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Imaging and Case Report

# Infradiaphragmatic Total Anomalous Pulmonary Venous Return With Congenital Diaphragmatic Hernia: The Gut or the Heart—A Surgical Conundrum

Aamisha Gupta, MD [a,b,*], Kanishka Ratnayaka, MD [a,b], John Nigro, MD [b,c], Howaida El-Said, MD [a,b]

[a] Department of Pediatrics, Division of Cardiology, Rady Children's Hospital, San Diego, California; [b] Department of Pediatrics, University of California, San Diego School of Medicine, San Diego, California; [c] Department of Cardiovascular Surgery, Rady Children's Hospital, San Diego, California

Total anomalous pulmonary venous return (TAPVR) with diaphragmatic hernia (DH) is rare, with a mortality rate of up to 80%.[1,2] In infradiaphragmatic (ID) TAPVR alone, there are previous reports of successful ductus venosus (DV) stenting to alleviate obstruction until definitive repair.[3,4] We present the unconventional management of left-sided DH and ID-TAPVR. Three-dimensional (3D) vascular reconstruction and multidisciplinary collaboration led to a staged approach with transcatheter wire securement and eventual stent implantations in the DV and a horizontal segment between the umbilical vein (UV) and hepatic portal venous egress (HPVE), followed by surgical decompression of the bowel, and later successful cardiac repair with an excellent outcome. To our knowledge, our case is the first transcatheter palliation of a patient with both DH and ID-TAPVR via this pathway.

## Case report

A 2.5-kg infant with left-sided DH and ID-TAPVR was taken to the cardiac catheterization laboratory on day of life (DOL) 5. The previously formatted 3D-imaging reconstructions were overlaid during pertinent procedural aspects. A 4F sheath was placed in the right femoral vein and an 8F sheath (bench-tested) was placed in the UV. Via the 8F sheath, a 0.014-inch guide wire (Ironman, Abbott Vascular) was positioned through the DV and anchored in the left innominate vein. A second guide wire (BMW, Abbott Vascular) and microcatheter (Renegade, Boston Scientific) were positioned through the same 8F sheath through the vertical vein and anchored in the distal right pulmonary vein, with a wedge angiogram used for confirmation. After the wire placement was secured in both the DV and vertical vein, DH decompression was performed by a general surgeon and the abdomen was temporarily closed with Integra-BioMesh (SurgiMend). Simultaneous kissing stents (Formula 418, Cook Medical) were implanted in the DV and HPVE (Figure 1A) to ensure no obstruction with future abdominal closure.

Over 2 weeks, stepwise abdominal approximation at 5-day intervals was followed by the final abdominal wall closure on DOL 15 by a general surgeon. Surgical ID-TAPVR repair was performed on DOL 27 by a cardiac surgeon (J.N.). At the time of ID-TAPVR repair, the DV stent portion that protruded into the right atrium was ligated. The patient was discharged home at 2.5 months of age, on enoxaparin injections for 6 months owing to initially observed dilation of the portal venous system and smoke appearance by angiogram, not for stent prophylaxis. The 6-month follow-up computed tomography scan demonstrated no residual flow through the remaining stent and a significant decrease in portal venous system dilation with unobstructed pulmonary veins (Figure 1B, Supplemental Video 1); therefore, anticoagulation was discontinued.

## Discussion

The concomitant diagnoses of DH and ID-TAPVR represent a challenge, given the mortality rate of up to 80%.[1,2] In our case, the true conundrum was that the ID-TAPVR was not yet obstructed but had the potential to be, with the DH complicating the picture of which comorbidity to first address for optimal success of both surgical repairs. Understanding vascular anatomy with 3D imaging was vital for transcatheter intervention. Multidisciplinary collaboration in the catheterization laboratory with wire placement within the DV and HPVE allowed for safe bowel decompression with line securement. Simultaneous stenting of the DV and HPVE was performed through the same 8F sheath in the UV to ensure patency and avoid potential jailing and potential future compression with abdominal wall closure. Formula 418 stents were used, as they are low-profile stents that could both fit through the 8F sheath over the previously placed 2 wires and be dilated in the future if needed.

*Keywords:* diaphragmatic hernia; infradiaphragmatic TAPVR; stent.

* Corresponding author: agupta@rchsd.org (A. Gupta).

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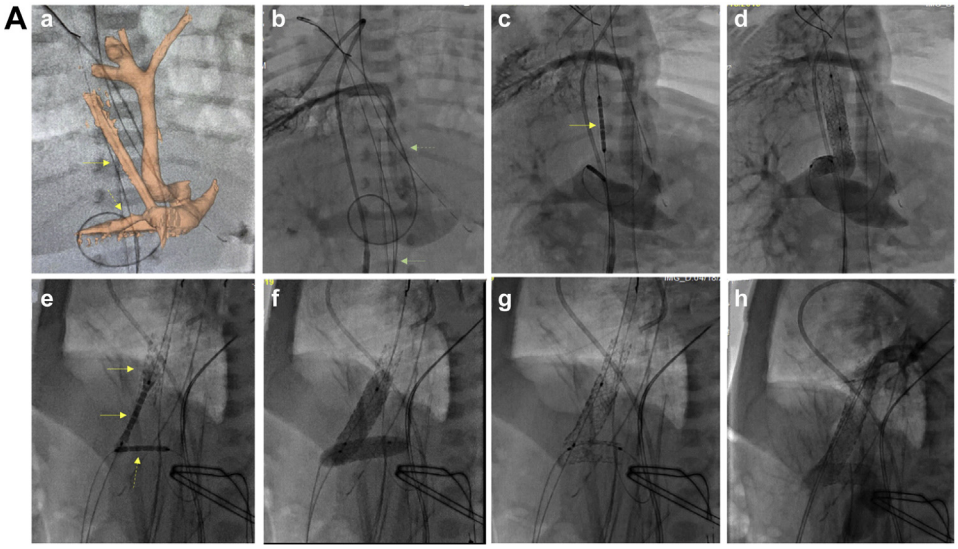


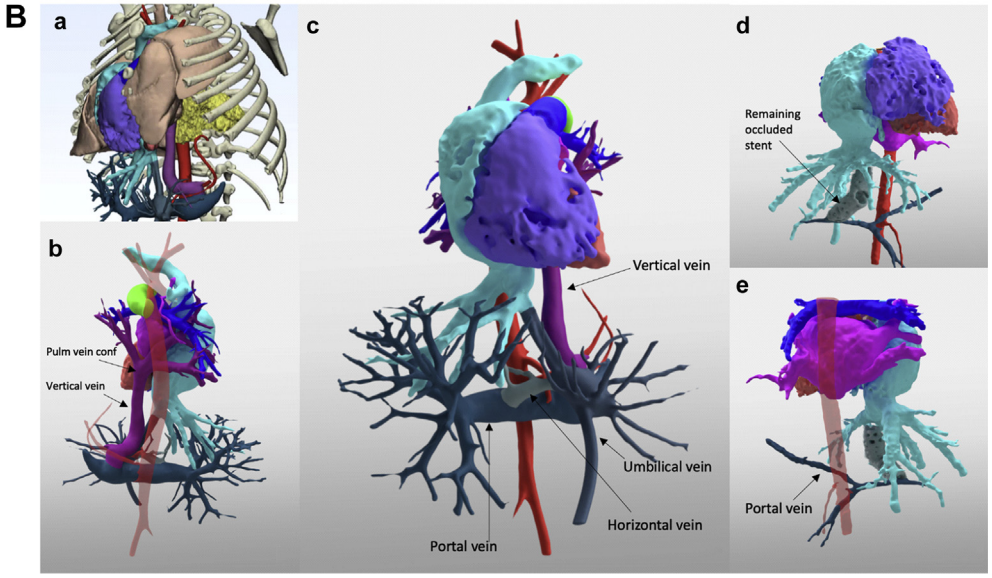


**Figure 1. (A) Angiograms and fluoroscopy**. (a) Overlaid computed tomography image to minimize contrast injections. (b) Wire through umbilical vein (green arrows) and through vertical vein during pulmonary vein angiogram. (c, d) 6 mm × 20 mm Formula 418 stent and overlying 7 mm × 20 mm Formula 418 stent in ductus venosus, 6 mm × 16 mm Formula 418 stent in hepatic portal venosus egress; all ballooned to 12 atm (e-h) lateral view. Yellow arrows point to wire placement. **(B) Three-dimensional segmentation and print**. (a) Left diaphragmatic hernia. (b) Posterior view and (c) anterior view of total anomalous pulmonary venous return with vertical vein entering dilated portal venous system. (d) Anterior view and (e) posterior view on follow-up, showing residual occluded stent in the ductus venosus; note significant decrease in the size of the portal vein.

Therefore, we solved our conundrum, with interventional stent placement in the DV and HPVE, which allowed for stabilized cardiac flow below the diaphragm during the final repair of the DH, allowing for ultimate relief of the intrathoracic lung space to optimize pulmonary and bowel physiology for successful ID-TAPVR surgery.

## Conclusion

Our unconventional management resulted in an excellent outcome in a scenario with historically high mortality. The role of 3D imaging demonstrates the importance of comprehending the anatomy before intervention. This remains a high-risk procedure, and each case should be evaluated closely. Consideration of this pathway provides a possible solution in these rare and complex cases.

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## Declaration of competing interest

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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## Ethics statement

This report adheres to the ethical guidelines put forward by Rady Children's Hospital and University of California, San Diego.

## Supplementary material

To access the supplementary material accompanying this article, visit the online version of the *Journal of the Society for Cardiovascular Angiography & Interventions* at https://doi.org/10.1016/j.jscai.2022.100390.

## References


1. Graziano JN. Cardiac anomalies in patients with congenital diaphragmatic hernia and their prognosis: a report from the Congenital Diaphragmatic Hernia Study Group. *J Pediatr Surg*. 2005;40(6):1045–1050. https://doi.org/10.1016/j.jpedsurg.2005.03.025
2. Montalva L, Lauriti G, Zani AL. Congenital heart disease associated with congenital diaphragmatic hernia: a systematic review on incidence, prenatal diagnosis, management, and outcome. *J Pediatr Surg*. 2019;54(5):909–919. https://doi.org/10.1016/j.jpedsurg.2019.01.018
3. Burkhardt BE, Stiller B, Grohmann J. Stenting of the obstructed ductus venosus as emergency and bridging strategy in a very low birth weight infant with infradiaphragmatic total anomalous pulmonary venous connection. *Catheter Cardiovasc Interv*. 2014;84(5):820–823. https://doi.org/10.1002/ccd.25560
4. Hope KD, Lahiri S, Qureshi AM. Venous duct stenting in a 940-gram infant with infradiaphragmatic total anomalous pulmonary venous return. *Cardiol Young*. 2020;30(10):1501–1503. https://doi.org/10.1017/S1047951120002188