CASE REPORT

Hypoplastic coronary artery disease: report of one case

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Hypoplastic coronary artery disease (HCAD) is a rare congenital abnormality with a high rate of sudden death and poor outcome. HCAD was revealed by myocardial infarction in a teenager with objective evidence of silent ischaemia on myocardial scintigraphy. After four years of follow up, he suddenly collapsed during exercise and subsequently died. Although HCAD is very uncommon, it should be actively excluded in children and young adults who experience sudden cardiac death. Aggressive treatment such as implantable cardioverter-defibrillator or heart transplantation may be indicated for this rare coronary abnormality.

Hypoplastic coronary artery disease (HCAD) is a rare congenital abnormality with a high rate of sudden death and poor outcome.1–7 We previously published the initial presentation of a child with HCAD revealed by acute myocardial infarction.1 We report here his subsequent evolution and outcome in, to our knowledge, the first documented and longest follow up of this syndrome.

CASE REPORT

An 11 year old boy with a history of asthma was admitted to the paediatric intensive care unit for a non-Q wave anterior myocardial infarction. During sustained exercise (a handball match), he suddenly collapsed and recovered after a brief cardiopulmonary resuscitation. He presented with chest pain, anterior ST segment depression on the ECG, and increased creatine kinase concentration (peak 2580 U/l; MB fraction 240 U/l). During the acute phase, he received anticoagulation medications, aspirin, and calcium inhibitors. Cardiac catheterisation was performed eight days after an uncomplicated intensive care unit course. Left ventriculography showed apical hypokinesia with a mildly depressed ejection fraction (55%). Selective coronaryography showed hypoplasia of the distal left anterior descending coronary artery (LAD) and right coronary artery (RCA) with no supply to the inferior aspect of the interventricular septum by a posterior branch of the RCA or left circumflex coronary artery. Additionally, no diagonal branches arising from the LAD but well developed collateral branches issuing from septal perforating arteries were documented. Intracoronary infusion of glyceryl trinitrate did not change the diameter or the morphology of the coronary arteries. Total cholesterol and high density lipoprotein (HDL) cholesterol, sedimentation rate, serological studies for connective tissue diseases, antithrombin III, proteins C and S, endogenous tissue plasminogen activator, and plasminogen activator inhibitor were normal. The final diagnosis was HCAD. The patient was discharged and prescribed diltiazem after a normal maximal exercise test 16 days after his admission. He was told to avoid any kind of sustained exercise.

One year after the initial presentation, the patient remained asymptomatic. Physical examination and ECG were normal. The echocardiogram showed an apical hypokinesia with normal ejection fraction and 24 hour Holter monitoring did not show silent ischaemia or arrhythmia. A stress test with thallium-201 imaging was performed and showed a perfusion defect involving the anteroseptal and apical segments with incomplete apical redistribution at rest (fig 1). A new cardiac catheterisation showed an apical dyskinesia on the ventriculogram and hypoplasia of the RCA and distal LAD with no diagonal branches (fig 2). A myocardial bridge, not recognised on the previous cardiac catheterisation, was documented at the distal LAD site. Close follow up with maintenance of calcium inhibitor and avoidance of any kind of physical exercise was the agreed course of action. A new stress test with thallium 201 imaging performed one year later showed persistence of the defects and ischaemia previously documented. The patient remained asymptomatic and physical examination, ECG, and 24 hour Holter monitoring were normal during the follow up. The left ventricular function on the echocardiogram did not worsen during follow up.

One year later, the diltiazem was interrupted because the patient had symptomatic orthostatic hypotension. Despite...
anomalies. In living patients the diagnosis is made by
incidence of 0.022% of hypoplastic proximal coronary artery
adults) is unknown.

DISCUSSION
HCAD was first reported in 1970 and refers to congenital
underdevelopment of one or more major branches of the
coronary arteries with greatly decreased luminal diameter or
The majority of the reported cases are isolated and
incidence of hypoplastic coronary arteries. Congenital anomalies of the coronary arteries. Am J Cardiol
In selected patients with proximal HCAD and a well
developed distal network, surgical treatment with coronary
to play a handball match a few days later. He suddenly
collapsed during the match, after he finished a run. Cardiac
monitoring during the unsuccessful cardiopulmonary
resuscitation showed ventricular fibrillation. He died on
June 2001 at age 15, four years after his initial presentation.

Figure 2  (A) Left anterior oblique left main coronary artery angiogram
showing a hypoplastic distal left anterior descending coronary artery
(arrows). Septal branches are well developed and supply the basal part
of the interventricular septum. In contrast, no diagonal branches are seen
on the anterolateral free wall of the ventricle and no posterior branch
issuing from the left circumflex artery. (B) Lateral view angiogram
showing hypoplastic right coronary artery without posterior descending
to the basal part of interventricular septum (arrows).

the recommendation to avoid exercise, the patient decided
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All authors, despite several reports, do not admit a
in the study of Zugibe et al. An additional deleterious role of
the myocardial bridge is possible in our patient.

In conclusion, HCAD is an uncommon abnormality rarely
diagnosed in living patients. It should be actively excluded in
children and young adults who experience sudden cardiac
death. The prognosis is poor and treatment options are
limited. In patients with evidence of myocardial ischaemia,
the implantation of a cardioverter-defibrillator should be
strongly considered.

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