Extreme right ventricular hypoplasia after relief of severe pulmonary stenosis

Use of balloon catheter occlusion of atrial septal defect in assessing right ventricular function

L COTTER, C D PUSEY, G A H MILLER

From Cardiac Department, Brompton Hospital, Fulham Road, London

SUMMARY A patient is described in whom extreme right ventricular hypoplasia and right-to-left shunting through an atrial septal defect occurred after relief of severe pulmonary stenosis. The ability of the hypoplastic right ventricle to deal with an increased volume load was assessed at cardiac catheterisation by occluding the atrial septal defect with a balloon tipped catheter.

Right-to-left interatrial shunting in the presence of normal right ventricular pressure is an unusual finding which has been described in rare congenital anomalies such as Uhl's syndrome and isolated right ventricular hypoplasia^{1,2} and rarely after complete relief of pulmonary valve stenosis.^{3,4} The shunting in these circumstances is attributed to an increased resistance to right ventricular filling. We describe a case with right-to-left interatrial shunting with normal right ventricular pressure and right ventricular hypoplasia after pulmonary valvotomy for pulmonary stenosis, in whom a balloon catheter was used to occlude the interatrial communication at cardiac catheterisation. This allowed assessment of the adequacy of the hypoplastic right ventricle to deal with an increased workload. Subsequent closure of a secundum atrial septal defect resulted in relief of the patient's cyanosis and effort intolerance.

Case report

A 26-year-old student presented with cyanosis and worsening effort intolerance. He had been cyanosed from birth and cardiac catheterisation at the age of 14 years had shown severe pulmonary stenosis with right-to-left shunting at atrial level. The right ventricular pressure was said to have been 280 mmHg. A pulmonary valvotomy was performed at that time giving some relief of symptoms for about a year, after which they recurred. On examination he was a well-developed man with grade IV cyanosis and clubbing of the fingers and toes. The pulse was 96 a minute, regular, and of normal

volume and character. The venous pulsation showed a prominent "a" wave, blood pressure 140/80 mmHg. There was no parasternal heave, the apex was impalpable. The first heart sound was normal, the second widely and fixedly split. There was a grade 1/6 soft mid-systolic murmur at the upper left sternal edge. There were no other abnormal physical signs. The haemoglobin was 22 g. Electrocardiograms showed sinus rhythm, with evidence of right atrial hypertrophy and high left ventricular voltage (R=31 mm in V5, S=23 mm in V1) with ST and T wave changes in leads V3 to V6. A chest x-ray film showed moderate cardiac enlargement with normal lung fields. At cardiac catheterisation a 14 mmHg right atrial "a" wave and a right ventricular pressure of 24/9 to 12 mmHg were shown with no significant gradient across the pulmonary valve. There was right-to-left shunting at atrial level, resulting in a systemic arterial saturation of 82 per cent, the ratio of systemic to pulmonary blood flows being 1.7:1. The interatrial defect was occluded by inflating a no. 7 balloon catheter in the left atrium and pulling it against the septum. This manoeuvre eliminated shunting and allowed the arterial oxygen saturation to reach 97 per cent with obvious relief of the cyanosis. This occlusion was maintained for 20 minutes with no adverse haemodynamic changes or conduction disturbances. A right ventricular angiogram indicated a small, thick-walled, poorly contracting right ventricle.

At operation (Mr C Lincoln) on cardiopulmonary bypass, a secundum atrial septal defect measuring

1 × 1 cm was closed by direct suture through a right atrial approach. The postoperative course was uneventful and the patient's symptoms were relieved. Repeat cardiac catheterisation showed normal haemodynamics at rest and on exercise, apart from a 15 mmHg right atrial "a" wave and a 10 mmHg gradient across the pulmonary valve; there was no evidence of intracardiac shunting. An electrocardiogram a month after the operation showed a decrease in P wave amplitude in lead II from 4 to 2 mm, with ST-T wave changes attributable to the pericardiotomy. He has remained well at follow-up to date, with a haemoglobin of 13 g, apart from one episode of supraventricular tachycardia which responded rapidly to carotid sinus massage.

Discussion

Pulmonary valvotomy is usually performed early in severe pulmonary stenosis, particularly if complications such as right ventricular failure have occurred. If it is performed before overt right ventricular failure, postoperatively right ventricular function may be normal,⁵ while if operation is delayed myocardial fibrosis and right ventricular systolic or diastolic dysfunction is likely. This continued right ventricular dysfunction postoperatively is probably multifactorial. Preoperative factors causing right ventricular dysfunction include poor coronary blood flow caused by extreme ventricular hypertrophy relative to the amount of coronary capillary circulation, the fixed size of the coronary ostia, the extremely high pressures in the right ventricular wall preventing any coronary blood flow during systole, and the decreased pressure gradient across the coronary bed produced by an increase in the right atrial pressure and right ventricular diastolic pressure⁶; operative factors particularly the efficacy of myocardial protection and in addition whether or not a ventriculotomy is performed are also important.^{7,8} If diastolic dysfunction is predominant, there may be continued "indistensibility" of the right ventricle because of poor compliance leading to a high right ventricular filling pressure.^{3,4} In the presence of an interatrial communication, either a patent foramen ovale or an atrial septal defect, the high right ventricular filling pressure, if it exceeds the left ventricular filling pressure, will cause right-to-left shunting at atrial level, regardless of the relative ventricular systolic pressures, with consequent arterial desaturation and, maybe, clinical cyanosis.

Right-to-left shunting at atrial level is not rare in severe pulmonary stenosis,⁹⁻¹¹ and is a cause of cyanosis in this condition. It is almost invariably relieved after pulmonary valvotomy when the right

ventricular filling pressure falls.¹¹ Continued right-to-left shunting after pulmonary valve surgery is, however, rare.^{3,4} Its incidence varies but Oakley *et al.*³ described it in six cases out of their series of 56 pulmonary valvotomies. When it occurs reoperation is recommended to close the interatrial shunt, whether a valve patent foramen ovale or an atrial septal defect, to avoid the possibility of paradoxical emboli and to avoid the sequelae of polycythaemia. The results of closing such a defect however, are not always predictable, as the right ventricle may not be able to cope with the increased workload, particularly if there is a degree of right ventricular hypoplasia, and right ventricular failure may result.¹²

Although right ventricular dysfunction is well recognised after relief of long-standing pulmonary stenosis, frank right ventricular hypoplasia which can also occur is very unusual and less well described.¹²⁻¹⁴ When it is present it may be helped by the volume loading which occurs if pulmonary regurgitation is caused at pulmonary valvotomy.¹³

The use of balloon tipped catheters to close interatrial communications has been advocated in patients with isolated right ventricular hypoplasia in order to assess the possible effects on the right ventricle of surgical closure of the defects.^{1,2} As far as we know, this is the first case of interatrial shunting after pulmonary valvotomy where this manoeuvre has been used to advantage in showing that, though the right ventricle was relatively hypoplastic, it could, nevertheless, cope with the increased workload.

We are grateful to Mr Christopher Lincoln for permission to report this case.

References

- ¹Van der Hauwaert LG, Michaelsson M. Isolated right ventricular hypoplasia. *Circulation* 1971; **44**: 466-74.
- ²Haworth SG, Shinebourne EA, Miller GAH. Right-to-left interatrial shunting with normal right ventricular pressure—a puzzling haemodynamic picture associated with some rare congenital malformations of the right ventricle and tricuspid valve. *Br Heart J* 1975; **37**: 386-91.
- ³Oakley CM, Braimbridge MV, Bentall HH, Cleland WP. Reversed interatrial shunt following complete relief of pulmonary valve stenosis. *Br Heart J* 1964; **26**: 662-70.
- ⁴de Castro CM, Nelson WP, Jones RC, Hall RJ, Hopeman AR, Jahnke EJ. Pulmonary stenosis, cyanosis, interatrial communication and inadequate right ventricular distensibility following pulmonary valvotomy. *Am J Cardiol* 1970; **26**: 540-3.
- ⁵Stone FM, Bessinger FB Jr, Lucas RV Jr, Moller JH. Pre- and postoperative rest and exercise hemodynamics

- in children with pulmonary stenosis. *Circulation* 1974; **49**: 1102-6.
- ⁶Schlant RC. In Hurst JW, ed. *The heart*. New York: McGraw-Hill, 1978: 822.
- ⁷Johnson AM. Impaired exercise response and other residua of pulmonary stenosis after valvotomy. *Br Heart J* 1962; **24**: 375-88.
- ⁸Finnegan P, Ihenacho HNC, Singh SP, Abrams LD. Haemodynamic studies at rest and during exercise in pulmonary stenosis after surgery. *Br Heart J* 1974; **36**: 913-8.
- ⁹Abrahams DG, Wood P. Pulmonary stenosis with normal aortic root. *Br Heart J* 1951; **13**: 519-48.
- ¹⁰Callahan JA, Brandenburg RO, Swan HJC. Pulmonary stenosis and interatrial communication with cyanosis; hemodynamic and clinical study of 10 patients. *Am J Med* 1955; **19**: 189-202.
- ¹¹Brock R. The surgical treatment of pulmonary stenosis. *Br Heart J* 1961; **23**: 337-56.
- ¹²Williams CB, Barrett-Boyes BG, Lowe JB. Under-developed right ventricle and pulmonary stenosis. *Am J Cardiol* 1963; **11**: 458-68.
- ¹³Rao PS, Liebman J, Borkat G. Right ventricular growth in a case of pulmonary stenosis with intact ventricular septum and hypoplastic right ventricle. *Circulation* 1976; **53**: 389-94.
- ¹⁴Rowe RD. Pulmonary stenosis with normal aortic root. In: Keith JD, Rowe RD, Vlad P, eds. *Heart disease in infancy and childhood* 3rd ed. New York: Macmillan, 1978: 762.

Requests for reprints to Dr L Cotter, Cardiac Department, Brompton Hospital, Fulham Road, London SW3 6HP.