present one. In retrospect, the singular appearance of an acute coronary syndrome in our patient most probably resulted from a transitory thrombogenic transformation of the atherosclerotic plaque in the middle segment of the RCA. According to the literature, all patients experienced their first anginal attacks during advanced adulthood (older than 40 years) despite the assumed congenital origin of the malformation. Therefore, the clinical and prognostic relevance of the suspected ischaemic syndrome in multiple coronary artery-left ventricular fistulae appears to be uncertain.⁵

Haemodynamic quantification of shunt flow might provide further insight into the pathophysiology of this anomaly. In the present case, total flow through the left and right epicardial coronary artery was calculated as 1660 ml/min by using a Doppler guidewire technique. In comparison, total myocardial blood flow in healthy adults has been determined as 150–300 ml/min at rest.⁸ Thus, total left ventricular output might be estimated as the sum of shunt flow (1400 ml/min) plus systemic flow (4800 ml/min). Accordingly, left-to-left shunt flow would amount to approximately 23% of total left ventricular output. It has been speculated that the considerable shunt volume draining into the left ventricle might result in diastolic overload imitating the haemodynamic situation of aortic regurgitation.⁹ In contrast, the estimated shunt fraction of 23% in our patient cannot be expected to have any negative impact on left

ventricular function for it would be classified as being mild even in subjects with aortic regurgitation. Accordingly, no left ventricular enlargement or reduction in ejection fraction were found. The clinical course of the patient was completely uneventful before and after the first anginal episode that led to coronary angiography and diagnosis. In view of these haemodynamic and clinical findings, the question remains whether this entity has any pathophysiological significance or has to be considered as a normal variation due to a prominent Thebesian system.^{5 10}

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SHORT CASES IN CARDIOLOGY

Intractable vasospastic angina

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A 48 year old woman with drug refractory vasospastic angina had been in our hospital for nine months. Electrocardiograms recorded during angina attacks showed marked elevation of ST segments in the precordial, inferior, and/or lateral leads, often with ventricular arrhythmia or atrioventricular block (fig 1). Coronary angiography revealed no atherosclerotic stenosis, and intracoronary injection of acetylcholine induced total occlusion of the left anterior descending and right coronary arteries despite continuation of high dose calcium antagonists (fig 2).

Calcium antagonists (nifedipine, nisoldipine, amlodipine, benidipine, diltiazem, and verapamil), nitrates (isosorbide dinitrate, isosorbide mononitrate, and nitroglycerin),

nicorandil or various combinations of drugs could not prevent her attacks. Blood concentrations of drugs such as diltiazem and nicorandil were much higher than normal therapeutic values. She did not have signs of a generalised vasospastic disorder such as Raynaud's phenomenon nor a family history of coronary artery disease or sudden death. The patient had stopped smoking after admission. Her attacks occurred not only between midnight and early morning but also in the daytime. The trigger of attacks such as emotional distress, exercise, cold, alcohol, or coffee could not be specified. Her regular menstrual cycle with normal variation of blood oestrogen concentrations was not related to the incidence of attacks.

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Figure 1 Electrocardiogram during one attack showing marked ST segment elevation in the precordial and inferior leads (1 mV = 10 mm).

Figure 2 Serial coronary angiograms performed two hours after administration of diltiazem (300 mg), benidipine (8 mg), and nicorandil (10 mg). (A) Total occlusion of proximal left anterior descending artery induced by acetylcholine. (B) No significant fixed stenosis of left coronary artery is shown after intracoronary injection of isosorbide dinitrate (ISDN). (C) Total occlusion of proximal right coronary artery induced by acetylcholine. (D) No significant fixed stenosis of right coronary artery is shown after intracoronary injection of ISDN.

In general, calcium antagonists are extremely effective in preventing coronary artery spasm, and simultaneous administration of two or three antagonists is rarely required.¹ In this patient, however, even an extremely high dose of calcium antagonists (diltiazem 1400 mg/day) could not prevent attacks, and further increase in the dosage could not be tolerated because of marked hypotension.

Medications other than conventional antianginal drugs were tried over the nine months in hospital: magnesium,² α_1 adrenergic blockers (prazosin and doxazosin),³ β_1 adrenergic agonist (denopamine),⁴ serotonin 2 receptor antagonist (sarpogrelate),⁵ prostaglandin I₂, thromboxane A₂ synthetase inhibitor (ozagrel), aspirin, heparin, angiotensin converting enzyme inhibitors (captopril and alacepril), prednisolone, biotin, and tranquillisers. These drugs also failed to control her attacks. For prevention of nitrate tolerance she was given N-ethylcystein after discontinuation of nitrate and nicorandil.

The patient had experienced more than 800 attacks over nine months. Coronary angiography was performed twice, nine months apart, and revealed no fixed stenosis. At least six patterns of ST elevation during attacks suggested that coronary spasm in various sites of arteries might be involved in her anginal attacks; therefore, we did not consider coronary bypass surgery or coronary stenting to be indicated.

Antianginal drugs were stopped because of lack of efficacy, the patient suffered frequent severe chest pain lasting more than 30 minutes; however, serum concentrations of creatine kinase did not exceed the normal upper limit. At present she is taking diltiazem (1200 mg/day), benidipine (16 mg/day), amlodipine (5 mg/day), isosorbide mononitrate (60 mg/day), nicorandil (20 mg/day), and N-ethylcystein (800 mg/day), and still suffers several attacks per day. We invite new strategies against these attacks from around the world.

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