



Ministry of Health Malaysia



SCIMITAR, THE CURVED BLADE. A MODERN SOLUTION TO AN ANCIENT PROBLEM

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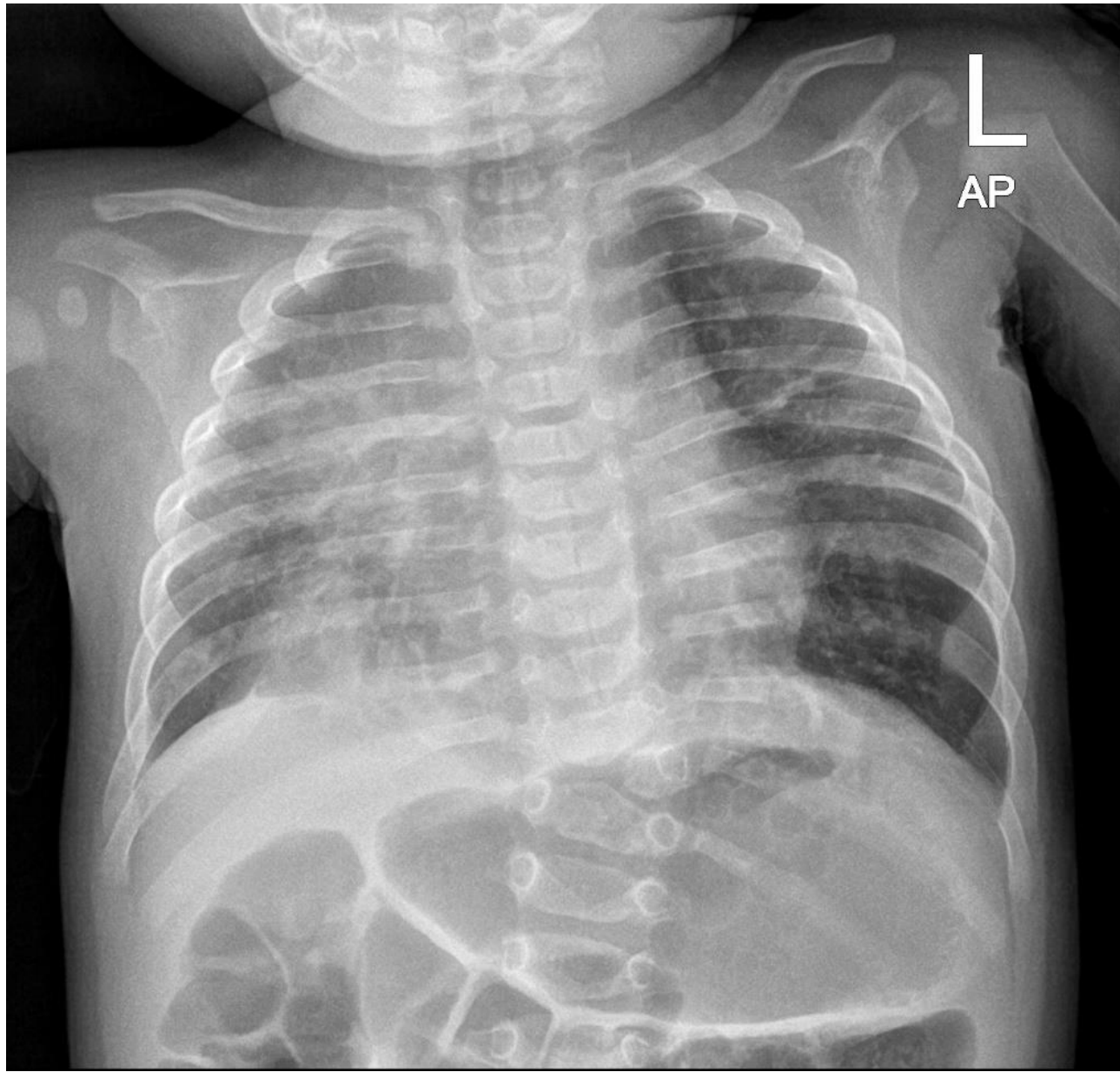
Background..

- Scimitar syndrome is a rare congenital anomaly. Described by *C A Neill*² in 1960
- Postulated : congenital veno-lobar malformation- partial anomalous venous return from the right lung into the systemic circulation.
- **Describes types;** (*Dupuis et al 1993*¹)
 - i) Infantile
 - ii) Adult
 - iii) Associated with Congenital Heart Condition
- Pulmonary Sequestration – Non-functional pulmonary tissue lacking communication to tracheo-bronchial tree. *Liechty KW*³
 - Association with Scimitar - rare
- Both pathologies are currently treated surgically in symptomatic patients. *Vida et al 2010*⁶
- In adult literature there are a few reported cases regarding Embolization for Symptomatic Scimitar's.

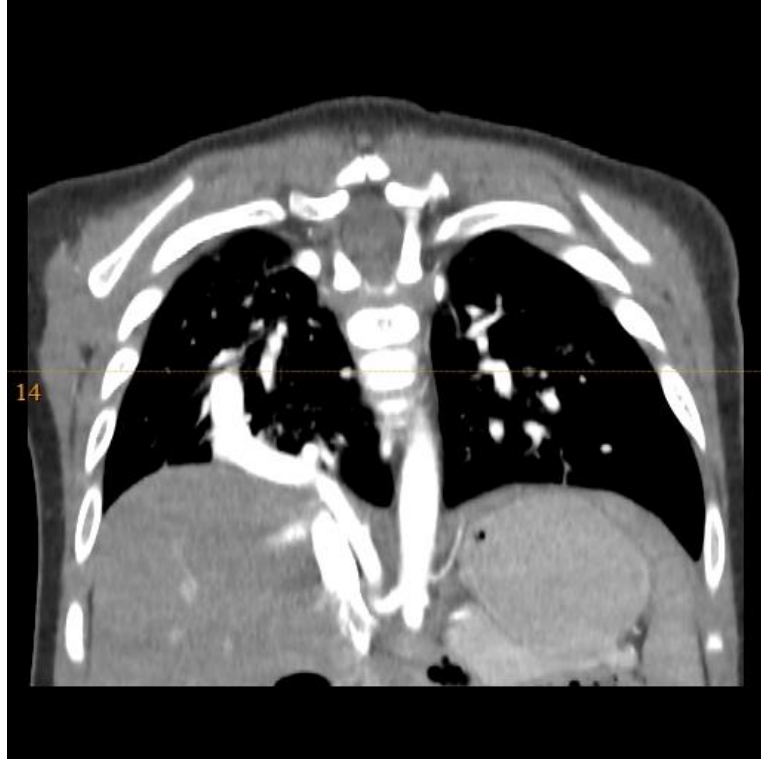


MS , 11 months old. Boy - 9.5kg

- Had recurrent episodes of chest infections during neonatal period, ages of 4 and 6 months of life. Requiring antibiotics and Nasal cannula O2.
- Clinical examination was unremarkable.
- Serial Chest Radiography revealed a patch of heterogenous opacity in the right middle + lower zone & hemivertebrae at T11.
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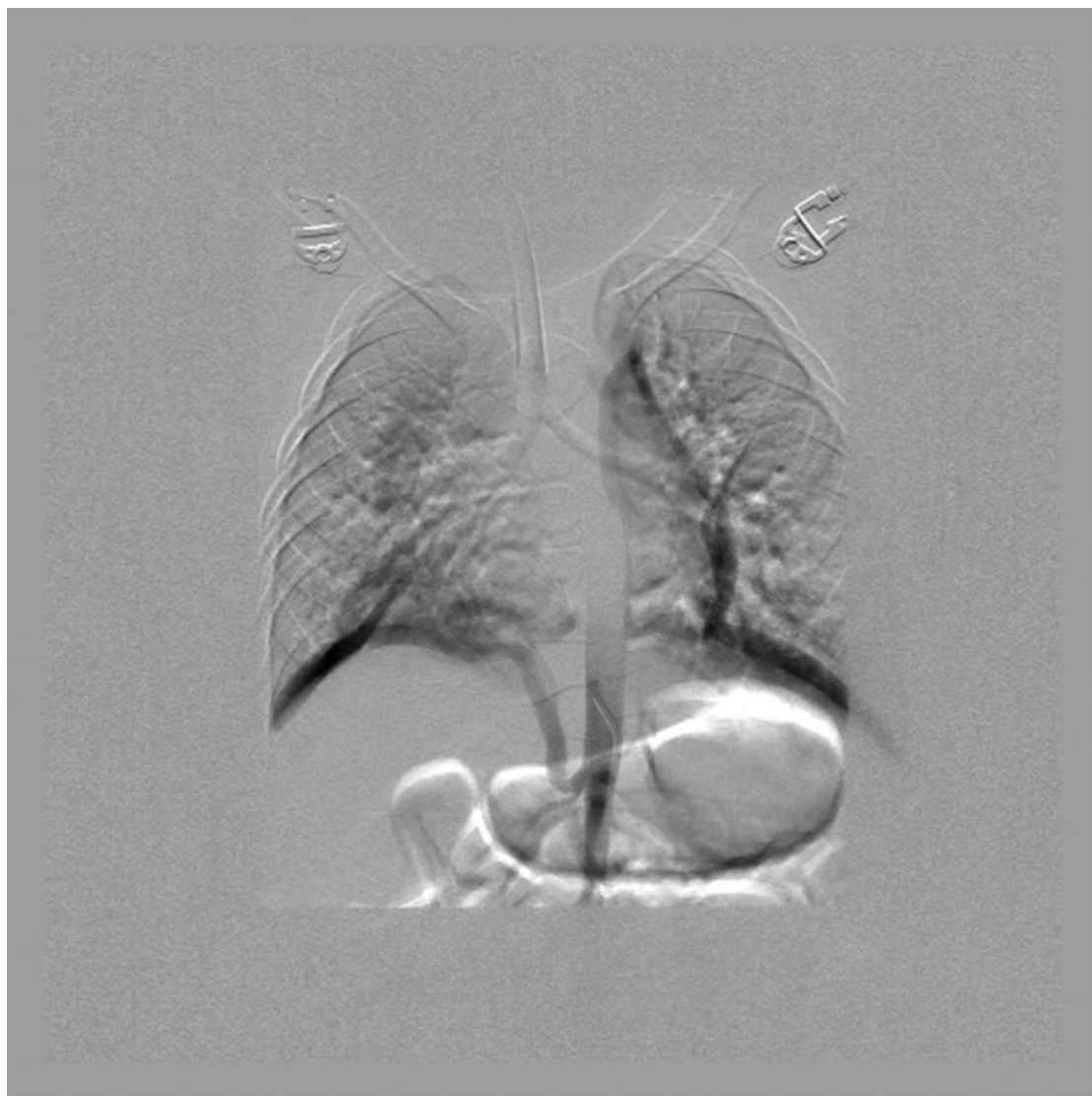


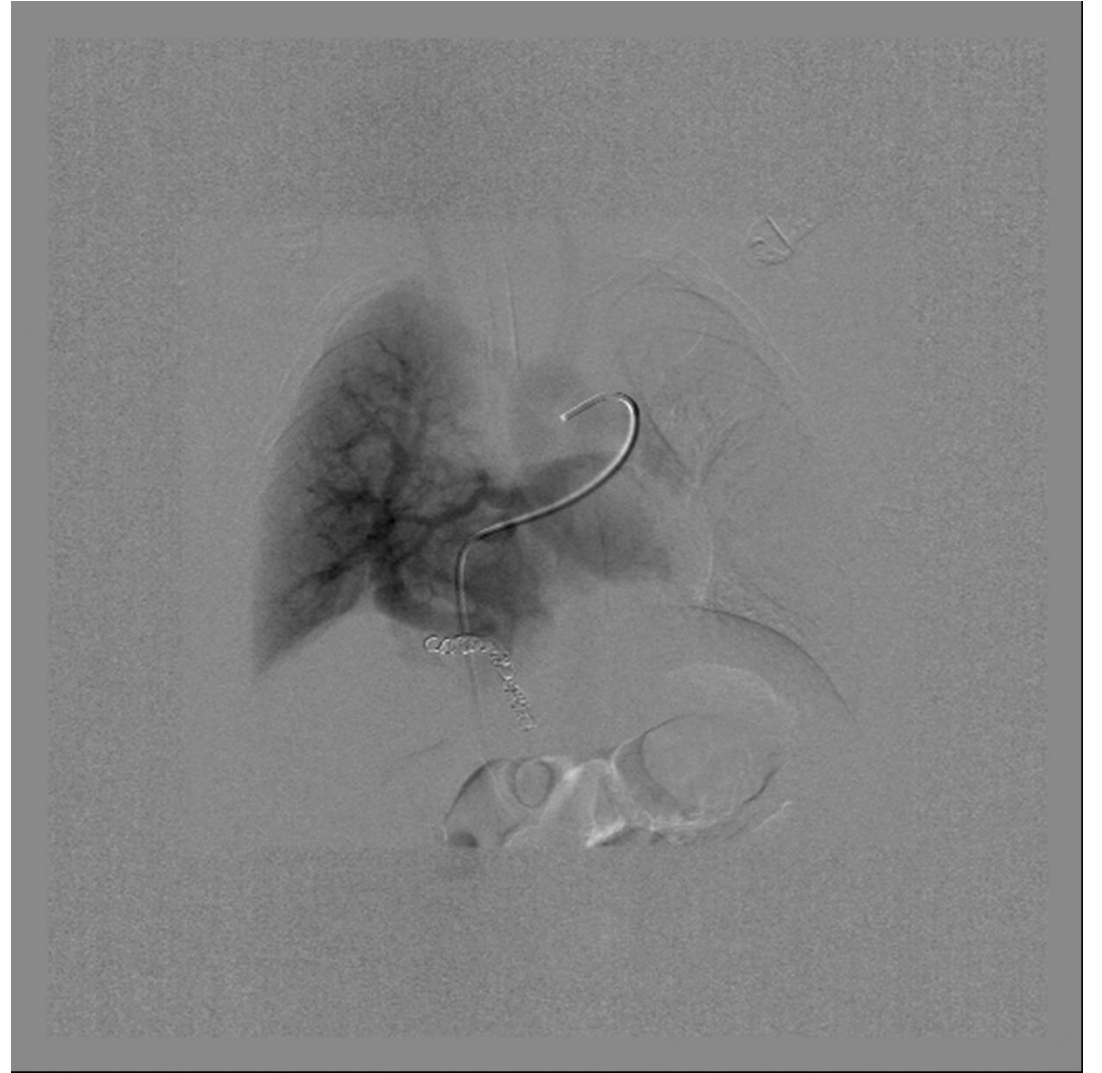




Stable Infantile Scimitar with Hybrid CPAM + Sequestration of the Right Lower Lobe

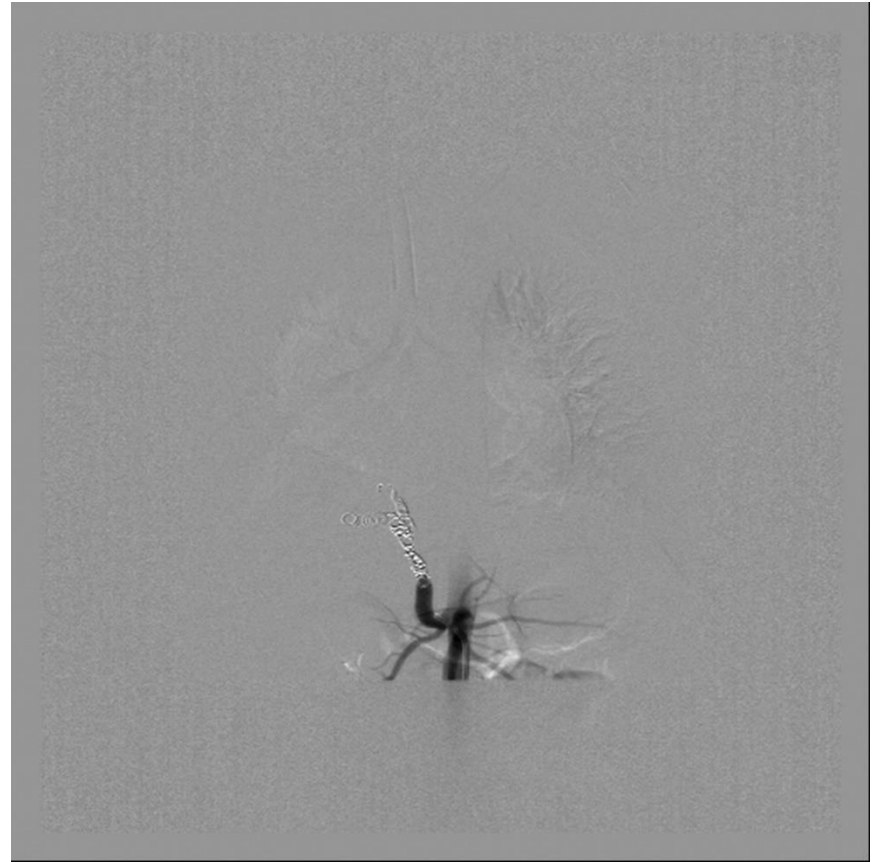
- MS was treated with Angiography + Embolization of the arterial branch to the sequestration in the right lower lobe.
- Post procedure recovery was uneventful. He was allowed home after 2 days.
- A repeat CT Thorax was done 1 month post procedure and depicted resolution of the sequestration with residual soft tissue posterior segment of the Right Lower lobe. CPAM component remains unchanged.
- MS remains asymptomatic till date.



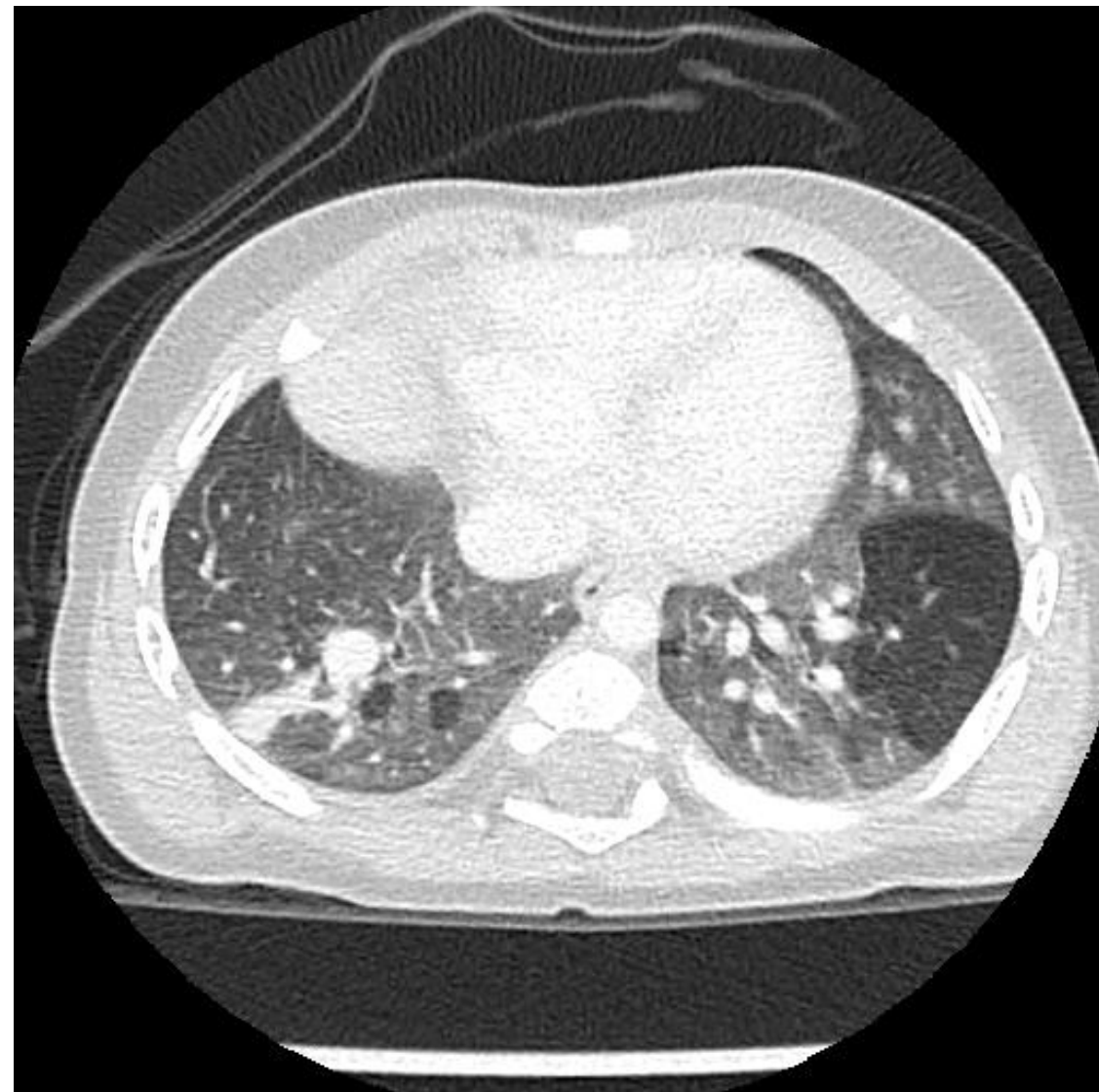


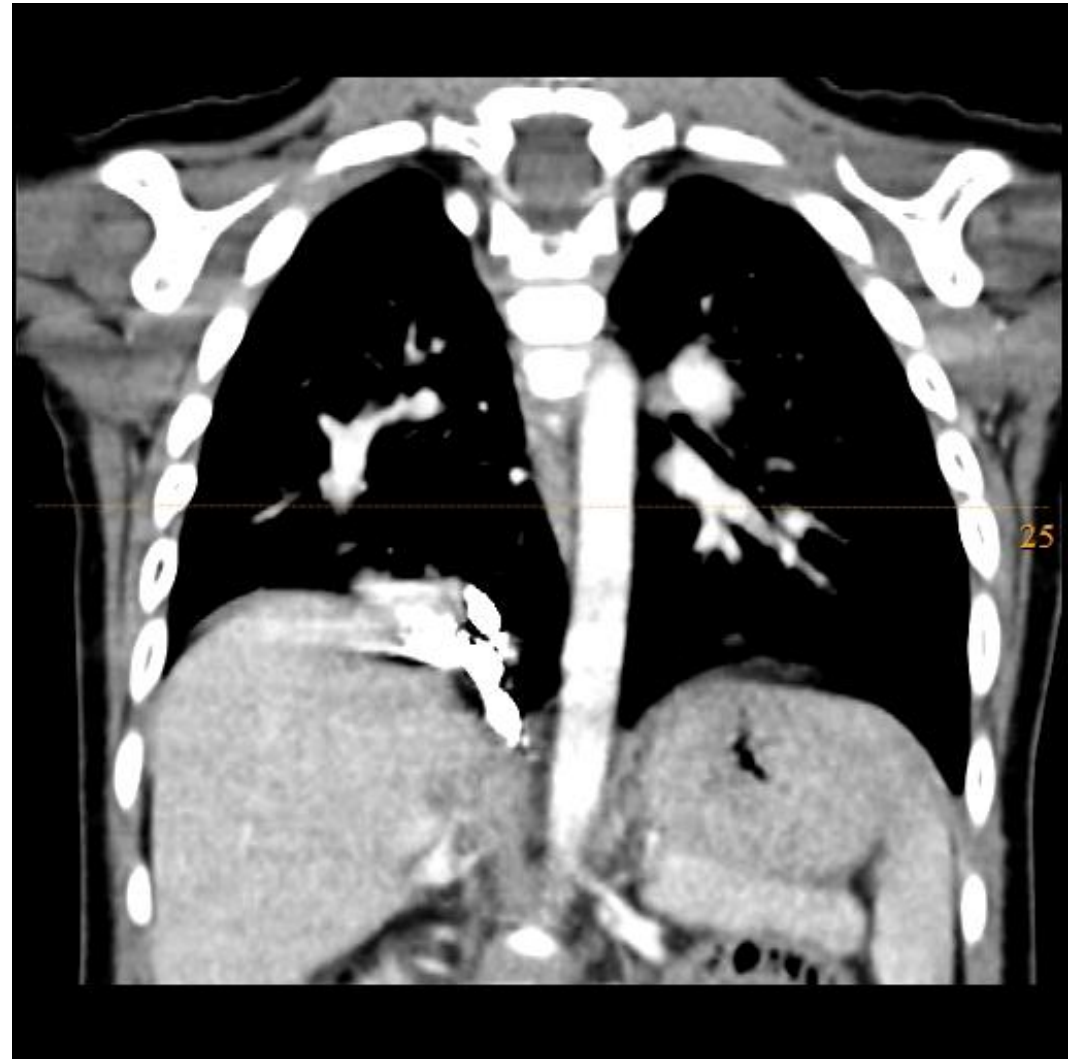
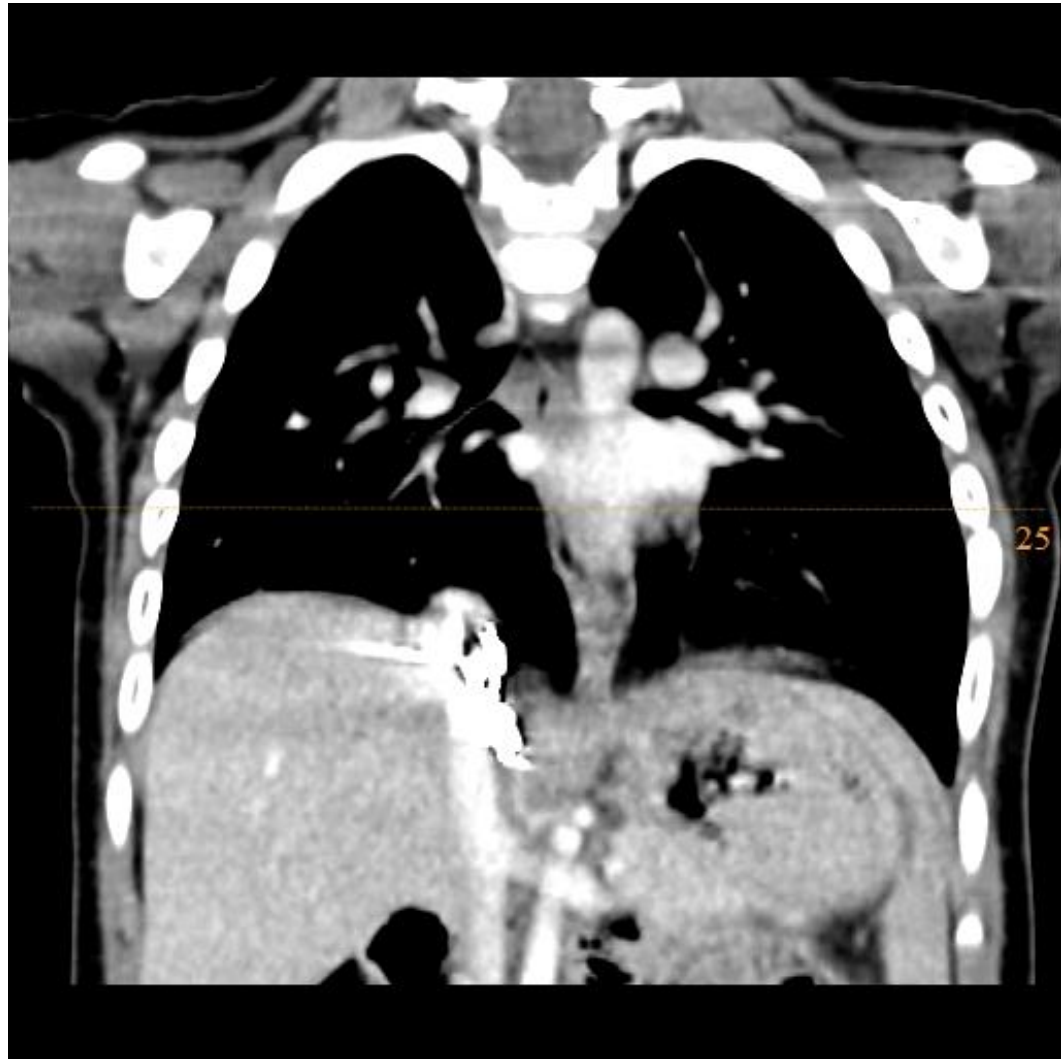


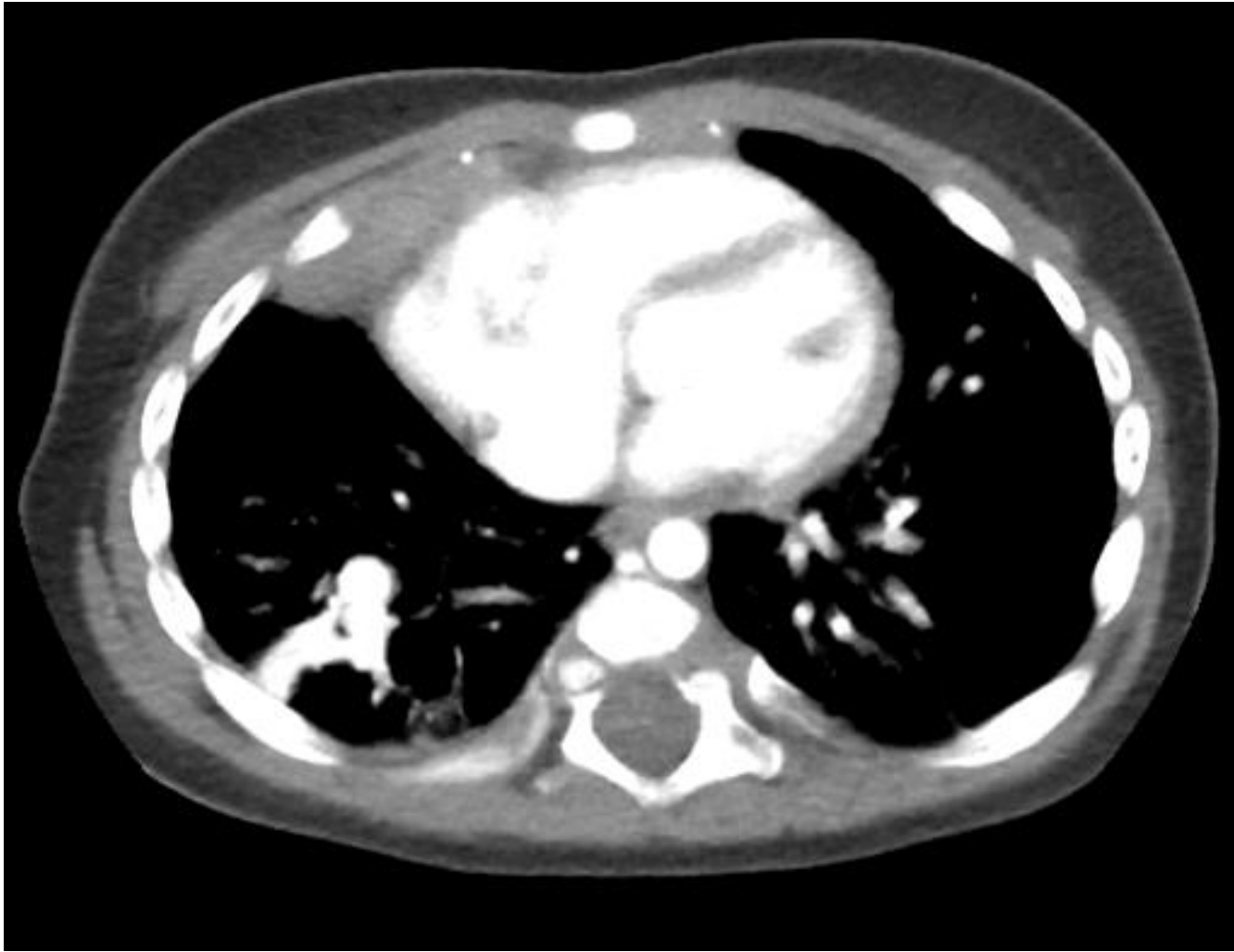
Pre Embolization



Post
Embolization







PRE Embolization



Post Embolization

Elements of Discussion

- Whilst published reports and guidelines on managing **Scimitars** or **Hybrid lesions** (CPAM + Sequestration) are available, they however discuss the management of the disease separately.
- Most Literature are mainly in the adult population. *Yamakawa et al*⁴, *Abe et al*⁵, *Vida et al*^{5,6}

There are limited publications describing management of both conditions simultaneously especially in the paediatric age group.

- Sequestration associated with scimitars - mainstay of treatment (excision of lesion with reconstruction of abnormal vessels in symptomatic patients).
- *Wang et al*⁸ described successful Embolization in a child with symptomatic Scimitars.

Conclusion

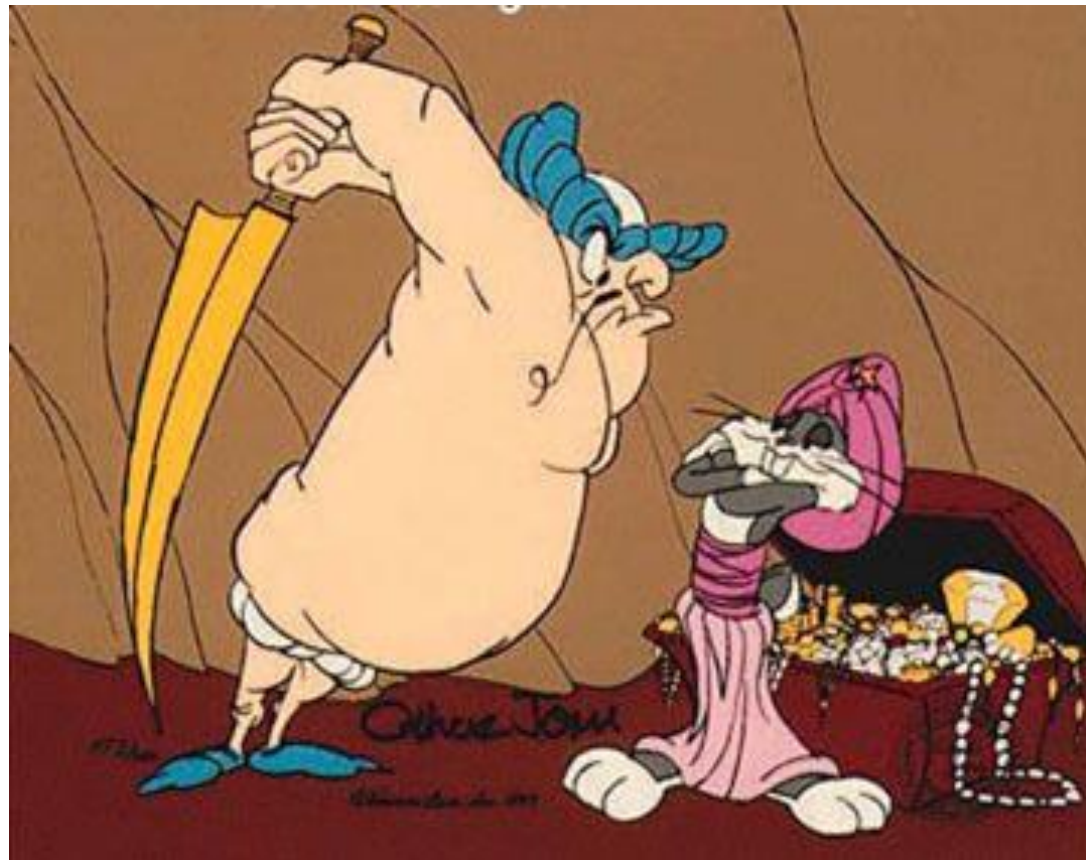
- We would like to highlight the complexity and challenges faced in managing this case.
- We anticipate difficulty in handling the hybrid lesion lying close proximity to the Scimitar vessel, hence surgery was not considered as the primary option.
 - This would require a Right Lower Lobectomy + Reconstruction of the Upper lobe vasculature.
- This case report also emphasizes the role of Interventional Radiology in assisting us Surgeons , providing an alternative to the classical operative approach for the mentioned problem.
- In tandem to modern advances in interventional radiological procedures, we have demonstrated that Angio-Embolization is a viable option in the paediatric population as well.

Long term Questions..

- We hope that the lower lobe lesion will eventually regress. Thus eliminating the need for surgical intervention altogether. However;
- What would be the long term outcome after embolizing the sequestration ?
- Will the CPAM lead to further problems ?
 - How frequent should we repeat imaging if the child remains asymptomatic?
 - Will this need excision ?
 - Risk of malignant transformation – Pleuropulmonary Blastoma.

References

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8. **Transcatheter intervention in a child with scimitar syndrome** [Wang Z](#) et al. Cardiovasc J of Afr May-June 2016



THANK YOU !