AUGMENTATION OF PULMONARY BLOOD FLOW WITH AN AXILLARY ARTERIOVENOUS FISTULA AFTER A CAVOPULMONARY SHUNT

Of 320 children with cyanotic congenital heart malformations who had previously undergone cavopulmonary shunt operations, 11 had increasing cyanosis and exercise intolerance and were considered unsuitable for definitive repair, a Fontan procedure, or other palliation. Eight had a previous Glenn shunt and three had a previous bidirectional cavopulmonary connection. To augment pulmonary blood flow, 10 patients underwent creation of an ipsilateral axillary arteriovenous fistula. Mean oxygen saturations were $80\% \pm 2\%$ before operation, $85\% \pm 2\%$ immediately after operation, and $84\% \pm 3\%$ at a mean follow-up interval of 7.4 years (range 0.1 to 15.5 years). Mean hemoglobin values were 202 ± 10 gm/L before operation, 177 ± 10 gm/L after operation, and 191 ± 14 gm/L at latest review. The only complication was mild swelling of the arm distal to the fistula in one patient. All patients reported improvement in exercise tolerance. Eight patients have continued evidence of fistula patency. Development of ipsilateral pulmonary arteriovenous fistulas has not been observed in any patient. Creation of an axillary arteriovenous fistula to augment pulmonary blood flow after a cavopulmonary shunt provides useful palliation for complex cyanotic heart disease when other options are limited. Such additional sources of pulmonary blood flow may influence the development of pulmonary arteriovenous fistulas. (J THORAC CARDIOVASC SURG 1996;111:176-80)

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Experimental bypass of the right side of the heart was initially performed by Carlon, Mondini, and de Marchi¹ in 1951. Further animal studies were performed by Glenn,² and in 1958 he reported the first clinical application of this concept, with a superior vena cava-right pulmonary artery end-to-end anastomosis bypassing the heart. Widely applied as palliation for a number of complex cardiac lesions not amenable to biventricular repair, the Glenn shunt has not remained the sole palliative treatment; for example, many lesions are now treatable with the more extended

total caval shunt proposed by Fontan and Baudet.³ More recently, an end-to-side superior vena cava-right pulmonary artery anastomosis (bidirectional cavopulmonary connection) has become a component of various treatment algorithms, allowing bilateral pulmonary flow, avoiding isolation of the right pulmonary artery,⁴ and acting as a preparatory step toward a Fontan procedure.⁵ In some patients, who have undergone previous Glenn or bidirectional cavopulmonary anastomoses, the immediate surgical mortality rate associated with definitive biventricular repair or creation of an atrium-dependent circulation is unacceptably high. In such situations, it is possible to manage increasing cyanosis by creation or revision of a systemic arterial shunt. Alternatively, pulmonary blood flow through the existing cavopulmonary shunt could be supplemented by creation of an axillary arteriovenous (AV) fistula, a concept first proposed by Glenn and Fenn⁶ in 1972. In this report, we review our experience with the creation of an axillary AV fistula in a subgroup of patients who had previously undergone a

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Table I. Patient clinical profiles

Pt. no.	Sex	Diagnosis	Age at CPS (yr)	Type of CPS	Age at fistula creation (yr)
1	F	DIRV, AVSD, TGA, PS	4	Glenn	9
2	F	TA	8	Glenn	20
3	M	TA, PA	2	Glenn	19
4	F	TGA, PA, straddling TV	6	Glenn	15
5	M	DILV, TGA, PA, straddling TV	1	Glenn	20
6	M	TA, PS	7	Glenn	14
7	M	DILV, TGA, PA	5	Glenn	10
8	F	DORV (SPVSD), PS	2	Glenn	—
9	M	TOF, AVSD	2	BCPS	7
10	F	TA	5	BCPS	11
11	M	DORV, PS	1	BCPS	4

CPS, Cavopulmonary shunt; DIRV, double-inlet right ventricle; AVSD, atrioventricular septal defect; TGA, transposition of the great arteries; PS, pulmonary stenosis; TA, tricuspid atresia; PA, pulmonary atresia; TV, tricuspid valve; DILV, double inlet left ventricle; DORV, double outlet right ventricle; SPVSD, subpulmonary ventricular septal defect; TOF, tetralogy of Fallot; BCPS, bidirectional cavopulmonary shunt.

cavopulmonary shunt and for whom other treatment options were limited.

Methods

Patient population. Between 1961 and January 1994, 320 cavopulmonary shunts were performed at the Hospital for Sick Children, Toronto, for palliation of cyanosis associated with complex heart lesions. Despite adequate early palliation with the caval shunt,⁷ 11 patients had progressive cyanosis and were considered poor candidates for definitive repair or a Fontan procedure. The underlying cardiac lesions are listed in Table I. The first eight patients had undergone previous Glenn shunts (end-to-end anastomosis of the superior vena cava to the right pulmonary artery), and the remainder had undergone bidirectional cavopulmonary shunts (end-to-side anastomosis of the superior vena cava to the right pulmonary artery). There were six male and five female patients; ages ranged from 4 to 20 years (median 12.5 years) and time from cavopulmonary shunt ranged from 3 to 18.5 years (median 6.4 years). Preoperative catheterization was performed in all patients, and one patient (number 8) was not operated on because of multiple pulmonary AV fistulas in the shunted lung, which were a cause of the progressive cyanosis. The remainder ($n = 10$) underwent creation of an axillary AV fistula between June 1978 and January 1994. One patient (number 2) was considered unsuitable for Fontan-type repair because of pulmonary vascular disease in the left lung after a Potts shunt. Five patients were considered unsuitable for further corrective repair because of inadequate left pulmonary artery size, three because of a previous Potts shunt (numbers 4, 6, and 7), one because of a left Blalock-Taussig shunt (number 3), and one because the native left pulmonary artery was hypoplastic (number 5). Patients 1 and 9 had atrioventricular valve regurgitation, and patient 9 also had poor ventricular function. In patients 10 and 11, a Fontan procedure had been performed but required takedown after 3 months because of a hypoplastic left pulmonary artery, and after 2 weeks because of poor ventricular function, respectively.

Procedure. The operative technique used was initially described by Glenn but modified by Trusler and coworkers.⁸ With the patient under general anesthesia, an incision is made 1 cm below and parallel to the lateral half of the clavicle; if necessary, it is extended along the course of the deltopectoral groove. The pectoralis major muscle is split in the direction of its fibers. The neurovascular bundle is identified, and the axillary artery and vein are isolated (Fig. 1). After systemic anticoagulation with 150 U/kg body weight heparin sulfate, a vascular clamp is placed across the artery and vein. A side-to-side anastomosis, 6 to 7 mm in length, is made using a 5-0 or 6-0 Prolene suture (Ethicon, Inc., Somerville, N.J.). The axillary vein is ligated distal to the fistula to create a functional side-to-end AV anastomosis. Postoperative anticoagulation therapy is not given.

Results

All procedures were accomplished uneventfully. The only complication was mild swelling of the arm distal to the fistula in the first patient in whom the distal vein was not ligated; she required reoperation after 19 days. After the operation, all patients had improvement of symptoms and increased exercise tolerance. On regular review, symptoms have been seen to be stable in all but three patients (numbers 1, 3, and 5), who had deterioration from New York Heart Association functional class 1 or 2 to class 3 or 4 after 14, 10, or 7 years. All patients have been seen during the past year, and the mean duration of follow-up has been 7.4 years (range 0.1 to 15.5 years). At the time of latest follow-up, all but two shunts (numbers 2 and 3) were patent. In patient 2, no murmur has been present since 1990 (12 years after the procedure), and a recent cardiac catheterization (1993) revealed that the fistula had occluded. Despite this occlusion, the patient's condition re-

Fig. 1. Diagram of operative technique used in creation of axillary AV fistulas.

mains stable, with flow through the preexisting Glenn shunt and some contribution to pulmonary blood flow to the contralateral lung through a central shunt. In patient 3, cardiac catheterization in 1991 revealed no flow in the fistula (12 years after procedure), and cardiac transplantation was attempted because of deteriorating cardiac clinical status. Unfortunately, transplantation was unsuccessful because of the presence of dense chest wall collateral vessels.

Oxygen saturation and hemoglobin values immediately before operation, after operation, and at latest follow-up are shown in Fig. 2. The mean oxygen saturation values were $80\% \pm 2\%$ before operation, $85\% \pm 2\%$ after operation, and $84\% \pm 3\%$ at latest follow-up. The mean hemoglobin values were 202 ± 10 gm/L before operation, 177 ± 10 gm/L after operation, and 191 ± 14 gm/L at latest follow-up. Latest saturation results are skewed by deterioration in patient 3, who had evidence of fistula occlusion as mentioned previously.

Clinical evidence of cardiac failure has not been detected in any patient. Among those with follow-up longer than 1 year ($n = 8$), the absence of pulmonary AV fistulas has been documented in seven by angiography, and in the other no change in arterial saturation value has occurred.

Discussion

Data from this⁷ and other institutions reveal that 6 to 8 years after a cavopulmonary shunt the signs and symptoms of hypoxemia usually reappear. Possible causes for the resurgence of cyanosis include the development of collateral venous channels between superior and inferior venae cavae, reduction of blood flow to the contralateral lung, development of pulmonary AV fistulas in the ipsilateral lung, and possibly competitive flow to the upper lobes of the shunted lung by ingrowth of systemic collateral vessels. The effects of these factors, alone or in combination, are compounded by the increase in blood viscosity caused by increasing hematocrit.

Fig. 2. A, Oxygen saturations (*O2 Sat*) before and after operation and at latest follow-up. B, Hemoglobin (*Hgb*) values before and after operation and at latest follow-up.

When a fistulous communication is created, there are increases in effective pulmonary blood flow and systemic oxygenation. This augmentation is afforded at low risk, is simple, and avoids distortion of the pulmonary arteries. The results from this and a previous series⁹ documented a modest improvement in arterial oxygen saturation, a drop in hemoglobin level, and a substantial improvement in effort tolerance. Follow-up data from this review suggest that most shunts remain patent for many years and do not require anticoagulation.

This approach to augmenting earlier palliation should be considered in any patient who requires improved oxygen saturation and is not considered a candidate for further, more definitive correction. Importantly, this small left-to-right shunt did not cause volume overload of the ventricular chamber, and no patient had symptoms of congestive heart failure.

The prevalence of pulmonary AV fistulas in the ipsilateral lung after Glenn anastomosis may be as high as 19.7%.⁹ This could preclude many patients from undergoing creation of an axillary AV fistula because it is believed that the increased flow would bypass alveoli for oxygen exchange and place volume load on the heart. Several theories have been proposed to explain the development of such pulmonary fistulas; these include nonpulsatile flow characteristics through the shunt^{10, 11} and the influence of an as yet unknown hepatic factor. Such

fistulas appear to occur only in patients who do not have any source of hepatic venous blood flow to the lungs, as with a Glenn anastomosis or bidirectional cavopulmonary connection with ligation or atresia of the main pulmonary artery. Pulmonary fistulas have not been reported in midterm to long-term follow-up after the Fontan procedure with inferior vena caval blood entering the pulmonary arteries,¹² but they have been observed when there is a Glenn shunt in the circuit and have been reported frequently after the Kawashima procedure (total caval diversion to the pulmonary arteries with hepatic veins draining into the heart).¹³ In the only two reported cases of pulmonary AV malformations after a modified Fontan procedure, it is of interest that hepatic venous blood was not conducted to one or both pulmonary arteries.¹⁴ Recent reports also link the presence of pulmonary AV malformations to left atrial isomerism with no other cardiac lesion and without evidence of hereditary telangiectasia.^{15, 16} It has been speculated that this may represent a problem with embryogenesis.

Systemic arterial flow from an axillary fistula could contribute to the prevention of pulmonary AV fistulas by adding pulsatility to the circuit or possibly by allowing the removal or addition of an unknown vasoactive hepatic substance.¹⁷ An interesting corollary is the hepatopulmonary syndrome, in which pulmonary AV fistulas develop in the presence of severe hepatic failure,¹⁸ which may regress after

allograft liver transplantation.¹⁹ It is interesting to note that none of the patients who have undergone a cavopulmonary shunt and axillary fistula creation had pulmonary AV malformations in the ipsilateral lung during the 7.4 years of follow-up, after having had the initial Glenn shunt for a total average of 6.4 years, a period when such patients are at greatest risk for the development of pulmonary AV fistulas.⁷ Although the failure of pulmonary AV malformations to develop in this series is not definitive proof that the axillary fistula is protective, the addition of such systemic arterial flow to either a Glenn or bidirectional cavopulmonary connection may reduce the tendency toward development of pulmonary fistulas or perhaps allow their resolution.

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