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Development of Superior Vena Cava to Pulmonary Vein Fistulae Following Modified Fontan Operation: Case Report of a Rare Anomaly and Embolization Therapy

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Abstract. A patient with double inlet single ventricle, L-transposition of the great arteries, and atrioventricular valve regurgitation developed progressive heart failure necessitating modified Fontan operation at 16 months of age. The procedure included replacement of the atrioventricular valve and pacemaker insertion. Eight years following the operation, the patient developed progressive cyanosis. Catheterization confirmed the presence of fistulae from the superior vena cava and innominate vein to the pulmonary veins as the cause of cyanosis. Coil embolization of several fistulae was performed successfully at catheterization. Contributing factors for fistula formation in Fontan patients are discussed, and therapy is reviewed.

Key words: Arteriovenous fistulae — Fontan surgery — Coil embolization

Superior vena cava to pulmonary artery anastomosis was first described by Carlton in 1951 [2] and Glenn in 1958 [4]. This palliative procedure fell into disfavor because of the frequent development of pulmonary arteriovenous fistulae and other communications within a few years of the operation [7–9]. With the successful application of the bidirectional cavopulmonary anastomosis and the Fontan operation, it was believed that pulmonary arteriovenous fistulae would be less likely to develop because of more equal blood flow to both lungs. However, arteriovenous fistulae have now been reported after both types of operations.[3, 8, 10, 12]

This report (1) describes a patient after Fontan operation who had developed a previously unreported type of fistulae between superior vena cava and pulmonary veins and from innominate vein to pulmonary veins, (2) discusses altered hemodynamics that may have contributed, and (3) describes successful embolization of the fistulae.

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Case Report

A newborn infant was referred to Children's Hospital of Michigan for evaluation of cyanosis and a heart murmur. Catheterization confirmed double inlet single ventricle with L-transposition of the great arteries, subaortic outflow chamber, pulmonary stenosis, and small patent ductus arteriosus. The patient developed progressive heart failure related to atrioventricular (AV) valve regurgitation initially that was treated with digoxin and diuretics. At 16 months, the patient underwent surgical closure of the cleft in the right AV valve and atrial septectomy. The patient again developed severe congestive heart failure. Repeat cardiac catheterization at 27 months of age confirmed the recurrence of severe right AV valvular regurgitation. At reoperation, it was evident that the previous right AV valve repair had broken down with marked loss of substance of the septal leaflet. The left AV valve was grossly incompetent and small. The left AV valve was closed, and the right AV valve was replaced with a 27 mm Bjork–Shiley prosthesis. A Fontan procedure was completed by constructing an anastomosis from the right atrial appendage to the confluence of the main and right pulmonary arteries and by partitioning of the common atrium.

Postoperatively, the patient developed complete AV block and atrial flutter. A permanent epicardial pacemaker lead and ventricular demand pacemaker were placed. Recurrent atrial flutter was controlled with antiarrhythmia agents. At 8 years of age, the patient was noted to be dusky on his routine office visit, with transcutaneous oxygen saturation at 82%. Echocardiography disclosed good ventricular function, no obvious patch leak, and normal prosthetic valve motion. A pulmonary perfusion study did not suggest perfusion abnormalities. Due to the progression of desaturation with time, the pulmonary perfusion scan was repeated, which suggested decreased flow to the left pulmonary artery.

Cardiac catheterization at 12 years of age disclosed a mean right atrial pressure of 9 mmHg without gradient across the Fontan anastomosis. The pulmonary artery wedge pressure was 7 mmHg. There was no gradient from the single ventricle into the aorta. The saturations in the pulmonary arterial system were 61%–65% and in the systemic arterial system, 85%. An angiogram in the main ventricular chamber confirmed an enlarged single ventricle with mildly depressed function and paradoxical motion of the inferior wall. There was no ventricular outflow obstruction. Aortic angiography demonstrated the absence of

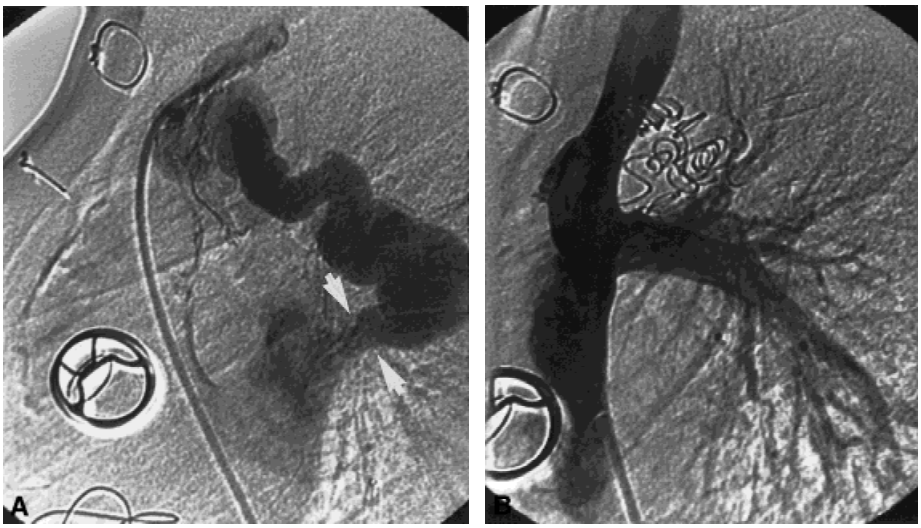

Fig. 1. (A) A large feeder vessel arises from the superior vena cava with fistulous connection to the pulmonary vein (*arrows*). **(B)** Contrast opacifies the superior vena cava, right atrium, inferior vena cava, and pulmonary arteries via the Fontan anastomosis. The fistulae from the superior vena cava are completely occluded with no residual filling of the pulmonary veins.

aortopulmonary collaterals. Right atrial angiography disclosed a widely patent Fontan anastomosis. Reflux of contrast from the right atrium into the superior vena cava revealed a plexus of veins arising from the superior vena cava at the right atrial junction with a fistulous connection to the right upper pulmonary vein (Fig. 1A). A series of injections was performed, which delineated three vessels that arose from the superior vena cava and fed the large fistulous connection. A total of 14 Cook Gianturco coils (0.038 in.) were delivered to the three feeder vessels using a 7 French Lehman catheter (USCI, Tewksburg, MA). Injections in the individual feeder vessels and in the superior vena cava disclosed complete occlusion of the fistula (Fig. 1B). The saturation remained 85%, and this level of desaturation was attributed to anesthesia. Follow-up perfusion scan 24 h later disclosed normal flow to the lungs.

One month later, the patient was still cyanotic with a saturation of 85%. At repeat catheterization, an injection performed in the innominate vein at its junction with the left jugular vein disclosed a large tortuous fistula from the innominate vein draining into the left upper pulmonary vein (Fig. 2A). Eight Cook Gianturco coils were delivered into this fistula with subsequent angiography confirming complete occlusion of the fistula (Fig. 2B). Follow-up angiograms in the innominate vein, right superior vena cava, and inferior vena cava disclosed a single residual fistula from the innominate vein to right pulmonary veins. This fistula could not be entered easily, and was left for a different approach in the future. The saturation following the case was 92%.

Discussion

The development of pulmonary AV fistula in patients after Glenn surgery leads to hypoxemia and shunt failure. The incidence of fistulae development in most series is around 20%–25% and increases with time [3, 7]. Glenn proposed that normally occurring tiny arteriovenous connections in the lung could enlarge to become fistulous and could cause precapillary shunting with arterial desaturation [5]. He noted that these communications most often occur in the lower lobe and speculated that the fistulae resulted from stagnation of flow. Others have suggested that reduced pulmonary flow may not be as significant a factor as nonpulsatile flow [3, 5, 7]. Maldistribution of pulmonary blood flow with more flow to the lower lobe has been proposed as an additional contributing factor for the development of fistulae, but either problem may be seen without the other [3].

The development of pulmonary arteriovenous fistulae has now been documented in patients following Fontan operation [3, 8, 10, 12]. Fontan patients have been shown to develop maldistribution of pulmonary flow as previously seen after the Glenn anastomosis [3, 5, 7, 8, 10]. In Fontan patients with atriopulmonary anastomosis, the flow velocity during atrial systole is reduced with half of the flow occurring passively during diastole [11]. With a more recent Fontan modification, total cavopulmonary anastomosis, all flow is passive. This raises concerns that even more Fontan patients will develop pulmonary arteriovenous fistulae in the future.

Our patient developed venovenous fistulae from the systemic veins to the pulmonary veins, leading to clinically significant arterial desaturation. The unusual location of the fistulae in this patient, from the SVC and innominate vein to the pulmonary veins, has not been previously reported in Fontan patients. Channels from the SVC to right atrium and IVC have been reported following the Glenn operation, and their mechanisms of development are unknown [9]. Embryologically, the pulmonary veins develop from angioblasts around the lung bud and developing bronchi [1]. Other nearby veins in the area of the splanchnic plexus develop and anastomose with the anterior cardinal system to become the systemic venous drainage of the mediastinum [1]. In addition, there are early communications between pulmonary veins and veins of the anterior cardinal system, which normally regress as the pulmonary veins become incorporated into the left atrium [1]. It is possible that some of these channels persisted or enlarged in our patient because of chronic elevation of systemic venous

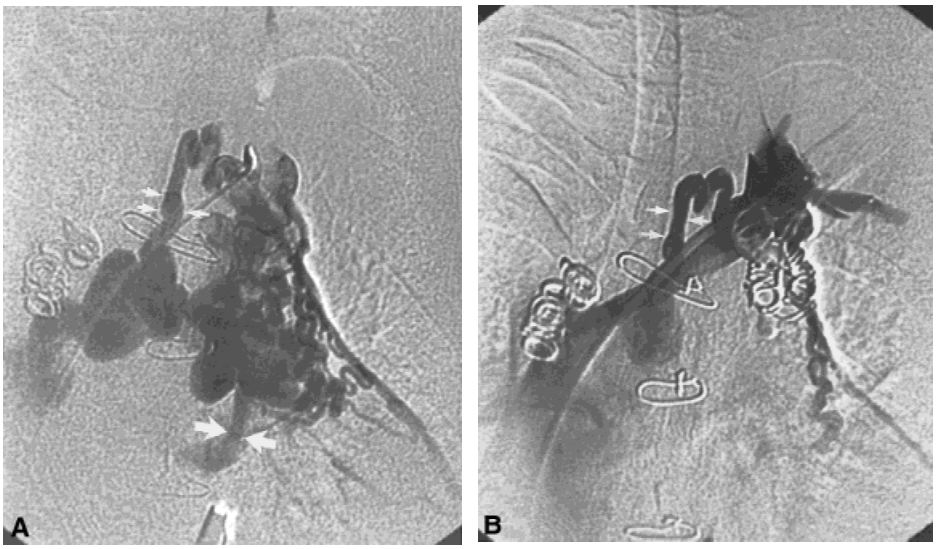

Fig. 2. (A) An injection of contrast at the mouth a group of feeder vessels arising from the innominate vein demonstrates two separate communications to the pulmonary veins (*small arrows* and *large arrows*). The coils on the patient's right are from the previous embolization (Fig. 1B). **(B)** Angiography of the innominate vein discloses complete occlusion by coil embolization of one of two fistulae. There is still a residual fistula to the right pulmonary veins (*arrows*).

pressure and/or pulmonary venous stasis. We did not find any discrete obstruction to innominate vein flow or SVC flow before or after coil embolization as a cause of elevated venous pressure to explain fistula development. However, the multiple sites of fistulae and our inability to improve the underlying hemodynamic status (chronic atrial flutter, borderline ventricular function, and prosthetic AV valve) make it likely that other fistulae will develop with time.

Several reports document the potential to perform coil embolization of pulmonary arteriovenous fistulae with good success and relative safety, regardless of the etiology of the fistulae [6, 10]. An additional case report was found describing coil occlusion of a venovenous (SVC to IVC) communication in a patient following Glenn anastomosis [7]. Ours is the first case report of successful coil occlusion of systemic venous to pulmonary venous fistulae following Fontan operation. Coil embolization can be performed with safety in this type of patient and should be considered the procedure of choice.

Clinical Implications

Systemic desaturation should be investigated when it occurs following Fontan operation. Contrast echocardiography and pulmonary perfusion scans may suggest the presence of fistula formation. The possibility of systemic venous to pulmonary venous fistulae should be considered, as well as pulmonary arteriovenous fistulae during diagnostic catheterization. Coil embolization is the treatment of choice.

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