SCIENTIFIC LETTER

Prevalence of haemoptysis in adults with pulmonary atresia and ventricular septal defect, and the role of mammary artery collateral vessels

K Greaves, P Bye, G Parker, D S Celermajer

Heart 2003;89:937-938

Recurrent or massive haemoptysis is a potentially lethal complication occurring in patients with congenital heart disease (CHD). In particular, it is most frequently described in patients with pulmonary vascular obstructive disease such as tetralogy of Fallot and pulmonary atresia with ventricular septal defect (PA-VSD). Although widely believed to be a rare complication of PA-VSD, the true prevalence of this complication and anatomic vessels involved are unknown.¹

Most of the patients who survive to adolescence or adulthood have had palliative or corrective surgery, and have developed an alternative blood supply to the lungs through systemic-to-pulmonary collateral vessels. These collateral vessels are prone to rupture and may result in serious and even fatal haemoptysis.¹ Selective embolisation of these systemic vessels is a widely accepted and successful technique used in the control of recurrent or massive haemoptysis, but identification of the culprit collateral(s) can be difficult.

We sought to determine the prevalence of haemoptysis in a cohort of adult patients with PA-VSD and report the collateral vessels involved.

METHODS

The records of all patients with PA-VSD regularly attending the adult CHD clinic at our tertiary referral centre (established in 1991) were examined to determine the number of patients who were recorded to have had a past history of haemoptysis. Twenty six patients, of whom eight were male, were identified with PA-VSD, mean age (at the time of writing) 29 years (range 18-54 years).

RESULTS

Three (12%) out of 26 patients had haemoptyses requiring hospital admission.

Case 1

A 23 year old woman presented with recurrent haemoptyses. She was known to have congenitally corrected transposition of the great arteries with PA-VSD, but had subsequently undergone shunt ligation, VSD closure, and conduit reconstruction of the pulmonary outflow tract. On admission, bronchoscopy demonstrated blood in the left lower lobe bronchus. Thoracic aortography showed collateralisation of the pulmonary artery vessels arising via the intercostal arteries. Arteriography of the left internal mammary artery (IMA) revealed an abnormal area of dilated vessels arising from a branch vessel which collateralised the left lower lobe and appeared to be the source of bleeding. Left bronchial arteriography showed a second abnormal collection of vessels. Successful embolisation was performed of both the left IMA and left bronchial artery using polyvinyl alcohol particles. No further haemoptysis has since occurred.

Case 2

A 22 year old man presented with massive haemoptysis. He was born with a double outlet right ventricle, transposed great

Figure 1 Left: Internal mammary artery (A) giving rise to a prominent branch (B), which collateralises the pulmonary vasculature with highly abnormal distal vessels. Right: Successful occlusion of branch vessel using two fibred platinum coils, which resulted in control of the haemoptysis.

arteries, PA-VSD, and override of the right atrioventricular valve. Previously he had had left and right Blalock-Taussig shunts, and a modified Fontan procedure. Admission chest x ray suggested bleeding in the right lower lung field, subsequently confirmed on bronchoscopy. A right IMA demonstrated an enlarged arterial side branch forming a prominent group of abnormal vessels (fig 1, left panel) supplying the right lower lobe. Two fibred platinum coils were inserted with successful cessation of flow in the culprit branch vessel only (fig 1, right panel). Similarly, an abnormal area of dilated vessels was found arising from the right bronchial artery (a branch of the left superior intercostal artery). This vessel was also occluded successfully with preservation of the left superior intercostal artery, and haemoptysis ceased.

Case 3

A man, born with a double outlet right ventricle with pulmonary atresia, multiple ventricular septal defects, and a straddling tricuspid valve in the setting of dextrocardia, presented with recurrent haemoptysis. His past medical

Abbreviations: CHD, congenital heart disease; IMA, internal mammary artery; NBC, non-bronchial systemic collaterals; PA-VSD, pulmonary atresia with ventricular septal defect

history included systemic-to-pulmonary artery shunts and total cavopulmonary connection using an intracardiac conduit. On admission, bronchoscopy demonstrated prominent vessels over the whole bronchial tree and several pinpoint areas of bleeding in both right and left main bronchi. Conservative management was initially undertaken but haemoptysis continued. Selective angiography was offered but the procedure was declined by the patient.

DISCUSSION

Haemoptysis is an important and sometimes fatal complication in patients with PA-VSD, occurring as a result of extensive systemic-to-pulmonary collateralisation that has developed as a compensatory mechanism to provide pulmonary blood flow.^{1,2} Although haemoptysis is widely believed to be a rare complication of PA-VSD, there are no reports to date that have investigated the prevalence of this complication.¹ This study investigated retrospectively a group of patients with PA-VSD and found that haemoptysis is a relatively frequent complication in adult survivors, occurring in approximately 12% of patients—a rate much higher than previously suspected.¹ Furthermore, IMA collaterals were responsible in at least two out of these three cases, a previously unreported finding in this condition.

The treatment for recurrent or massive haemoptysis can be either medical, surgical or embolotherapy. Embolotherapy is indicated for those patients who continue to have recurrent haemoptysis in spite of conservative therapy, and is widely accepted as a successful treatment of massive haemoptysis. Surgical intervention can also be effective but is associated with a high operative mortality (> 15%).^{2,3} Embolotherapy has been shown to produce immediate control of bleeding in up to 90% of patients,⁴ but carries risks such as spinal cord ischaemia caused by anterior spinal artery occlusion; benefit may be transient as 20% of patients will rebleed within six months, with approximately 50% experiencing further significant haemoptyses on longer term follow up.⁴ In order to achieve successful control of haemoptysis, it is paramount that the operator has a thorough understanding of bronchial anatomy and recognises that other non-bronchial arteries may be involved. These non-bronchial systemic collaterals (NBC) originate from the phrenic, intercostal, internal mammary, thyrocervical, and other branches of the subclavian and axillary arteries.⁵ Our study found that the arteries responsible in at least 2 of 3 cases were non-bronchial and is the first to demonstrate the IMA as a cause of haemoptysis in PA-VSD. Furthermore, although the majority of non-cardiac related haemoptysis originates from the bronchial arteries, the source of bleeding is just as likely to originate from NBC in the context of CHD. Indeed, on reviewing all cases of CHD complicated by haemoptysis receiving embolotherapy (written in English), bronchial and NBC arteries appear to be culprit vessels in approximately equal proportions.

Haemoptysis is a common complication in patients with repaired/palliated PA-VSD. Embolotherapy can be a life saving treatment in this condition, provided the operator is aware that NBC arteries are often culprit vessels and, in particular, the internal mammary artery.

Authors' affiliations

K Greaves, *D S Celermajer, Department of Cardiology, Royal Prince Alfred Hospital, Sydney, New South Wales, Australia
P Bye, Department of Respiratory Medicine, Royal Prince Alfred Hospital
G Parker, Department of Radiology, Royal Prince Alfred Hospital

*Also Department of Medicine, University of Sydney

Correspondence to: Professor David S Celermajer, Department of Cardiology, Royal Prince Alfred Hospital, Page Chest Pavilion Building, Missenden Road, Camperdown, Sydney, NSW 2050, Australia; david.celermajer@email.cs.nsw.gov.au

Accepted 7 April 2003

REFERENCES

- 1 Haroutunian LM, Neill CA. Pulmonary complications of congenital heart disease: hemoptysis. *Am Heart J* 1972;**84**:540–59.
- 2 Crocco JA, Rooney JJ, Fankushen DS, et al. Massive hemoptysis. *Arch Intern Med* 1968;**121**:495–8.
- 3 Sehhat S, Oreizie M, Moinedine K. Massive pulmonary hemorrhage: surgical approach as choice of treatment. *Ann Thorac Surg* 1978;**25**:12–15.
- 4 Uflacker R, Kaemmerer A, Picon PD, et al. Bronchial artery embolization in the management of hemoptysis: technical aspects and long-term results. *Radiology* 1985;**157**:637–44.
- 5 Vujic I, Pyle R, Hungerford GD, et al. Angiography and therapeutic blockade in the control of hemoptysis. The importance of nonbronchial systemic arteries. *Radiology* 1982;**143**:19–23.

WEB TOP 10

www.heartjnl.com

These articles scored the most hits on *Heart's* website during May 2003

1 Non-invasive coronary imaging: computed tomography or magnetic resonance imaging?

CR Peebles

June 2003;**89**:591–4. (Editorial)

2 Resuscitation

R Vincent

June 2003;**89**:673–80. (Education in Heart)

3 A rational basis for selection among drugs of the same class

MJ Brown

June 2003;**89**:687–94. (Education in Heart)

4 What level of physical activity protects against premature cardiovascular death? The Caerphilly study

S Yu, JWG Yarnell, PM Sweetnam, L Murray

June 2003;**89**:502–6. (Cardiovascular medicine)

5 Drug treatment of hypertension: implications of ALLHAT

B Williams

June 2003;**89**:589–90. (Editorial)

6 Predicting mortality in patients with heart failure: a pragmatic approach

ML Bouvy, ER Heerdink, HGM Leufkens, AW Hoes

June 2003;**89**:605–9. (Cardiovascular medicine)

7 Indications for percutaneous and surgical revascularisation: how far does the evidence base guide us?

PM Schofield

May 2003;**89**:565–70. (Education in Heart)

8 Ambulatory blood pressure monitoring in the management of hypertension

E O'Brien

May 2003;**89**:571–6. (Education in Heart)

9 Microarray analysis: a novel research tool for cardiovascular scientists and physicians

C Napoli, LO Lerman, V Sica, A Lerman, G Tajana, F de Nigris

June 2003;**89**:597–604. (Review)

10 Stress and myocardial infarction

JS Chi, RA Kloner

May 2003;**89**:475–6. (Editorial)

Visit the *Heart* website for hyperlinks to these articles, by clicking on "Top 10 papers"

www.heartjnl.com