Creation of a Brachial Arteriovenous Fistula for Treatment of Pulmonary Arteriovenous Malformations After Cavopulmonary Anastomosis

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Background. **Pulmonary arteriovenous malformations (PAVMs) occur in approximately 20% of patients after unidirectional superior cavopulmonary anastomosis (CPA), and frequently after bidirectional CPA in patients with polysplenia syndrome. It is hypothesized that exclusion of a growth-modulating factor produced in the liver may predispose to PAVM formation. Resolution of PAVMs after inclusion of hepatic venous effluent into the cavopulmonary circulation has been reported. An upper extremity systemic arteriovenous (AV) fistula may be created to augment pulmonary blood flow and improve oxygenation in hypoxemic patients with CPA, but there has been no systematic investigation of the effects of such fistulas on PAVMs after CPA.**

Methods. **We studied 11 patients with PAVMs who underwent creation of a brachial AV fistula a median of 11 years after CPA.**

Results. **Eight patients had discontinuous pulmonary arteries or unilateral flow of a bidirectional CPA and were not considered good candidates for Fontan completion; the other 3 patients had polysplenia and unilateral hepatic venous streaming after Fontan completion. Three patients died of progressive complications of their heart disease 4 to 18 months after AV fistula creation. Pulmonary arteriovenous malformations resolved after creation of a brachial AV fistula in 4 of 5 surviving patients with unilateral flow of a superior CPA, but in none of 3 patients with polysplenia who had unilateral hepatic venous streaming after Fontan completion and PAVMs in the contralateral lung.**

Conclusions. **These findings are consistent with the "hepatic factor" hypothesis, according to which the development of PAVMs is facilitated when an unidentified factor produced or metabolized in the liver does not reach the pulmonary circulation before traversing another capillary bed. Patients with unilateral superior CPA flow and PAVMs who are not considered candidates for Fontan completion may benefit from a brachial AV fistula.**

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Pulmonary arteriovenous malformations (PAVMs) causing significant hypoxemia occur in as many as 20% of patients after unidirectional superior cavopulmonary anastomosis (CPA [ie, a classic Glenn procedure]) in the lung receiving flow from the CPA [1, 2]. Pulmonary arteriovenous malformations may also develop after a Kawashima procedure (bidirectional CPA) in patients with polysplenia syndrome, interruption of the inferior vena cava, and lower extremity venous return to the superior vena cava (SVC) through an azygous or hemiazygous vein [3–11].

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Although the cause of PAVMs is unknown, there is increasing evidence to support the theory that the development of PAVMs is facilitated when an unidentified factor produced or metabolized in the liver does not reach the pulmonary circulation before traversing another capillary bed [3, 4, 9, 12]. The "hepatic factor" hypothesis is supported by reports of PAVMs apparently resolving in patients with a bidirectional CPA after diversion of hepatic venous effluent to the pulmonary arteries, including a recent series from our center [4, 7–9]. The hepatic factor hypothesis is also consistent with the observation that PAVMs may persist in one lung after Fontan completion in patients with a prior Kawashima circulation if there is streaming of hepatic venous blood to the other lung, a finding that suggests resolution of PAVMs only in the lung receiving hepatic venous effluent [4, 13].

Creation of an upper extremity systemic arteriovenous (AV) fistula (usually brachial or axillary) to augment pulmonary blood flow and systemic oxygenation in patients with a CPA was first proposed by Glenn in 1972 [14]. Since that report, the clinical effects of this procedure have been described only in a few small series and case reports, which have demonstrated modest improvement in hypoxemia after AV fistula creation in patients with a CPA who are not considered candidates for Fontan completion [2, 15–17]. However, there is no published information on the effect of an upper extremity systemic AV fistula on PAVMs in

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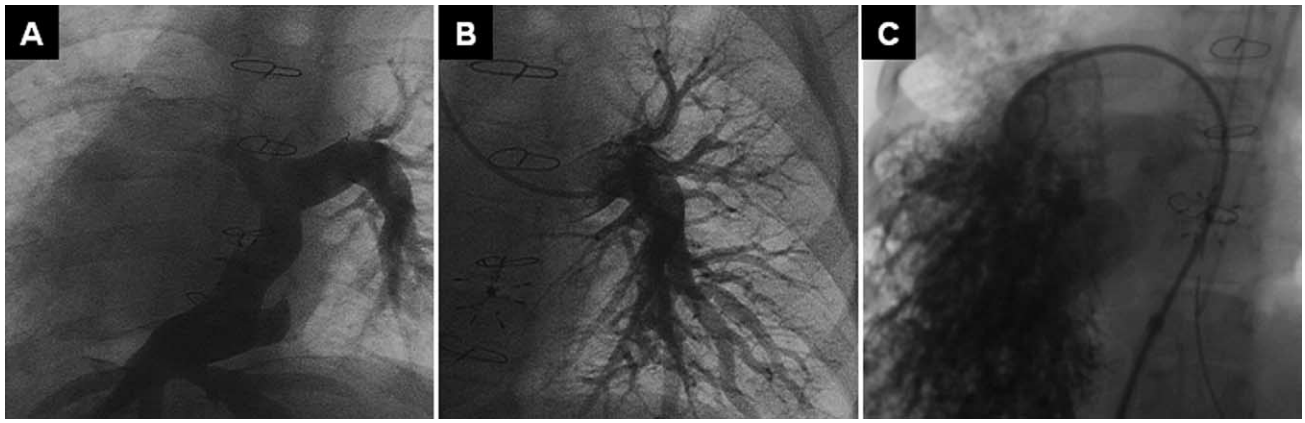

Fig 1. Angiograms in a patient with polysplenia syndrome and unilateral pulmonary arteriovenous malformations (PAVMs) in the right lung after cavopulmonary incorporation of hepatic venous effluent, which streams almost exclusively to the left lung. (A) Angiogram in the hepatic vein-pulmonary artery pathway demonstrates essentially all flow traveling to the left lung. (B) Angiogram in the left pulmonary artery demonstrates normal pulmonary arteries, with no angiographic evidence of PAVMs. (C) Angiogram in the right pulmonary artery demonstrates diffuse PAVMs.

patients with a CPA. In the Toronto series reported by Magee and colleagues [15], for example, patients with PAVMs after CPA were specifically excluded from creation of an axillary AV fistula owing to concerns that augmentation of blood flow to the lung with PAVMs might lead to increased intrapulmonary right-to-left shunting [15]. We have created a brachial AV fistula in selected patients with a CPA and hypoxemia, in most cases as an attempt to deliver hepatic venous blood to the affected lung in patients with PAVMs. Our hypothesis has been that an upper extremity systemic AV fistula will allow systemic arterial blood containing the putative hepatic factor to bypass the systemic capillary network and perfuse the PAVM-containing lung supplied by the CPA.

Patients and Methods

Between 1990 and 2004, 17 patients underwent creation of a brachial AV fistula at Children's Hospital, Boston, after prior unidirectional or bidirectional CPA. Six patients in whom an AV fistula was created to increase pulmonary blood flow and systemic oxygenation, but who did not have documented PAVMs at the time of AV fistula creation, were not included in this study. The remaining 11 patients, all of whom underwent AV fistula creation between 1997 and 2002, constitute the study group for this investigation. Three of these patients were included in prior reports [3, 4].

Results

Eight of the 11 patients were considered poor candidates for Fontan completion owing to the presence of significant PAVMs and the following considerations: 3 had persistent unilateral CPA flow after conversion of a unidirectional CPA to bidirectional CPA, 2 had systemic-to-PA collaterals as the sole source of flow to the lung contralateral to the CPA, 1 had atresia of the PA contralateral to the CPA, 1 had pulmonary vascular disease in the lung contralateral to the CPA (supplied by a systemic-to-PA shunt), and 1 had undergone a prior failed Fontan procedure that was taken down to a bidirectional CPA and had almost all CPA flow to the right lung. The other 3 patients had undergone Fontan completion 3.4 to 5.8 years before AV fistula creation, consisting of incorporation of the hepatic veins into the

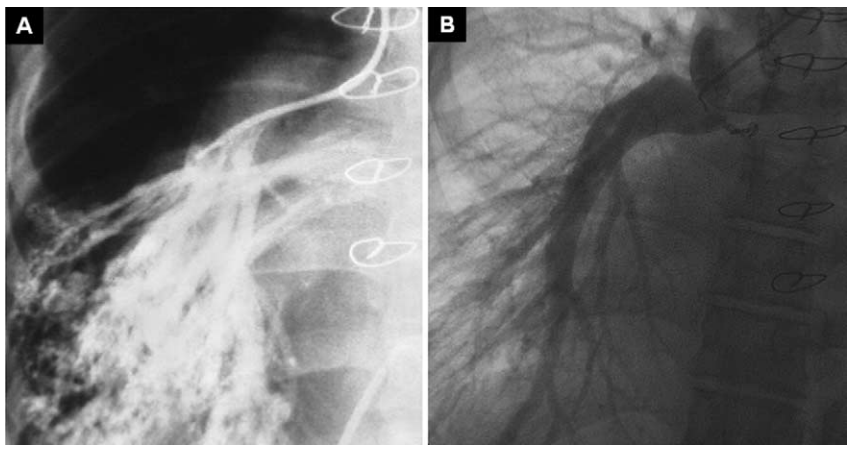

Fig 2. Angiograms in a patient with unilateral flow of a bidirectional cavopulmonary anastomosis and pulmonary arteriovenous malformations (PAVMs) in the right lung. (A) Angiogram in the right pulmonary artery before creation of a brachial arteriovenous fistula demonstrates diffuse, prominent PAVMs. (B) Angiogram in the right pulmonary artery 5.7 years after creation of a brachial arteriovenous fistula demonstrates a normal right pulmonary artery system, with no angiographic evidence of PAVMs.