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Rare association of right upper lobe agenesis, dextrocardia, and aberrant hepatic arterial supply

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A 33-year-old woman with previously diagnosed dextrocardia and no other significant medical history attended our center for a second opinion regarding the previously diagnosed dextrocardia. She provided prior imaging studies, including a cardiac MRI (Fig. 1 A), which confirmed dextrocardia with dextroapex and situs solitus. A thoracoabdominal CT revealed right upper lobe agenesis (Fig. 1 B and 1 C), hypoplasia of the right upper pulmonary artery and anomalous arterial supply to the right lower lobe from the hepatic artery (Fig. 1 D), without evidence of pulmonary sequestration. The CT scan also showed agenesis of the intrahepatic inferior vena cava with azygos vein drainage and normal pulmonary venous drainage. Lung abnormalities, such as lung hypoplasia or agenesis are common findings in patients with dextrocardia and Scimitar Syndrome[1]. Aberrant systemic lung arterial supply, although described in pulmonary malformations and variations of Scimitar Syndrome, is a highly unusual finding when accompanied by dextrocardia and upper lobe agenesis [2,3]. Our case, remarks the importance of a complete evaluation of patients with dextrocardia or other congenital cardiopulmonary abnormalities, as unexpected vascular variations may alter diagnostic or management strategies.

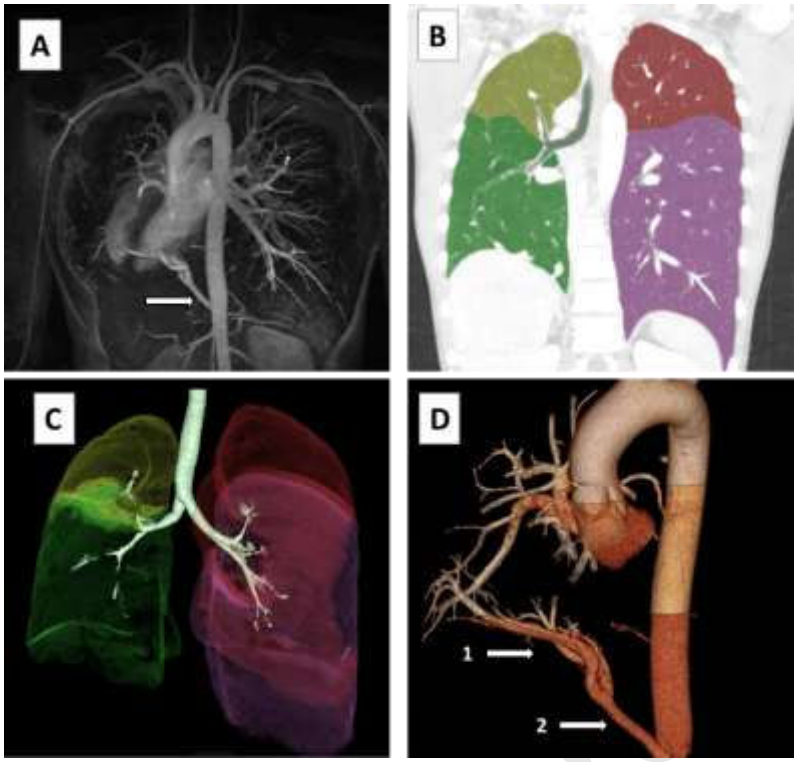

Figure 1. Multimodal Imaging Findings. A) Cardiac MRI showing dextrocardia and an anomalous systemic arterial supply to the right lower lobe (arrow). B) Coronal computed tomography image of the chest with automated pulmonary lobe segmentation, highlighting the agenesis of the right upper lobe, with the right middle and lower lobes displayed in yellow and green, respectively. The left upper and lower lobes are fully visualized and color-coded in red and purple. C) 3D reconstruction of the tracheobronchial tree revealing the absence of the right upper lobe bronchus. Both B and C were obtained using Synapse 3D® (Fujifilm). D) 3D vascular reconstruction from the thoracic CT scan detailing the anomalous systemic arterial supply to the right lower lobe (1), originating from the hepatic artery (2)

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Ethics in publishing

1. Does your research involve experimentation on animals?:

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No

3. Does your study include a clinical trial?:

No

4. Are all data shown in the figures and tables also shown in the text of the Results section and discussed in the Conclusions?:

Yes

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