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RESEARCH

Extracardiac Fontan Operation with Prebuilt Fenestration: Preparing the Road for Future Atrial Access

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Abstract

The extracardiac Fontan procedure is the most widely adopted surgical approach for definitive palliation of single-ventricle anomalies, offering improved hemodynamics and reduced arrhythmogenic risk. However, limited access to the pulmonary venous atrium during follow-up poses significant challenges for electrophysiologic and interventional procedures. We describe a simple technical modification of the extracardiac Fontan designed to preserve these advantages while facilitating future percutaneous access.

Introduction

The extracardiac conduit Fontan procedure is widely regarded as the preferred approach for definitive palliation of single ventricle anomalies by many surgeons and centers [1]. This technique offers several advantages, including improved hemodynamics, technical simplicity, possible avoidance of aortic cross-clamping, elimination of intracardiac prosthetic materials (thereby reducing thrombogenic risk), a lower incidence of arrhythmias due to fewer atrial sutures, and reduced pulmonary vein compression.

Limitations of this technique persist include the difficulty of creating and maintaining a patent fenestration and the limited venous access to the atrium for pacing or intracardiac interventional procedures.

Possible approaches include traversing an existing fenestration, performing a transbaffle or transcaval puncture, or employing a retrograde aortic approach with remote magnetic navigation.

The presence of a prior Fontan fenestration does not guarantee persistent patency, as fenestrations across extracardiac conduits are prone to spontaneous closure with the time.

Having a defined anatomical landmark for fenestration can therefore be highly advantageous.

We present herein a technical modification of the standard extracardiac Fontan procedure, incorporating a prebuilt fenestration specifically designed to facilitate future access to the right atrium by interventional cardiologists or electrophysiologists, improving procedural flexibility and long-term management options.

Since January 2016, we have adopted this technique, which has now become the standard practice in all Fontan procedures performed at our institution.

Surgical Technique

Cardiopulmonary bypass is established via direct cannulation of the aorta and both caval veins. The procedure is routinely conducted on a beating heart under normothermia. Alternatively, depending on the underlying cardiac anatomy, the Fontan procedure may be conducted under cardioplegic cardiac arrest while maintaining normothermia.

A polytetrafluoroethylene (PTFE) conduit is interposed between the inferior vena cava and the pulmonary artery confluence. To facilitate future percutaneous access, a radiopaque marker is positioned around the fenestration.

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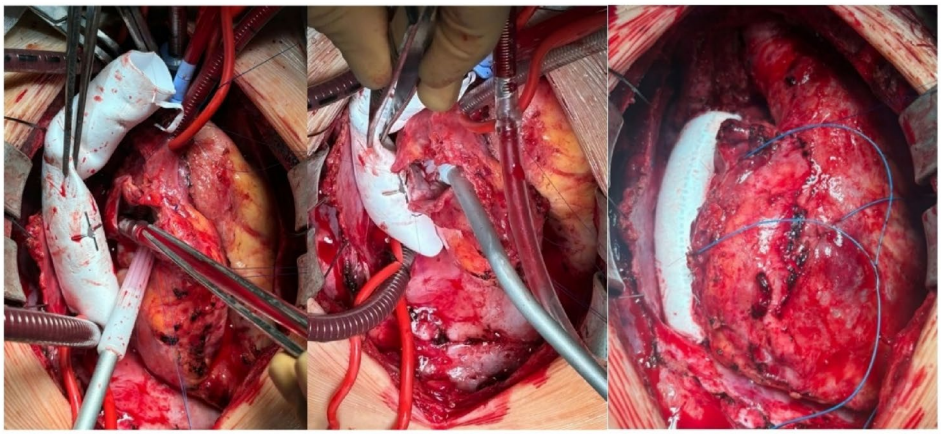

Fig. 1 Surgical steps of the extracardiac fenestrated Fontan procedure: creation of a radio-opaque landmark around the fenestration; incorporation of the landmark into the suture line; final result of the completed extracardiac fenestrated Fontan

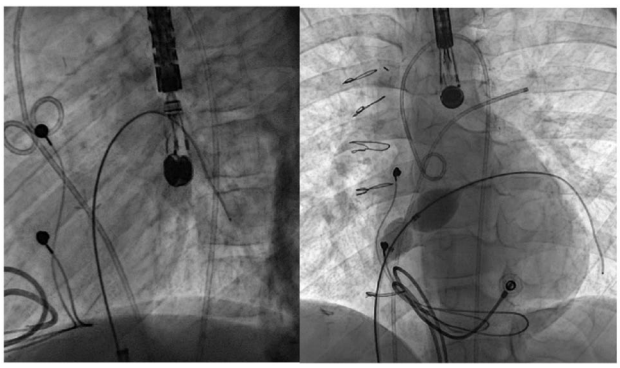

Fig. 2 Intra-procedural fluoroscopic views showing successful placement and radiographic detectability of our developed marker

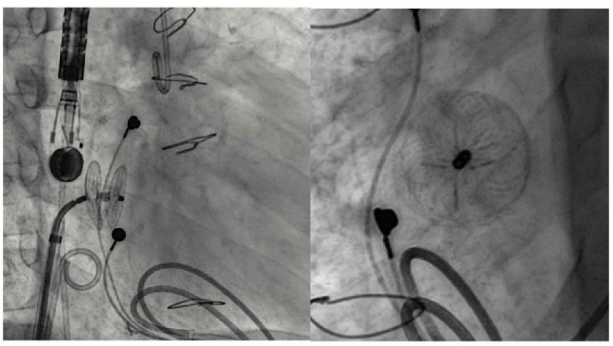

Fig. 3 Fluoroscopic views showing the deployed 18 mm Amplatzer Talisman PFO occluder

The fenestration can be created either by applying four metallic clips at the cardinal points or by placing circular metallic sutures. The right atrial free wall is anastomosed to the conduit surrounding the fenestration using a continuous running suture, leaving a small margin around the orifice to prevent inadvertent closure. The fenestration itself remains untouched by the suture line, with the marker fully incorporated within it (Fig. 1).

The same technique is applied even in cases where direct fenestration is not performed, with the marker being left in place and sutured to the atrial wall to facilitate potential future procedures.

All patients had an uneventful postoperative course and were discharged on oral anticoagulation therapy for one year, after which it's replaced with aspirin.

During follow-up, the clinical utility of this technique was demonstrated in a patient with unbalanced complete atrioventricular canal, who underwent a fenestrated extracardiac Fontan in 2020. The patient presented with recurrent syncopal episodes and severe exercise-induced desaturation over the preceding year. Cardiac catheterization was indicated to assess the potential closure of the fenestration.

CT scan or MRI imaging was not required.

The procedure involved right femoral venous access for device delivery, left femoral venous access for contrast injection and pressure sampling, and right femoral arterial access for hemodynamic monitoring.

The radio-opaque marker enabled immediate localization of the fenestration (Fig. 2).

An 18 mm AGA balloon occlusion test was performed, resulting in a marked increase in oxygen saturation from 75% to 85–90%, without any rise in pulmonary artery pressure. The fenestration was then accurately measured, allowing for selection of an appropriately sized closure device.

An 18 mm Amplatzer Talisman PFO occluder was successfully implanted (Fig. 3).

Notably, incorporation of the radio-opaque marker was associated with a significant enhancement in technical simplicity compared with similar procedures conducted without it. The patient was discharged the following day with complete fenestration closure and no complications.

Discussion

The extracardiac Fontan procedure remains the preferred option for definitive palliation of single-ventricle anomalies, offering excellent long-term outcomes. However,

arrhythmias continue to represent one of the major complications of the Fontan circulation, albeit less frequent with the extracardiac than with the intracardiac approach [2].

Several approaches have been reported to access arrhythmias in patients with extracardiac Fontan. Transthoracic percutaneous approach was reported in patients with lateral tunnel or in extracardiac Fontan under Computed Tomography guidance [3, 4]. Hybrid procedures have been proposed to reduce the risk of pneumothorax or haemorrhage by reopening the chest and performing the ablation through a small atriotomy incision [5].

Few anecdotical reports describe transvenous transbaffle access through the extracardiac conduit [6]. The difficulties to cross the Gore-Tex conduit with possible deformation of the radiofrequency needle, and the risk of bleeding from the conduit and/or the pulmonary venous atrium, make this technique still under evaluation.

Finally, a retrograde aortic approach has been described in intra-atrial conduits by remote magnetic-guided ablation, but the experience is very limited, and available in few centres [7].

A creative solution has been reported by the UCLA Group with the so-called trap-door modification of extracardiac Fontan [8].

We propose a simple, reproducible modification of the extracardiac Fontan procedure designed to facilitate future electrophysiological or interventional access while preserving all the advantages of the standard technique.

Our approach addresses a frequent limitation of non-marked fenestrations, in which prolonged fluoroscopy and contrast injection are often required to identify the orifice.

This modification enables straightforward transcatheter access through the fenestration, using the radio-opaque landmark to guide a transseptal needle for precise entry. The same configuration is also used to create a late-functioning fenestration in patients with hemodynamic deterioration, facilitating controlled decompression through stent placement - even in non-fenestrated Fontan circuits.

Although our early experience demonstrates the technical feasibility and safety of this approach, none of the patients in our current cohort have yet required atrial access for ablation or fenestration creation.

Nonetheless, the modification has been adopted as routine practice at our institution and represents a refinement of previously described fenestration techniques [9].

This modification offers a practical, low-cost solution that balances surgical simplicity with future accessibility. It may reduce the need for complex hybrid or trap-door procedures and improve the long-term management and adaptability of patients with a Fontan circulation.

While fenestration improves early postoperative outcomes, it also introduces long-term challenges, particularly cyanosis and exercise intolerance in selected patients.

Our modification aims to facilitate interventional procedures after extracardiac Fontan, including both electrophysiological and interventional cardiology procedures.

Conclusions

The described technical modification to the extracardiac Fontan procedure - incorporating a prebuilt, radio-opaque-marked fenestration - enhances the ability to localize and access the fenestration during interventional procedures. This low-cost, reproducible approach facilitates future catheter-based interventions, reduces procedural time, and may improve long-term management of Fontan patients.

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Author Contributions All authors contributed equally to the conception, development, and writing of this manuscript.

Data Availability No datasets were generated or analysed during the current study.

Declarations

Competing Interests The authors declare no competing interests.

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