

## Images in Congenital Heart Disease

# Reversal of shunting in pulmonary hypertension after treatment with oral Sildenafil

Ageliki A. Karatza,<sup>1</sup> Andrew Bush,<sup>2</sup> Alan G. Magee<sup>1</sup>

<sup>1</sup>Department of Paediatric Cardiology; <sup>2</sup>Department of Paediatric Respiratory Medicine, Royal Brompton Hospital, London, UK

**A**N 11-YEAR-OLD GIRL WITH PULMONARY vascular disease associated with an atrial septal defect and a patency of the arterial duct was treated with oral Sildenafil at a dosage of 0.5 mg/kg 4 hourly. The dose was subsequently

increased at monthly intervals to 1 mg/kg, and then to 2 mg/kg.

Before treatment was started, she had clubbing of the toes, a saturation of oxygen of 80% in the toes, and saturations of oxygen of 90–93% in the fingers.

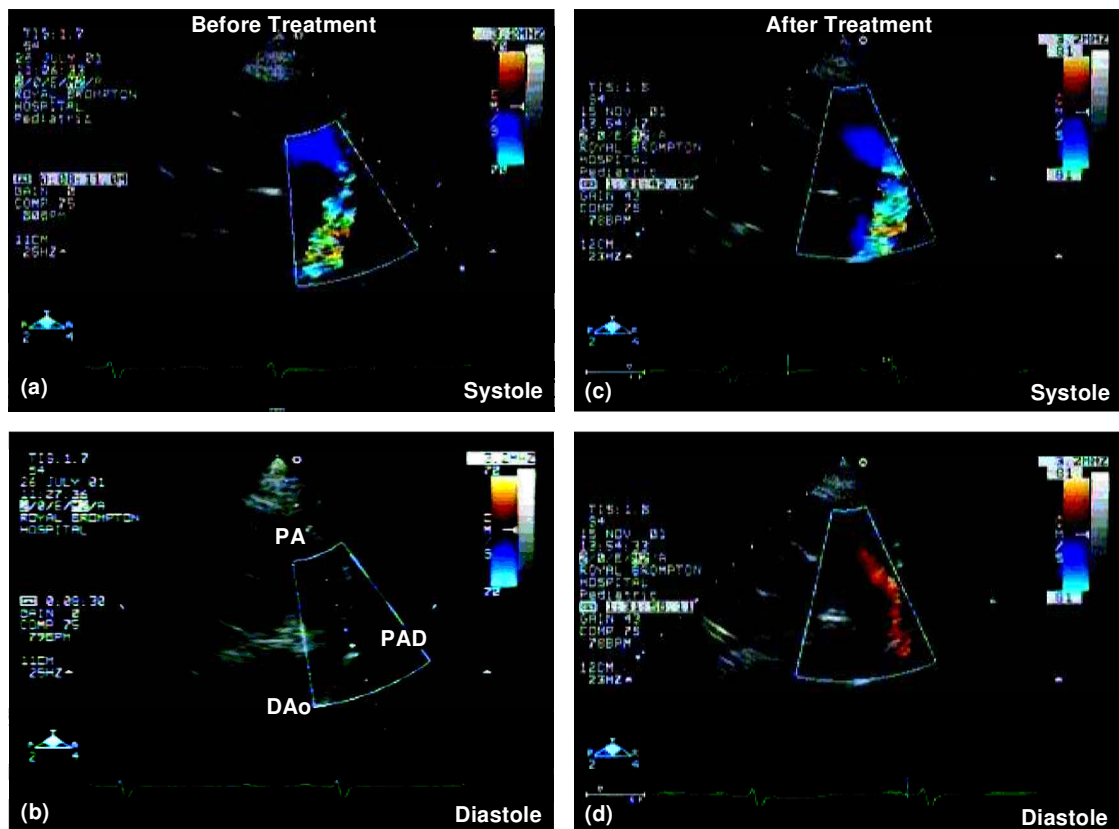


Figure 1.

Correspondence to: Alan G. Magee, MD, FRCP, Consultant Paediatric Cardiologist, Royal Brompton Hospital, Sydney Street, London SW3 6NP, UK. Tel: 020 7351 8152; Fax: 020 7351 8547; E-mail: a.magee@rbh.nthames.nhs.uk

AAK was funded by the Royal Brompton Clinical Research Committee. Sildenafil Citrate was supplied by Pfizer Pharmaceuticals, Sandwich, Kent, UK

Accepted for publication 12 July 2002

Parasternal short axis echocardiographic views at the base of the heart showed right-to-left shunting through the arterial duct (PAD), from the pulmonary artery (PA) to the aorta (DAo), during systole, with no shunting in diastole (Fig. 1a, b). Tricuspid regurgitation was also present, with a peak Doppler velocity of 5.3 m/s, giving an estimated pressure gradient of 132 mmHg.

After initiation of treatment, the patient reported gradual improvement in exercise capacity, with improvement in the functional classification of the New York Heart Association from grade III to grade

I/II. No side effects were noted. After 16 weeks of treatment, the saturations of oxygen remained unchanged in the fingers, but those in the toes had risen to 90%. Echocardiography showed that shunting through the arterial duct was now from right-to-left in systole (Fig. 1c), and left-to-right in diastole (Fig. 1d), although the estimated right ventricular pressure remained suprasystemic.

Sildenafil is a selective inhibitor of phosphodiesterase 5 that may have a role in treating some patients with pulmonary vascular disease associated with congenital cardiac defects.