Cardiology in the Young

cambridge.org/cty

Coexistence of transposition of the great arteries, coarctation of the aorta, and bilateral pulmonary artery hypoplasia

Akif Kavgacı[1], Semiha Terlemez[1], Osman Tunç[2] and Serdar Kula[1]

[1]Department of Pediatric Cardiology, Faculty of Medicine, Gazi University, Ankara-Turkey and [2]BTech Innovation, Ankara-Turkey

Brief Report

Cite this article: Kavgacı A, Terlemez S, Tunç O, and Kula S (2023) Coexistence of transposition of the great arteries, coarctation of the aorta, and bilateral pulmonary artery hypoplasia. *Cardiology in the Young* **33**: 819–821. doi: 10.1017/S1047951122002803

Received: 3 July 2022
Accepted: 1 August 2022
First published online: 30 August 2022

Keywords:
Transposition of the great arteries; pulmonary artery hypoplasia; coarctation of the aorta

Author for correspondence:
Akif Kavgacı, Department of Pediatric Cardiology, Faculty of Medicine, Gazi University, Ankara-Turkey.
E-mail: akifkavgaci@gmail.com

Abstract

Transposition of the great arteries is the most common cyanotic CHD in newborns. This CHD, in which the aorta arises from the right ventricle and the pulmonary artery from the left ventricle, is often accompanied by one or several defects such as atrial septal defect or patent foramen ovale, ventricular septal defect and patent ductus arteriosus, which allow the transition between both parallel circulations. Rarely, the disease may be accompanied by left ventricular outflow tract obstruction (subpulmonary obstruction) and coarctation of the aorta.

We present a highly complicated and unusual transposition of the great arteries patient with critical aortic coarctation and hypoplastic pulmonary arteries with abnormal outflow and course.

Case report

The patient, who was born with a weight of 1675 g in 31 + 6 weeks of gestation, was hospitalised in the neonatal ICU due to cyanosis. The aorta emerged from the right ventricle in the front, and the pulmonary artery arose from the left ventricle in the back, according to the echocardiogram. It was accompanied by small muscular ventricular septal defect, atrial septal defect (secundum, small), and a large patent ductus arteriosus at its classic site. Although the main pulmonary artery was observed to be wide, there was no classical bifurcation and pulmonary artery branching. Although the ascending aorta was mildly hypoplastic, the arcus aorta was determined to be extremely hypoplastic. Through the patent ductus arteriosus, the flow direction was observed from the aorta to the pulmonary artery. The patient was started on prostaglandin infusion. Oxygen saturation of the patient was around 70%, and balloon atrial septostomy was performed at the 8th postnatal hour because the transition from atrial septal defect was restrictive. The catheter angiography performed during septostomy revealed that pulmonary artery branches emerged as two separate hypoplastic branches from different levels of the pulmonary artery. The surgical treatment strategy was then determined using CT imaging. The pulmonary trunk was wider than typical on CT with the transposition of the great arteries, and the pulmonary artery branches were hypoplastically separated from the right and left sides of the more distal part of the pulmonary artery, though at slightly different levels (Fig 1). The diameter of the right pulmonary artery was 2.2 mm, and the diameter of the left pulmonary artery was 3.5 mm. After leaving the left subclavian artery, the arcus aorta was measured to be 2.8 mm in diameter and was classified as tubular hypoplastic (Fig 2). CT images were reconstructed as 3D PDF (Appendix). Within the scope of the study, 3D planning and modelling were carried out using Mimics Innovation Suite 22.0 (Materialize, Leuven, Belgium) software. With the 3D PDF document rendered, hypoplastic pulmonary artery branches were evaluated interactively in detail with their courses and lumen structures (Video). The surgical correction could not be performed because both the patient's weight was 1660 g and the accompanying complicated pathologies were present. It was planned to monitor the patient with palliative approaches until adequate weight gain. The prostaglandin infusion was ceased on the tenth day of the follow-up. Despite the fact that the ductus opening had been thinned, it had not been entirely closed, and the patient had not been desaturated. However, findings of respiratory distress emerged during the patient's follow-up. CT angiographic imaging was performed again to clarify respiratory distress in the patient with normal chest X-ray findings and unremarkable infection parameters in the laboratory. It was observed that the large main pulmonary artery compressed the trachea, and the left pulmonary artery compressed the left bronchus. The patient died due to respiratory distress, acidosis, and related complications before an intervention could be planned.

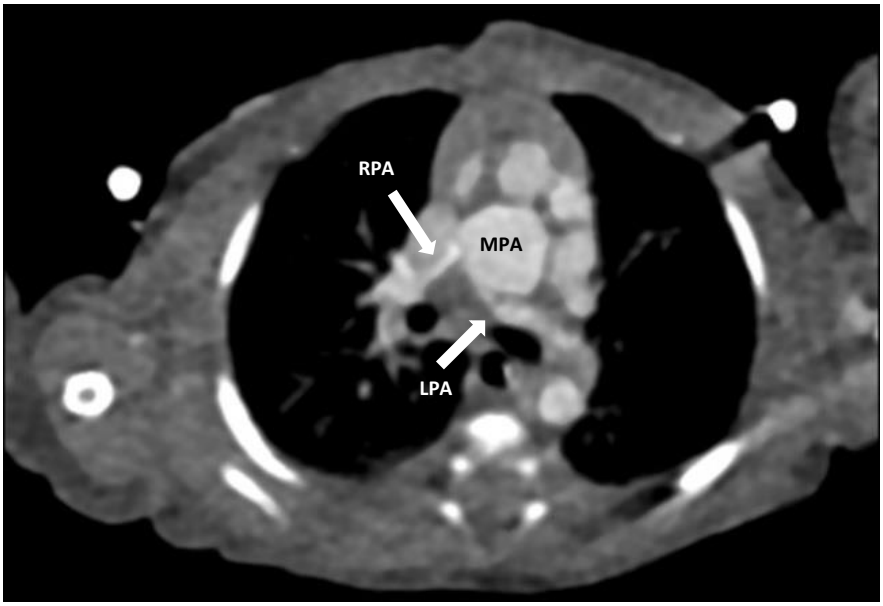

Figure 1. Aneurysmatic appearance in the main pulmonary artery and bilateral peripheral pulmonary stenosis in the evaluation by CT angiography.

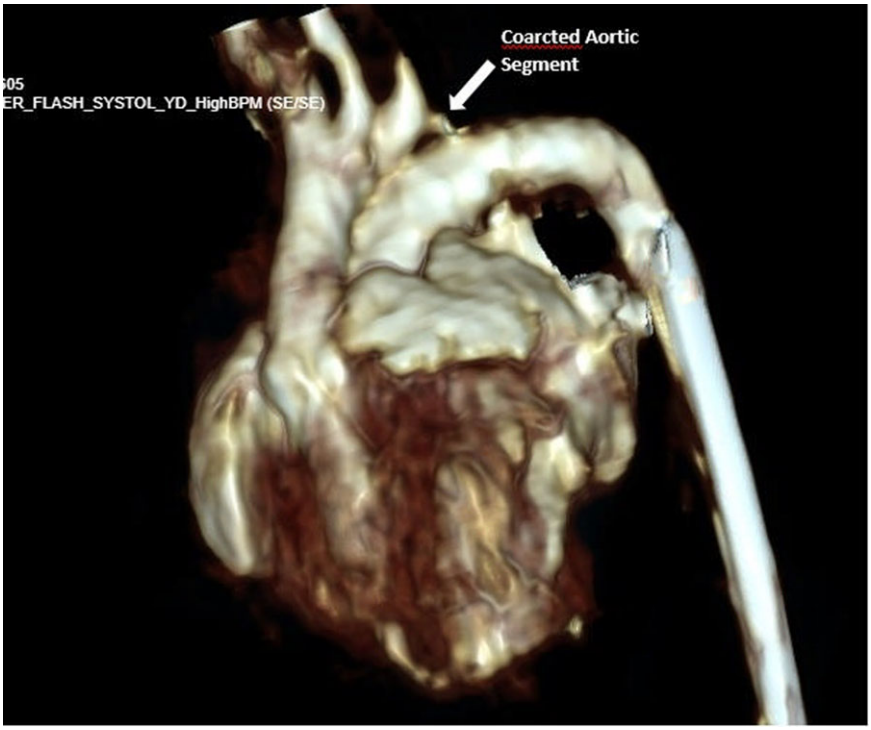

Figure 2. On the image obtained by the 3D reconstruction of CT angiography, the coarcted region in the distal part of the left subclavian artery.

Discussion

Transposition of the great arteries is a heart disease that accounts for about 5–7% of all CHDs and has a very low survival if not treated. Untreated, approximately 30% of infants die within the first week and 90% at the first age.[1]

In 50% of the cases, ventriculoarterial discordance is an isolated finding and this is called simple transposition. However, in complex transposition, many different anomalies such as ventricular septal defect, left ventricular outflow tract obstruction, aortic arch anomalies, and anomalous venous return may accompany.

Although the location and size of ventricular septal defects vary, it is the most common comorbid defect to transposition of the great arteries. In these cases, aortic hypoplasia, coarctation, and even aortic interruption can be detected. In addition, if the outlet septum deviates backwards and to the left, this can cause subpulmonary stenosis.[2] The general approach is to correct any concomitant cardiac anomalies during the surgical correction. In the literature, we did not find a transposition case with both aortic arch anomaly and pulmonary artery anomalies. In our patient, especially, the fact that the pulmonary artery branches had an outflow anomaly and were hypoplastic was the primary factor that made surgical correction impossible (Fig 3). However, the very low weight of the patient led us to consider palliative approaches rather than corrective surgery. Because the patient had significant hypertensive values as a result of coarctation, it was decided to use

https://doi.org/10.1017/S1047951122002803 Published online by Cambridge University Press

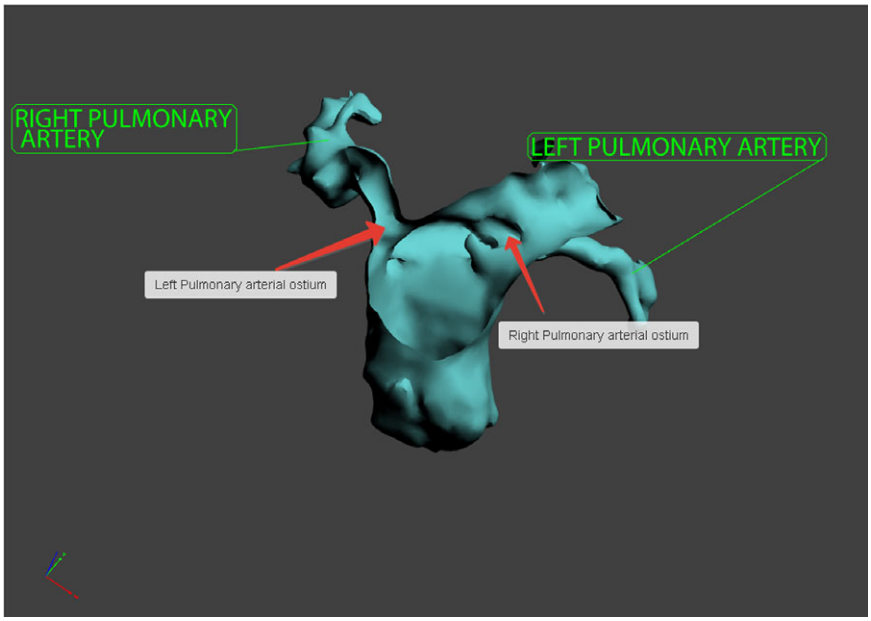

Figure 3. Hypoplastic pulmonary artery branches and ostiums on the image obtained by rendering CT angiography into a 3D PDF.

balloon angioplasty to treat the coarctation. However, since the patient started to bleed intracranially during this period, the procedure was postponed by considering anticoagulant applications during the procedure. Then, bronchial compression and respiratory distress findings emerged. All this caused us to lose the patient without the possibility of treatment. We could have intervened sooner if we had recognised bronchial compression in our patient earlier. We should have intervened with the coarctation at that point. However, we are not sure whether curative treatment could be possible.

Transposition of the great arteries is considered a disease with very satisfying results if surgical correction is achieved. Mortality has significantly decreased with the experience of corrective surgery. Surgical treatment is usually possible in accompanying additional cardiac pathologies. However, as in our patient, some patients may be excluded from all these generalisations. Concomitant cardiac anomalies can be severe, complicated, and unusual. It may be necessary to evaluate each patient individually and make separate decisions in their management.

Supplementary material. To view supplementary material for this article, please visit https://doi.org/10.1017/S1047951122002803

Acknowledgements. The 3D PDF file mentioned in this study is provided as an attachment (Appendix). This document requires Adobe Reader to view it. If no Adobe Reader is available, please click here to download it to your desktop. Then, right-click on the 3D PDF file and open using Adobe Reader.

Financial support. This research received no specific grant from any funding agency, commercial, or not-for-profit sectors.

Conflict of interest. None.

References

1. Wernovsky G. Transposition of the great arteries and common variants. Pediatr Crit Care Med 2016; 17: S337–S43.
2. Martins P, Castela E. Transposition of the great arteries. Orphanet J Rare Dis. 2008; 3: 1–10.