▲ Romero-Corral A, Montori VM, Somers VK, *et al.* Association of bodyweight with total mortality and with cardiovascular events in coronary artery disease: a systematic review of cohort studies. *Lancet* 2006;**368**:666–78.

Familial aortic dissection syndromes ► Dissection of the aorta may occur spontaneously even in normal sized aortas. A familial link is present in some cases. The Loeys-Dietz syndrome is a recently described autosomal dominant aortic aneurysm syndrome with widespread systemic involvement. The disease is characterised by the triad of arterial tortuosity and aneurysms, hypertelorism, and bifid uvula or cleft palate, and is caused by heterozygous mutations in the genes encoding transforming growth factor (TGF)- β receptors 1 and 2 (*TGFBR1* and *TGFBR2*, respectively). On the mild end, the mutations have been found in association with a presentation similar to that of Marfan's syndrome or with familial thoracic aortic aneurysm and dissection. From a study of a large cohort of patients and their families, the authors suggest that the natural histories of Loeys-Dietz syndrome type I and type II differ considerably from those of other connective tissue disorders. The median survival in their cohort was 37 years, as compared with 48 years among patients with vascular Ehlers-Danlos syndrome and 70 years among patients with Marfan syndrome who have been treated. The mean age at the first major vascular event in our group with Loeys-Dietz syndrome type II (29.8 years) is similar to that among patients with vascular Ehlers-Danlos syndrome caused by a deficiency of type III collagen (24.6 years). In both the Loeys-Dietz syndrome and vascular Ehlers-Danlos syndrome, dissection can occur without pronounced arterial dilatation. However, the incidence of fatal complications during or immediately after vascular surgery is about 45% in vascular Ehlers-Danlos syndrome but only 1.7% in Loeys-Dietz syndrome overall and 4.8% in type II. Thus, genotyping is beneficial in patients who present with features of vascular Ehlers-Danlos syndrome or Marfan syndrome. At least in mice with a Marfan-type syndrome, TGF- β signalling is returned to normal with losartan. A trial in humans is awaited.

▲ Loeys BL, Schwarze U, Holm T, *et al.* Aneurysm syndromes caused by mutations in the TGF-receptor. *N Engl J Med* 2006;**355**:788–98.

Ross procedure is safe long-term ► The use of the Ross procedure (pulmonary autograft replacement of the aortic valve) in adults remains controversial. Two hundred and sixty-four patients undergoing the Ross procedure in London and Rotterdam were followed up for a total of 1634 patient years; of these, 203 were men and 61 women, with a mean age of 35 years ranging from 18–66 years. The need for the procedure was due to congenital (52%), degenerative (22%) and rheumatic (8%) disease, and 21% of patients had undergone a prior aortic valve replacement. Overall 30-day mortality was 2.3% ($n = 6$), and four more patients died during follow up (mean follow up 6.2 years). Cumulative survival at five years was 96.8%, and at 10 years 95.4%. Ten patients had to undergo a further operation on the valve due to progressive dilatation and aortic regurgitation, and one due to arterial dissection of the wall of the homograft. Overall freedom from pulmonary homograft reoperation was 94.9% after 10 years, and for autograft reoperation was 92.9%. No risk factors for early or late mortality or reoperation were detected. The authors suggest that these excellent retrospective results mandate a prospective randomised trial to see whether their observations truly reflect the potential advantages of the Ross procedure, or whether they are caused by patient selection.

▲ Yacoub MH, Klieverik LM, Melina G, *et al.* An evaluation of the Ross operation in adults. *J Heart Valve Dis* 2006;**15**:531–9.

Journals scanned

American Journal of Medicine; American Journal of Physiology: Heart and Circulatory Physiology; Annals of Emergency Medicine; Annals of Thoracic Surgery; Archives of Internal Medicine; BMJ; Chest; European Journal of Cardiothoracic Surgery; Lancet; JAMA; Journal of Clinical Investigation; Journal of Diabetes and its Complications; Journal of Immunology; Journal of Thoracic and Cardiovascular Surgery; Nature Medicine; New England Journal of Medicine; Pharmacoeconomics; Thorax

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IMAGES IN CARDIOLOGY

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Percutaneous closure of a coronary to pulmonary artery fistula

A 54-year-old woman presented with a three year history of dyspnoea and chest pain on minimal exertion. Physical examination revealed a continuous murmur over the precordium. Background history included a cavernous haemangioma of the right lobe of the liver incidentally detected 10 years previously. Coronary angiography demonstrated a large fistula connecting the left anterior descending coronary artery (LAD) with the pulmonary artery (panel A). Very poor opacification of the LAD with dye injection was observed (see arrow in panel A). It was postulated that symptoms arose from a coronary steal effect and this was supported by myocardial perfusion imaging performed with adenosine stress which showed a reversible perfusion abnormality in the anterior wall of the myocardium. The fistula was occluded percutaneously by the deployment of 12 platinum coils (Trufill) into the fistula via the LAD. An instant improvement in dye opacification of the LAD was observed following the occlusion (see arrow in panel B) and corresponded with an immediate and dramatic improvement in symptoms.

(A) Left anterior descending to pulmonary artery fistula. (B) Fistula occluded and flow restored in anterior descending artery.

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