KREP

2025

KARNATAKA RADIOLOGY EDUCATION PROGRAM

CASE PRESENTATION

Case of Scimitar syndrome

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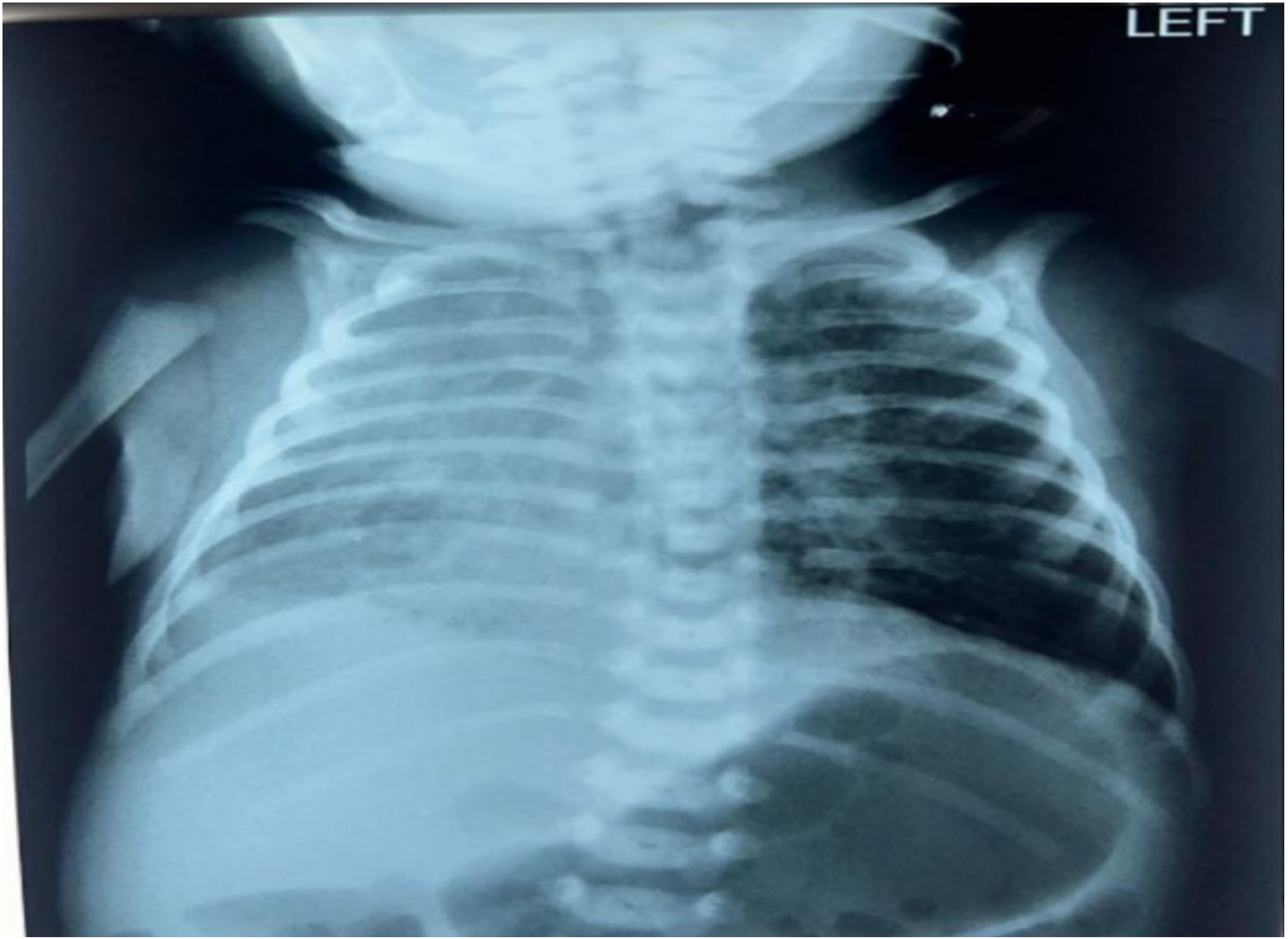
- A 7 day old neonate presented with history of respiratory difficulty and drop in oxygen saturation.
- Patient was advised for chest x ray


LEFT

XRAY FINDINGS

- Triangular opacity extending from hilum to diaphragm
- Obscuration of medial aspect of right hemi diaphragm
- Obscured right descending interlobar pulmonary artery
- Elevation of right hemidiaphragm
- Crowding of right sided ribs
- Ipsilateral mediastinal shift

Features suggestive of collapse of right lower lobe with ipsilateral mediastinal shift

CT Pulmonary Angiogram was advised for the baby

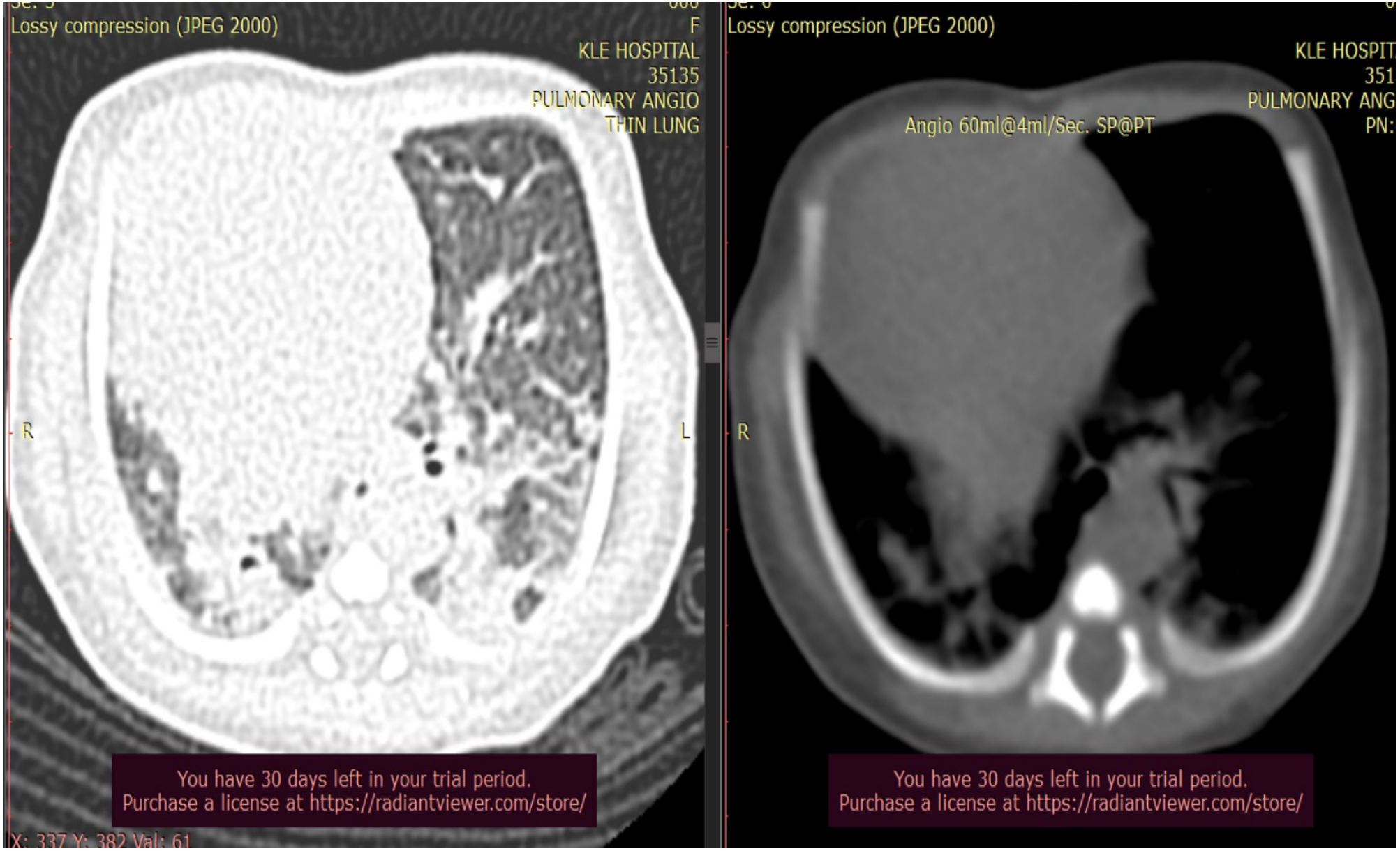
CT PULMONARY ANGIOGRAM FINDINGS

- Hypoplasia of right lower lobe
- Hypoplasia of ipsilateral pulmonary artery
- Partial anomalous pulmonary venous return
- Anomalous systemic blood supply to right lower lobe from aorta

Features suggestive of SCIMITAR SYNDROME

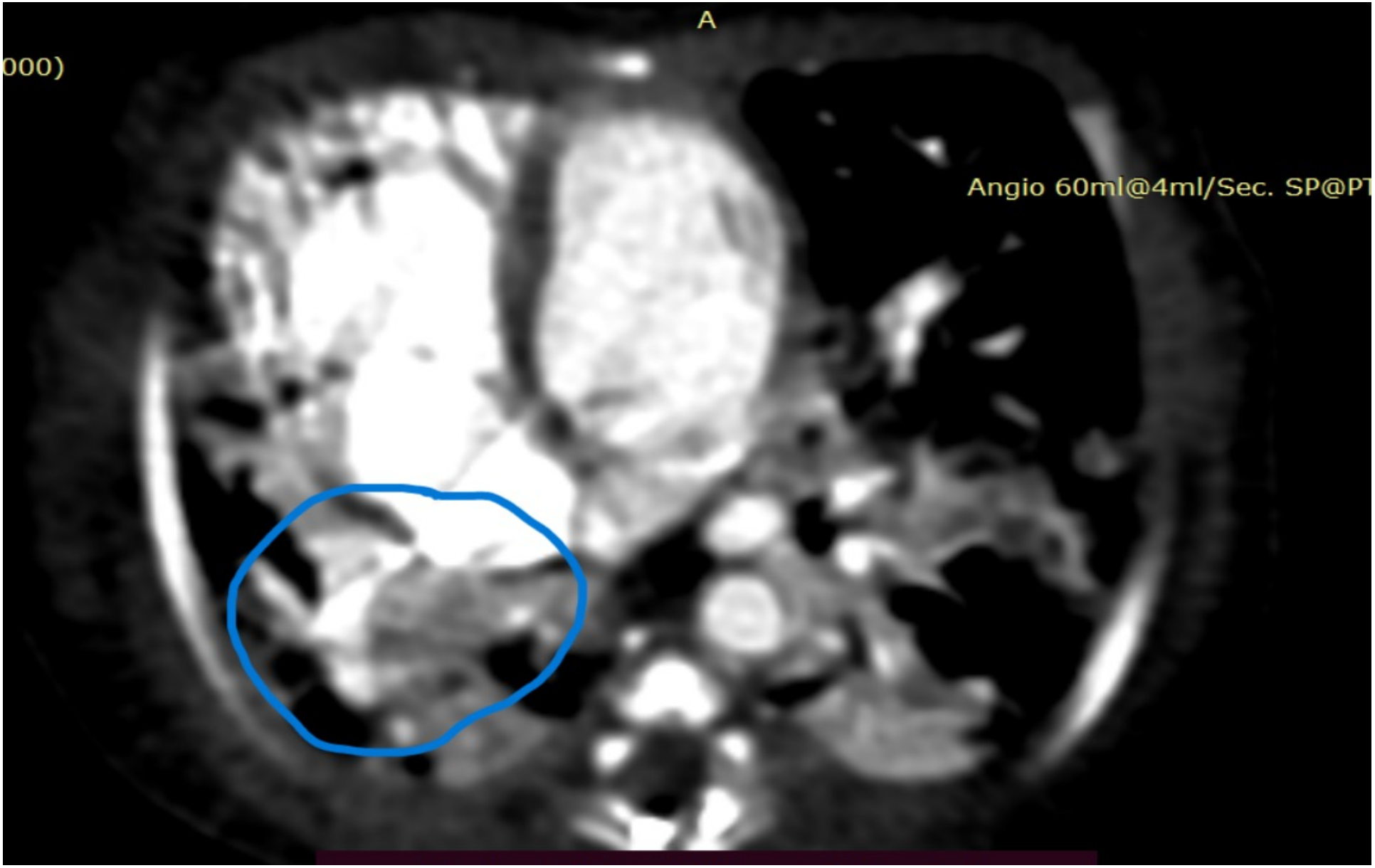
With associated features

- Atrial septal defect
- Ventricular septal defect
- Diaphragmatic herniation

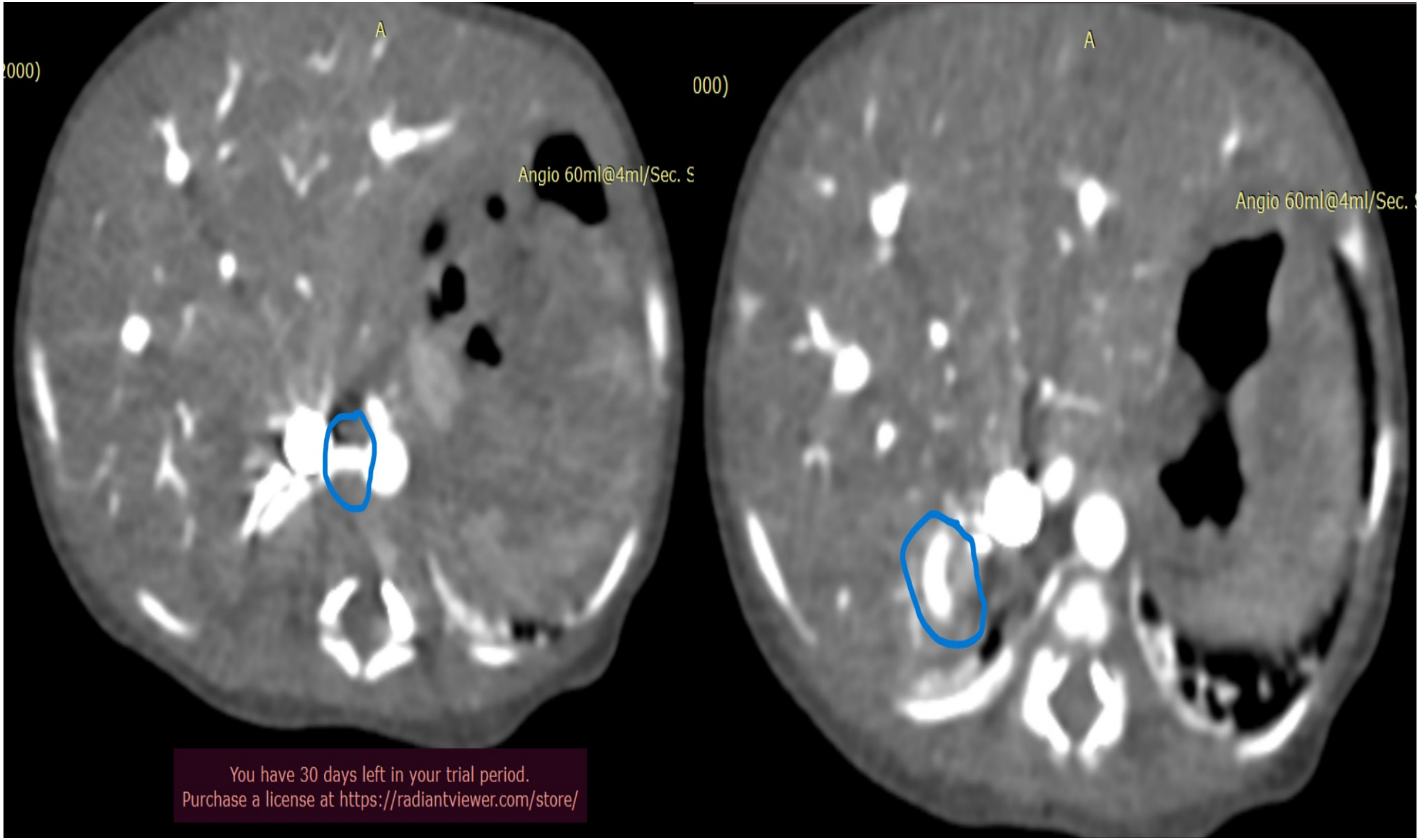


Hypoplasia of right lower lobe with ipsilateral mediastinal shift

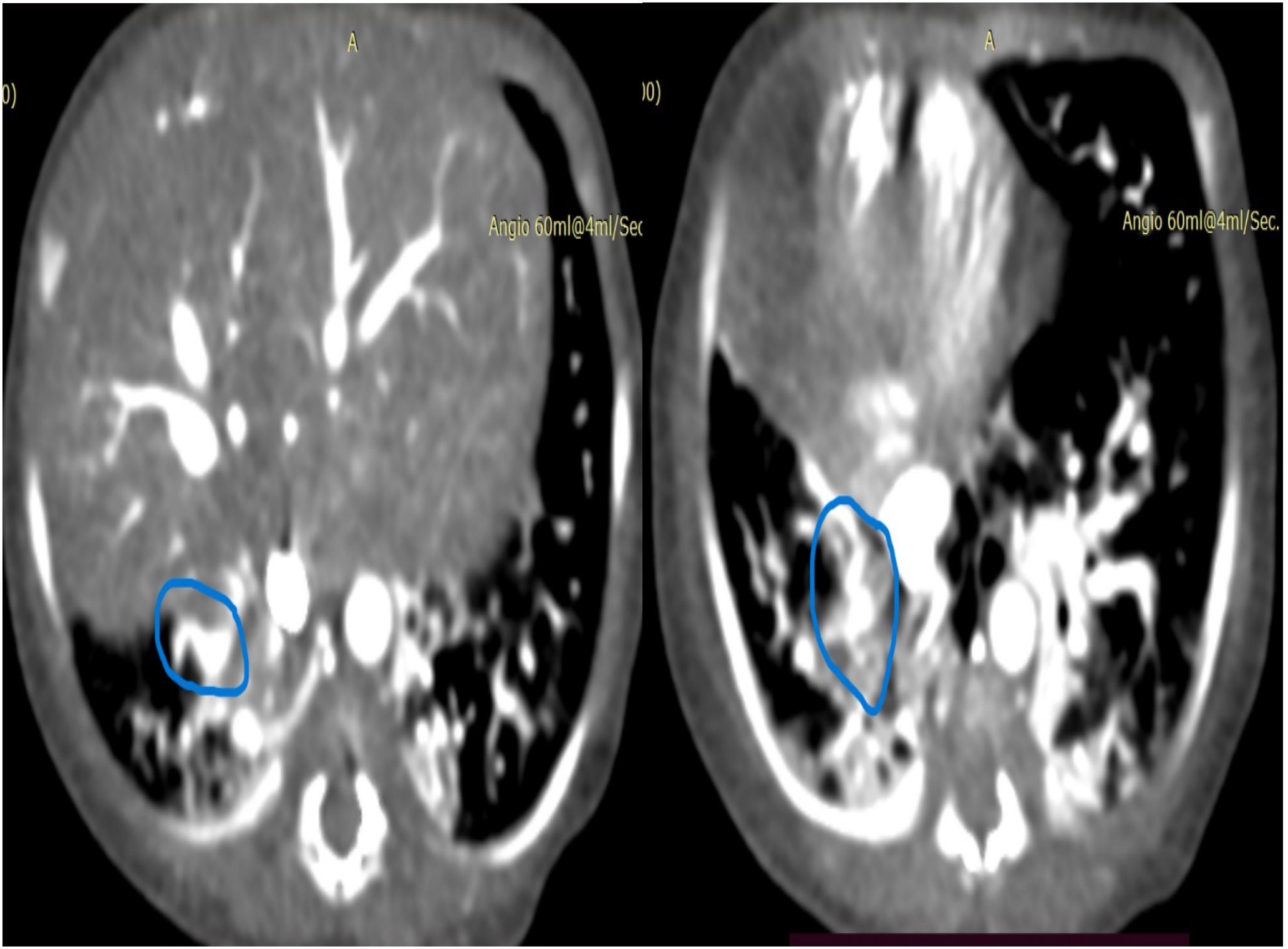
Hypoplasia of ipsilateral pulmonary artery



Partial anomalous pulmonary venous return – right inferior pulmonary vein is seen to drain into IVC



Anomalous systemic blood supply to right lower lobe from aorta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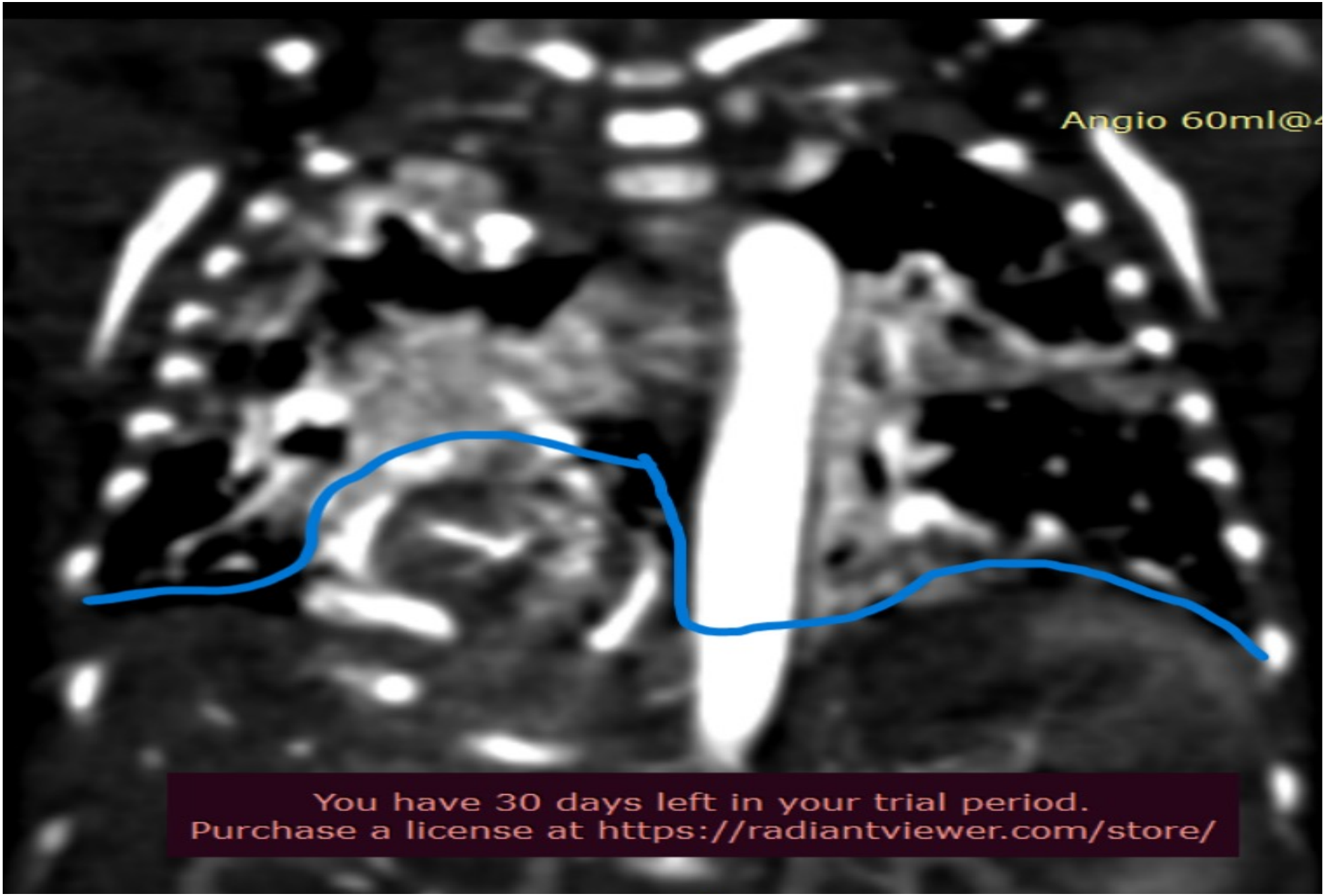

Diaphragmatic herniation

FOLLOW UP

- Currently baby is discharged from hospital and follow up will be done after 6 months
- Based on growth of lung parenchyma and right pulmonary artery baby will be planned for further management

- **Scimitar syndrome**, also known as **hypogenetic lung syndrome**, is characterised by a hypoplastic lung that is drained by an anomalous pulmonary vein into the systemic venous system. It is a type of partial anomalous pulmonary venous return and is one of the several findings in congenital pulmonary venolobar syndrome.
- **Associations**
- Congenital heart diseases: Ex- ASD, VSD, TOF and PDA
- Ipsilateral diaphragmatic anomalies: e.g. accessory diaphragm, diaphragmatic hernia
- Localised bronchiectasis
- Horseshoe Lung
- vertebral anomalies: Ex: Hemivertebrae
- Genitourinary tract abnormalities
- Pulmonary Sequestration

Clinical presentation

- Infants present with signs of heart failure, which may be due to associated congenital heart disease. Adults are most frequently asymptomatic but may have recurrent pulmonary infection or dyspnoea on exertion.

Pathology

- It is essentially a combination of pulmoanry hypoplasia and PAPVR. It almost exclusively occurs on the right side.
- Haemodynamically, there is an acyanotic left-to-right shunt. The anomalous vein usually drains into the:
- Inferior Venacava: most common
- Right Atrium
- Portal Vein
- In some variants, the abnormal lung segment may receive arterial blood supply from the aorta or its branches. Some cases may show the anomalous systemic arteries supplying an area of associated pulmonary sequestration

Treatment and prognosis

- Surgical correction should be considered in the presence of significant left-to-right shunting and pulmonary hypertension. This involves the creation of an interatrial baffle to redirect the pulmonary venous return into the left atrium.
- Alternatively, the anomalous vein can be re-implanted directly into the left atrium.

Thank you