Systolic excursion of the leaflets of the truncal valve: An unusual mechanism for pulmonary stenosis in common arterial trunk

Saurabh Kumar Gupta1, Abhinav Aggarwal1, Gurpreet S Gulati2, Sivasubramanian Ramakrishnan1, Shyam S Kothari1, Anita Saxena1, Sanjiv Sharma2, Balram Airan3, Robert H Anderson4

1Department of Cardiology, All India Institute of Medical Sciences, New Delhi, India, 2Department of Cardiovascular Imaging and Endovascular Intervention, All India Institute of Medical Sciences, New Delhi, India, 3Department of Cardiothoracic and Vascular Surgery, All India Institute of Medical Sciences, New Delhi, India, 4Institute of Genetic Medicine, University of Newcastle, Newcastle upon Tyne, UK

INTRODUCTION

Common arterial trunk is a rare congenital cardiac malformation characterized by the presence of a single arterial trunk exiting the ventricular mass through a common ventriculo-arterial junction. The arterial trunk gives rise directly to the systemic, pulmonary, and...
coronary circulations.\textsuperscript{[1-3]} In this setting, the pulmonary vasculature is exposed to systemic systolic pressures. A lower vascular resistance in the lungs permits a relatively higher flow of blood to the lungs. The relentless excessive flow of blood to the lungs predisposes to the rapid development of pulmonary vascular disease, usually rendering the patients inoperable early in childhood. Based on the relative sizes of its components, the common arterial trunk can be classified as having either aortic or pulmonary dominance.\textsuperscript{[3,4]}

In some patients, obstruction in the pulmonary outflow can protect the pulmonary vasculature. In the majority of such cases, the obstruction is due to the narrowing of the orifice of the pulmonary arterial (PA) segment, while in others, the right or left pulmonary arteries may be stenosed. In our prospective study of 70 patients with common arterial trunk, we identified 27 patients with aortic dominance and sinusal origin of the PA segment. Among these patients, 5 had a truncal valvar leaflet obstructing the flow of blood to the lungs.\textsuperscript{[4]} In this study, we have sought to clarify morphologic characteristics leading to this novel mechanism of protecting the pulmonary vasculature.

**METHODS**

We prospectively studied 70 patients with common arterial trunk from July 2015 to May 2018 following clearance from the institute ethics committee. From the 70 patients in our initial study,\textsuperscript{[4]} we selected 27 patients with an aortic dominant common arterial trunk and sinusal origin of the PA segment for this analysis. A detailed account of methods used for physiologic and morphologic analysis is published elsewhere.\textsuperscript{[4]}

All patients underwent echocardiography and computed tomographic (CT) angiography. In selected cases, we also performed cardiac catheterization for hemodynamic assessment. The anatomy of common arterial trunk was assessed on CT angiographic datasets, which were analyzed using a syngo.via workstation (Siemens, Germany). The relative positions and measurements of the truncal root, and the origin of the pulmonary arteries, were assessed after obtaining a standard double-oblique view.\textsuperscript{[5,6]} This double-oblique technique provides an “enface” view of the truncal valve as seen from the feet of the patient. Having obtained the double-oblique view, we designated a clock face such that the position of noon and 6 o’clock corresponded to anterior and posterior locations, while 3 and 9 o’clock positions represented left and right locations, respectively. On this basis, the clock face was divided into the right anterior, left anterior, left posterior, and right posterior quadrants. In all cases, the analysis was supplemented by virtual dissection using Horos (Pixmeo, Switzerland), a Mac-based open source software, thus permitting us to achieve enhanced three-dimensional intracardiac visualization.\textsuperscript{[7]}

The number and orientation of the sinuses were assessed using the clock face designation. Each hour of the clock was taken to be equivalent to 30° of the overall circle. Sinuses were deemed asymmetric if their size differed by more than 30° of the circle. We named the sinuses according to their spatial orientation within the truncal root.

**RESULTS**

Among the 27 patients with aortic dominance and sinusal origin of the PA segment, 18 patients had trisinusate, 7 patients had quadrisinusate, and 2 had bisinusate truncal valves. In 5 (19%) of the patients with trisinusate valves, the pulmonary orifice was obstructed by the systolic excursion of a truncal valvar leaflet [Figure 1 and Videos 1, 2]. In all these patients, the valvar sinuses were asymmetric, with the left sinus being the largest, occupying from 1 to 2 o’clock to 6–7 o’clock in the circumference of the trunk. In all, the orifice of the PA segment arose from 2 to 4–6 o’clock and was confined to the largest sinus [Figure 2]. The pulmonary orifice was obstructed by the systolic excursion of the relatively large leaflet of the largest truncal valvar sinus producing a gradient of up to 50 mmHg across the orifice of the pulmonary outflow [Table 1].

None of the cases with quadrisinusate and bisinusate truncal valves had pulmonary stenosis. Among the other 13 patients having trisinusate truncal roots without obstruction...
pulmonary stenosis, the sinuses were symmetrical in 6, while in 7, the pulmonary orifice arose from a smaller sinus with a relatively smaller leaflet, thus leaving it unobstructed [Figures 2 and 3].

Our first patient in whom we detected this finding was a 6-year-old girl. The obstruction to the pulmonary orifice was demonstrated both by echocardiography and CT angiography [Figures 1 and 4]. Her oxygen saturation in room air was 88%. Cardiac catheterization, performed because of her age, revealed a systolic gradient of 37 mm of mercury (mmHg) across the PA orifice. The pressures in the pulmonary arteries measured 70/42/56 mmHg, compared to aortic pressures of 107/39/69 mmHg. The Qp/Qs ratio was 2.3, while the estimated pulmonary vascular resistance indexed to body surface area was 7.8 WU.m² [Table 2]. The findings were confirmed during surgical repair, which was achieved by using a 16-mm aortic homograft to connect the PA segment to the right ventricle and closure of interventricular communication. At 24 months of follow-up, she is in New York Heart Association functional Class I and has good effort tolerance.

Echocardiography has revealed normal biventricular function. The right ventricular systolic pressure, as predicted based on the tricuspid regurgitation, was normal. Cardiac catheterization confirmed normalization of the PA pressures at 35/12/18 mmHg [Table 2]. The other 4 patients were younger than 1 year at the time of investigation and were deemed suitable for surgical repair without cardiac catheterization. The surgical repair, however, could not be performed as the parents declined consent.

**DISCUSSION**

In most of the patients with common arterial trunk, unrestricted flow of blood to the lungs leads to the rapid development of pulmonary vascular disease. In some patients, obstructions within the pulmonary outflow protect the pulmonary vasculature. The obstruction is

**Table 1: Demographic and morphologic characteristics of five patients with sinus origin of pulmonary artery segment having pulmonary stenosis induced by truncal valvar leaflet**

| Case | Case 2 | Case 3 | Case 4 | Case 5 |
|------|--------|--------|--------|--------|
| Age (months) | 72 | 3 | 9 | 4 | 5 |
| Sex | Female | Male | Female | Male | Female |
| Oxygen saturation (%) | 88 | 90 | 90 | 92 | 92 |
| Arrangement of atriums | Usual | Usual | Usual | Usual | Usual |
| Interventricular communication | Subtruncal | Subtruncal | Subtruncal | Subtruncal | Subtruncal |
| Aortic arch | Left | Left | Right | Right | Right |
| Morphology of truncal valve | Number of sinuses | Three | Three | Three | Three |
| Size of the sinuses | Asymmetric | Asymmetric | Asymmetric | Asymmetric | Asymmetric |
| Largest sinus | Left | Left | Left | Left | Left |
| Orientation of sinuses* | 1-6, 6-9, 9-1 | 1-6, 6-9, 9-1 | 1-6, 6-9, 9-12 | 2-6, 6-10, 10-2 | 2-7, 7-10, 10-2 |
| Pulmonary outflow | Origin | Common | Common | Common | Common |
| Site of origin | Left sinus | Left sinus | Left sinus | Left sinus | Left sinus |
| Location of the orifice* | 2-5 o’clock | 2-6 o’clock | 2-5 o’clock | 2-6 o’clock | 2-4 o’clock |
| Gradient across orifice | 37 mmHg | 45 mmHg | 40 mmHg | 50 mmHg | 40 mmHg |

*o’clock position based on a multiplanar assessment of CT angiography in short-axis projection of the truncal root after obtaining a double-oblique view of the truncal root.
mostly the result of a narrowed pulmonary orifice or stenosis in the peripheral pulmonary arteries.

Although not included in commonly-used systems for classification,\(^1\)\(^-\)\(^3\) the variant of common trunk in which the PA segment arises from a truncal valvar sinus is becoming recognized with increasing frequency.\(^4\)\(^-\)\(^10\) The origin of the PA segment from a truncal sinus brings the leaflet supported by the sinus closer to the PA orifice. As highlighted in all our cases, when the pulmonary segment was arising from the largest sinus, the excursion of its leaflet narrowed the pulmonary outflow sufficiently to cause obstruction. We found that such obstruction to flow existed only if the valvar leaflet supported by a truncal sinus was relatively larger than the PA orifice arising from it. Thus, we believe that when the sinuses are small and symmetrical, a smaller leaflet relative to a large PA orifice prevents hemodynamically relevant obstruction to the pulmonary outflow. A relatively small sinus supporting the pulmonary component also explains the lack of pulmonary stenosis in all 7 patients with quadrisinusate truncal roots despite the presence of sinusal origin of the PA segment. In the patients with bisinusate truncal roots, however, the lack of stenosis is more likely related to the relative orientation of the sinus and the PA segment.

Although sinusal origin of pulmonary arteries is being recognized with increasing frequency, to the best of our knowledge ours is the first report of obstruction to pulmonary flow produced by a truncal valvar leaflet.\(^4\)

As revealed by the management of one of our patients, such obstruction may prevent early development of pulmonary vascular disease.

CONCLUSION

In patients with common arterial trunk, origin of the PA segment from a truncal valvar sinus brings the valvar leaflet close to the pulmonary orifice. When large, such a leaflet can obstruct the pulmonary orifice, thus protecting the lung vasculature. These findings have obvious implications for the optimal management of these patients, particularly if they present beyond infancy.

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Conflicts of interest

There are no conflicts of interest.

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**Table 2: Cardiac catheterization at presentation and 24 months after surgical repair**

| At presentation | 24 months follow-up |
|-----------------|---------------------|
| **Saturations** |                     |
| Room air        | Postoxygen SaO\(_2\) (PaO\(_2\)) | |
| SCV             | 63                  | 76 (50) | 68 |
| RA              | 62                  | 94 (70) | 68 |
| RV              | 80                  | 100 (275) | 99* |
| PA              | 88                  | 99 (230) | 99 |
| LA              | 99                  | 10 (38)  | 99 |
| LV              | 88                  | 107 ed 15 | 115 ed 10 |
| Aorta           | 107/39 | 110 ed 15 | 115/40/73 |
| **Pressures**   |                     |
| RA              | Mean 7             | Mean 7 | Mean 5 |
| RV              | 102 ed 10          | 110 ed 10 | 38 ed 8 |
| PA              | 70/42/56           | 70/38/54 | 35/12/18 |
| LA              | Mean 15            | Mean 15 | Mean 10* |
| LV              | 107 ed 15          | 110 ed 15 | 115 ed 10 |
| Aorta           | 107/39/69          | 110/42/73 | 115/50/75 |
| **Calculated variables** |         |
| Qpi             | 4.73               | 9.29    | 2.05 |
| Qsi             | 2.8                | 2.49    | 2.05 |
| Qpi/Qsi         | 2.3                | 3.73    | 1.0  |
| PVRI (WU.m\(^2\)) | 7.8        | 4.2     | 2.92 |
| SVRI            | 28.7               | 26.5    | 34.15 |
| PVRI/SVRI      | 0.29               | 0.16    | 0.08 |

\(\ast\)assumed. *Based on following variables: At presentation: Weight 15 kg, height 118 cm, Hb 13 g/dL, and BSA 0.74 m\(^2\); indexed assumed indexed VO\(_2\) max=137 ml/min for an 8-year-old girl having heart rate 100/min. At 24-month follow-up: Weight 18 kg, height 122 cm, Hb 12 g/dL, and BSA 0.80 m\(^2\); assumed indexed VO\(_2\) max=137 ml/min for an 8-year-old girl having heart rate 90/min. BSA: Body surface area; Hb: Hemoglobin; SCV: Superior caval vein; PA: Pulmonary artery; RA: Right atrium; RV: Right ventricle; LA: Left atrium; LV: Left ventricle; PVRI: Pulmonary vascular resistance index; SVRI: Systemic vascular resistance index
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