

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

ABSTRACT

- Background** : Pulmonary stenosis in patients with common arterial trunk protects the pulmonary vasculature. In our recently published prospective study of common arterial trunk, some patients with sinusal origin of the pulmonary arterial segment had pulmonary stenosis induced by systolic excursion of a truncal valvar leaflet. We aimed to determine the detailed morphologic characteristics of this unusual finding.
- Methods and Results** : All 70 patients underwent echocardiography and computed tomographic angiography as per predefined study protocol. In selected cases, we also performed cardiac catheterization. Among 27 patients with aortic dominance, we found sinusal origin of the pulmonary arterial segment. In 5 of these patients, pulmonary stenosis was induced by systolic excursion of a truncal valvar leaflet. In all these patients, the truncal valve was trisinusate, albeit with asymmetric sinuses. The pulmonary arterial segment arose from the largest left sinus with its relatively large leaflet obstructing the pulmonary orifice during systole. In the remaining 22 patients, without pulmonary stenosis but with sinusal origin of pulmonary arterial segment, the truncal valve was quadrisinusate in 7, bisinusate in 2, and trisinusate in 13. None of the patients with quadrisinusate and bisinusate truncal valves had pulmonary stenosis. Among the 13 patients with trisinusate valves, the sinuses of the truncal valve were symmetrical in 6, while in 7, the pulmonary orifice originated from a smaller asymmetric sinus.
- Conclusion** : Pulmonary stenosis, produced by a relatively large leaflet of an asymmetric truncal sinus, may prevent early development of pulmonary vascular disease. Timely recognition of this unusual mechanism of pulmonary stenosis is important for optimal management.
- Keywords** : Common arterial trunk, computed tomography, virtual dissection

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

Videos Available on: www.annalspc.com

Access this article online

Quick Response Code:



Website:

www.annalspc.com

DOI:

10.4103/apc.APC_22_20

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Gupta SK, Aggarwal A, Gulati GS, Ramakrishnan S, Kothari SS, Saxena A, et al. Systolic excursion of the leaflets of the truncal valve: An unusual mechanism for pulmonary stenosis in common arterial trunk. *Ann Pediatr Card* 2020;13:194-8.

Address for correspondence: Dr. Saurabh Kumar Gupta, Room No. 9, 8th Floor, Department of Cardiology, Cardio-Thoracic Centre, All India Institute of Medical Sciences, New Delhi - 110 029, India. E-mail: drsaurabhmd@gmail.com

Submitted: 17-Feb-2020

Revised: 17-Mar-2020

Accepted: 25-Apr-2020

Published: 29-Jun-2020

coronary circulations.^[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.^[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.^[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,^[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.^[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.^[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.^[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

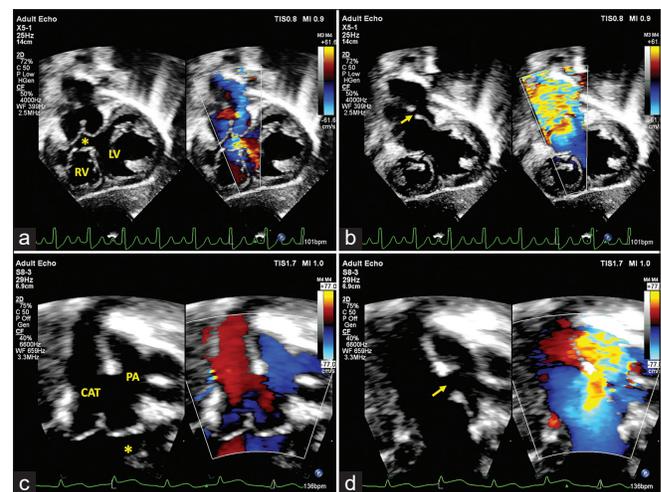


Figure 1: The transthoracic echocardiographic findings from first patient (a and b) and second patient (c and d). In both, with sinusal origin of pulmonary arteries, the pulmonary orifice is widely patent during diastole (a and c) but obstructed (yellow arrow) during systole (b and d) by a truncal valvar leaflet. Note the large subtruncal interventricular communication (*). CAT: common arterial trunk; LV: Left ventricle; PA: pulmonary arterial segment; RV: Right ventricle. Note that the echo images of the first patient are published as Figure 6 in our previous publication.^[4]

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].^[5]

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.

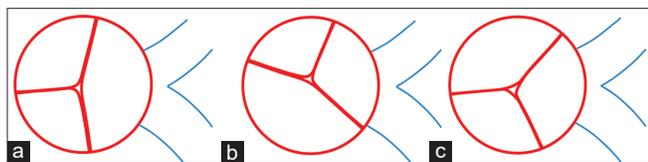


Figure 2: The drawing shows the truncal root as viewed in its short axis in our patients with common arterial trunk, sinusal origin of the pulmonary arterial segment, and a trisinusate valve. Despite a similar circumferential location of the pulmonary arterial segment, from 2 to 5 o'clock as shown in the blue color, the pulmonary outflow is obstructed only if it arises from the largest asymmetric left sinus, as shown in (a). The pulmonary outflow remains unobstructed, if the pulmonary arterial segment arises from a smaller asymmetric sinus (b), or if the valvar sinuses are symmetrical (c)

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

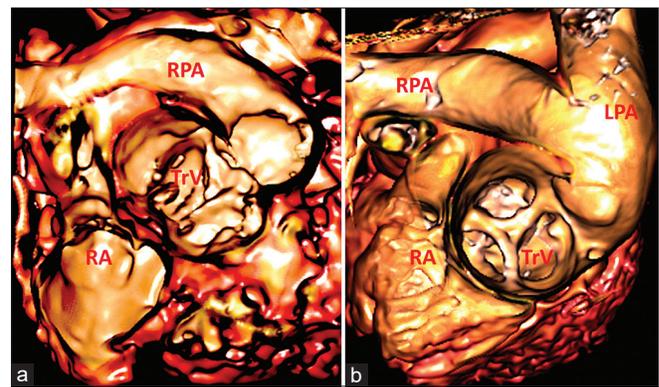


Figure 3: Virtual dissection obtained from diastolic phase of CT angiogram shown in short-axis plane and seen from head end of the patient. (a) Asymmetric trisinusate truncal valve with the pulmonary arterial segment arising from the largest left sinus in patient 1. (b) A trisinusate valve but with symmetric sinuses in a patient with no pulmonary obstruction. LPA: Left pulmonary artery; RA: Right atrium; RPA: Right pulmonary artery; TrV: truncal valve

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

	Case 1	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	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	12-6, 6-9, 9-12	2-6, 6-10, 10-2	2-7, 7-10, 10-2
Pulmonary outflow					
Origin	Common	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

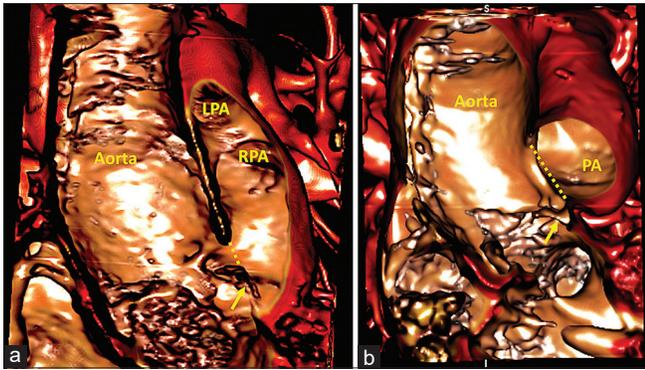


Figure 4: Virtual dissection obtained from systolic phase of computed tomography angiogram shown in anteroposterior projection. (a) Sinusal origin of the pulmonary arterial segment and narrowing of the pulmonary outflow (yellow dashed line) by a truncal valvar leaflet (yellow arrow) in patient no. 1. (b) Unobstructed pulmonary orifice despite sinus origin of the pulmonary arterial segment in the patient shown in right hand panel of Figure 3. LPA: Left pulmonary artery; PA: Pulmonary arterial segment; RPA: Right pulmonary artery

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

	At presentation		24 months follow-up
	Room air	Postoxygen SaO ₂ (PaO ₂)	
Saturations			
SCV	63	76 (50)	68
RA	62		
RV	80		
PA	88	94 (70)	68
LA	99	100 (275)	99*
LV	88		
Aorta	88	99 (230)	99
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

*assumed. [#]Based on following variables: At presentation: Weight 15 kg, height 118 cm, Hb 13 g/dL, and BSA 0.74 m²; Indexed assumed indexed VO₂ max=142 ml/min for a 6-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 m²; assumed indexed VO₂ 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

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.^[4,8-10] 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 sinus 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 sinus 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.

Financial support and sponsorship

The study was funded by the Pediatric Cardiac Society of India and the Indian Council of Medical Research.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

- Collett RW, Edwards JE. Persistent truncus arteriosus; a classification according to anatomic types. *Surg Clin North Am* 1949;29:1245-70.
- Van Praagh R, Van Praagh S. The anatomy of common aorticopulmonary trunk (truncus arteriosus communis) and its embryologic implications. A study of 57 necropsy cases. *Am J Cardiol* 1965;16:406-25.

3. Russell HM, Jacobs ML, Anderson RH, Mavroudis C, Spicer D, Corcrain E, *et al.* A simplified categorization for common arterial trunk. *J Thorac Cardiovasc Surg* 2011;141:645-53.
4. Gupta SK, Aggarwal A, Shaw M, Gulati GS, Kothari SS, Ramakrishnan S, *et al.* Clarifying the anatomy of common arterial trunk: A clinical study of 70 patients. *Eur Heart J Cardiovasc Imaging* 2019;jez255. doi: 10.1093/ehjci/jez255.
5. Parodi J, Berguer R, Carrascosa P, Khanafer K, Capunay C, Wizauer E. Sources of error in the measurement of aortic diameter in computed tomography scans. *J Vasc Surg* 2014;59:74-9.
6. Asch FM, Yuriditsky E, Prakash SK, Roman MJ, Weinsaft JW, Weissman G, *et al.* The need for standardized methods for measuring the aorta: Multimodality core lab experience from the GenTAC registry. *JACC Cardiovasc Imaging* 2016;9:219-26.
7. Gupta SK, Spicer DE, Anderson RH. A new low-cost method of virtual cardiac dissection of computed tomographic datasets. *Ann Pediatr Cardiol* 2019;12:110-6.
8. Adachi I, Uemura H, McCarthy KP, Seale A, Ho SY. Relationship between orifices of pulmonary and coronary arteries in common arterial trunk. *Eur J Cardiothorac Surg* 2009;35:594-9.
9. Gupta SK, Kothari SS, Gulati GS, Nair VV, Rajashekar P, Airan B. The trunk with a twist: Right sinus origin of pulmonary arteries in a child with common arterial trunk. *World J Pediatr Congenit Heart Surg* 2014;5:615-9.
10. Phelps CM, Da Cruz E, Fagan T, Younoszai AK, Tissot C. Images in cardiovascular medicine. Anterior origin of the main pulmonary artery from the arterial valvar sinus: Unusual truncus arteriosus. *Circulation* 2009;119:624-7.