A previously healthy 13-year-old boy presented with cardiac arrest during a soccer match. After experiencing a few minutes of chest discomfort, he suddenly lost consciousness and collapsed. Resuscitation was initiated and continued until paramedics performed electric defibrillation of the ventricular fibrillation (VF; Panel A). Spontaneous respiration resumed. ECG showed sinus rhythm with ST-segment depressions and prolonged QTc of 489 ms (Panel B). The patient was intubated and hypothermia treatment initiated.

The patient had no history of typical angina despite an active lifestyle. Two exercise-provoked incidents of dyspnoea and greyout had been attributed to asthma, and no further evaluation had been performed.

Upon admission, the ST depressions had regressed. QTc was 491 ms (Panel C). Echocardiography showed normal cardiac function but difficulties visualizing the coronaries.

CT-angiography and CAG revealed right-sided origin of the left main coronary artery (LMCA) from the right coronary sinus (Panels D and E, dotted arrow) with an inter-arterial course between the pulmonary artery and the aorta (Panels D and E, full arrow) causing ischaemia. A cardiac stress test to confirm exercise-induced ischaemia was not performed, as the VF during the soccer match was considered diagnostic.

Beside amnesia for the incident and temporary impaired short-term memory, the patient was neurologically intact.

After an uneventful surgical re-implantation of the LMCA, the patient received a prophylactic ICD scheduled for removal after 5 years. Today, 16 months later, the patient has resumed sports activities and there have been no cardiac symptoms or incidences of VF. The asymptomatic older brother was screened with echocardiography and CT, finding normal cardiac function and LMCA anatomy.