- expression during cardiac growth. *Int Rev Cytol* 1991;124:137-86.
- 21 Karam R, Lever HM, Healy BP. Hypertensive hypertrophic cardiomyopathy or hypertrophic cardiomyopathy with hypertension? A study of 78 patients. *J Am Coll Cardiol* 1989;13:580-4.
 - 22 Gilligan D, Cleland J, Oakley C. The clinical and genetic heterogeneity of hypertrophic cardiomyopathy. *Current Medical Literature—Cardiovascular Medicine* 1990;9:155-61.
 - 23 Maron BJ, Wolfson JK, Ciro E, Spirito P. Relation of electrocardiographic abnormalities and patterns of left ventricular hypertrophy identified by two dimensional echocardiography in patients with hypertrophic cardiomyopathy. *Am J Cardiol* 1983;51:189-94.
 - 24 McKenna W, Stewart JT, Nihoyannopoulos P, McGinty F, Davies MJ. Hypertrophic cardiomyopathy without hypertrophy; two families with myocardial disarray in the absence of increased myocardial mass. *Br Heart J* 1990;63:287-90.
 - 25 Panza JA, Maron BJ. Relation of electrocardiographic abnormalities to evolving left ventricular hypertrophy in hypertrophic cardiomyopathy during childhood. *Am J Cardiol* 1989;63:1258-65.

ABSTRACTS IN CARDIOLOGY

Prevention of root dilatation in Marfan's syndrome

Prophylactic replacement of a dilating aortic root to forestall dissection is an accepted form of management in Marfan's syndrome but there is now clear evidence, in the article by Shores *et al*, that the need for surgery can be reduced by long-term β adrenergic blockade. Such evidence was needed. The doses used to achieve a negative inotropic effect, and theoretically reduce risk, are high. To expose

young people to long-term side effects without proven benefit is not justifiable.

The problem of dissection in Marfan's syndrome is not totally solved however. In some patients dissection occurs before the aorta dilates and even in those on β blockade aortic dilatation continues, albeit at a slower rate. We are, however, closer to rational therapy.

M J DAVIES

Progression of aortic dilatation and the benefit of long-term β -adrenergic blockade in Marfan's syndrome

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Background—The aortic root enlarges progressively in Marfan's syndrome, and this enlargement is associated with aortic regurgitation and dissection. Long-term treatment with β -adrenergic blockade, by reducing the impulse (i.e., the rate of pressure change in the aortic root) of left ventricular ejection and the heart rate, may protect the aortic root.

Methods—We conducted an open-label, randomized trial of propranolol in adolescent and adult patients with classic Marfan's syndrome (32 treated and 38 untreated [control] patients). Aortic-root dimensions and clinical end points (aortic regurgitation, aortic dissection, cardiovascular surgery, congestive heart failure, and death) were monitored for an average of 9.3 years in the control group and 10.7 years in the treatment group. All 70 patients were included in the analysis according to the intention-to-treat principle.

Results—The dose of propranolol was individualized: the mean ($\pm$ SE) dose was 212 ± 68 mg per day. The mean slope of the regression line for the aortic-root dimensions, which reflect the rate of dilatation, was significantly lower in the treatment group than in the control group (0.023 v 0.084 per year, $P < 0.001$). Clinical end points were reached in five patients in the treatment group and nine in the control group. The Kaplan-Meier survival curve for the treatment group differed significantly from that for the control group during the middle years of the trial and remained better for the treatment group throughout the study.

Conclusions—Prophylactic β -adrenergic blockade is effective in slowing the rate of aortic dilatation and reducing the development of aortic complications in some patients with Marfan's syndrome. (*N Engl J Med* 1994;330:1335-41).