting electrocardiogram showed incomplete right bundle branch block. B: Echocardiogram raised the suspicion for a hypoplastic left main coronary artery (arrow) and an ectatic right coronary artery (arrow). C: Axial, oblique coronal and oblique sagittal reformatted and D–E volume rendered color coded images of the cardiac computed tomography angiography revealed a diffusely hypoplastic left coronary system (arrows for LAD) and an ectatic right coronary artery (arrow for RCA and PDA) giving rise to large collateral vessels to the hypoplastic left anterior descending artery and obtuse marginal artery (small black arrowhead). F. Functional assessment of this coronary anomaly with exercise stress N13 positron emission tomography revealed no evidence of stress-inducible ischemia.