Cardiology in the Young

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Totally anomalous "double drainage" of pulmonary venous confluence: case report with embryological aspects

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Brief Report

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Abstract

Double drainage of the confluence of all four pulmonary veins is extremely rare. We present the image findings in a child with double drainage of the pulmonary venous confluence into the coronary sinus and left superior caval vein with co-existent right superior caval venous stenosis.

Total anomalous pulmonary venous connection accounts for less than 5% of all congenital cardiac anomalies.[1] It has been classified into supra-cardiac, cardiac, infra-cardiac, and mixed types.[2] Double drainage of the confluence of all four pulmonary veins is extremely rare and has previously been scarcely reported in literature. We present the image findings in a child with double drainage of the pulmonary venous confluence into the coronary sinus and left superior caval vein with co-existent right superior caval venous stenosis and briefly describe the embryological aspects of pulmonary venous development.

Case report

Three-month male infant was brought to the emergency with breathing difficulty and bluish discolouration for 1 week. On examination, the child had central cyanosis, tachypnoea, and laboured breathing. His chest radiograph showed cardiomegaly with septal lines. Transthoracic echocardiography showed enlarged right-sided chambers, pulmonary trunk, left and right pulmonary arteries. Ostium secundum type of interatrial communication was seen with small-sized left atrium. The coronary sinus was dilated with flow directed into the right atrium. Drainage of pulmonary veins into the left atrium was not seen. His cardiac CT showed pulmonary veins from both sides joining to form a common pulmonary venous confluence in the sagittal body plane, which drained into left superior caval vein and coronary sinus (Figs 1 and 2). The right superior caval vein showed significant stenosis at its insertion into the right atrium.

Discussion

The development of pulmonary veins begins with the development of intrapulmonary venous plexuses in the bifurcating tracheo-bronchial tube which develops from the ventral aspect of primitive foregut.[3] A midline venous channel (in sagittal body axis) develops within the mediastinal tissues at 5 weeks of gestation and drains the developing intrapulmonary venous plexuses from both lungs. This pulmonary venous confluence (also labelled as common pulmonary vein in literature) located within dorsal mesocardium canalises to open into the atrial cavity, committing the pulmonary venous drainage between the pulmonary ridges into the primitive atrium (later into left atrium after development of primary atrial septum).

Total anomalous pulmonary venous connection results from complete failure of connection between the primitive intrapulmonary venous plexus and the common pulmonary vein (pulmonary venous confluence) or common pulmonary vein and left atrium. As a result, there is persistence of connection between the primitive pulmonary venous system, common pulmonary vein (pulmonary venous confluence), and cardinal venous system (which later develops into systemic veins). Mixed type of total anomalous pulmonary venous connection (defined as drainage of pulmonary veins into systemic venous side at multiple levels) is rare, accounting for less than 5% of patients with total anomalous pulmonary venous connection.[4] The embryological basis of mixed drainage can be possibly explained by an insult at earlier period in gestation, resulting in persistence of aberrant connections at more than one level.[5]

An extremely rare variant of total anomalous pulmonary venous connection, which is not separately classified in widely used "Darling" system, is "double drainage" type (being currently classified as a variant of mixed total anomalous pulmonary venous connection). In this type,

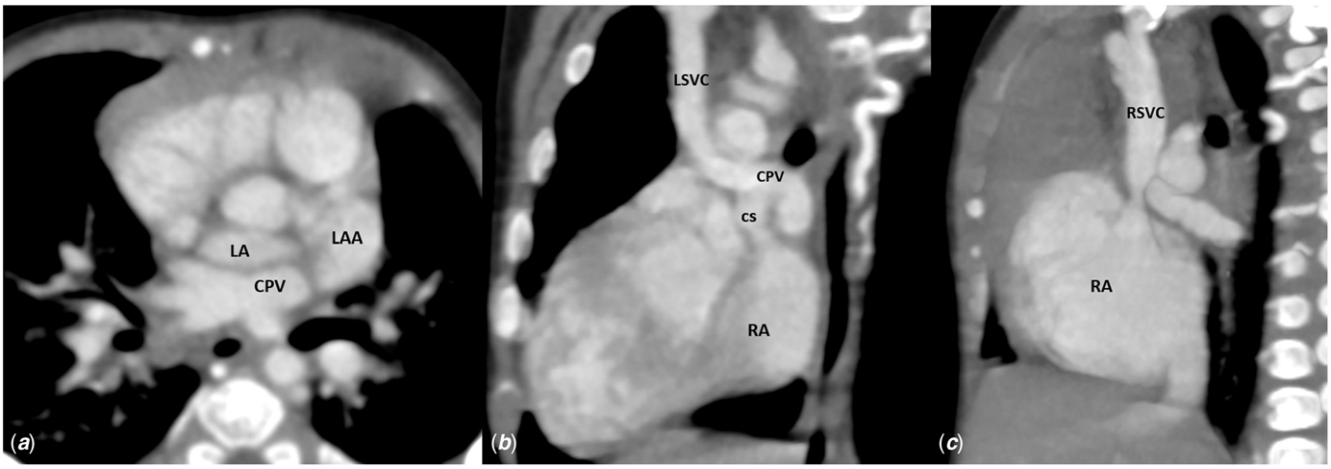

Figure 1. Cardiac CT (***a***, axial section, ***b***, oblique sagittal section, ***c***, sagittal section at right atrial aspect) showing all four pulmonary veins joining to form common pulmonary vein (marked as CPV, pulmonary venous confluence) draining into left superior vena cava (LSVC, left superior caval vein) and coronary sinus (CS). Right superior vena cava (RSVC, right superior caval vein) shows significant anatomic stenosis close to its drainage into right atrium (RA). LA = left atrium; LAA = left atrial appendage.

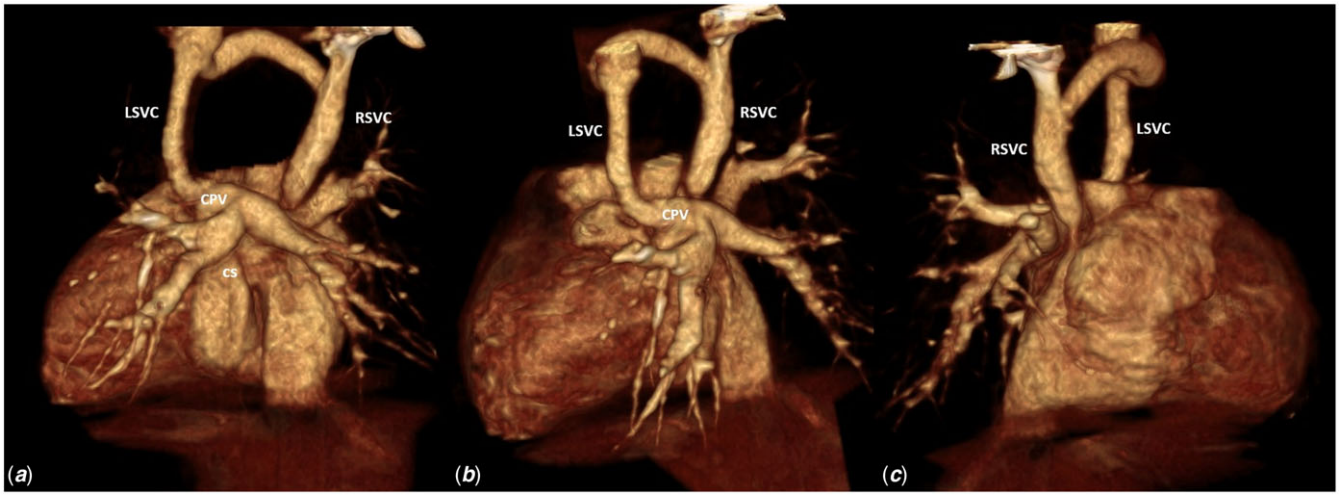

Figure 2. Cardiac CT (Volume rendered images seen from ***a***, posterior aspect, ***b***, left posterior oblique aspect, ***c***, right anterior oblique aspect) showing all four pulmonary veins joining to form common pulmonary vein (marked as CPV, pulmonary venous confluence) draining into left superior vena cava (LSVC, left superior caval vein) and coronary sinus (CS). Right superior vena cava (RSVC, right superior caval vein) shows significant anatomic stenosis close to its drainage into right atrium.

which is extremely rare, all four pulmonary veins are traditionally described to drain into a common confluence, which further drains into two or more sites in systemic venous side. Dual drainage could be explained embryologically on the basis of initial collateral communications with the developing superior cardinal veins on both sides of the fetus. Careful observation of prior reported cases in literature shows that the vascular channel described in these patients as a common confluence is not a true single tubular channel in the sagittal body axis, which could have developed from an embryological confluence.[4–7] Our patient had double drainage of a single vascular channel in sagittal body axis, draining all four pulmonary veins (a pattern most fitting to the embryologically described common pulmonary vein) into the left superior caval vein and coronary sinus.

The most common sites of double drainage reported in literature are coronary sinus and superior caval vein through left innominate vein via left vertical vein.[8] Drainage has also been reported to other sites like left brachiocephalic vein and right superior caval vein; portal vein and right superior caval vein and via two orifices into left superior caval vein.[4,7,9] Our patient also had a co-existent significant right superior caval vein stenosis close to its entry into the right atrium, thereby leading to obstruction to drainage at supra-cardiac level.

Despite its rare occurrence, it is vital to preoperatively diagnose dual drainage to prevent post-operative persistent left to right shunt and cyanosis after successful surgical repair of one of the sites of total anomalous pulmonary venous connection. Echocardiography which is the initial imaging investigation may fail to identify double drainage of pulmonary veins. Cardiac CT angiography helps in comprehensive evaluation of pulmonary venous drainage including demonstration of length of the confluent channel and presence of morphologic obstruction.[4] Surgical repair aims to establish wide and non-restrictive connection between all pulmonary veins and the left atrium. Variability in drainage sites in mixed type of total anomalous pulmonary venous connection requires an individualised approach for management

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based on sites of drainage. This approach is however different from surgical repair in "double drainage" types and in patients having pulmonary venous confluence. In most of the cases in literature with a similar pattern of dual drainage, the surgical approach included anastomosis of confluence of pulmonary veins to the left atrium with ligation of communication with coronary sinus and left vertical vein.[4,8] Coil embolisation of the vertical vein can also be performed after primary surgical repair.

Conclusion

Totally anomalous "double drainage" of a common pulmonary venous confluence is a rare type of total anomalous pulmonary venous connection with a morphological pattern different from mixed total anomalous pulmonary venous connection. Comprehensive preoperative imaging evaluation with cardiac CT or MR is essential for optimal surgical planning. Surgical repair needs to be individualised based on the pattern of pulmonary venous drainage.

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Conflicts of interest. None.

References

1. Dillman JR, Yarram SG, Hernandez RJ. Imaging of pulmonary venous developmental anomalies. Am J Roentgenol 2009; 192: 1272–1285.
2. Craig JM, Darling RC, Rothney WB. Total pulmonary venous drainage to the right side of the heart: report of 17 autopsied cases not associated with other major cardiovascular anomalies. Lab Invest 1957; 6: 44–64.
3. Anderson RH, Brown NA, Moorman AF. Development and structures of the venous pole of the heart. Dev Dyn 2006; 235: 2–9.
4. Siddharth CB, Yadav M, Bhoje A, Hote MP. Dual drainage total anomalous pulmonary venous connection : a rare mixed variant. Asian Cardiovasc Thorac Ann 2018; 26: 305–307.
5. Krishna MR, Gnanappa GK, Mervis J, Forsey J, Ayer JG, Orr Y. Double drainage of total anomalous pulmonary venous connection: a rare variant of mixed total anomalous pulmonary venous connection. Ann Pediatr Card 2020; 13: 100–101.
6. Arciprete P, McKay R, Watson GH, Hamilton DI, Wilkinson JL, Arnold RM. Double connections in total anomalous pulmonary venous connection. J Thorac Cardiovasc Surg 1986; 92: 146–152.
7. Lee M-L, Wang J-K, Wu M-H, Chu S-H, Lue H-C. Unusual form of total anomalous pulmonary venous connection with double drainage. Pediatr Cardiol 1995; 16: 301–303.
8. Tanabe S, Nakasato M, Suzuki H, Ishikawa A, Hayasaka K. A new form of total anomalous pulmonary venous connection with double drainage. Pediatr Int 2000; 42: 369–371.
9. Nagao H, Tominaga K, Kamei N, Tanaka T. Double drainage of total anomalous pulmonary venous connection revealed after surgical repair of a supracardiac total anomalous pulmonary venous connection. Clin Case Reports 2020; 8: 3012–3016.

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