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Original
Transcatheter Closure of a Secundum Atrial Septal Defect with Deficient Aortic Rim Through the Left Internal Jugular Vein in a Child with Situs Inversus and Interrupted Inferior Vena Cava: Device's Choice Matters / Piccinelli, E., Bautista-Rodriguez, C., Aw, T., Bates, O., Fraisse, A.. - In: PEDIATRIC CARDIOLOGY. - ISSN 0172-0643. - 42:8(2021), pp. 1879-1881. [10.1007/s00246-021-02708-9]

Availability:
This version is available at: 11583/2970583 since: 2022-08-10T15:18:03Z

Publisher:
SPRINGER

Published
DOI:10.1007/s00246-021-02708-9

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11 October 2026

Pediatric Cardiology
https://doi.org/10.1007/s00246-021-02708-9

CASE REPORT

Transcatheter Closure of a Secundum Atrial Septal Defect with Deficient Aortic Rim Through the Left Internal Jugular Vein in a Child with Situs Inversus and Interrupted Inferior Vena Cava: Device's Choice Matters

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Received: 9 June 2021 / Accepted: 17 August 2021
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Abstract
Percutaneous closure of secundum atrial septal defect (sASD) in children with interrupted inferior vena cava is challenging, especially in case of deficient aortic rim. Trans-jugular access is generally preferred in this scenario. Patients with situs inversus and sASD also carry technical difficulties for transcatheter closure because of the orientation of the atrial septum. We report a successful case of percutaneous closure of a sASD with deficient aortic rim using an occlutech figulla flex II ASD device through the left internal jugular vein in a child with situs inversus, dextrocardia, and interrupted IVC. This case was facilitated by the absence of left-sided hub of the Occlutech device to provide stable opening of the device into the left atrium, whereas the ball-connection of the delivery system allowed an angle of almost 180 degrees between the device and the atrial septum.

Keywords Interrupted inferior vena cava · Secundum atrial septal defect · Occlutech ASD occluder · Transjugular approach

Introduction

Percutaneous closure is the treatment of choice for secundum atrial septal defect (sASD) [1]. Generally, the procedure is performed using femoral venous access; however, in the case of interrupted inferior vena cava (IVC) with azygos continuation to the superior vena cava (SVC) this approach may be difficult. Alternative accesses such as transhepatic, jugular, or femoral via the azygos vein have been described [2–5]. Patients with situs inversus and sASD are rarely encountered and transcatheter closure is also feasible but carries technical difficulties due to the orientation of the atrial septum [6, 7]. We report a challenging case of percutaneous closure of a sASD in a child with interrupted IVC, situs inversus, and deficient aortic rim using an Occlutech Figulla Flex II ASD device (Occlutech; Helsingborg, Sweden) through the left internal jugular vein.

Case Report

A 5-year-old, 20 kg girl with situs inversus, dextrocardia, interrupted IVC, and history of percutaneous closure of a patent ductus arteriosus at the age of 7 months was under follow-up for a sASD. Although asymptomatic, the last transthoracic echocardiogram (TTE) in the outpatient clinic demonstrated a large defect with deficient aortic rim and right ventricular volume overload. The PDA device was in a good position without residual shunt or left pulmonary artery stenosis. It was then decided to perform elective percutaneous closure of her sASD.

Under general anesthesia and transoesophageal echocardiography (TOE) guidance a 15 mm defect with aortic rim deficiency was seen [Fig. 1]. Through left internal jugular vein (LIJV) access, a 5F left internal mammary catheter

Enrico Piccinelli and Carles Bautista-Rodriguez have contributed equally to this work.

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Published online: 25 September 2021

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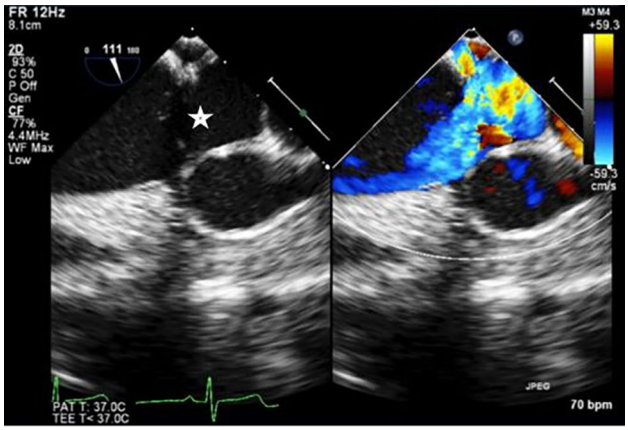

Fig. 1 Transoesophageal echocardiogram confirmed large secundum atrial septal defect (white star) measured at 15 mm with deficient aortic rim

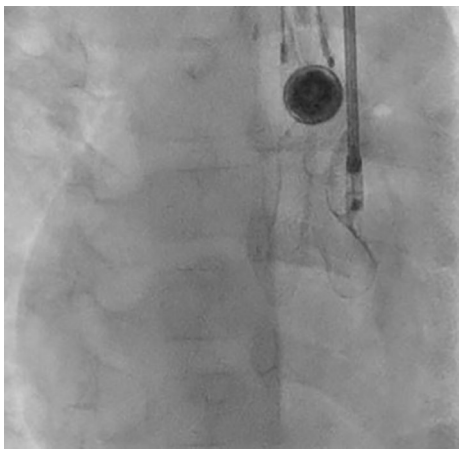

Fig. 2 A 16.5 mm Occlutech Figulla Flex II ASD device was successfully positioned. Note the almost parallel angulation of the device and the Flex Pusher anchored to the Flex II hub

was successfully used to cross the sASD and a semi-rigid 0.035″ diagnostic "J" tip guidewire (Medline International B.V., Arnhem, Netherlands) was positioned in the left ventricle (LV). Because the long sheath of the delivery system (Occlutech; Helsingborg, Sweden) was felt to be too rigid for this case, a 9 Fr Cook flexor sheath (Cook Medical; Bloomington, U.S.A.) was advanced in the LV over the wire while a 16.5 mm Occlutech Figulla Flex II (FFII) ASD device (Occlutech; Helsingborg, Sweden) was loaded in an 9 Fr short sheath. The device was advanced at the distal end of the long sheath. The long sheath was then gently pulled into the left atrium (LA) while the LA disk was opened and then the right atrium (RA) disk was positioned over the defect. The ball-connection between the pusher and the device hub allowed achieving the correct orientation of the left and right atrial disks with the atrial septum [Fig. 2]. Further TOE and TTE images showed a good position of the device that was successfully released after confirmation of stability by pull–push motion [Fig. 3]. Post procedure echocardiography confirmed the excellent procedural result and the patient was discharged home on aspirin.

Discussion

Percutaneous closure of sASD is generally performed via the femoral venous route. However, this may be very challenging in case of interrupted IVC with azygos continuation. Although transcatheter closure through the azygos vein has been reported [5], this may not be possible in cases with large defects and deficient rims. Transhepatic approach is feasible but can be associated with major complications, especially in young patients, such as retro/intraperitoneal bleeding, liver injury, and hemobilia [2]. Consequently, the jugular access is the preferred route to close such sASD due

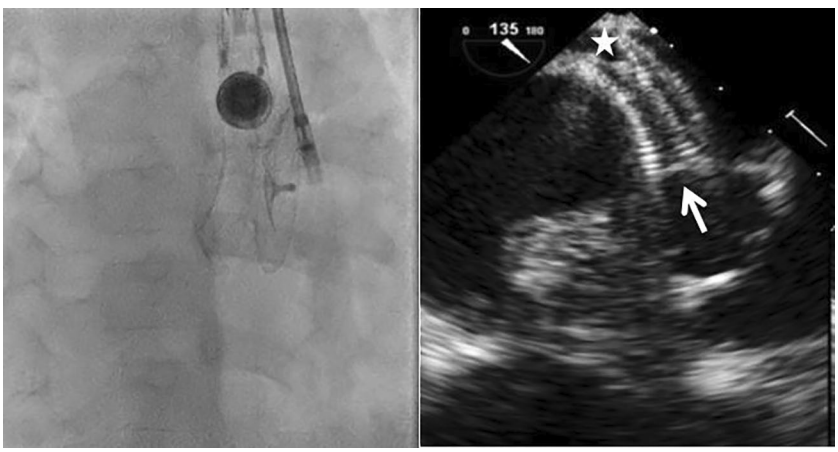

Fig. 3 On fluoroscopy ASD device successfully released after confirmation of stability by pull–push motion. On transoesophageal echocardiogram device in situ with clear posterior rim between the disks (white star) and stable position toward the deficient aortic rim (white arrow)

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to the possibility of accommodating relatively large sheaths and the favorable orientation to the atrial septum [3, 5, 6]. Several types of devices have been implanted using a transjugular approach in children and adults such as Amplatzer Septal Occluder (St. Jude Medical, Inc., Minnesota, USA) [6], Blockaid ASD Occluder (Shanghai shape memory alloy limited Shanghai, China) [4], and Ceraflex Septal Occluder (Lifetech, Shenzen, China) [8]. The main difficulty encountered with this type of procedure is to ensure the correct orientation and alignment of the device with the atrial septum in order to avoid its prolapse in the right atrium. This technical difficulty is even more important in cases with associated situs inversus. Sreekanth Yerram et al. have addressed this problem in a patient with interrupted IVC and situs inversus by opening the left atrial disk in the left ventricle inflow, taking into account the risk of damaging mitral valve leaflets [8]. In the present case, the technical difficulty was almost abolished by using the FFII (Occlutech; Helsingborg, Sweden). The lack of a left-sided hub enabled the FFII to open in a stable way and a round shape during left atrial deployment, without the device prolapsing across the atrial septum. Moreover, the ball attachment connected with a bioptome-like system, allowed a tilted angle of 180 degrees without any stress or tension on the implant and optimal septum alignment [9].

This case report demonstrates that percutaneous closure of sASD with a deficient aortic rim in case of situs inversus, dextrocardia and interrupted IVC with azygos continuation is feasible through transjugular approach. The characteristics of the Occlutech FFII device and its specific delivery set facilitated the successful implantation.

Author Contributions EP, CBR, and AF contributed in the drafting of the manuscript and final approval of the manuscript submitted. TCA and OB revised critically the manuscript and gave final approval of the manuscript submitted.

Declarations

Conflict of interest Alain Fraisse is consultant and proctor for Abbott and for Occlutech. The other four authors have nothing to declare.

Ethical Approval Yes.

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