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Fontan Fenestration Closure Prior to Posterior Spinal Fusion in Patients with Single Ventricle Heart Disease

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Abstract

Study Design: Case series

Objective: To describe transcatheter closure of the Fontan fenestration prior to posterior spinal fusion in 2 children to prevent paradoxical venous air embolism during the operation.

Summary of Background Data: Scoliosis is common amongst patients with single ventricle congenital heart disease who have undergone Fontan operation and spinal surgery can offer physiologic benefit. Venous air embolism is a rare, but important reported complication during spinal surgery performed in the prone position. Patients with Fontan circulation can have significant right to left shunting via a patent Fontan fenestration that can increase the risk of paradoxical systemic embolization of any entrained venous air.

Methods: We retrospectively reviewed the charts of 2 patients with single ventricle congenital heart diasease who had undergone fenestrated Fontan operation and underwent transcathter fenestration closure prior to spinal fusion.

Results: Two patients with Fontan circulation underwent successful transcatheter fenestration closure with Amplatzer Ductal Occluder II devices. Five to 6 months after closure, both underwent uncomplicated posterior spinal fusion.

Conclusions: Transcatheter closure of the Fontan fenestration prior to spinal fusion in 2 with Fontan circulation and scoliosis is a rare, but important indication for fenestration closure that warrants emphasis.

Key Words: Fontan, scoliosis, fenestration, venous air embolism

Level of Evidence: N/A

Introduction

Scoliosis is common amongst patients with congenital heart disease who have undergone Fontan operation (1). Venous air embolism (VAE) is a rare complication of spinal surgery performed in the prone position (2). Patients with Fontan circulation can have significant right to left shunting via a Fontan fenestration that can increase the risk of paradoxical systemic embolization of venous air. We report transcatheter fenestration closure prior to posterior spinal fusion in 2 children with significant scoliosis to prevent paradoxical VAE during the operation.

Cases

Two adolescents with single ventricle heart disease who had undergone fenestrated Fontan operation also developed significant thoracolumbar scoliosis and were in need of posterior spinal fusion. The patient details are described in Table 1. Imaging associated with Patient 1 is summarized in Figures 1-2(A,B,C) and imaging for Patient 2 is summarized in Figures 3-4(A,B,C). Both underwent cardiac catheterization prior to spinal fusion with closure of the Fontan fenestration. In both, the mixed venous saturation improved and Fontan conduit pressure remained unchanged during temporary fenestration occlusion. Five to 6 months after catheterization, both exhibited evidence of complete fenestration occlusion based on aortic saturation and echocardiogram and both underwent uncomplicated posterior spinal fusion without evidence of VAE.

Discussion

In 2 adolescents with a Fontan circulation, we report closure of the fenestration prior to posterior spinal fusion to eliminate right to left shunting and decrease risk of paradoxical VAE during the operation. This is an important, but uncommon indication for fenestration closure that warrants emphasis.

VAE has been reported in over 20 cases of spine surgery performed in the prone position in patients with a normal heart, many of which were pediatric patients (2-12). Some led to catastrophic embolic events (2,3,5,7-10,12). VAE occurs as air from the operative field enters the venous vasculature. During spine surgery, decorticated bone is directly exposed in the operative field and air may enter venous channels within the spine. For air to enter the channels, a negative pressure gradient must exist between the venous channel exposed to air and the central venous pressure (2,3). For spine surgery in prone position, the operative field is above the heart and a gravitational gradient can exist that favors entrainment of air. Central venous pressure can further drop during inspiration, with significant blood loss, or with vasodilation (2-4).

Patients with single ventricle heart disease undergo a staged surgical palliation that creates a Fontan circulation in which both the superior and inferior vena cavae are directly connected to the pulmonary arterial circulation. At the Fontan operation, the inferior vena cava is connected directly to the pulmonary artery using an extracardiac conduit or using an intraatrial baffle; frequently a communication is made between the Fontan conduit or baffle and the right atrium (Figure 5). This communication is termed the Fontan fenestration and is associated with decreased post-operative morbidity (13). This fenestration is a right to left shunt which could allow paradoxical embolization of air to the systemic circulation in setting of VAE.

Scoliosis is common amongst patients with a Fontan circulation and can negatively affect Fontan physiology by leading to restrictive lung disease (14,15). Scoliosis surgery has been described in patients with Fontan circulation and most series do not describe the status of the fenestration (1,16-18). Significant complications have been reported including an operative death in the setting of massive bleeding (1,16-18). VAE has not been specifically noted in these series.

In patients with Fontan circulation, we feel it is important to eliminate significant right to left shunting prior to spine surgery. A patent Fontan fenestration is the most common type of right to left shunt. Patients may also have veno-venous collaterals connecting systemic veins to pulmonary veins. Both of our patients underwent catheterization prior to spinal surgery; veno-venous collaterals were not observed and fenestration closure was undertaken. Transcatheter fenestration closure is commonly performed to eliminate symptomatic hypoxemia and/or to decrease risk of paradoxical embolization (19-21). Both of our patients showed favorable oximetric/hemodynamic changes during temporary occlusion. We used the Amplatzer Duct Occluder II (St. Jude Medical, MN) to occlude the fenestration as has been reported (20).

Conclusion

Scoliosis is common in patients with Fontan circulation. VAE is a rare, but potentially catastrophic, complication of scoliosis surgery. Right to left shunting via a Fontan fenestration may increase risk of paradoxical embolization of air during scoliosis surgery in this population. We report transcatheter closure of the Fontan fenestration in 2 patients prior to spinal operation. This is an uncommon indication for fenestration closure that warrants highlighting.

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Figure Legends

Figure 1

AP chest and abdominal x-ray showing severe thoracolumbar scoliosis in 14 year-old with single ventricle congenital heart disease who has undergone fenestrated, extracardiac Fontan operation.

Figure 2 (A-C)

A: AP view of inferior vena cava (IVC) angiogram showing the Fontan conduit that connects to the inferior vena cava. Right to left flow from the conduit to the atrium via the Fontan fenestration is seen. **B:** AP view of IVC angiogram following placement of an Amplatzer Ductal Occluder II (ADO II) across the fenestration. There is trivial residual flow through the device and it has not yet been released from the delivery cable. **C:** Echocardiogram showing apical view 3 months after transcatheter fenestration closure. The device is seen across the fenestration with no residual flow by color Doppler interrogation.

Figure 3

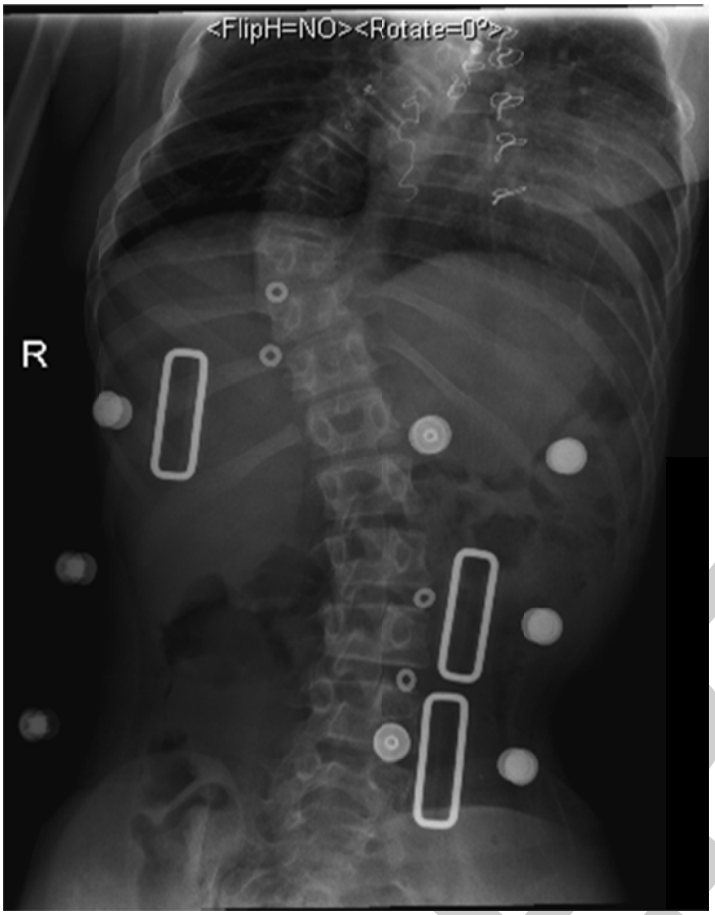
AP chest and abdominal x-ray showing severe scoliosis in a 13 year old with single ventricle congenital heart disease who has undergone fenestrated, extracardiac Fontan operation.

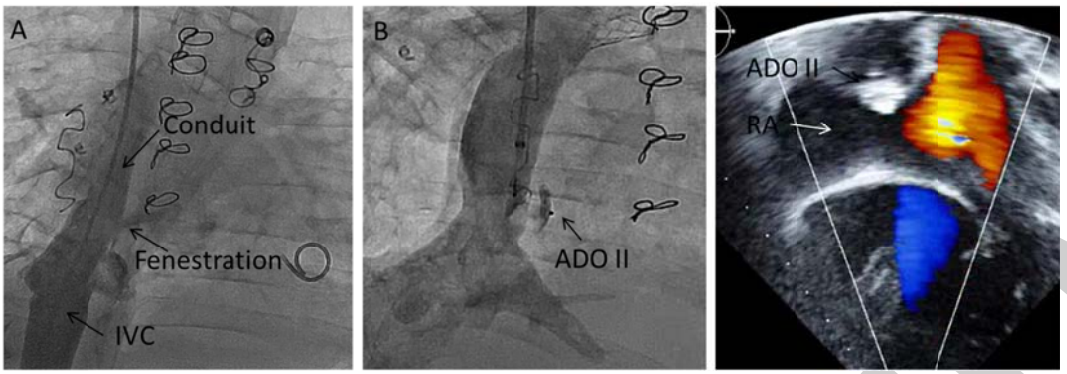
Figure 4

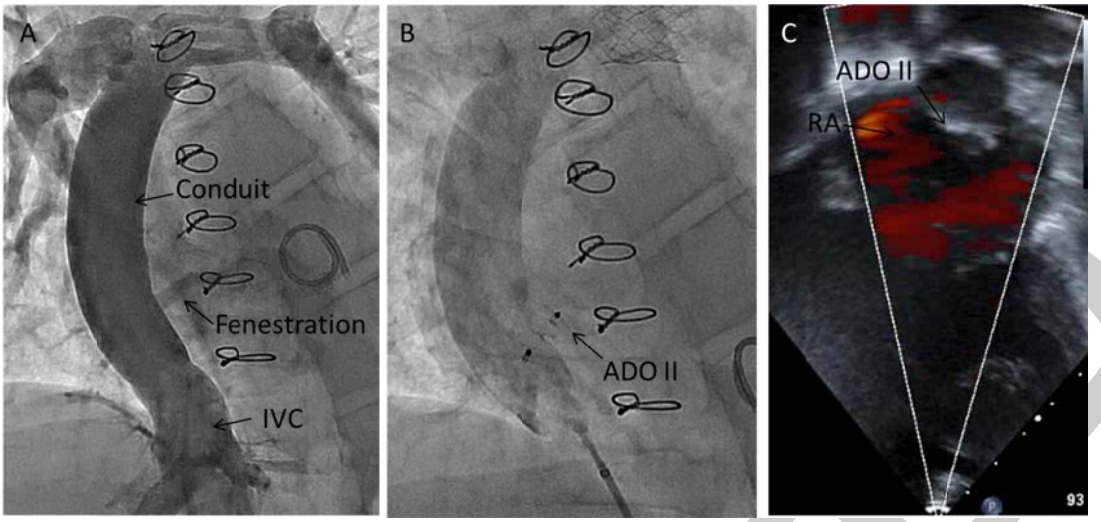
A: AP view of inferior vena cava (IVC) angiogram showing the Fontan conduit connecting the IVC to the right pulmonary artery. Right to left flow from the conduit to the atrium via the fenestration is seen. **B:** AP view of IVC angiogram following placement of an Amplatzer Ductal Occluder II (ADO II) across the fenestration showing no residual flow across the fenestration. **C:** Apical view from an echocardiogram performed 3 months after fenestration closure showing no residual flow across the fenestration.

Figure 5

Diagram showing an extracardiac fenestrated Fontan circuit in a patient with hypoplastic left heart syndrome. Inferior vena cava (IVC) return is directed to the pulmonary arteries via the extracardiac Fontan conduit and the fenestration provides a communication between the conduit and heart which could facilitate systemic air embolus in setting of venous air entrainment.







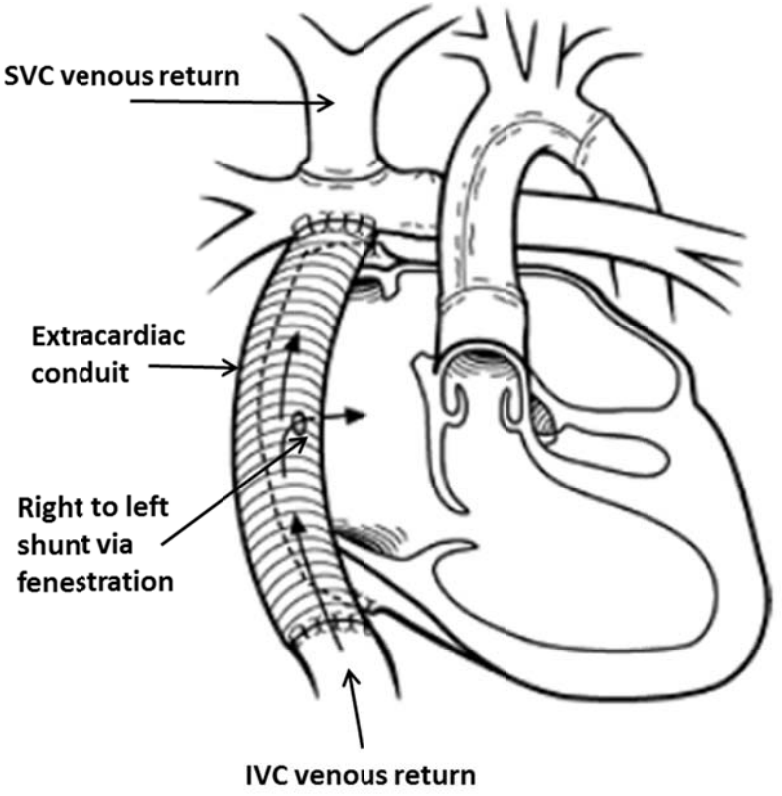


Table 1. Summary of 2 patients with Fontan circulation who underwent transcatheter fenestration closure prior to posterior spinal fusion to prevent complications possible venous air embolism.

Patient	Cardiac Anatomy	Type of Fontan	Age at Fenestration Closure	Aortic saturation Pre/Post Closure	Device Used for Fenestration Closure	Time Between Fenestration Closure and Surgery	Orthopedic Operation	Post-operative LOS
1	Tricuspid stenosis	EC, fenestrated	14 years	87%/95%	5/6 ADO II	6 months	Posterior T2-L1 fusion	4 days
2	Mitral atresia, DORV	EC, fenestrated	13 years	88%/96%	5/4 ADO II	5 months	Posterior T2-L3 fusion	4 days

ADO: Amplatzer Ductal Occluder II, DORV: double outlet right ventricle, EC: extacardiac, LOS: length of stay