Obstruction of the central pulmonary artery after shunt operations in patients with pulmonary atresia

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SUMMARY The central pulmonary artery was studied angiographically 1-10 years after a palliative operation in 56 patients (mean age 6 years, range 1-20) with congenital heart disease and pulmonary atresia. All of them had originally had a central pulmonary artery that was common to both the right and left pulmonary arteries. Juxtaductal obstruction of the left pulmonary artery had developed in 70% of the patients; in 40% the obstruction was atretic unilaterally. Atresia developed at the right junction to the ductus where there was a shunt to the right pulmonary artery and at the left junction where there was a shunt to the left pulmonary artery. Stenosis causing a reduction of more than 50% of the inner diameter developed at the anastomotic site in 10% of 30 patients with a functioning Blalock shunt. In contrast, there was occlusion of the pulmonary artery or its upper branch in 73% of 11 patients with a non-functioning anastomosis.

Surgical techniques such as the Rastelli^{1,2} or the Fontan procedure^{3,4} have been developed to treat patients with congenital heart disease, pulmonary atresia, and a central pulmonary artery that is common to both the right and left pulmonary arteries. Palliative operation may be important when further intracardiac repair is planned. We found that juxtaductal obstruction of the left pulmonary artery developed in 67% of 21 patients with pulmonary atresia without a preceding operation.⁵ In patients who have had a palliative operation additional stenosis may develop in the central pulmonary artery at the point of surgical intervention. The development and clinical application of our new preformed catheter for selective angiography of the central pulmonary artery in 1981⁶ means that atresia of the central pulmonary artery can be visualised more clearly than before. We have investigated obstruction of the central pulmonary artery developing after palliative operation in patients with pulmonary atresia.

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Patients and methods

From November 1981 to November 1984, 56 consecutive patients with congenital heart disease and pulmonary atresia were admitted to our institute for cardiac catheterisation 1-10 years after palliative operation. At preoperative aortography contrast medium passed into the central pulmonary artery through the ductus arteriosus. The precise arrangement of juxtaductal pulmonary arteries was not displayed before operation because selective angiography was not available. During the same period 21 similar patients who had not had a palliative operation were studied by selective pulmonary angiography. These were the subject of another study of the course of juxtaductal obstruction of the left pulmonary artery in pulmonary atresia not treated by palliative operation.⁵

We reviewed angiograms and surgical records of the 56 patients who had had a palliative operation. Intracardiac anatomy was diagnosed by cross sectional echocardiography and ventriculography. Table 1 lists the previous operations and intracardiac diagnosis in these 56 patients (aged 1 to 20 years, mean 6). The interval between the previous operation and the present study ranged from one to 10 years (mean 4). Patients with bilateral ductus

Table 1 Intracardiac anatomy and previous operations in 56 patients with pulmonary atresia

Intracardiac anomaly	No operation	Palliative operation				Total
		Left Blalock	Right Blalock	Brock	Others	
Fallot's tetralogy	11	11	4	1	8	35
Intact ventricular septum	2	2		2		6
Tricuspid atresia	1	2				3
Univentricular heart	2	4	4		2	12
Ventriculoarterial discordance	3	6	2		2	13
Atrioventricular and ventriculoarterial discordance	1	3	3			7
Atrioventricular septal defect	1					1
Total	21	28	13	3	12	77

arteriosus or major aortopulmonary collateral arteries⁷ were excluded from this study.

Angiography of the central pulmonary artery was performed as follows. After ventriculography or aortography, the ductus arteriosus or the surgically created anastomosis was catheterised and selective pulmonary angiography was performed with biplane cinefilm or cut-film equipment. Since November 1981 we have successfully used a preformed catheter⁶ for selective injection of contrast medium through the ductus. Pulmonary vein wedge angiography⁸⁻¹⁰ was performed to visualise occult pulmonary arteries.

In this article we use the term central pulmonary

artery to describe the pulmonary trunk (main pulmonary artery) and the right and the left pulmonary arteries proximal to the hilar portion. In three patients with situs inversus the terms right pulmonary artery or left pulmonary artery relate to the morphology of the vessels. Pulmonary artery stenosis was defined as a reduction of the inner diameter to less than 50% of its largest diameter. The term atresia was applied to obstructions where angiograms showed a sharply demarcated obstruction without wash out of contrast (figs 1 and 2). When the pulmonary artery was generally smaller than normal, this was termed hypoplasia. Two types of pulmonary atresia were identified on the angiograms. In

Fig 1 Pulmonary arteriograms of a two year old boy with tetralogy of Fallot, pulmonary valve atresia, ductus arteriosus, and a left Blalock shunt. Injection at the orifice of the anastomosis showed the left pulmonary artery with an atretic junction to the ductus (a). Selective injection through a preformed catheter into the ductus visualised only the right pulmonary artery and a well developed pulmonary trunk ((b), frontal; (c), lateral). Ao, aorta; RPA, right pulmonary artery; PT, pulmonary trunk; BT, Blalock-Taussig shunt.

Fig 2 Pulmonary arteriograms of a 10 year old girl with tetralogy of Fallot, pulmonary trunk atresia, ductus arteriosus, and a right modified Blalock shunt. Injection into the anastomosis visualised the right pulmonary artery and pulmonary trunk ((a), frontal; (b), lateral). Juxtaductal atresia in the left pulmonary artery was diagnosed on the basis of this angiogram. Injection into the ductus, which visualised only the left pulmonary artery (not shown), confirmed the diagnosis. See legend to fig 1 for abbreviations.

pulmonary valve atresia the pulmonary trunk was long and tubular and sinuses of Valsalva were seen at its anterior end (fig 1). In truncal atresia, the pulmonary trunk was short and cone shaped (fig 2). The ductus was patent at catheterisation in 40 of the 56 patients. The ductus arteriosus was left sided in all 40 patients and originated from the left innominate artery in the 10 patients with right aortic arch.

STATISTICAL ANALYSIS

Differences in proportions were tested by the χ^2 test¹¹ and a p value of 0.05 was regarded as significant.

Results

Stenosis or atresia in the central pulmonary artery developed either at the junction of the ductus (juxtaductal obstruction⁵) or at the site of the surgical shunt. We did not find stenosis or atresia at any other part of the central pulmonary artery other than the point of surgical intervention. The influence of each surgical intervention on the development of the obstruction in the central pulmonary artery was

different and we give the results for each surgical group.

LEFT BLALOCK SHUNT

This group of 28 patients includes those with the original Blalock shunt and a modified Blalock shunt with a polytetrafluoroethylene (PTFE) graft. We found eight patterns of ductus arteriosus and obstructions in the central pulmonary artery (fig 3). Only six (21%) of 28 did not have any obstruction in the central pulmonary artery, 79% had juxtaductal obstruction—in 12 (43%) patients this was atretic. In all but one of these 12 patients juxtaductal atresia was left sided. In this exceptional case the left Blalock shunt was becoming occluded and blood flow through the shunt was much reduced. Blood flow to the left pulmonary artery was maintained from the ductus, and juxtaductal atresia developed in the right side as it did in those patients without previous shunt operation.⁵

In two patients the anastomosis was occluded and the left pulmonary artery was not visualised by contrast medium coming through either the ductus or the anastomosis. Pulmonary vein wedge angio-

Fig 3 Patterns of the central pulmonary artery after a left Blalock-Taussig (BT) shunt in 28 patients with congenital heart disease and pulmonary atresia in whom the central pulmonary artery had originally been common to both the right and left pulmonary arteries. Numbers of patients and their ages are given. A, aorta; PA, pulmonary artery; PDA, ductus arteriosus; LPA, left pulmonary artery.

Fig 4 Patterns of the central pulmonary artery after a right Blalock-Taussig (BT) shunt in 13 patients with congenital heart disease and pulmonary atresia in whom the central pulmonary artery had originally been common to both the right and left pulmonary arteries. Numbers of patients and their ages are given. See legend to fig 3 for abbreviations.

Fig 5 Frequency of occlusion of the pulmonary artery or its branches at the anastomotic site of Blalock shunt.

graphy confirmed the patency of the peripheral left pulmonary artery in both patients.

RIGHT BLALOCK SHUNT

There were 13 patients in this group and six patterns of arrangement of the ductus and obstructions in the central pulmonary artery (fig 4). Eight (62%) had juxtaductal obstruction and six of these had atresia. Juxtaductal atresia of the left pulmonary artery always developed to the right of the ductus in this group. A closed ductus with juxtaductal obstruction in the left pulmonary artery was found in only one

patient. No patient showed significant obstruction of the right pulmonary artery at the anastomosis.

Figure 5 summarises the type of obstruction that we found in the pulmonary artery at the point of anastomosis in these two groups of patients with Blalock shunts. At the junction of the anastomosis the contour of wall of the pulmonary artery was uneven and the inner diameter was often reduced slightly (fig 2). Stenosis or occlusion developed in only three out of 30 patients with functioning subclavian to pulmonary artery anastomosis, however. In the 11 patients with non-functioning Blalock shunt, there was obstruction of the pulmonary artery or its upper branch (fig 6) in eight (73%) patients. In the remaining three patients we could not visualise this part of the pulmonary artery.

WATERSTON SHUNT

We studied six patients with a Waterston shunt. Three had associated ductus arteriosus and two had a left Blalock shunt. One had had bilateral non-functioning Blalock shunt before Waterston shunt was done. Six patterns of the central pulmonary artery were observed (fig 7). Juxtaductal obstruction of the left pulmonary artery was present in all six,

Fig 6 Pulmonary arteriograms of a seven year old girl with ventriculoarterial discordance, right aortic arch, pulmonary trunk (PT) atresia, ductus arteriosus (PDA), and a non-functioning right Blalock shunt. Injection into the ductus visualised the central pulmonary artery except for its right upper branch which was atretic at the site of the occluded Blalock shunt. (a), frontal; (b), lateral.

Fig 7 Patterns of the central pulmonary artery after a Waterston shunt in six patients with congenital heart disease and pulmonary atresia in whom the pulmonary artery had originally been confluent. Numbers of patients and their ages are given. See legend to fig 3 for abbreviations. LBT, left Blalock-Taussig.

Fig 8 Pulmonary arteriograms of an eight year old boy with tetralogy of Fallot, pulmonary atresia, non-functioning Blalock shunts, and functioning Waterston shunt. Selective injection into the right pulmonary artery showed occlusion of the upper branch (a). Injection into the right subclavian artery showed well developed collateral vessels and occluded anastomosis (b). Injection into the wedge position of the upper pulmonary vein (PVW) with a transarterial catheter showed atresia of the upper branch of the right pulmonary artery (c), followed by extravasation into the sputum and the appearance of contrast in the bronchi a few seconds later (d).

Fig 9 Patterns of the central pulmonary artery after Brock operation or central shunt in four patients with congenital heart disease and pulmonary atresia in whom the central pulmonary artery had originally been common to both the right and left pulmonary arteries. Numbers of patients and their ages are given. See legend to fig 3 for abbreviations.

and in three of these there was atresia. At the anastomosis the proximal right pulmonary artery was obstructed completely in three patients (fig 8), and partially in the other three patients.

A complication of pulmonary vein wedge angiography occurred in an eight year old boy with tetralogy of Fallot, pulmonary atresia, bilateral non-functioning Blalock shunts, and a functioning Waterston shunt. Selective right pulmonary arteriography showed complete occlusion of the upper branch and occlusion at the proximal right pulmonary artery. Power injection into the right upper pulmonary vein was associated with intra-

Fig 10 Frequency of juxtaductal obstructions of the left pulmonary artery in each surgical group and the group not treated by operation. LBT, left Blalock shunt; RBT, right Blalock shunt.

Table 2 Numbers and percentages of juxtaductal obstructions in each type of pulmonary atresia in 56 patients 1-10 years after palliative operation

Juxtaductal obstruction	Pulmonary valve atresia (%)	Pulmonary trunk atresia (%)	Type not diagnosed (%)
No obstruction	10 (48)	5 (28)	0
Stenosis	9 (43)	7 (39)	3 (18)
Atresia	2 (9)	6 (33)	14 (82)
Total	21 (100)	18 (100)	17 (100)

pulmonary injection, the appearance of contrast in the bronchi, and bloody sputum. Blood disappeared from the sputum within 24 hours and he recovered without any noticeable residual lesion.

BROCK OPERATION AND CENTRAL SHUNT

We studied three patients with Brock operation and another patient with a central shunt (ascending aortopulmonary artery graft). Figure 9 shows the four patterns of the central pulmonary artery that we found in these patients. Two had juxtaductal stenosis in the left pulmonary artery.

Figure 10 shows the frequencies of juxtaductal stenosis and atresia in each surgical group. We compared these results with data collected from a group of 21 patients who had not had a palliative operation. Juxtaductal stenosis or atresia developed in every group. It was milder in Brock's operation and central shunt, but the small number of patients in this group precludes any definite conclusion.

Table 2 shows how juxtaductal obstruction in these 56 patients correlated with the type of pulmonary atresia. In 17 patients the pulmonary trunk could not be visualised and the type of pulmonary atresia remained unknown. Atresia of the pulmonary valve was associated with less frequent and milder juxtaductal obstruction of the left pulmonary artery than atresia of the pulmonary trunk.

Discussion

In this study and an earlier one,⁵ juxtaductal obstruction in the left pulmonary artery was the most common and most major obstruction found in the central pulmonary artery in patients with congenital heart disease and pulmonary atresia in whom the central pulmonary artery had been common to both the right and left pulmonary arteries. This obstruction, however, was modified by the preceding palliative operation. In those patients who had not had a palliative operation⁵ with a right Blalock or Waterston shunt, juxtaductal atresia developed to the right of the ductus. In patients with left Blalock shunt, juxtaductal atresia developed on the left side of the ductal junction. In patients who had

not had a palliative operation juxtaductal obstruction developed exclusively in those with truncal atresia; no patient with an atretic pulmonary valve had juxtaductal obstruction.⁵ This was not true of the 56 patients reported in the present study.

Progressive juxtaductal obstruction was common both in patients with a shunt operation and in those without.⁵ Angiography showed a cone-shaped stenosis on both sides of the junction of the ductus and in the left pulmonary artery. In those without shunts this stenosis progressed to atresia after the age of four.⁵ In some of those with shunts, however, juxtaductal atresia developed at the age of two or three years. This suggests that a shunt operation may accelerate juxtaductal obstruction of the left pulmonary artery.

In patients with an anastomosis between the left subclavian artery and pulmonary artery, atresia was more common at the left junction. Juxtaductal atresia developed at the left junction in all four patients who had both a ductus arteriosus and had had a well functioning left Blalock shunt for more than a year. The mechanism that causes the development of atresia at the left junction in this particular group is not clear. Presumably, blood flow became turbulent at this point after the establishment of a shunt to the left pulmonary artery and this caused a jet lesion to develop there. The same mechanism could account for the development of atresia at the right junction to the ductus after a shunt to the right pulmonary artery. This mechanism could also explain why juxtaductal obstruction was mild in patients after a Brock operation or central shunt.

We showed a clear correlation between the patency of the anastomosis between the subclavian artery and pulmonary artery and occlusion of the pulmonary artery at the anastomotic site. Significant occlusion at the anastomotic site rarely developed in the pulmonary artery while the anastomosis was functioning, whereas when the anastomosis was not functioning there was invariably occlusion of the pulmonary artery or its branch at this site. This emphasises the importance of a good anastomosis in patients with pulmonary atresia in whom further intracardiac repair is planned.

In patients with atresia of a pulmonary artery at the site of a non-functioning anastomosis, the patency of the peripheral pulmonary artery can be shown by pulmonary vein wedge angiography.⁸⁻¹⁰ This technique is not without risk, however. Pulmonary vein wedge angiography should be tried only after careful selection of the injection site, dose of contrast material, and injection speed. We recommend a hand injection of contrast material to visualise an occult upper branch of a pulmonary artery. In

patients with one occult pulmonary artery, we recommend injection into the pulmonary vein of the middle or lower lobe. Power injection should only be used after hand injection of a small amount of contrast material has established the presence of a patent peripheral portion of the pulmonary artery.

A Waterston shunt is often associated with occlusion of the right pulmonary artery at a point proximal to the anastomosis.^{12 13} This was confirmed in the present study. Obstructive lesions of this type will require careful treatment by angioplasty at intracardiac repair. Occlusion of the right pulmonary artery proximal to the anastomosis also curtails the supply of blood shunted to the left pulmonary artery and this may inhibit development of the vascular bed in the left lung. These aspects should be considered when palliative operations are selected for patients in whom intracardiac repair is planned.

Brock's operation, if technically feasible, is probably the most suitable operation for patients with atresia of the pulmonary valve. A Blalock shunt or modified Blalock shunt is the standard operation for patients with atresia of the pulmonary trunk. There is no known method of preventing the development of juxtaductal obstruction in the left pulmonary artery; however, juxtaductal obstruction in the left pulmonary artery was generally mild in those patients with a spontaneously closed ductus arteriosus. Ligation of the ductus at palliative operation may prevent progress of juxtaductal obstruction in the left pulmonary artery in the neonatal period.

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