

Recurrent syncope in the child – don't forget the coronary arteries. Atresia of the left coronary artery in a 9 years-old boy.

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Background. Syncope is one of the most frequent reasons for referral for evaluation in pediatric cardiology. Whereas vaso-vagal syncope are usually harmless, potentially life-threatening aetiologies, including arrhythmias, structural heart disease, anomalies of the coronary arteries (CA) and myocardial ischemic disease need to be ruled out.

Methods. Case report and review of the literature

Results. This boy presented with recurrent syncope following physical activity since the age of 5. Repetitive cardiac evaluations including clinical examination, electrocardiogram (ECG), 24h-ECG, stress ECG, event recorder, echocardiogram and stress echocardiogram were unremarkable. Neurological and gastroenterological evaluations were normal. At the age of 9 years the boy experienced sudden cardiac arrest during a soccer game. Cardiopulmonary resuscitation was immediately started and was successful after threefold defibrillation of ventricular fibrillation. Again all non-invasive examinations were unremarkable, except a slightly diminished ventricular function, which quickly normalized under milrinone. Invasive electrophysiological study was normal. Coronary angiography showed a prominent right CA and retrograde perfusion of the left sided CA system, with small left anterior descending (LAD) CA and ramus circumflexus (Figure). The ostium and the first segment of the left main CA were missing and atresia of the left coronary artery was diagnosed. Myocardial perfusion studies consisting of SPECT and perfusion magnetic resonance showed no ischemia under adenosine stress and absence of scarring. Prophylactic medication with metoprolol was started and surgical revascularisation with a left internal mammary artery to proximal LAD bypass grafting performed without complications. The patient was discharged 10 days postoperatively in good clinical conditions with acetylsalicylic acid.

Conclusions. CA anomalies are rare, but a frequent cause of sudden cardiac death in adolescents and young adults. In patients with exercise related syncope, CA anomalies need to be ruled out. Atresia of one CA is exceedingly rare with only 28 reported cases. This is usually an isolated lesion, but can be related to other cardiac defects in up to 30% of the cases. Surgical revascularization using internal mammary artery is the therapy of choice providing good results and growth of the left CA system.



Figure: selective right CA angiography