Hour-glass deformity of the pulmonary valve: a third type of pulmonary valve stenosis

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SUMMARY In the 12 years from 1975 to 1987, 55 patients had open pulmonary valve surgery for isolated congenital stenosis of the pulmonary valve. Three types of pulmonary stenosis were seen: (a) dome-shaped pulmonary stenosis (34 patients); (b) dysplastic pulmonary valves with thick, cauliflower-like cusps (12 patients), and (c) hour-glass deformity of the pulmonary valve, with "bottle-shaped" sinuses (nine patients). This third type has not been described before.

Preoperative identification of the valve structure is important because the choice of treatment (balloon dilatation for some dome-shaped valves and excision for dysplastic and hour-glass valves) depends on the type of stenosis.

Isolated pulmonary valve stenosis is the most common cause of right ventricular outflow tract obstruction and accounts for 8-10% of all congenital cardiac anomalies.¹ It was first described by Morgagni in 1761, and Meckel reported anatomical features in 1817.²

In the most common form of stenosis, the pulmonary valve is conical as a result of fusion of the valve leaflets, and it protrudes superiorly into the pulmonary artery. Its shape and opening are influenced by the degree of thickness and rigidity of the fused cusps. Its appearance on angiography has led to the descriptive term dome-shaped pulmonary stenosis. In another type of pulmonary stenosis fusion of the commissures is slight, but there is considerable obstruction to flow caused by noticeably thickened, immobile cusps composed of disorganised myxomatous tissue; this distinctive type of pulmonary stenosis is associated with a rather narrow valve annulus and has been termed "pulmonary valvular dysplasia."^{3,4} Patients with Noonan's syndrome have pulmonary valves of this type.⁵

We report a third type of pulmonary valve deformity, which, because of its angiographic appearance,

we have called hour-glass deformity of the pulmonary valve with bottle-shaped sinuses.

Patients and methods

Between 1975 and 1987, 55 patients (30 male and 25 female, aged from 6 weeks to 32 years) had open pulmonary valve operation for isolated stenosis in the Department of Thoracic and Cardiovascular Surgery at the Sheba Medical Centre and the Department of Cardiac Surgery at the Rambam Medical Centre. Isolated pulmonary valve stenosis was diagnosed by clinical, electrocardiographic, and Doppler echocardiographic criteria and was confirmed by cardiac catheterisation and right heart angiography. We also used the criteria of Koretzky *et al*³ and the radiographic features of Jeffrey *et al*⁴ to identify dysplastic valves.

Operation was indicated if the right ventricular systolic pressure was >75 mm Hg and there were electrocardiographic signs of severe right ventricular hypertrophy. The preoperative identification of cauliflower or hour-glass valves improved as we became more experienced in interpreting the angiograms.

The pulmonary valve was examined during operation; the structural characteristics were noted, including thickness and type of the cusps, degree of commissural fusion, form of the valve sinus, and the effective size of the valve orifice.

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We did not study patients with bicuspid valves, unicuspid valves, and valves with little or no commissural development. We focused on the different structural types of abnormalities of the tricuspid pulmonary leaflets.

All patients are being followed clinically, radiographically, electrocardiographically, and echocardiographically for long term assessment.

Results

We identified three distinct structural types of pulmonary valve stenosis under direct vision during operation:

THE DOME-SHAPED VALVE (FIG 1B)

Thirty four of the 55 patients had pulmonary stenosis caused by a dome-shaped valve. The commissures were fused and their ridges extended towards the centre of the valve (fig 2a).⁶ The sinuses were moderately deep and wide open from commissure to commissure. The cusps were thicker than normal, but smooth on both sides. The valve orifice was usually round and central. The angiographic picture (fig 2b) clearly showed that the stenotic pulmonary valve was dome-shaped. The diameter of the pulmonary artery at the level of the commissures was normal and neither enlarged nor narrowed. Post-stenotic dilatation of the main pulmonary artery increased with age. None of the 34 patients with dome shaped pulmonary stenosis showed features of Noonan's syndrome.⁵

These patients were treated by commissurotomy extending to the arterial wall, with unhinging of the cusps for better mobilisation. Balloon dilatation, first described by Kan *et al*⁷ in 1982 and subsequently reported by others,⁸⁻¹⁰ should now be considered as an alternative treatment in patients with a dome-shaped valve.

THE DYSPLASTIC PULMONARY VALVE (FIG 1C)

Twelve patients had dysplastic valves (fig 1c) with thick, cauliflower-like cusps. The firm and verrucous tissue and the uneven distribution of the residual primitive myxomatous material of the cusps obstructed the flow through the pulmonary valve. There was either little or no fusion of the commissures, but the valve sinuses were to a large extent occupied by the dysplastic material of the cusps, making it difficult for them to deviate laterally in systole and allow unobstructed forward blood flow. None the less, the sinuses were well developed with wide open orifices (fig 3a). Once we knew what to look for, this type of valve was easily recognised on angiography (fig 3b) and verified on operation. Various grades of post-stenotic dilatation were present in these patients,

both on angiography and at operation, while at the valve level the pulmonary artery showed no particular changes.

In our first operation on this type of valve, in 1975, we performed partial cusp resection and trans-valvar patching. This approach was not satisfactory, and we found that complete removal of all the obstructing material within the valve by partial or complete valvectomy allowed free flow into the pulmonary system. There was no need for additional trans-valvar patching, as the obstruction was at the level of the cusps rather than the annulus.

All patients with Noonan's syndrome had valves of this type, but the reverse was not true.

HOURLASS DEFORMITY OF THE PULMONARY ARTERY WITH BOTTLE-LIKE VALVE SINUSES (FIG 1D)

Nine patients presented with this newly described type of stenosis. Patients showed varying degrees of post-stenotic dilatation of the pulmonary artery, distal to the plane of the commissures and orifice. At

Fig 1 Appearance in diastole of normal pulmonary valve (a), dome-shaped pulmonary stenosis (b), dysplastic pulmonary valve with cauliflower-like cusps (c), and hour-glass deformity of the pulmonary artery with deep bottle-like sinuses (d).

Fig 2 Photograph of a dome-shaped stenotic pulmonary valve showing (a) fusion of the commissures resulting in a funnel with an almost circular valve orifice (reprinted with permission from Parker⁶). Note the wide orifices of the sinuses. (b) Angiogram (lateral view) in end systole. Fusion of the three cusps gives the appearance of a dome.

this level the pulmonary artery was narrower than it was at its origin from the right ventricular outflow tract or at the post-valvar segment, and thus it resembled an hour-glass (fig 1d). The commissures were either not fused or only slightly fused. The cusps were thicker than those of the dome-shaped valves, but their surface was smooth on both sides; the outflow edges were thickened and rolled in towards the valve sinuses, forming relatively narrow, asymmetrical orifices to each. The pulmonary artery at the level of the commissures and orifice was markedly thickened, with a ring of sinus and orifice tissue on its inner side (fig 1d). This was soft in the younger patients and firm and scar-like in older patients. The sinuses resembled bottles, being deep and dilated with narrow orifices (fig 4a). The valve outlet was obstructed both by the hour-glass deformity and by the thickened cusps and their rolled-in edges. Preoperative diagnosis of hour-glass deform-

mity could be made angiographically by recognition of the narrowing at the plane of the commissures and orifice and by the bottle-like sinuses on a right ventricular angiogram (fig 4b).

We tried both commissurotomy and partial valve resection before we found that total pulmonary valvectomy was the most successful treatment. Trans-valvar patching was not required, because the pulmonary artery was widened sufficiently by relief of the inward tension at the level of the sinus orifices.

None of the 55 patients died nor were there major complications.

Discussion

Congenital pulmonary valve stenosis includes both the well-known dome-shaped pulmonary stenosis and the more recently described dysplasia of the pulmonary valve,^{3,4} with its special relation to Noon-

Fig 3 Photograph of a dysplastic pulmonary valve (DPV) showing the cauliflower-like cusps. The commissures are not fused. The sinus orifices are wide. The obstruction to flow in this valve is caused by excess dysplastic tissue. There is considerable post-stenotic dilatation of the pulmonary artery (PA). RVOT, right ventricular outflow tract. (b) Angiogram (lateral view) after injection of contrast into the right ventricle. Irregular filling defects (arrows) of contrast material at the valve level indicate smooth, thicker than normal, cauliflower-like masses on the cusps, that obstruct flow through the valve. There is considerable post-stenotic dilatation.

an's syndrome.⁵ In the present study we found a third form of pulmonary stenosis, the hour-glass pulmonary stenosis, which, to our knowledge, has not been recognised before. Koretzky *et al*, who described dysplasia of the pulmonary valve in 1969, found supra-valvar pulmonary stenosis in two of their six specimens.³ Jeffrey *et al* described the radiographic features of supra-valvar pulmonary stenosis and presented cases in which there was pronounced annular hypoplasia and valve thickening.⁴ Their case 10 demonstrated bottle-like sinuses in diastole. Among patients operated on for isolated pulmonary stenosis, Utley and Roe described one case in which the stenosis of the valve was associated with narrowing of the proximal pulmonary artery at the distal attachment of the valve cusps.¹¹ We believe that these

workers described the narrowing at the plane of the commissures and orifice that we saw in hour-glass deformity.

We used the term hour-glass deformity of the pulmonary valve to describe both the external and internal shape of the malformation; it has its own distinct features and its own relevance to surgical repair.

The structure of these three types of pulmonary stenosis can be identified by angiography: (a) doming of the valve (fig 2) is the usual finding in pulmonary stenosis; (b) filling defects at the valve level (fig 3) as well as irregular thickening of the cusps are highly suggestive of dysplastic pulmonary valve with cauliflower deformation of the cusps, while (c) narrowing at the distal cusp level with deep

Fig 4 (a) Photograph showing hour-glass deformity of the pulmonary valve. The commissures are not fused. The orifices of the three sinuses are very narrow (bottle-necks). Distal to the commissure-orifice plane, there is considerable post-stenotic dilatation of the pulmonary artery. (b) Angiogram (lateral view) of end systole showing the hour-glass deformity of the pulmonary artery and the bottle-shaped sinuses. The filling defects at the valve level indicate smooth cusps. There is considerable post-stenotic dilatation of the pulmonary artery.

“bottling” of the sinuses (fig 4) indicates hour-glass pulmonary stenosis.

Radiographic and direct surgical examination showed various degrees of post-stenotic dilatation of the pulmonary artery in all types of pulmonary stenosis. We found that the degree of post-stenotic dilatation depended not on the type of valve obstruction, but rather on its duration and severity. Others, however, found post-stenotic dilatation only with dome-shaped valves.^{1 4 12}

Preoperative diagnosis of the type of pulmonary valve stenosis is important. Only the most common type, the dome-shaped valve, is structurally amenable to simple balloon dilatation or to the double balloon technique recently reported by Al Kasab *et al*,¹³ which eliminate the need for surgery. Although it may seem reasonable to assume that, as with surgical intervention, balloon dilatation results in valvotomy along the fused commissures, this has not

yet been shown to be the case. Unfortunately, the balloon technique is less satisfactory when combined infundibular and pulmonary valve stenosis are present¹⁴ or when the valve is either of the cauliflower^{15 16} or of the hour-glass type, because fused commissures are not the major problem in these two types. Patients with dysplastic cauliflower valves are most likely to benefit from excision of the obstructive cusps. In patients with the hour-glass valves excision of the cusps frees the constriction of the sinuses and allows the valve to dilate to nearly its physiological diameter at the commissural level thus eliminating any need for transvalvar patching.

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