**INTRODUCTION**

Congenital pulmonary stenosis (PS) is the most common cause of right ventricular outflow tract (RVOT) obstruction. The morphology of the pulmonary valve (PV) can vary between typical pliable doming valve to significantly dysplastic valve having a very complex morphology.\(^1\) Balloon pulmonary valvuloplasty (BPV) is the standard of care for the management of PS irrespective of valve morphology. However, the success rate of BPV in severe dysplastic PV is inferior to a pliable PV. Here, we report a case of successful BPV in a very severe dysplastic PV restenosis with difficulty in negotiating the balloon across the valve.

**CASE REPORT**

A 2-year-old male child was diagnosed as a case of severe PS and referred to us for management. His two-dimensional echocardiography showed severe PS, peak gradient of 112 mmHg, and annulus diameter of 9 mm with normal right ventricular (RV) function. BPV was performed using valver balloon (Balton®, Poland) of size 10 mm × 40 mm over 0.018” wire without any complications. However, there was a residual gradient of 40 mmHg at the end, and it was decided to have clinical and echocardiographic follow-up of the patient at 6-month interval. At his first follow-up, there was severe valvular restenosis with a peak gradient of 92 mmHg. A detailed echocardiography was performed which showed a significantly thickened and dysplastic PV, a noncapacious muscle bound RV, and no significant poststenotic dilation of the pulmonary artery (PA). Thus, options of surgery versus redo-BPV were considered. After a detailed discussion with the child’s family and after taking informed consent, the baby was taken up for redo-BPV.
RV angiography revealed dysplastic PV (thickness 4.8 mm), annulus of 9 mm without dilatation of distal main PA [Figure 1a]. A 7 Fr access was obtained in the right femoral vein. The PV was crossed with a straight tip Radifocus® guidewire (Terumo, Japan) with 5 Fr MPA diagnostic catheter. The Radifocus® wire was exchanged with 0.035” Amplatz super stiff (Cook® Medical, USA) guidewire which was placed in the distal branch of the right PA. A TYSHAK-II® balloon (NuMed, USA) 12 mm x 40 mm was advanced over the super stiff wire. However, due to the dysplastic valve anatomy and nondilated main PA along with the fibrosis because of restenosis, the balloon could not be crossed across the valve. Multiple attempts with a balloon of smaller size (10 mm x 40 mm) and minor modifications in technique including changing the wire position, failed to cross the valve. Caution was undertaken to prevent entering into RV through the tricuspid valve apparatus, which could prevent tracking the balloon over the wire. Any attempt to push the balloon led to complete assembly falling back in RV, and thus, it was decided to gain extra support to the wire. A 7F AMPLATZER™ TorqVue™ 180° patent ductus arteriosus (PDA) device delivery sheath (St. Jude Medical, USA) as an off-label use was tracked over the wire across the valve and placed in the proximal PA. The TYSHAK-II 12 mm x 40 mm balloon was advanced through the sheath and was placed at its distal end. The sheath was then withdrawn back below the PV into the RVOT, thus positioning the balloon across the PV. The balloon was inflated, but post-procedure, there was a residual gradient of 40 mmHg. Thus, another TYSHAK-II balloon size 14 mm x 40 mm was placed across the PV [Figure 1b] and inflated till the total disappearance of the waist but lead to balloon rupture [Figure 1c and Video 1]. The residual gradient across the valve was 10 mmHg with RV systolic pressure of 65 mmHg and PA systolic pressure of 55 mmHg. The residual gradient on day 3 of the procedure was 36 mmHg, which was predominantly infundibular. Currently, at 1 year of follow-up, he is doing fine with residual RVOT gradient of 30 mmHg with mild pulmonary regurgitation (PR).

DISCUSSION
Dysplastic PV accounts for 10%–20% of the PS patients,[2] The success rate of BPV is lesser in dysplastic valves due to various factors such as myxomatous and thickened valve, relatively lesser commissural fusion, and associated hypoplastic pulmonary annulus and main PA. Nevertheless, considerable success has been achieved in such cases despite the above reasons with case series reporting up to 65% success.[3,4] We consider our case to be unique in view of the following findings. An extremely thickened (4.8 mm) PV, noncapacious muscle bound RV, and no poststenotic dilation PA made the procedure very difficult as the super stiff 0.035” wire could not provide the support for tracking the desired size balloon across the valve. After trying all possible ways to do the procedure, we had to use a novel way (PDA device delivery sheath) to provide support to our BPV balloon for crossing PV. As the sheath is braided and the dilator has a tapered distal end, it was able to negotiate the PV with some difficulty only making our procedure successful.

We used the final size of the balloon for dilation to be 14 mm x 40 mm, as we did not get an optimum result after dilation with 12 mm x 40 mm size balloon (residual gradient of 40 mmHg), and a balloon of size 13 mm x 40 mm was unavailable. We wanted to size the balloon/annulus ratio to be 140%[5] but actually oversized the balloon/annulus ratio to 155%. It could be seen best in Figure 1c wherein as compared to Figure 1a, which shows with respect to PA, the size of the balloon is much larger. This may lead to RVOT rupture or dissection in PA, which may be catastrophic. However, no such complication occurred in our patient, and to our surprise, there was no PR immediately after the procedure. The proximal edge of the balloon was abutting the distal tip of the sheath used for supporting the balloon. At the peak of inflation of balloon, the distal end of the sheath might have led to the rupture of the balloon. The sheath could have been withdrawn into the RV cavity but at the possible cost of the losing support to balloon position which was not preferred. However, this did not lead to any complication such as PA dissection, balloon material distal embolization, or inability to retrieve the balloon into the sheath.

We also understand that such improvisation may sometimes lead to complications, but as there was no other option left for the operator, the above mentioned technique and balloon sizing was used.

CONCLUSION
In cases with severe dysplastic PV, BPV can be significantly challenging because of extremely thickened PV and other associated anatomical abnormalities. We used a PDA device delivery sheath to obtain support to track
our balloon across such an extremely dysplastic PV and also used a balloon/annulus ratio of 155% to obtain a successful result of BPV.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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