Pulmonary artery stenosis after systemic-to-pulmonary shunt operations

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Received 14 April 1998; revised version received 9 June 1998; accepted 16 June 1998

Abstract

Objective: Systemic-to-pulmonary shunt operations are still required for palliation of certain congenital heart defects. The aim of this study was to analyze the incidence and etiology of the development of pulmonary artery stenosis after these procedures. Methods and results: Pre- and post-operative angiograms of 59 patients who underwent 54 peripheral and 12 central shunt operations were analyzed retrospectively. Patients without prior cardiovascular interventions (group I, n = 47) were differentiated from patients with prior interventions (group II, n = 12). In group I, all peripheral shunts were inserted contralaterally to the ductus arteriosus. Follow-up for all patients was 1.8 years (4 days–7.8 years). Pulmonary artery stenosis was diagnosed in 12/59 patients (20.3%, group I 12/40 = 30%). The stenoses were located ipsilaterally to the shunt in 7/12 and contralaterally in 5/12. Statistical analysis did not show any impact of age, weight, sex, shunt type or size, pulmonary artery diameters, Nakata and McGoon indices and prior interventions on the development of pulmonary artery stenosis. However, a patent ductus arteriosus and administration of Prostaglandin E1 had a significant impact on the development of pulmonary artery stenosis on the side of the ductus arteriosus. Conclusion: Pulmonary artery stenosis is not a rare event after systemic-to-pulmonary shunt operations. A patent ductus arteriosus with or without administration of Prostaglandin E1 is related to pulmonary artery stenosis on the side of the ductus arteriosus. Surgical technique, increased intimal proliferation, or pulmonary artery kinking. Treatment depends on severity of cyanosis and on further surgical plans. © 1998 Elsevier Science B.V. All rights reserved

Keywords: Systemic-to-pulmonary shunt operation; Congenital heart disease; Pulmonary artery stenosis; Heart surgery

1. Introduction

Surgical correction of congenital heart disease is currently performed as early as possible in life. Yet, systemic-to-pulmonary shunt operations are still required for palliation of some congenital heart malformations. One complication of these techniques is the development of pulmonary artery stenosis (PAS). Although systemic-to-pulmonary shunt operations have been performed for decades, the incidence and etiology of this event has been rarely discussed [1,2]. Recently, additional spontaneous acquisition of discontinuity of pulmonary arteries at the site of the ductus arteriosus has been described [3]. To assess the incidence and etiology of PAS after systemic-to-pulmonary shunt operations, a retrospective analysis of patients with classical or modified Blalock–Tausig shunts and central aorto-pulmonary shunts was performed.

2. Material and methods

Since 1986, a total number of 77 patients underwent 89 systemic-to-pulmonary shunt operations at our institution. Fifty-nine of those had both, pre- and post-operative angiograms and were included in this study. They underwent 66
systemic-to-pulmonary shunt operations. Fifty-seven of those had reduced pulmonary blood flow due to pulmonary atresia (n = 30), pulmonary stenosis (n = 25) or pulmonary artery banding (n = 2). In the remaining two patients the shunt procedure was performed in combination with a Damus–Kay–Stansel operation. Pathologic anatomical diagnoses were univentricular heart (n = 20), tetralogy of Fallot (n = 14), pulmonary atresia with ventricular septum defect (n = 8) or with intact ventricular septum (n = 5), double outlet right ventricle (n = 6), truncus arteriosus communis (n = 1), transposition of the great arteries (n = 1), congenitally corrected transposition of the great arteries (n = 1) and other complex cardiac malformations (n = 3). The pre-operative and post-operative cineangiograms were analyzed to detect PAS on either side. The pulmonary artery diameters were determined and Nakata as well as McGoon indices were calculated.

The technique of classical or modified Blalock–Taussig shunt construction may influence the development of acquired PAS, therefore our operative procedure is described below.

Surgical access is obtained by a lateral thoracotomy in the bed of the fourth rib. The subclavian artery is dissected and an elastic loop is placed around it without grasping the vessel. In classical Blalock–Taussig shunts the first branches are ligated and divided. The subclavian artery is ligated, divided beyond these branches, and mobilized to its origin.

Subsequently, the pulmonary artery is dissected from the upper lobe artery to its central origin. Elastic loops are placed centrally around the pulmonary artery and around its major branches without grasping of the vessels. Opening of the pericardium is avoided if possible, as entering of blood my cause intrapericardial adhesions. Ligation of the ayzygos vein is rarely necessary.

Systemic heparinization (1 mg/kg) is performed. In modified Blalock–Taussig shunts the length of the graft is estimated. The pulmonary end of the graft is cut obliquely, taking the course of the subclavian artery into account. Usually, a shunt diameter of 5 mm is taken; a 4 mm graft is used rarely if very small vessels are present, 6 mm grafts are preferred for older infants.

A delicate side-biting clamp is placed on the subclavian artery without distorsion. The artery is incised longitudinally. The anastomosis is made with a running 6–0 polypropylene suture, the clamp on the graft is opened temporarily for de-airing. The anastomosis is completed, the loops are cut and the clamps are removed. It is ascertained that a palpable continuous thrill in the pulmonary artery and shunt is present. Heparin is antagonized.

A pulmonary artery stenosis was defined as a reduction in diameter to less than 50% in angiography or requiring urgent, intermediate, or long-term intervention. The calculation was based on the diameter of the ipsilateral pulmonary artery on the site of the branching to the upper lobe. By this method, miscalculation due to general hypoplasia was avoided. Patients with stenosis of less than 50%, who did not require any intervention, were excluded.

To assess the impact of other previous surgical and non-surgical interventions on the development of PAS, the patients were divided into two groups. Patients of group I (n = 47) had no previous intervention on the cardiovascular system and had their first shunt operation in our institution. Patients with previous systemic-to-pulmonary shunt operation (n = 6), Glenn-anastomosis (n = 1), pulmonary valve balloon dilatation (n = 1), pulmonary artery banding (n = 3), or repair of aortic coarctation (CoA) (n = 1) who underwent subsequent additional systemic-to-pulmonary shunt operations were defined as group II (n = 12).

The mean follow-up time for all patients was 1.8 ± 1.5 years (range: 4 days–7.8 years). Group I was followed up for 1.9 ± 1.3 years (range: 4 days–5.4 years) and group II for 1.6 ± 2.3 years (9 days–7.8 years) (ns).

The mean age of patients in group I (n = 47) at the time of operation was 5.8 ± 11.7 months (range: 4 days–5.5 years); 42 patients were under one year of age, including 17 neo-nates. The mean weight was 4.9 ± 2.8 kg (range: 2.2–14.5 kg). The ductus arteriosus was patent (PDA) in 25 patients, Prostaglandin (PGE1 ) had been given in 21 patients A modified Blalock–Taussig shunt operation was performed in 37 cases (PTFE graft sizes: 4 mm = 2, 5 mm = 28, 6 mm = 7), a classical Blalock–Taussig shunt in three and a central aorto-pulmonary shunt in seven cases (graft-sizes: 5 mm = 6, 1 Waterston–Cooley-anastomosis). All Blalock–Taussig shunts were placed contralaterally to the ductus arteriosus.

Six of these 47 patients required a second shunt operation after a mean time interval of 1.0 year (range 7 days–3.9 years) due to shunt stenosis (n = 3), to increased cyanosis because of patient growth (n = 2), and to PAS at the site of the shunt (n = 1). One patient developed shunt stenosis after the second shunt and needed a third shunt operation 28 days later.

At the time of operation, patients of group II (n = 12) were significantly older (4.8 ± 5.0 years; range: 6.8 months–18.6 years; P ≤ 0.001) and had a significantly higher body weight (16.4 ± 12.5 kg; range: 6.2–48.9 kg; P ≤ 0.001) in comparison to patients of group I. The ductus arteriosus was closed in all cases. In this group a modified Blalock–Taussig shunt was performed in two cases (graft...
sizes: 6 mm = 2), a classical Blalock–Taussig shunt in one and a central aorto-pulmonary shunt in nine cases (graft sizes: 5 mm = 4, 6 mm = 2, 8 mm = 1, without graft material = 2). Thus, in group II, the number of patients with a central aorto-pulmonary shunt (75%) was significantly higher compared to patients of group I (14.9%) (P ≤ 0.001). Pre-operative pulmonary artery diameters and indices in patients of group I and II are listed in Table 1.

Statistical analysis was performed with the Statistical Package for Social Sciences (SPSS, Chicago, IL). Data were expressed, where appropriate, as mean ± SD and range. Differences between two groups were assessed using the χ² and Mann-Whitney U-Wilcoxon rank sum test. Multivariate analysis was performed using logistic regression. A P-value of less than 0.05 was considered significant.

3. Results

Twelve of 59 patients (20.3%) developed stenosis (n = 9) or discontinuity (n = 3) of the pulmonary arteries after 66 (18.2%) systemic-to-pulmonary shunt operations. Pre- and post-operative data are listed in Table 2. This observation was only made in patients of group I (12/47 = 25.5%) and particularly only after Blalock–Taussig shunts (one classical, 11 modified) operations (12/40 = 30%).

The diagnosis of PAS was made 2.2 ± 1.5 years (range: 4 days–5.3 years) after the shunt operation. In nine patients the stenosis was discovered during a routine cardiac catheterization 1–5.3 years after the palliation. The other three patients showed continuous decrease of oxygen saturation and subsequently had cardiac angiograms 4 days, 6 months and 3.7 years after the shunt operation.

The stenoses were localized ipsilaterally to the Blalock–Taussig shunt (Fig. 1) in seven patients (14.9%) and contralaterally to the Blalock–Taussig shunt on the site of the ductus arteriosus (Fig. 2) in five patients (10.7%) of group I. Whereas ipsilateral PAS is related to a Blalock–Taussig shunt, contralateral PAS can occur in central and peripheral shunts. Although none of the seven patients with a central systemic-to-pulmonary shunt in group I developed PAS, the incidence of contralateral PAS was not significantly higher for patients with Blalock–Taussig shunts (P = 0.322). Taking all patients of group I and II into account, the incidence was not significantly increased, either (P = 0.154).

The ductus arteriosus had been patent in eight of the twelve patients with PAS and in all five cases with stenosis at the site of the ductus arteriosus. Four of these five patients had had administration of PGE1 for 2.6 ± 1.8 months (range: 6 days–4.2 months) whereas only one of the seven patients with PAS on the ipsilateral site had had PGE1 (12 days) pre-operatively. Univariate statistical analysis revealed a significant impact of a PDA (P = 0.038) and administration of PGE1 (P = 0.023) on the development of PAS on the site of the ductus arteriosus.

Pre-operative cardiac catheterization had shown normal pulmonary arteries in seven of the twelve patients with PAS, and hypoplastic pulmonary arteries without localized stenosis in the remaining five. Statistical analysis did not reveal any impact of hypoplasia of the pulmonary arteries on the development of PAS on either side (P = 0.368). Pulmonary artery diameter on the side of a Blalock–Taussig shunt had no impact on the development of ipsilateral PAS (P = 0.164). Additionally, the diameter of the pulmonary artery on the side of the ductus in patients with a PDA and administration of PGE1 had no influence on the development of contralateral PAS (P = 0.607). Except for PDA and PGE1, no other investigated parameter, including diagnosis, sex, and tube graft size, had a statistical impact on the development of this complication (Table 3).

The clinical picture of patients with PAS varies strongly. Although symptoms may be severe, in our series only one patient needed emergency pulmonary artery balloon dilatation 4 days after the shunt procedure. In nine patients, the PAS was asymptomatic and was discovered in a routine angiogram. In addition to the emergency pulmonary artery balloon dilatation in one patient, the PAS was surgically treated in eight patients during the correction operation. One patient underwent a second shunt operation with reconstruction of the stenosed pulmonary artery and two patients are clinically stable and not yet corrected.

4. Discussion

Congenital obstructive malformations of the pulmonary artery pathways in patients with stenosis or atresia of the right ventricular outflow tract have been described and investigated in detail [4,5]. Despite the fact that systemic-to-pulmonary shunt operations have been performed frequently in the past and are still required for palliation of some malformations, the incidence and etiology of post-operatively acquired stenosis or discontinuity of the pulmonary arteries have been discussed rarely [1–3]. Odim and colleagues [1] report an incidence of 16% of pulmonary

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Group I (n = 47)</th>
<th>Group II (n = 12)</th>
<th>P-value</th>
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</thead>
<tbody>
<tr>
<td>Mean ± SD (range)</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>L-PA diameter (mm)</td>
<td>4.9 ± 2.0</td>
<td>8.1 ± 2.5</td>
<td>0.0002</td>
</tr>
<tr>
<td>R-PA diameter (mm)</td>
<td>5.3 ± 2.5</td>
<td>7.4 ± 2.7</td>
<td>0.0165</td>
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<tr>
<td>McGoon index</td>
<td>1.6 ± 0.4</td>
<td>1.8 ± 0.7</td>
<td>0.3637</td>
</tr>
<tr>
<td>Nakata index</td>
<td>169 ± 93</td>
<td>217 ± 158</td>
<td>0.6258</td>
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artery distortion after insertion of a modified Blalock–Taussig shunt via thoracotomy and a 10% incidence via sternotomy approach. Tamisier and colleagues [2] found this complication in 19% of patients after Blalock–Taussig shunt operation via thoracotomy in infants under 3 months of age. Shimazaki and colleagues [5] reported an incidence of 23% of PAS in patients with tetralogy of Fallot, who had undergone a previous palliative operation. Recently, Waldman and colleagues [3] retrospectively investigated 52 patients with congenital pulmonary atresia who had undergone systemic-to-pulmonary shunt operations and found acquired pulmonary artery discontinuity or stenosis in 15 cases (29%). In the present study, twelve of 59 (20.3%) patients developed post-operative stenosis (15.2%) or discontinuity (5.1%) of the pulmonary arteries. Considering the 47 patients who underwent their first shunt operation, the incidence increases to 25.5%. Thus, PAS after systemic-to-pulmonary shunt operations is not a rare event and deserves further investigation.

Contralateral stenoses were clearly related to a PDA or the administration of PGE1. This phenomenon was first described in 1996 by Waldman and colleagues [3] and was attributed to abnormal extension of ductile tissue beyond the ductus itself, and damage of the ductile tissue by PGE1 with a potential for acquired obstruction. Early post-operative PAS seems to be related to active contraction of the ductile tissue and delayed PAS to progressive edema, fibrosis, and calcification of the ductile tissue [6] with intimal proliferation leading to fibrocellular hypertrophy [7]. Based on the observation that none of the patients with a central systemic-to-pulmonary shunt developed PAS on the side of the ductus, it was assumed that this type of shunt may offer hemodynamic advantages preventing PAS. However, this complication was described in other series in patients with central shunts [3]. Besides a PDA and administration of PGE1, none of the other clinically, and angiographically, obtained parameters had a significant impact on the development of contralateral PAS.

In contrast to this, the pathogenesis of ipsilaterally located PAS has to be discussed under different aspects.
Firstly, inappropriate surgical technique may lead to pulmonary artery distortion [1,2], stenosis of the shunt-to-pulmonary artery anastomosis, thrombosis due to intimal lesions caused by clamp injuries, or kinking of the pulmonary artery particularly if the shunt length was estimated too short. In modified Blalock–Taussig shunts, the sternotomy approach is discussed as being associated with fewer shunt failures than the thoracotomy approach, being technically more challenging [1]. Probably, this approach reduces acquired PAS in infants with small pulmonary arteries. A potential disadvantage of sternotomy is the presence of adhesions in sternal re-entry for subsequent procedures. Secondly, extensive intimal proliferation due to altered hemodynamics [7] is discussed as an independent factor which applies to both peripheral and central shunts. Thirdly, kinking of the pulmonary artery after a long duration has to be attributed to the growth of the child. Thus, the development of ipsilateral PAS seems to have various mechanisms.

In the present series, none of the investigated parameters turned out to be a risk factor for the development of ipsilateral PAS; particularly neither pulmonary artery diameters nor McGoon and Nakata indices had a significant impact. Ipsilateral PAS did not occur in patients of group II; this may be related to the small number of patients (n = 3) with Blalock–Taussig shunts.

We assumed that previous interventions on the cardiovascular system may increase the risk of PAS by leading to a modified anatomic situs and post-operative adhesions. Our results do not confirm this theory; patients of group II did not show a higher incidence of PAS.

In contrast to our findings, in the study of Waldman and colleagues [3] 12 of 15 patients (80%) were symptomatic. Consequently, in his series, diagnosis of this complication was made earlier (4 days–1.5 years after the shunt procedure). He describes two different patterns of clinical symptoms: progressive decrease in oxygen saturation and lung hypoperfusion in some cases within days after the operation, or gradual decrease in oxygen saturation over months.

The treatment of PAS depends on the severity and the time of onset of symptoms as well as on the planned future procedures for a patient. In the case of an early onset of symptoms (within hours or days), emergency angiography is usually advisable. If a ductus-related PAS is detected, pulmonary blood flow can occasionally be partially restored by administration of PGE1 in patients who received this drug pre-operatively [3]. Alternatively, pulmonary balloon dilatation or stent implantation can be tried. This therapy applies to any kind of symptomatic ipsilateral pulmonary artery stenosis. However, it is generally not recommended in infants and small children because of concerns with limited stent redilatation when patients grow, and with surgical treatment on stented pulmonary arteries. Yet, if this complication is accompanied by a significant shunt stenosis, shunt revision is the treatment of choice. In the case of late onset of symptoms, correction of the basic lesion has to be discussed. In asymptomatic patients, usually no immediate intervention is necessary unless the PAS interferes with planned operative procedures (e.g. Fontan). In these cases, dilatation with or without stenting or another shunt implantation might be indicated to improve the anatomic situation before correction.

In conclusion, PAS appears to be a frequent event after systemic-to-pulmonary shunt operations. The presence of a PDA and the administration of PGE1 has an impact on the development of PAS on the side of the ductus arteriosus. PAS ipsilateral to a Blalock–Taussig shunt may be due to...
inappropriate surgical technique, increased intimal proliferation, or pulmonary artery kinking due to patient growth. Previous surgical interventions do not have an impact on the development of PAS. Treatment of PAS depends on the severity of the symptoms and on further surgical plans with regard to the basic malformation.

References