diomyocytes of the carrier biopsy. Dystrophin deficient cardiomyopathy cytes are able to maintain sarcolemmal conjunction with the remaining dys-cardiac muscle of MDX mice. In diomyocytes in this patient, as has deficiency in more than 50% of the car-cytes (Fig. 1). This finding suggests that utrophin may compensate, at least partially, for the dystrophin de-ficiency in all the car-dytes (Fig. 1). Therefore, utrophin may compensate, at least partially, for the dystrophin de-ficiency in all the car-dytes (Fig. 1). Therefore, utrophin may compensate, at least partially, for the dystrophin de-ficiency in all the car-dytes (Fig. 1). Therefore, utrophin may compensate, at least partially, for the dystrophin de-ficiency in all the car-dytes (Fig. 1). Therefore, utrophin may compensate, at least partially, for the dystrophin de-ficiency in all the car-dytes (Fig. 1). Therefore, utrophin may compensate, at least partially, for the dystrophin de-ficiency in all the car-dytes (Fig. 1). Therefore, utrophin may compensate, at least partially, for the dystrophin de-ficiency in all the car-dytes (Fig. 1). Therefore, utrophin may compensate, at least partially, for the dystrophin de-ficiency in all the car-dytes (Fig. 1). Therefore, utrophin may compensate, at least partially, for the dystrophin de-ficiency in all the car-dytes (Fig. 1). Therefore, utrophin may compensate, at least partially, for the dystrophin de-ficiency in all the car-

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Percutaneous stent implantation in an adult with left pulmonary artery stenosis and absent right pulmonary artery

We encountered a rare case of progressive dyspnoea of 3 years duration in a middle aged woman. Tricuspid regur-gitation (TR) was grade 2/3 with a right ventricular systolic pressure of 70 mmHg at two-dimensional echocardiography and Doppler study. There was significant narrowing of the proximal left pulmonary artery (LPA) and the right pulmonary artery (RPA) could not be seen. A radionuclide perfusion scan with 99mTc-Technetium showed no perfusion of the right lung and a first pass gated blood pool study revealed a right ejection fraction (EF) of 33% and a left ventricular (LV) EF of 60%. Cardiac catheterization revealed a mean right atrial pressure of 12 mmHg; RV pressure was 70/12 mmHg with a gradient of 45 mmHg across the LPA stenosis. A pulmonary artery angiogram revealed a 15 mm long 70% concentric stenosis of the proximal LPA, absent RPA and poor RV contractility.

The patient was therefore subjected to balloon dilatation and possible stenting of the LPA stenosis. With a 7 F Judkins right coronary catheter as a guide, a 0.035 inch angled glide wire (Terumo Corporation, Tokyo, Japan) was used to cross the stenosis and the catheter was then tracked distally into the left pulmonary artery. A 0.038 inch double length Amplatz extra stiff exchange guide wire (Cook Inc., Bloomington, Indiana) was positioned in the left lower lobe pulmonary artery. A 18 mm × 3 cm balloon catheter (Mansfield Inc., Boston) was positioned at the stenosis. The balloon was hand-inflated across the stenotic segment until disappearance of the waist. A repeat RV angiogram revealed only marginal improvement in the minimal internal diameter of the LPA (6 mm pre-dilation to 7 mm post dilatation). Since the lesion was stretchable with an elastic recoil, we decided to implant a stent to maintain dilatation of the LPA stenosis. A 14 F Mullins sheath was introduced over the guide wire and past the stenosis. A Palmaz iliac stent, which is 0.076 mm thick with a deflated profile of 3.4 mm and length of 3.0 cm was manually crimped on a 18 mm × 4.0 cm 8.5 F balloon (Cook Inc., Bloomington, Indiana) and the whole assembly was advanced into the Mullins sheath. The balloon-mounted stent was advanced up to the LPA stenosis and then hand inflated to deploy the stent. After successfully implanting the stent, RV angiograms (Fig. 1), taken in various projections showed a marked improve-ment in the minimal internal diameter (15 mm post stent implantation) and a minimal residual lesion. There was a significant improvement in RV contractility. Ten thousand units of heparin were given during the procedure and heparin was continued for 24 h. Subsequent-ly she was advised to take aspirin orally 75 mg, day~1 for 3 months. Pre-discharge Doppler study showed no TR and a gradient of 10 mmHg across the LPA.

Figure 1 Immunohistochemical up-regulation of utrophin in all the cardiomyocytes of the carrier biopsy.

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As the sole blood supply to the lungs in this case was through the LPA, its stenosis assumed a very critical role and the RV failure made it imperative to relieve the stenosis. The pulmonary arteries stretch before the intima and media tear during balloon dilatation, and under these conditions a stent proves advantageous since it prevents elastic recoil and leads to permanent vessel enlargement[41]. Also, lesser vessel wall trauma may reduce the chances of restenosis. It seems safe to balloon dilate and stent a branch pulmonary artery even if the other branch is absent; however, safety is limited in cases with complications.

B. V. DALVI
A. M. VORA
D. NARULA
H. KULKARNI
Department of Cardiology, King Edward VII Memorial Hospital, Parel, Bombay, India

Reference

Mitril, aortic and tricuspid valvular heart disease associated with ergotamine therapy for migraine

Ergotamine has been associated with numerous vascular complications but only rarely with valvular heart disease. We present a patient whose ergotamine abuse resulted in claudication of the legs followed by gangrene as well as valvular heart disease; the latter led to triple-valve replacement.

A 45-year-old woman presented with non-progressive weakness, orthopnea, and ankle oedema. Her pertinent medical history included the use, for more than 5 years, of Cafergot suppositories (ergotamine tartrate 1 mg, caffeine 100 mg), up to 10 per day for migraine headaches. There was no childhood history suggesting scarlet or rheumatic fever.

At the time of writing, 18 months ago the patient underwent mitral valve replacement for fourth degree mitral regurgitation. Two months later a de Vega tricuspid annuloplasty was necessary due to tricuspid regurgitation, and 6 months ago the patient underwent aortic and tricuspid re-placement due to tricuspid stenosis and second to third degree aortic regurgitation. Furthermore, a permanent pacemaker (VVI) was implanted as a result of symptomatic bradycardia absoluta. After tricuspid replacement and relocation of the epicardial lead, the first lead was left on site.

On admission, we found typical signs of congestive heart failure: tachycardia, dyspnoea, weakness, ankle oedema and radiological signs of oedema of the lung. Echocardiography demonstrated left atrial (LAD 42 mm) and right ventricular dilatation (end-diastolic diameter 38 mm), but normal left ventricular size (LVEDD 48 mm) and function. The diastolic pulmonary artery pressure measured 45 mmHg, but there was no sign of haemodynamic relevant valvular or prosthetic dysfunction.

Due to claudication of the leg and progressing gangrene, angiography of the lower extremity was performed. It demonstrated severe diffuse narrowing of the iliac and femoral arteries without any localized stenosis. The right foot was amputated. The specific medical history revealed abuse of ergotamine suppositories used in the treatment of migraine for more than 5 years.

We re-examined the pathological and histopathological findings acquired by other institutions. The excised mitral valve displayed severe diffuse leaflet and chordal thickening with commissural fusion. No appreciable calcification was evident. Proliferation of fibroblasts and smooth muscle cells was identified by standard light microscopy; features included abberant nuclear shape and cytoplasmatic eosinophilia. These cells formed a thick coating that surrounded the normal leaflet and chordal element. There were no signs of acute inflammation.

All three aortic valve cusps demonstrated mild thickening, without commissural fusion or calcification. The tricuspid valve showed commissural fusion, but no calcification. The histopathological findings were similar to, but less severe than, those of the mitral valve.

Although it cannot be proven conclusively that ergotamine caused the valvular lesions described in this patient, it is the most likely aetiological agent. The patient had no history suggestive of rheumatic fever, scarlet fever, congenital heart disease, or infective endocarditis. The patient developed the signs of claudication and of congestive heart failure concomitantly while using Cafergot suppositories (ergotamine tartrate 1 mg, caffeine 100 mg). Angiography showed vasocostriction but no localized stenosis. Vasculitis was not detected in any specimen.

The pathogenesis of ergotamine-associated proliferative valvar process is still unclear[2-5]. The cardiac valvular lesions resemble those described for carcinoid heart disease. Among the patients with carcinoid heart disease, levels of circulating serotonin are elevated markedly, and serotonin and other vasoactive amines seem to be instrumental in the formation of carcinoid plaques. It has been speculated that the serotonin antagonist, cyproheptadine, may also have agonistic properties in the cardiovascular system.

The valvular dysfunction, e.g. tricuspid and aortic regurgitation, most likely resulted from reduced mobility of the thickened valves. In addition, a reduced valve area resulting from scarring is possible.

In summary, although valvular heart disease has only rarely been associated with chronic ergotamine toxicity[41], chronic ergotamine abuse must be considered as an aetiologi agent[52], especially in the case of mitral regurgitation.

A. WILKE
H. HESSE
G. HUFNAGEL
B. MAISCH
Philips-University, Center of Internal Medicine, Department of Cardiology, Marburg, Germany

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Eur Heart J, Vol. 18, April 1997