Pulmonary hypertension frequently complicates mitral stenosis. Increased pulmonary artery pressure results from raised left atrial pressure, pulmonary arteriolar constriction, and obliterative changes in the pulmonary vascular bed, and usually responds to surgical relief of mitral stenosis. However, severe pulmonary hypertension may persist after surgical treatment of mitral stenosis. We describe a patient whose severe pulmonary hypertension following mitral valve replacement was treated successfully with continuous intravenous epoprostenol.

CASE REPORT

A 46 year old woman, diagnosed at the age of 11 with acute rheumatic fever, underwent placement of a #26 Duran ring for mitral regurgitation in 1991. Pulmonary artery pressures were normal although mild mitral stenosis was noted postoperatively. She was lost to follow up.

In 1999 she sought medical attention because of dyspnoea and orthopnoea. Cardiac catheterisation showed severe pulmonary hypertension (mean pulmonary artery pressure 48 mm Hg) with a 14 mm Hg gradient across the mitral valve (pulmonary capillary wedge pressure 18 mm Hg). In August 1999 she underwent mitral valve replacement with a 23 mm St Jude prosthesis. Postoperative transoesophageal echocardiography showed good function of the new prosthetic valve, but severe pulmonary hypertension persisted.

In June 2001 she again sought medical attention because of dyspnoea with the slightest activity. Echocardiography showed a normally functioning prosthetic mitral valve and severe pulmonary hypertension. Diagnostic workup did not suggest another aetiology for pulmonary hypertension. Cardiac catheterisation showed severe pulmonary hypertension without evidence of mitral stenosis (table 1). Continuous intravenous epoprostenol (Flolan) was begun. Over the next 4 months the dose was increased from 7 ng/kg/min to 23 ng/kg/min and the patient noted improvement in her exercise capacity. Six and 17 months after beginning epoprostenol repeat right heart catheterisation showed significant and sustained reductions of pulmonary artery pressure and pulmonary vascular resistance (table 1).

DISCUSSION

Pulmonary hypertension is a common complication of severe mitral valve disease. Raised pulmonary artery pressure results initially from increased left atrial pressure, pulmonary arteriolar vasoconstriction, and ultimately obliterative changes in the pulmonary vascular bed. Wood studied 500 cases of critical mitral stenosis, 12% of whom had extremely high pulmonary vascular resistance (>10 units) and 16% in whom it was moderately high (6–10 units). Injection of 1 mg acetylcholine lowered pulmonary artery pressures in 14 of 16 cases, but in two cases with extremely high pulmonary vascular resistance there was no response, presumably because of obliterative pulmonary vascular disease.

Mitral valve replacement generally alleviates pulmonary hypertension, but a small subgroup of patients have persistent severe pulmonary hypertension in spite of relief of mitral stenosis by closed valvuloplasty or mitral valve replacement. Other patients with extreme preoperative increases in pulmonary vascular resistance do not survive valvuloplasty or valve replacement because of pulmonary vascular disease. Failure to respond to surgical relief of mitral stenosis often reflects pathological changes in the pulmonary arteries that resemble those seen with other causes of pulmonary arterial hypertension. Excessive thickening of the media and intimal fibrosis of small muscular pulmonary arteries are typical of long standing mitral

| Pulmonary hypertension frequently complicates mitral stenosis. Increased pulmonary artery pressure results from raised left atrial pressure, pulmonary arteriolar constriction, and obliterative changes in the pulmonary vascular bed, and usually responds to surgical relief of mitral stenosis. However, severe pulmonary hypertension may persist after surgical treatment of mitral stenosis. We describe a patient whose severe pulmonary hypertension following mitral valve replacement was treated successfully with continuous intravenous epoprostenol. |

<table>
<thead>
<tr>
<th>Table 1 Haemodynamic variables 2 years after mitral valve replacement (baseline) and after long term epoprostenol treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Baseline</strong></td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td>Mean systemic arterial pressure (mm Hg)</td>
</tr>
<tr>
<td>Mean right atrial pressure (mm Hg)</td>
</tr>
<tr>
<td>Mean pulmonary artery pressure (mm Hg)</td>
</tr>
<tr>
<td>Cardiac output (l/min)</td>
</tr>
<tr>
<td>Pulmonary artery occlusion pressure (mm Hg)</td>
</tr>
<tr>
<td>Left ventricular end diastolic pressure (mm Hg)</td>
</tr>
<tr>
<td>Pulmonary vascular resistance (dyne.s/cm⁵)</td>
</tr>
</tbody>
</table>

* Epoprostenol dose 24 ng/kg/min. † Epoprostenol dose 26 ng/kg/min. † Thorbosis of the anterior leaflet of the St Jude valve led to mild increases in pulmonary artery occlusion pressure at 6 months and 17 months after epoprostenol was started.
Epoprostenol after mitral valve replacement

Sion,10 and suggests that epoprostenol may have a role in the dynamic response of our patient was typical of the response mechanism for this effect remains uncertain. The haemodynamic after successful mitral valve replacement.

treatment of patients with sustained pulmonary hypertension.89 Sometimes hypertension is difficult, particularly when the pulmonary artery wedge pressure is only modestly raised.89 Sometimes there are clues—such as the identification of another family member with primary pulmonary hypertension—which help to distinguish these two conditions but, in spite of a careful search, we were unable to identify such a clue in this case. We cannot exclude the possibility that the patient represents the extremely unusual coexistence of primary pulmonary hypertension with mitral stenosis, although this possibility seems less likely in the setting of established mitral stenosis.

The present case illustrates a favourable response to epoprostenol of a patient with severe pulmonary hypertension following successful mitral valve replacement. Epoprostenol improves survival as well as haemodynamics in primary pulmonary hypertension,10 although the exact mechanism for this effect remains uncertain. The haemodynamic response of our patient was typical of the response described for patients with primary pulmonary hypertension, and suggests that epoprostenol may have a role in the treatment of patients with sustained pulmonary hypertension after successful mitral valve replacement.

Authors’ affiliations

C G Elliott, Departments of Medicine, Pulmonary and Critical Care Divisions, LDS Hospital and the University of Utah School of Medicine, Salt Lake City, USA

H I Palevsky, Department of Medicine, Pulmonary and Critical Care Division, Presbyterian Medical Center, University of Pennsylvania Health System, USA

Correspondence to: Professor C G Elliott, Pulmonary Division, LDS Hospital, Salt Lake City, UT 84143; ldgellio@ihc.com

Received 11 April 2003
Accepted 14 August 2003

REFERENCES

8 Cheng TD. Differentiation between mitral stenosis and coexisting primary pulmonary hypertension. Thorax 2000;55:807
9 Langleben D, Schlesinger R. Differentiation between mitral stenosis and coexisting primary pulmonary hypertension: authors’ reply. Thorax 2000;55:807
Treatment with epoprostenol of pulmonary arterial hypertension following mitral valve replacement for mitral stenosis
C G Elliott and H I Palevsky

Thorax 2004 59: 536-537
doi: 10.1136/thx.2003.008193

Updated information and services can be found at:
http://thorax.bmj.com/content/59/6/536

These include:

References
This article cites 8 articles, 4 of which you can access for free at:
http://thorax.bmj.com/content/59/6/536#BIBL

Email alerting service
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Topic Collections
Articles on similar topics can be found in the following collections
Pulmonary hypertension (205)

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/