Cardiology in the Young

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Partially unroofed coronary sinus in an infant with pulmonary vein stenosis: mixed blessing

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Brief Report

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Abstract

Transcatheter intervention on the pulmonary vein often requires performing transseptal puncture, which is a risky procedure. We describe a rare association of a partially unroofed coronary sinus with pulmonary vein stenosis causing desaturation in a young infant where the presence of the unroofed coronary sinus allowed for a novel interventional approach to the stenotic left pulmonary vein, avoiding the high risk of transseptal puncture.

Case presentation

A 3-month-old male weighing 4.8 kg was born with truncus arteriosus, interrupted aortic arch, and persistent left superior caval vein draining into the right atrium via what was initially thought to be fully roofed coronary sinus. He underwent surgical repair on day 10 of life consisting of end-end arch repair, ventricle septal defect closure, and placement of right ventricle-pulmonary artery homograft. Immediately after repair, stents were placed in the proximal right and left pulmonary arteries due to severe stenosis. Due to the persistent low oxygen saturation (in the mid-80s), a bubble study echocardiogram was performed which revealed that the coronary sinus was partially unroofed. A cardiac CT scan 3 weeks post-repair showed stenosis of the left pulmonary artery proximal to the stent, as well as normal size common right and common left pulmonary veins. An echocardiogram at 3 months of age revealed increased estimated right ventricle systolic pressure, increased flow velocity in the branch pulmonary arteries, flow turbulence in the common left pulmonary vein, no atrial septal defect, dilated coronary sinus ostium, and unroofed distal coronary sinus with the unroofed segment measuring 7 mm in length (Figs 1 and 2). He was taken to the cardiac catheterisation lab for possible intervention on the stented pulmonary arteries. The right femoral vein and artery were accessed using a 5-Fr sheath and a 20-gauge catheter, respectively. Haemodynamic measurements showed elevated distal pulmonary artery pressure with a mean of 26 mmHg and an elevated calculated pulmonary vascular resistance (9.8 Wood units $\times$ m^2). Balloon angioplasty was then performed in the right and left pulmonary artery stents, along with the placement of an additional stent in the proximal left pulmonary artery to treat the aforementioned stenosis. The left pulmonary artery wedge angiogram showed severe ostial stenosis of the common left pulmonary vein (Fig 3A). Attempts to cross the atrial septum (probing the foramen ovale) were unsuccessful. Knowing the patient had an unroofed coronary sinus, we elected to attempt left atrial access via the coronary sinus. A coaxial system consisting of a 5-Fr JR4 guide catheter (Medtronic), 4-Fr JR 2 catheter (Cook Medical), and 0.035" angled glide wire (Terumo) was inserted through the right femoral vein sheath into the coronary sinus. The catheters were oriented superiorly and rightward. The wire was advanced to cross the coronary sinus into the left atrium. The catheters were then advanced over the wire inside the left atrium. The angled glide wire was exchanged for a 0.014" Choice wire (Boston Scientific). The catheters were turned toward the common left pulmonary vein. The Choice wire was manipulated to cannulate the common left pulmonary vein. The JR2 catheter was then advanced over the wire into the left lower pulmonary vein (Fig 2). Pressure measurements revealed a gradient of 16 mmHg across the stenosis. Angiograms showed severe stenosis of the left pulmonary vein just distal to the ostium and the normal pulmonary vein measuring 4.2 mm (Fig 3B). The Choice wire was exchanged for a stiffer coronary wire [0.014" Asahi Grand Slam wire (Abbott)] to provide more support for pulmonary vein stent placement. Then, the 5-Fr guide catheter was advanced over the JR2 catheter inside the vein and the JR2 catheter was removed. A 4 $\times$ 8 mm Synergy everolimus-eluting stent (Boston Scientific) was advanced over the wire inside the guide catheter and centred across the stenotic portion. The stent was deployed by inflating the balloon to 18 atmospheres (the rated burst pressure) to achieve a stent diameter slightly larger than 4 mm to match the normal distal pulmonary vein segment. Angiography revealed a well-positioned stent (Fig 3C). Pressure measurement showed no gradient across the stent. No significant improvement in his oxygen saturation was noticed at the end of the procedure. The

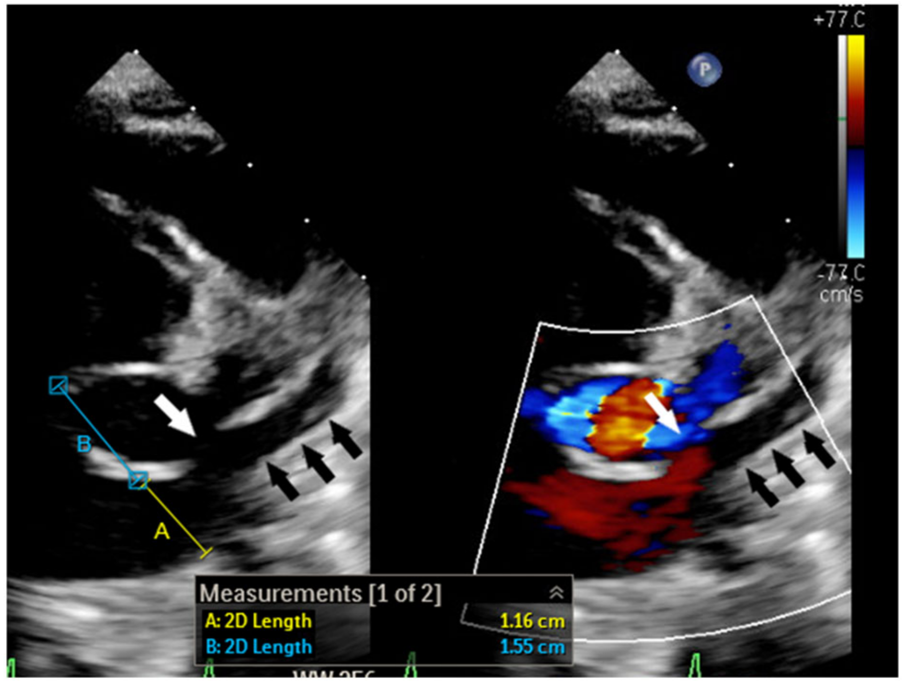

Figure 1. 2-Dimensional echocardiogram with and without colour shows dilated coronary sinus ostium (**A**) with short septal length (**B**) and partially unroofed coronary sinus (black arrows). The unroofed segment is depicted with the white arrow.

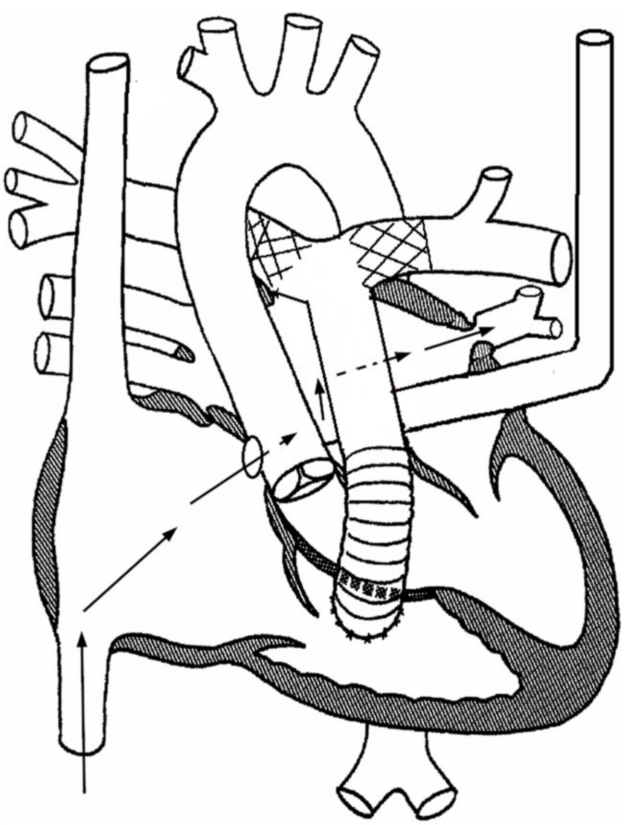

Figure 2. Schematic diagram explaining the anatomy. Black arrows depict the course that was utilised to access the stenotic pulmonary vein. The catheters were advanced from the right atrium coronary sinus ostium unroofed coronary sinus left atrium left upper pulmonary vein.

patient was discharged home on the same home dose of aspirin and started on Sildenafil. Over the next few weeks following the procedure, his oxygen saturation has increased to 93–95%.

Discussion

Despite the advancement in surgical and transcatheter treatments, the prognosis of extensive primary pulmonary vein disease continues to be poor with an estimated mortality rate of 40–50% at 1 year.[1–4] When the atrial septum is intact, transcatheter pulmonary vein intervention requires creating an atrial septal defect which is challenging and carries substantial risks, especially in young infants.[5] Moreover, the presence of the dilated coronary sinus further complicates the safety of the procedure as the septal length becomes significantly shorter (Fig 1).[6] In the described case, the coaxial system of the 5-Fr JR 4 guide, 4-Fr Jr 2 catheter, and guidewires allowed us to cross the unroofed coronary sinus into the left atrium and the stenotic common left pulmonary vein, with relative ease. In addition, the 5-Fr guide catheter (with an outer diameter of 1.67 mm) enabled us to deploy the stent without the need to exchange for a long sheath. Due to the antiproliferative effect, drug-eluting stents slow the progression of pulmonary vein disease when compared with bare-metal stents.[4] Although exclusive balloon angioplasty is an attractive treatment option, the likelihood of complete occlusion is higher when compared with stent placement.[7] As our patient has a common left pulmonary vein, occlusion would result in complete loss of the physiologic function of the entire left lung which could lead to persistent pulmonary hypertension. Additionally, the presence of a stent will ease transcatheter recanalization should future occlusion occurs as the stent is easily visualized by fluoroscopy. For the future management of the patient, we are planning to perform serial dilation, and we anticipate the need to intentionally fracture the stent to accommodate for somatic growth if a larger diameter is needed.[4]

Many features make our case unique. The association of pulmonary vein stenosis in patients with truncus arteriosus and unroofed coronary sinus has never been described in the literature.[2,3] Additionally, the exact mechanism of pulmonary vein stenosis is unknown. However, it falls under the category of primary pulmonary vein stenosis as it affected a vein that has never been operated on.[1] The presentation of pulmonary vein disease was late, as imaging

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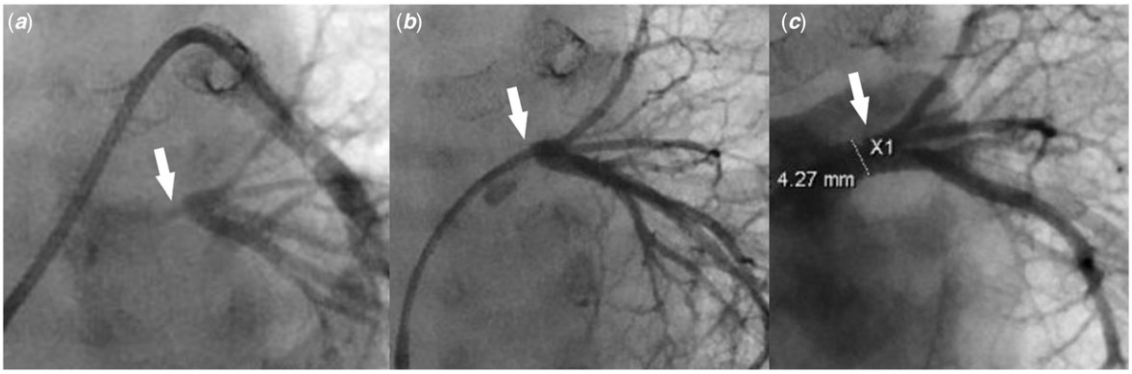

Figure 3. (***a***) Pulmonary artery wedge angiogram shows severe left common pulmonary vein ostial stenosis (white arrow). (***b***) Left common pulmonary vein angiogram shows severe left common pulmonary vein stenosis (white arrow) with well-developed distal pulmonary vein branches. (***c***) Left pulmonary vein angiogram post-stent placement shows a well-positioned stent (white arrow) inside the left common pulmonary vein with good expansion, resolution of the stenosis, and no evidence of vascular injury or jailing of the segmental left pulmonary veins.

3 weeks later post-repair showed no pulmonary vein stenosis. Primary pulmonary vein stenosis in patients with associated CHD is usually either identified prior to or immediately after the surgical repair and is usually attributed to the technique of repair.[2,3] Furthermore, the presence of an unroofed coronary sinus allowed for safe access to the left atrium without the need to create a new atrial septal defect. To our knowledge, this is the first time such an approach has been described.

Conclusion

Trans-coronary sinus access in patients with an unroofed coronary sinus should be considered for intervention on the left atrium or pulmonary vein if no other atrial communication is present. Large-scale studies are needed to guide the management and improve the outcome of pulmonary vein stenosis in children

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References

1. Vanderlaan RD, Rome J, Hirsch R, Ivy D, Caldarone CA. Pulmonary vein stenosis: treatment and challenges. J Thorac Cardiovasc Surg 2021; 161: 2169–2176.
2. Feins EN, Ireland C, Gauvreau K, et al. Pulmonary vein stenosis: anatomic considerations, surgical management, and outcomes. J Thorac Cardiovasc Surg 2021; 5223: 1492–1496.
3. Kalfa D, Belli E, Bacha E, et al. Outcomes and prognostic factors for postsurgical pulmonary vein stenosis in the current era. J Thorac Cardiovasc Surg 2018; 156: 278–286.
4. Khan A, Qureshi AM, Justino H. Comparison of drug eluting versus bare metal stents for pulmonary vein stenosis in childhood. Catheter Cardiovasc Interv 2019; 94: 233–242.
5. Srinivas BC, Singla V, Reddy B, Nagesh CM, Nanjappa MC. Percutaneous transseptal mitral commissurotomy in a patient with absent right superior vena cava and aneurysmally dilated coronary sinus. Cardiovasc Interv Ther 2013; 28: 419–421.
6. Muller MJ, Backhoff D, Schneider HE, et al. Safety of transseptal puncture for access to the left atrium in infants and children. Pediatr Cardiol 2021; 42: 685–691.
7. Buiatti A, von Olshausen G, Martens E, et al. Balloon angioplasty versus stenting for pulmonary vein stenosis after pulmonary vein isolation for atrial fibrillation: a meta-analysis. Int J Cardiol 2018; 254: 146–150.