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.

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the recommendation to avoid exercise, the patient decided 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.

**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 length. The majority of the reported cases are isolated and often diagnosed at necropsy. Ogden et al. found an incidence of 0.022% of hypoplastic proximal coronary artery in a study of 224 necropsy cases of congenital artery anomalies. In living patients the diagnosis is made by coronary angiography. In a recent analysis of 58,023 coronary angiographies performed in adult patients, coronary artery anomalies were diagnosed in 257 patients (0.44%) with congenital atresia in only 16 patients (atretic RCA in two patients; atretic left circumflex in 14). However, the true incidence of HCAD in the general population (children and adults) is unknown.

HCAD is often revealed by sudden death, especially in young adults and athletes. Myocardial infarction is an unusual presentation pattern, although in our patient HCAD was found because of a probably life threatening arrhythmia, since the patient collapsed at initial presentation.

All authors, despite several reports, do not admit a correlation between HCAD and ischaemia. In our patient, detailed follow up is available with nuclear cardiological documented evidence of persistent myocardial ischaemia despite no clinical symptoms (fig 1). The apical and anteroseptal defects during stress with incomplete redistribution at rest on thallium scintigraphy result from silent ischaemia or the presence of hibernating myocardium. In healthy people, collateral branches issuing from the RCA or the distal LAD supply this myocardial area. In our patient, as the RCA and distal LAD were underdeveloped, collateral vessel flow to the left ventricle was insufficient, leading to inadequate stress perfusion.

Circumstances surrounding the death of our patient are comparable with previously reported deaths caused by HCAD (ventricular arrhythmia during effort), consistent with a sudden and total occlusion of the LAD. The mechanisms involved may be a coronary artery spasm reflecting abnormal vasodilator mechanisms and endothelial dysfunction or intramural thrombus as reported for four of seven patients in the study of Zugibe et al. An additional deleterious role of the myocardial bridge is possible in our patient.

In selected patients with proximal HCAD and a well developed distal network, surgical treatment with coronary artery bypass can be proposed. Heart transplantation may also be considered for patients who develop ischaemic cardiomyopathy with end stage heart failure. In our patient, coronary artery bypass was not indicated because of the diffuse and distal coronary hypoplasia. Since he never experienced clinical heart failure and his left ventricular function was normal on the follow up echocardiograms, heart transplantation was not considered. However, although the ECG and 24 hour Holter monitoring were normal during follow up, an implantable cardioverter-defibrillator could have been proposed, since there was persistent myocardial ischaemia on the scintigraphy and sudden death is a frequent issue in this coronary abnormality.

Unfortunately in our centre, implantable cardioverter-defibrillators were not routinely used in the paediatric population at the time of the patient’s presentation and follow up.

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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**Authors’ affiliations**

N Amabile, A Fraisse, J Quilici, Département de cardiologie, Hôpital de la Timone, Marseille, France

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Correspondence to: Dr Alain Fraisse, Département de cardiologie, Hôpital de la Timone, 264 rue St Pierre, 13385 Marseille Cedex 05, France; alain.fraisse@ap-hm.fr

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