CASE REPORT

An unconventional valvuloplasty: severe pulmonary stenosis with 'candy cane' inferior vena cava

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SUMMARY

Femoral vein is the usual approach to balloon pulmonary valvuloplasty (BPV). However, alternative access may be mandated in some peculiar situations. We hereby report a 10-year-old patient with symptomatic severe valvular pulmonary stenosis with interrupted inferior vena cava which was successfully treated with transjugular BPV. Technical considerations for this approach as well as other possible options for such patients are discussed.

BACKGROUND

Isolated pulmonary stenosis is a common diagnosis and percutaneous valvuloplasty is the dominant mode of management in most patients.¹ The usual approach to balloon pulmonary valvuloplasty (BPV) is via the femoral vein.¹ Infrequently, we may encounter associated anomalies or special situations where an alternative access is warranted. Herein we present a 10-year-old patient with symptomatic severe valvular pulmonary stenosis with interrupted inferior vena cava (IVC) which was successfully treated with BPV via right internal jugular vein (IJV) approach.

CASE PRESENTATION

A 10-year-old girl presented to us with a history of insidious onset and gradually progressive intolerance to outdoor playing activities with peer group since last 5 years. Clinically, she was acyanotic with left parasternal heave, a loud pulmonary ejection click and a grade 4 late peaking ejection systolic murmur at base. Transthoracic echocardiogram confirmed the diagnosis of valvular pulmonary stenosis (PS) with peak gradient of 90 mm Hg across the pulmonary valve (PV) and annulus measuring 16 mm in diameter (figure 1A). There were no other associated structural heart defects noted. She was planned for percutaneous BPV.

Under aseptic conditions and conscious sedation, the right femoral arterial access was taken with a paediatric 5 Fr sheath. The left ventriculogram ruled out any ventricular septal defect. The right femoral venous access was taken with a paediatric 6 Fr sheath to introduce a 6 Fr Berman catheter (Arrow, USA). The catheter could not enter the right atrium (RA), rather it kept going towards the left side of the superior vena cava (SVC) until meeting an obstruction. Angiography in the IVC revealed an interrupted IVC with azygous continuation draining into the right SVC (figure 2A), which we had missed on echocardiogram. No wire, end-hole

or balloon-tipped catheter could even reach the PV despite repeated manipulation (figure 2B) due to multiple severe bends enroute right ventricular outflow tract (RVOT) from the femoral vein and severe right ventricular hypertrophy. BPV by an alternative approach was mandated.

TREATMENT

A transjugular approach to BPV was contemplated. The right IJV was accessed through a 7 Fr sheath under ultrasonographic guidance. The pulmonary wedge catheter (6 Fr, Arrow) easily floated with a balloon from the RA to the RV apex, but a 180° turn from the RA to the RVOT posed difficulty in advancement; finally, the catheter could be manoeuvred across the RVOT with a wire support and a deflated balloon. A J-tip, 0.035 in (0.89 mm), 260 cm, extra stiff wire (Amplatz ES, Cook Medical) was parked in the left pulmonary artery (PA) (figure 3A). An anteroposterior and lateral RV angiogram was done by monorailing the 5 Fr multitrack catheter (Numed) (figure 3B). A 17×30 mm Tyshak II balloon was gently passed from the RA to cross the PV annulus. Repeated balloon slippage necessitated multiple inflations till the desired achievement of inflation to the disappearance of waist (figure 3C). The multitrack catheter confirmed fall of the RVOT gradient from 96 to 22 mm Hg post-dilation. Haemodynamics remained stable throughout the procedure. The post-procedure echo showed a significant fall in the RVOT gradient, mild pulmonary regurgitation, without any harm to the tricuspid valve apparatus (figure 1B).

OUTCOME AND FOLLOW-UP

The patient was discharged on the second post-cath day. On outpatient follow-up at 4 weeks, the mother reported substantial symptomatic improvement and repeat echo was similar to the postoperative echo. A clinical/echocardiographic follow-up is planned after 6 months.

DISCUSSION

Prevalence of isolated PS is 7%–9% of all congenital heart disease.¹ Kan *et al* revolutionised the management of this disorder by performing first percutaneous balloon pulmonary valvuloplasty almost 35 years ago.² Since then, BPV is considered treatment of choice for most patients with isolated PS with surgery being reserved for rare cases where BPV is not successful.¹

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Figure 1 Prevalvuloplasty mean gradient of 94 mm Hg across the pulmonary valve (A), which fell to 29.5 mm Hg post-BPV (B). Dilated HV draining into an enlarged RA misinterpreted as a normal venous drainage (C). BPV, balloon pulmonary valvuloplasty; HV, hepatic veins; RA, right atrium.

The absence of hepatic segment of IVC with azygous continuation in the right or left SVC is a rare anomaly seen in 0.6%–2% of congenital heart defects, most commonly associated with visceral heterotaxy syndromes (up to 86%) and rarer when situs is normal.³ Transthoracic/transoesophageal echocardiography is usually sufficient for the diagnosis of this disorder, but the presence of dilated hepatic veins directly draining into the RA may mislead the echocardiographer like in our case (figure 1C), specially, if subxiphoid positioning of aorta and IVC relative to spine, and colour doppler, are not carefully checked.

Chaara *et al* reported the right IJV access for pulmonary valvuloplasty in severe RV failure.⁴ Transjugular balloon dilation of the PV⁵ and the PA⁶ has since been reported. Rarely, transaxillary⁷ and transhepatic⁸ approaches have been used for BPV. The most frequent indication for an alternative route access was severe RV failure, others being femoral vein occlusion, postoperative Glenn shunt and inability to enter the PA via a femoral route.^{4–8} Kim and colleagues have reported transumbilical BPV in a newborn with critical PS and an interrupted IVC.⁹ Our case is first such report in literature where transjugular BPV was mandated due to an interrupted IVC. The more sinuous course via the axillary vein, risk of intraperitoneal bleeding with a transhepatic access, plus familiarity of the jugular venous access for catheterisation, made us to choose this approach. The risk of pneumothorax, hydrothorax, vascular injury, and nervous injury, is minimised by ultrasound guidance.^{5,6}

Figure 2 Interrupted IVC with azygous continuation to the SVC giving rise to the characteristic 'candy cane' appearance (A). Tortuous course and RV hypertrophy made the RVOT access difficult (B). IVC, inferior vena cava; RV, right ventricle; RVOT, right ventricular outflow tract; SVC, superior vena cava.

Figure 3 Successful transjugular access to BPV—extra-stiff wire parked in the pulmonary artery (A). Lateral RV angiogram showing the pulmonary leaflet doming and landmarks for BPV (B). Balloon dilation till the disappearance of waist (C). BPV, balloon pulmonary valvuloplasty; RV, right ventricle.

There were multiple challenges in this procedure. First, safe IJV catheterisation was achieved under ultrasonographic guidance. Second, the passage of a wire/balloon-tip catheter across the RVOT was difficult due to a 180° loop, RV hypertrophy and dilation—timely deflation of the balloon just as the catheter tip faces the RVOT followed by careful steering across the RVOT, worked after a few trials. Third and technically most significant, was balloon instability, despite using an extra-stiff wire, because balloon control was difficult due to the wire loop and catheter in right heart. Backward traction shortened the wire loop rather than preventing distal balloon slippage, unlike infemoral approach. This difficulty was also faced by Joseph *et al* in their series,⁵ but as proposed by them, taking a longer balloon size or a stiffer balloon type was not a feasible option in our small-sized patient.

Learning points

- Absence of hepatic segment of inferior vena cava (IVC) with azygous continuation in the right or left superior vena cava is a rare congenital anomaly, and is rarer when situs is solitus.
- Transthoracic/transoesophageal echocardiography is helpful for its diagnosis, but the presence of dilated hepatic veins directly draining into the right atrium may mislead the echocardiographer to interpret it as a normal venous drainage. Subxiphoid positioning of aorta and IVC relative to spine, and course of IVC, must be carefully checked.
- Right internal jugular vein approach for balloon pulmonary valvuloplasty is a safe, effective, and viable alternative to the conventional transfemoral access.
- This approach may be considered as the preferred auxiliary route or even first choice in situations like iliofemoral obstruction, IVC obstruction/interruption, severe right ventricular failure with gross tricuspid regurgitation, difficult pulmonary valve crossing, postoperative Glenn shunt or post-atrial switch in transposed great arteries.

Competing interests None declared.

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