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Commentary: Congenital absence of a pulmonary valve cusp: A rare entity with too many solutions

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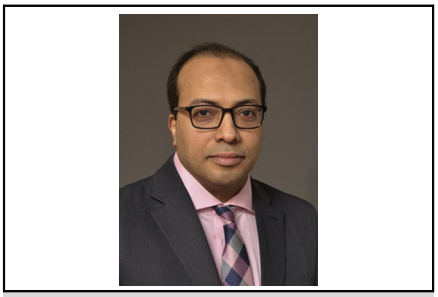

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CENTRAL MESSAGE

Congenital absence of a single semilunar cusp is a rare entity that can occur in isolation but more commonly with other heart defects. Several treatment options are available.

Another face of a congenitally bicuspid pulmonary valve (PV) is congenital absence of a PV cusp. This is a truly rare entity as an isolated anomaly, but it is most commonly associated with other cardiac defects such as ventricular septal defect in the setting of tetralogy of Fallot with absent PV syndrome, where the cusps of the PV are actually present albeit rudimentary and the pulmonary artery and its branches are quite dilated. This has been described in association with atrial septal defect,[1] and patent ductus arteriosus[2] as well, and has been described in the pediatric and the adult populations. There are very limited reports, however, in the literature describing this rare anomaly in isolation and it is also most commonly associated with some degree of dysplasia or abnormalities in the other PV cusps. The same abnormality has been reported with the aortic valve where one cusp can be absent, resulting in significant aortic regurgitation.[3]

Inoue and colleagues[4] reported an interesting case of a 66-year-old woman presenting with right-sided heart failure. A thorough evaluation revealed moderate PV regurgitation secondary to isolated absence of a PV cusp that was discovered intraoperatively. The patient also had severe mitral valve regurgitation. The authors performed mitral valve replacement and PV repair by creating a third leaflet using bovine pericardium.

Several points are worth mentioning about this case:

1. An interesting finding is the presence of completely well-developed 2 PV cusps and lack of any evidence of a third cusp such as rudimentary tissue or nubbin at the annulus.
2. The late presentation: it is difficult to know if the pulmonary regurgitation resulting from absence of a cusp resulted in the patient's symptoms as the degree of regurgitation was only moderate, but it could be exacerbated by concomitant severe mitral stenosis, which may further explain the late presentation.
3. One would expect the ability to identify this PV abnormality preoperatively rather than intraoperatively, especially with the current advances in imaging modalities. This is always helpful for surgical planning and preoperative discussion with the patient.
4. The long-term durability of a single PV cusp reconstruction is unknown.

Despite the rarity of this anomaly, several techniques have been described to address it surgically and vary between repair or replacement. Different materials have been also described to reconstruct the missing cusp, such as bovine pericardium, homograft monocusp,[5] and autologous tissue such as the pulmonary artery wall.[6] The long-term outcomes of these different repair strategies, however, remain to be determined.

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