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Fenestrated Fontan Procedure: Evolution of Technique and Occurrence of Paradoxical Embolism

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Abstract. The Fenestrated Fontan procedure (FFP) has improved outcome in high risk patients. The technique is evolving, however, and complications are not fully known. Over a 3-year period 13 patients (mean age 35 ± 29 months) underwent an FFP in our institution. In the first two patients the fenestration had to be created because of high right atrial pressure and low cardiac output; in 11 patients the FFP was planned. In three patients the sutures for the adjustable fenestration were crossing the defect. In 10 patients, purse-string sutures were placed around but not across the defect. Because large fenestrations were created in 11 patients (8–12 mm) Glenn shunts were performed to improve arterial saturation. The postoperative course was relatively uneventful, with chest tubes being removed 1–8 days (mean 4 ± 3 days) postoperatively and the hospital stay ranging from 7 to 27 days (mean 14 ± 6 days). One patient had bleeding and another had a mediastinal abscess. The first patient died (7.6%) because of hemodynamic instability due to prolonged cardiopulmonary bypass from the creation and enlargement of the fenestration. One patient had a paradoxical cerebral embolism from clots that formed on the sutures crossing the fenestration. Because of this problem the remaining patients were placed on salicylates while awaiting closure of their fenestration. All 12 patients had their fenestrations closed, performed under local anesthesia in 9, at mediastinal abscess drainage in 1, and spontaneously in 2. We conclude that creation of large fenestrations in combination with Glenn shunts and the use of adjustable fenestrations are viable modifications of the FFP. The use of purse-string sutures around the fenestration and antiplatelet drugs can probably minimize the occurrence of paradoxical embolism.

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Key words: Fontan procedure—Univentricular hearts—Paradoxical embolism

The Fontan operation has been applied successfully for tricuspid atresia with ventriculoarterial concordance and single ventricle with pulmonic stenosis [2, 7, 8, 16]. However, for other single ventricle equivalents and in the presence of other risk factors such as increased pulmonary artery pressure and resistance, left-sided obstruction (subaortic stenosis, coarctation), distorted and scarred pulmonary arteries, vascular adhesions, and aortopulmonary collaterals, the morbidity remains high [5, 10–12].

A significant number of patients with one or more risk factors undergoing the Fontan operation develop transient systemic venous hypertension leading to low cardiac output states during the immediate postoperative period [4]. Leaving a fenestration in these patients allows right-to-left shunting and decompresses the right atrium, improving results. The fenestration is either fixed (which can be closed in the cardiac catheterization laboratory with occlusive devices) or adjustable (which can be closed in the operating room under local anesthesia) [1, 13, 14].

The size of the fenestration affects the degree of systemic venous hypertension and therefore cardiac output during the immediate postoperative period. Small fenestrations tend to be associated with high systemic venous pressure and hemodynamic instability, whereas large ones tend to cause significant arterial desaturation. To circumvent these problems, we have created large fenestrations in combination with Glenn shunts. Additionally, because we encountered paradoxical embolism from clots developing from sutures crossing the atrial fenestration, we started using purse-string sutures around the fenestration. Antiplatelet drugs were also given to the patients while awaiting closure of their fenestrations. This paper reports our experience.

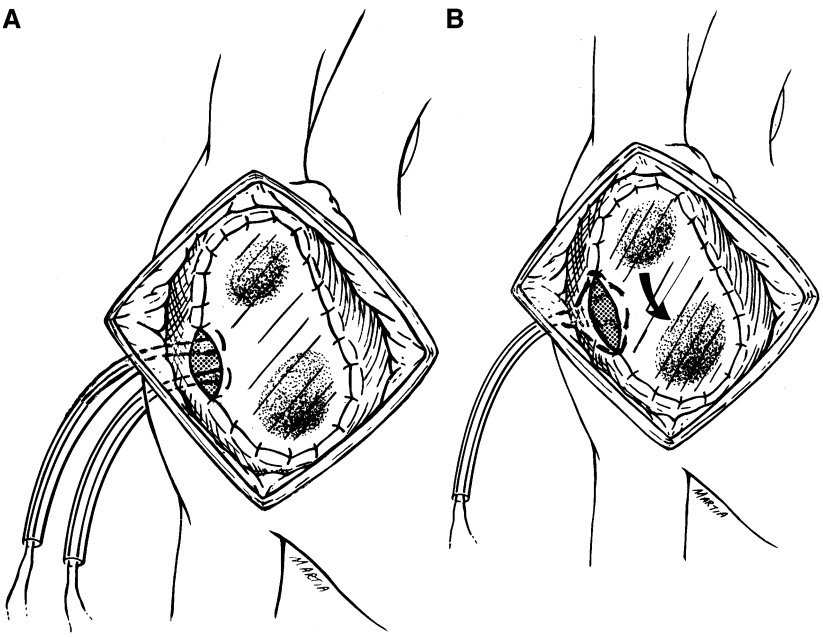

Fig. 1. (A) Two sutures cross the atrial fenestration used in three patients, one of whom developed a paradoxical embolism. Atrial partitioning with the tricuspid valve communicating with the left atrium used in the patient who had embolization is shown. **(B)** Purse-string suture was used in 10 patients around but not across the fenestration. The suture was passed through a Silastic snare and placed subcutaneously, to the right of the sternum.

Materials and Methods

Over a 3-year period ending in August 1993 a total of 13 patients underwent a fenestrated Fontan procedure in our institution. The age ranged from 13 months to 10 years (mean 35 ± 29 months). The diagnosis included tricuspid atresia ($n = 5$), double-outlet right ventricle with hypoplastic left ventricle ($n = 4$), pulmonary atresia with intact ventricular septum ($n = 2$), and single ventricle ($n = 2$). There were 19 previous operations, including 14 modified Blalock-Taussig shunts (bilateral in four patients), 3 pulmonary artery banding, 1 main pulmonary artery-to-aorta connection, and 1 classic Glenn shunt.

In the first two patients, the fenestrations had to be created because of high right atrial pressure and low cardiac output with persistent metabolic acidosis and oliguria. In the remaining 11 patients the fenestration was planned. In three of the first four patients the sutures for the adjustable fenestration were crossing the defects (Fig. 1A). Because one of these three patients had paradoxical embolism from clots that formed on the sutures (Fig. 2), purse-string sutures, with none crossing the defect, were placed in the next nine patients (Fig. 1B). The purse-string suture was on a Silastic snare and placed subcutaneously, to the right of the sternum.

Because we created a large fenestration to effectively decompress the right atrium in 11 patients (8–12 mm, mean 9.5 ± 1.0 mm), obligatory Glenn shunts were performed to improve arterial saturation [3]. The Glenn shunt was to the right pulmonary artery in three patients, to the left pulmonary artery in six patients, and bidirectional with intact pulmonary valve in two patients in whom a right atrium to right ventricle connection was performed. Four patients who had the Glenn shunt to the left lung had bilateral superior vena cava connected to the left pulmonary artery. In one patient the fenestration was 4 mm; and in the other patient, who had a previous classic Glenn shunt, a 6 mm fenestration was created.

Results

Early Results

The first patient in this series died 24 hours postoperatively. The death was due to hemodynamic instability and bleeding after a prolonged operation caused by repeated bypass from creation and subsequent enlargement of the atrial fenestration. One patient required reexploration for bleeding, and another patient underwent drainage of a mediastinal abscess 10 days postoperatively, at which time the atrial fenestration was also closed by tying the purse-string suture. The Silastic snare was removed. The chest tube was removed 1–8 days (mean 4 ± 3 days) postoperatively, and the length of hospital stay ranged from 7 to 27 days (mean 14 ± 6 days).

Late Results

One patient had paradoxical embolism involving the basal ganglia with right hemiparesis 32 days postoperatively due to clots developing on the sutures that crossed the atrial fenestration. The patient was anticoagulated with heparin, and 3 days later closure of the fenestration was performed by pulling on the purse-string suture on the snares under local anesthesia. Neurologically, the patient recovered completely. Following the occurrence of paradoxical embolism in this patient, the remaining nine patients were placed on salicylates at 10 mg/kg per day in addition to avoiding sutures crossing the atrial fenestration.

Among the remaining 10 patients, 8 had closure of the atrial fenestration 24–150 days (mean 51 days ± 46 days) postoperatively by simply pulling the purse-string suture against the Silastic snare under local anesthesia. One patient was found to have spontaneously closed a 4

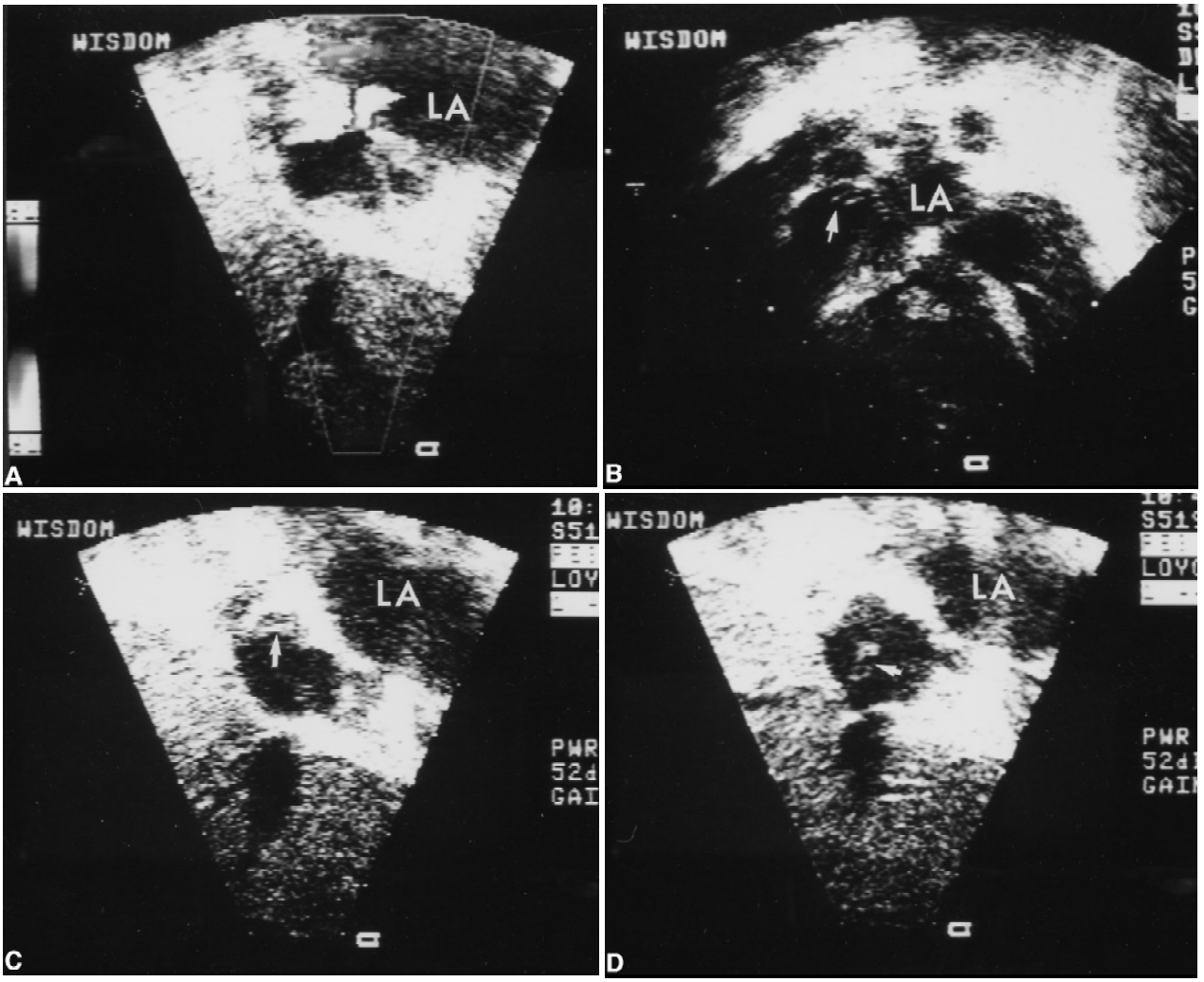

Fig. 2. (A) Doppler echocardiogram shows the right-to-left shunt through the atrial fenestration. **(B)** Sutures for the fenestration (*arrow*). **(C)** Clot (*arrow*) is attached to the sutures. **(D)** Another piece of clot, probably an extension of the clot to the right atrium, originates from the sutures. *LA*, left atrium.

mm fenestration 4 months postoperatively. The remaining patient, whose 1 cm fenestration was purposely not closed early because of problems of bronchospasms and oxygen requirement preoperatively, was found to have a trivial fenestration 2 years postoperatively.

Discussion

Staged operations, such as establishing a Glenn shunt followed by the Fontan operation later, have improved the outcome in high risk patients [4, 9, 15]. This approach, however, entails two major operations and anesthesia, which can be avoided by introduction of the fenestrated Fontan procedure. Moreover, immediate relief of the ventricular volume overload is also accomplished.

The surgical technique for the fenestrated Fontan procedure is evolving, including the right size of the fenestration. Generally, 4 to 6-mm fenestrations [1, 13, 14] have been created that can be adjusted with a snare and can be closed later under local anesthesia [17]; or fixed fenestrations are created that can be closed with an occlusive device in the catheterization laboratory [1, 13].

Because some patients with 4 to 6-mm fenestrations would still have high systemic venous pressure and low cardiac output postoperatively [14], we have created larger fenestrations and maintained reasonable arterial saturation with obligatory Glenn shunts [3]. The incidence of effusion is minimal, with the chest tubes being removed early and the hospital stay short.

There is a disproportion of flow in classic Glenn shunts, which has been a disadvantage. The superior vena caval flow, which is approximately 45% of the total systemic venous return, goes to the right lung, which has

approximately 55% of the total pulmonary vascular bed. This discrepancy is eliminated in most of our patients by creating a Glenn shunt to the left lung or a bidirectional Glenn shunt in the presence of an intact pulmonary valve and a right atrium to right ventricle connection.

Although we could find no other reports of cerebral embolism with a fenestrated Fontan procedure, the presence of a right-to-left shunt with the fenestration makes paradoxical embolism a real problem. It is possible that the problem will be more common with adjustable fenestration than with the fixed fenestration because of the presence of sutures across or around the defect with adjustable fenestration. The adjustable fenestration has an advantage in that it can be tightened or closed based on changes in hemodynamics during the immediate postoperative period. Placement of a purse-string suture around but not across the fenestration and use of antiplatelet therapy while awaiting closure can probably minimize the occurrence of paradoxical embolism with the fenestrated Fontan procedure.

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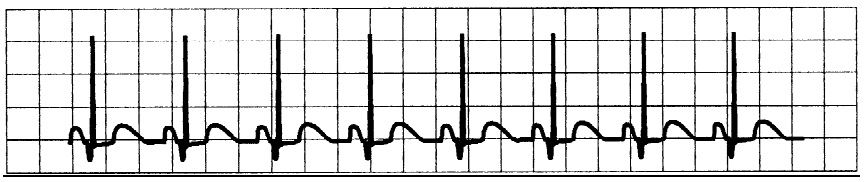
Shortening of PR and QRS Durations

Lidocaine and phenytoin typically cause no change to the electrocardiogram. However, in high doses they may reduce the PR and QRS intervals through increasing V_{max} of the action potential.

Ra-id Abdulla



Short PR interval and narrow QRS complex.

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