Sildenafil Use in Patients With the Eisenmenger Syndrome

To the Editor:

Michelakis et al report encouraging results with the use of sildenafil in patients with pulmonary hypertension. The authors included a patient with repaired congenital heart disease (and a small residual ventricular septal defect) who has pulmonary vascular disease, but also labeled him as having the Eisenmenger syndrome. This is unfortunate, as it gives the impression that sildenafil is therefore effective in patients with the Eisenmenger syndrome when it has yet to be demonstrated. The Eisenmenger syndrome refers to patients with pulmonary vascular disease associated with unrestrictive shunting, commonly at ventricular level, who are usually cyanosed and polycythaemic. Sildenafil in patients with the Eisenmenger syndrome may potentially cause a reduction in pulmonary blood flow and an increase in cyanosis as a result of even mild systemic vasodilation and consequent increased right-to-left shunting. Although on going trials may yet show that sildenafil (and other pulmonary dilators such as bosentan) are safe and effective in patients with the Eisenmenger syndrome, this article should not lead to widespread use of sildenafil in these patients.

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Response

We thank Dr Rosenthal for his letter and agree that our article should not encourage the use of sildenafil in patients with Eisenmenger’s syndrome. Although in our patient sildenafil was beneficial, this is only an isolated case. Most importantly, our trial was not placebo-controlled, and until blinded and placebo-controlled data are available, sildenafil should not be used routinely in patients with Eisenmenger’s syndrome or any form of pulmonary arterial hypertension (PAH). Our pilot data should only encourage research in this area and perhaps inclusion of patients with congenital heart disease and PAH in larger clinical trials; unfortunately, patients with PAH and congenital heart disease are often excluded from PAH trials. The modern management of PAH dictates referral to physicians specializing in PAH or to multidisciplinary pulmonary hypertension clinics in major tertiary care centers.

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Circulation. 2004;109:e197
doi: 10.1161/01.CIR.0000127113.72093.45

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