

CONGENITAL HEART DISEASE

IMAGING VIGNETTE: CLINICAL VIGNETTE

Intravascular Lithotripsy in an Adult With Fontan Pathway Obstruction



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ABSTRACT

Fontan pathway obstruction is a potentially serious complication characterized by an anatomical or functional narrowing anywhere in the cavo-pulmonary pathways. Here, we report the first case in the literature where an innovative Fontan conduit rehabilitation procedure with intravascular lithotripsy was used achieving a dramatic increase in the pathway size. (J Am Coll Cardiol Case Rep 2024;29:102296) © 2024 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

A 28-year-old man with history of tricuspid atresia treated with a fenestrated lateral tunnel Fontan palliation and subsequent Fontan obstruction treated with bare-metal stenting at age 12 years, presented for evaluation of syncope, ascites, and varicose veins. Echocardiographic evaluation revealed a mildly reduced single ventricular function, and abdominal magnetic resonance imaging revealed a cirrhotic liver morphology with stage 3 to 4 fibrosis. Cardiac magnetic resonance imaging raised concerns for Fontan conduit stenosis. Cardiac catheterization confirmed a heavily calcified Fontan conduit with diffuse obstruction, with a minimum diameter of 12 mm. The inferior vena cava pressure was 15 mm Hg, and the superior vena cava pressure was 12 mm Hg, consistent with Fontan pathway obstruction.¹

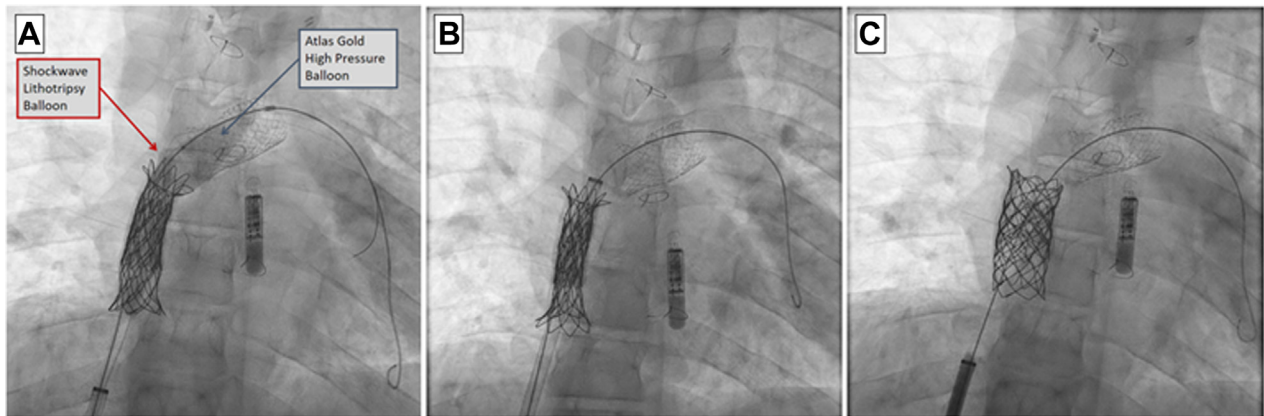
A complex Fontan conduit rehabilitation was performed, sequentially placing an 8-zig 4.5-cm Covered Cheatham-Platinum stent, an 8-zig 5.0-cm Covered Cheatham-Platinum stent, and a bare-metal Palmaz XL 4010 stent without expansion of the Fontan pathway beyond 12 mm because of severe calcification. An 18-mm Atlas Gold high pressure balloon was then used for dilation without success. At this juncture, it was hypothesized that deployment of a shockwave lithotripsy balloon could modulate the heavily calcified wall. To implement this approach, a coronary wire was advanced alongside the existing 18-mm Atlas balloon, over which a 7.0 × 60-mm diameter shockwave lithotripsy balloon was advanced. Subsequently, 3 independent dilations of the balloon system were executed. After removal of the shockwave balloon, the Atlas balloon was redilated to 16 atm with successful conduit expansion to 21 mm (**Figure 1**).

Fontan pathway obstruction is a potentially reversible cause of Fontan circulatory failure. Clinical manifestations range from reduced exercise tolerance to Fontan circulatory failure. Given that the Fontan operation is often performed at a very early age, utilizing conduits that may not be sufficiently sized to support adequate venous flow in adulthood, a proactive approach with frequent assessments with cross-sectional imaging should be considered in these patients to assess and define possible stenotic conduits. A recent retrospective

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the [Author Center](#).

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FIGURE 1 Intravascular Lithotripsy in an Adult With Fontan Pathway Obstruction

(A) Angiography demonstrating the 7.0 × 60-mm diameter shockwave lithotripsy balloon positioned alongside the 18-mm Atlas high-pressure balloon during modulation of the heavily calcified conduit. (B) Fontan stent after high-pressure dilation with unsuccessful expansion beyond a diameter of 12 mm caused by severe calcification. (C) Final appearance following complete Fontan conduit rehabilitation with lithotripsy and dilation, obtaining a 75% increase in the pathway size, to a diameter of 21 mm.

study² demonstrated that percutaneous Fontan stenting is associated with a significant reduction in Fontan gradients with normalization of original conduit diameters and improvement in NYHA functional class in one-half of symptomatic patients.

Although percutaneous stenting and surgical pathway replacement are the current treatment options, the emergence of intravascular lithotripsy as a novel technique for the treatment of vascular calcification presents a promising adjunct to the current therapies. The successful use of lithotripsy shockwave balloon in our case highlights the potential efficacy of intravascular lithotripsy in treating calcific Fontan pathway obstruction and suggests that this technology could potentially be applied in other areas of structural and congenital intervention for larger, heavily calcified conduit rehabilitation. This case presents an exciting direction for future options in the percutaneous treatment of calcific Fontan pathways and highlights the synergistic merging of interventional therapies to treat unique patient populations.

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