Myocardial infarction caused by an
anerysm of the left main coronary
artery without evidence of Kawasaki disease

Sirs,—We read the case report from Pfafferott and coworkers’ about atypical Kawasaki syn-
drome with great interest. The authors report the case of a 20 year old woman with acute
myocardial infarction and a huge aneurysm of the left main coronary artery that was
occluded by a large thrombus. Because other vascular and systemic diseases were ex-
cluded, atypical Kawasaki syndrome was diagnosed. Diagnostic criteria of Kawasaki
disease include fever for five or more days unresponsive to antibiotics, bilateral conges-
tion of the conjunctiva, peripheral limb changes including indurative oedema, and
erythema followed by membranous desqua-
mation of the fingertips, erythema of oral and pharyngeal mucosa and a strawberry tongue, a
polymorphous exanthema of the trunk, and
cervical lymphadenopathy. Furthermore, the
diagnosis is accepted when coronary aneu-
rysms are present in addition to four of these
major symptoms. 2 Thrombosis of coronary aneu-
rysms, however, often leads to myocar-
dial infarction in this disease and therefore
has major influence on long term prognosis. 2

In November 1996, a 19 year old male
patient was transferred from a primary care
care hospital to our institution. The day before he
had had an acute anterior myocardial infarc-
tion. He had been treated with intravenous recombinant tissue plasminogen activator (rt-
PA), and the chest pain and ST segment elevation vanished. Some hours later a new ST segment elevation in the anterior leads was
diagnosed accompanied by severe chest pain.
Therefore, further thrombolysis with rt-PA was
given and, owing to haemodynamic insta-
Bility, the patient was transferred to our insti-
tution by helicopter. Coronary angiography
revealed an increase of the known left main
anerysm whilst the other arteries were un-
changed (fig 1). The patient was haemody-
namically stable but nuclear magnetic reso-
nance imaging of the brain revealed severe
hypoxic encephalopathy.

This case of a young patient with huge left
main coronary aneurysm combined with APC
resistance raises the question whether Pfaff-
erott and coworkers screened their patient for haemostatic abnormalities. We do not believe
that our patient suffered from atypical Kawa-
saki syndrome as his medical history was
negative for typical signs and symptoms of this
disease. Although our case is very similar to
that of Pfafferott et al we think that Kawasaki
disease should not be diagnosed when there is
coronary aneurysm without other diagnostic
criteria of this disease.

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Inappropriate management of
polycythaemia in adults with cyanotic
congenital heart disease

Sirs,—The editorial by Thorne 1 highlights
misconceptions in the management of poly-
cythaemia in adults with congenital heart
disease. Inappropriate venesection is not only
countered in patients with inoperable cyanotic heart disease. Venesection with home
nonresponsive to nonphosphate, with radioactive phosphorus are also used in patients with operable
congenital heart disease. Two recent cases are
illustrative.

Case 1 is a 38 year old man with double
outlet right ventricle, a straddling tricuspid valve, and pulmonary atresia who had a clas-
sis Blalock-Taussig shunt at age 11 years.
After many years of venesection for his “inoperable congenital heart disease” he suf-
f the question of whether he had a
eront operation with an increase in his exercise tolerance. He is currently awaiting transcatheter occlusion of the atrial
fenestration.

Case 2 is a patient with classic tricuspid atresia, a restrictive ventricular septal defect, and
pulmonary stenosis. After repeated venesections, he was referred to our unit at the age of 45 years, after he had become
unwell during a venesection. He had a fenes-
trated Fontan operation, which was followed
by transcatheter occlusion of his fenestration.
Four years later he had oxygen saturations of
95% and a greatly improved exercise toler-
ance. (On a lighter note, his aged but
m sprightly mother “complains” that she is no
longer able to keep up with him when walking—a reversal of the situation before surgery.)

These cases are typical of patients who have
been “lost to follow-up” from congenital heart
disease clinics and are seen only in general
dical, adult cardiology, or haema-
tology departments. Adults with congenital
heart disease should be managed in conjunc-
tion with congenital heart disease units. In
particular, patients labelled as “inoperable” or even “complex” congenital heart disease
need to be reviewed in light of modern approaches to treatment.

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