

# CASO DE AUTOPSIA



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# HISTORIA CLÍNICA

Sexo masculino, **30 años**

➤ AP: Asma bronquial

## ➤ Historia Hematológica

▪ **2007: Linfoma de Hodgkin clásico** estadio IB, masa Bulky

ABVD x 6 ciclos y Radioterapia → Respuesta completa

▪ **2009: Recaida**, estadio IIIA

DHAP x 3 ciclos / trasplante autólogo → Buena respuesta

▪ **2010: 2da Recaida**

GEMOX x 6 ciclos → Refractario

Brentuximab → Respuesta completa

# HISTORIA CLÍNICA

- **2012:** Trasplante Médula Ósea Alogénico ( Hermano HLA idéntico )



EICH aguda cutánea grado III

Eritema y Prurito en cuello y palmas

- **2013-2016:** Recidiva Linfoma

**EICH** crónica cutánea

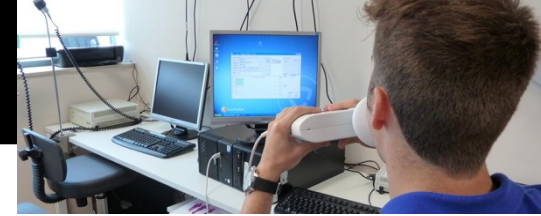
**IRC** a los **11 meses post-trasplante**

Episodios de reagudizaciones con necesidad de ingresos

Oxígeno domiciliario

# PRUEBAS RESPIRATORIAS

DISNEA



**2013**

- **CPT**= 5,2 Litros (70% N)
- **VR**= 1,52 Litros (86% N)

- **DLCO**= 48% N



**2015**

- **CPT**= 2,84 Litros (39% N)
- **VR**= 1,37 Litros (77% N)

- **DLCO**= 29% N

CPT  
VR  
DLCO



**ALTERACIÓN VENTILATORIA  
RESTRICTIVA PARENQUIMATOSA**

**+**

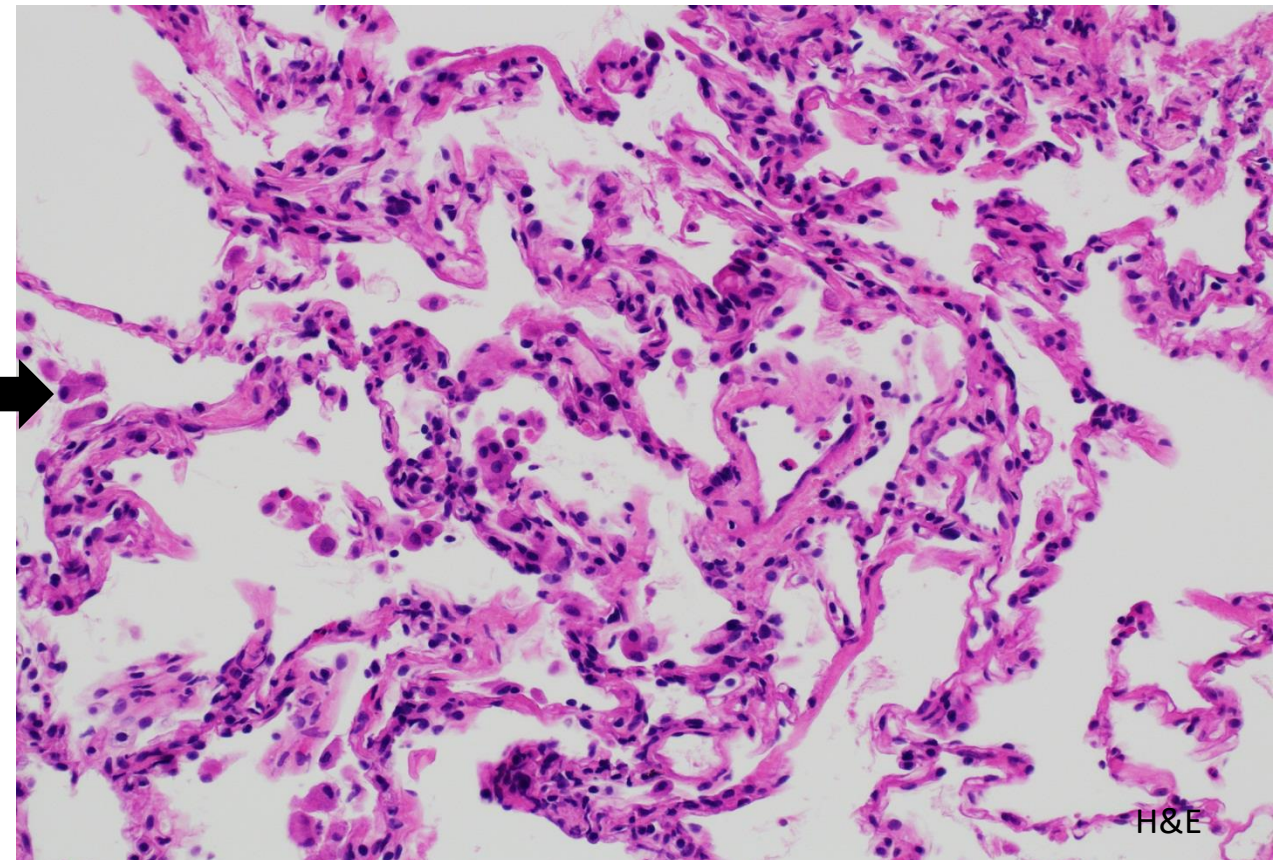
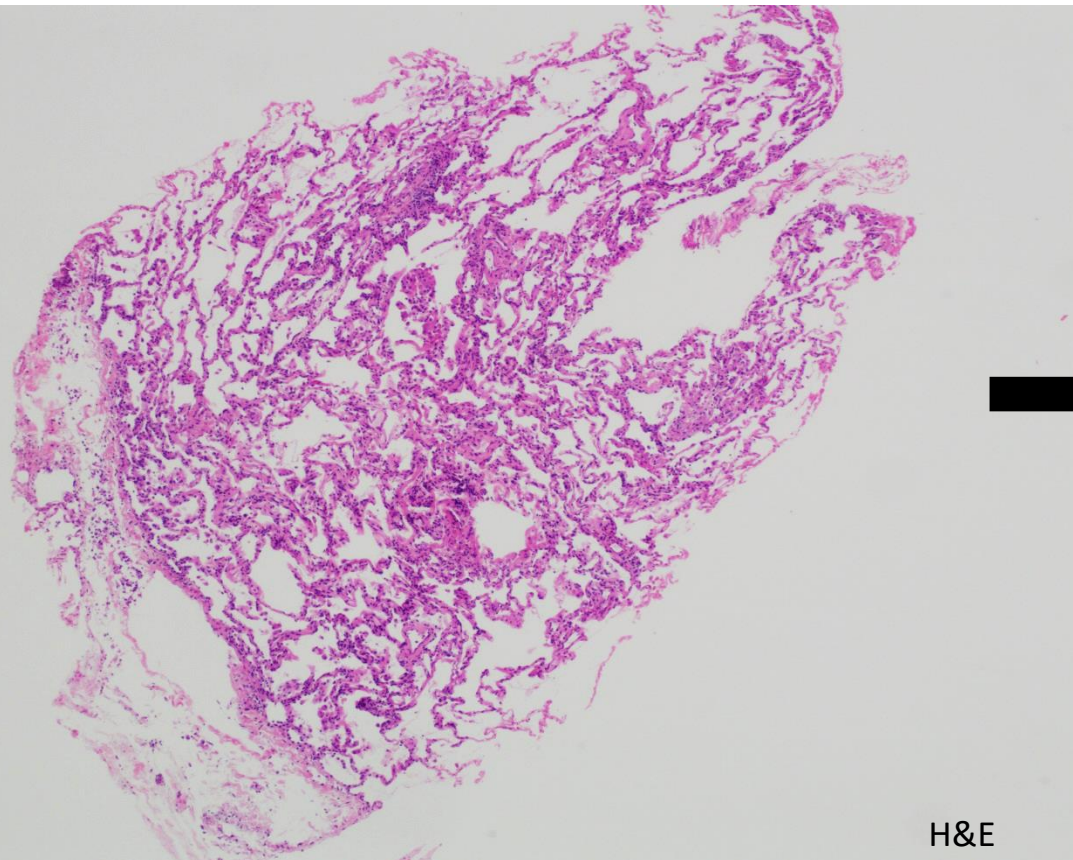
**Intensa Disminución de la Difusión CO**



# BIOPSIA PULMONAR TRANSBRONQUIAL 2013

➤ INFILTRADO INFLAMATORIO CRÓNICO, INTERSTICIAL, PARCHEADO, CON EOSINÓFILOS, Y CAMBIOS REACTIVOS DE LOS NEUMOCITOS

**DG:** CAMBIOS INFLAMATORIOS INTERSTICIALES INESPECÍFICOS

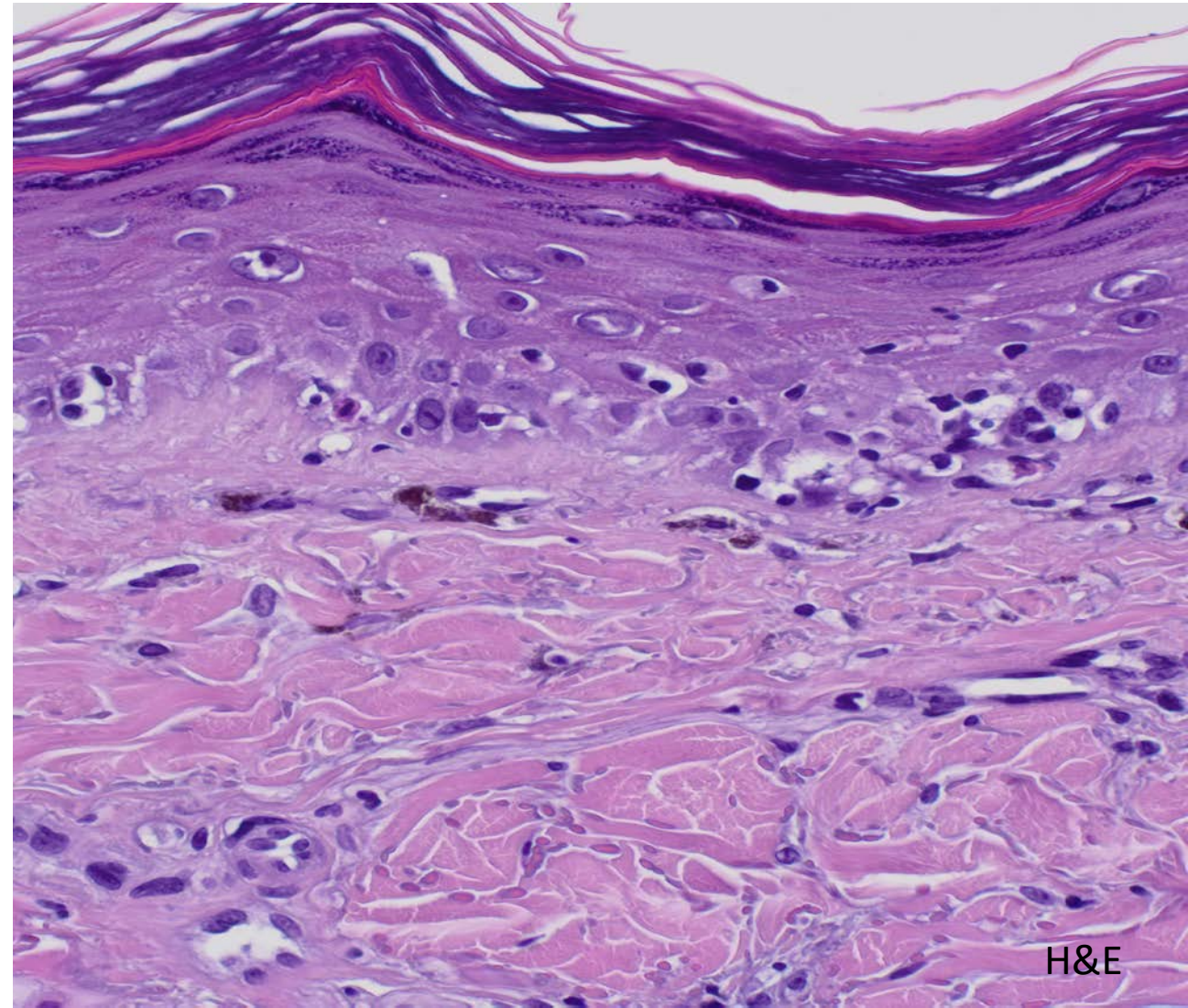
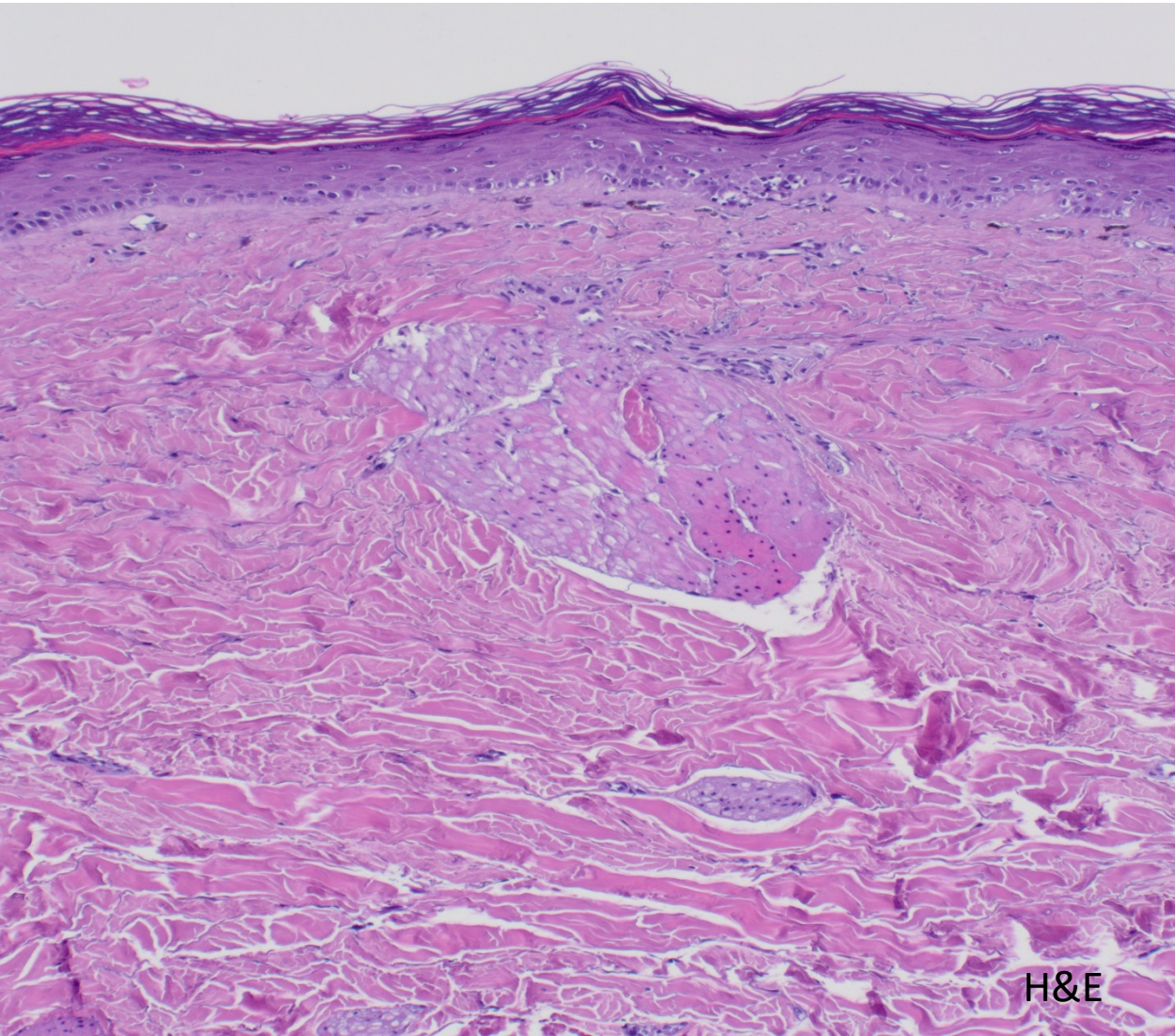




# PUNCH REGIÓN LUMBAR 2014

COMPATIBLE CON ENFERMEDAD DEL INJERTO CONTRA HUESPED

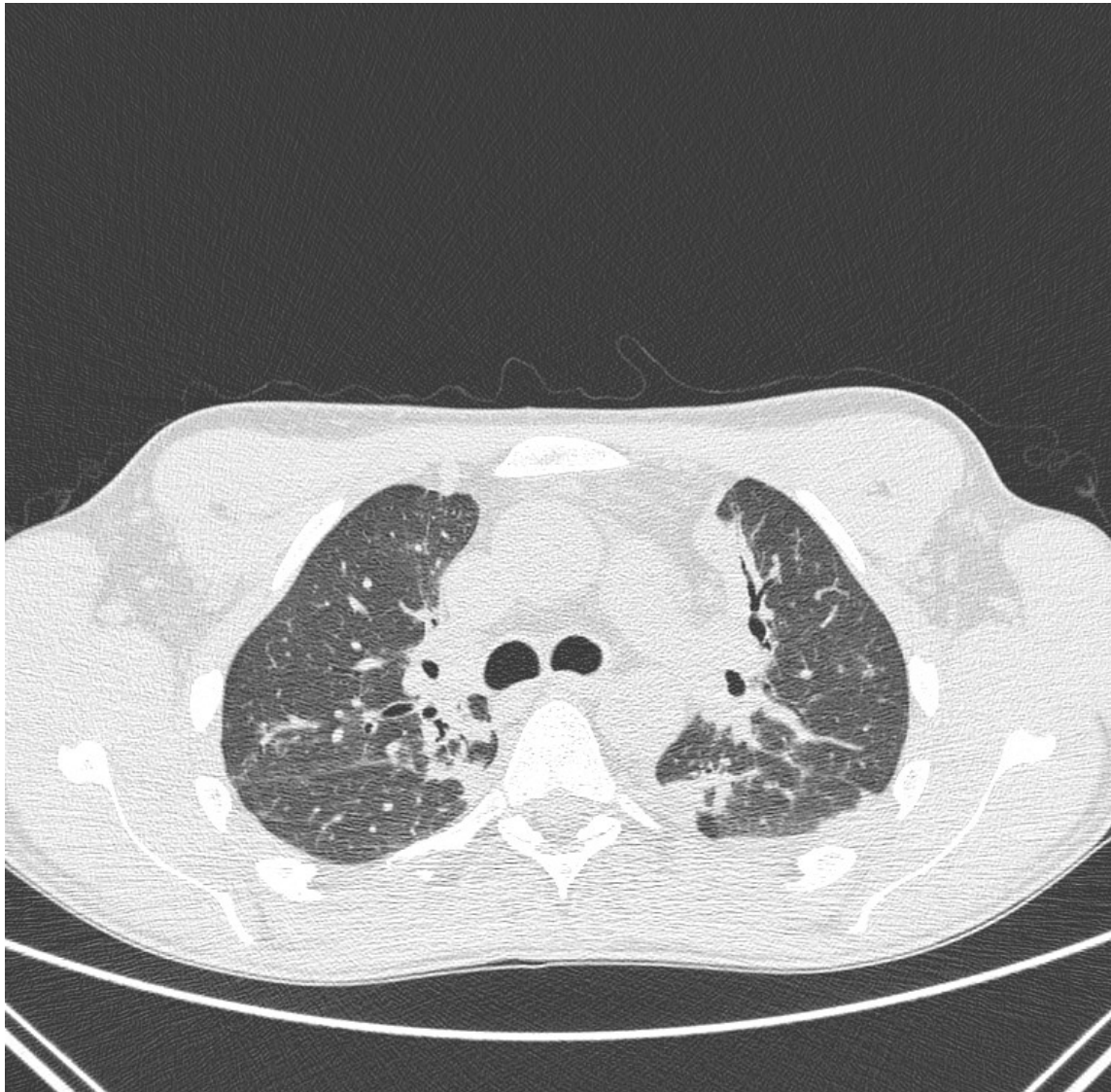
TIPO ESCLERODERMIFORME



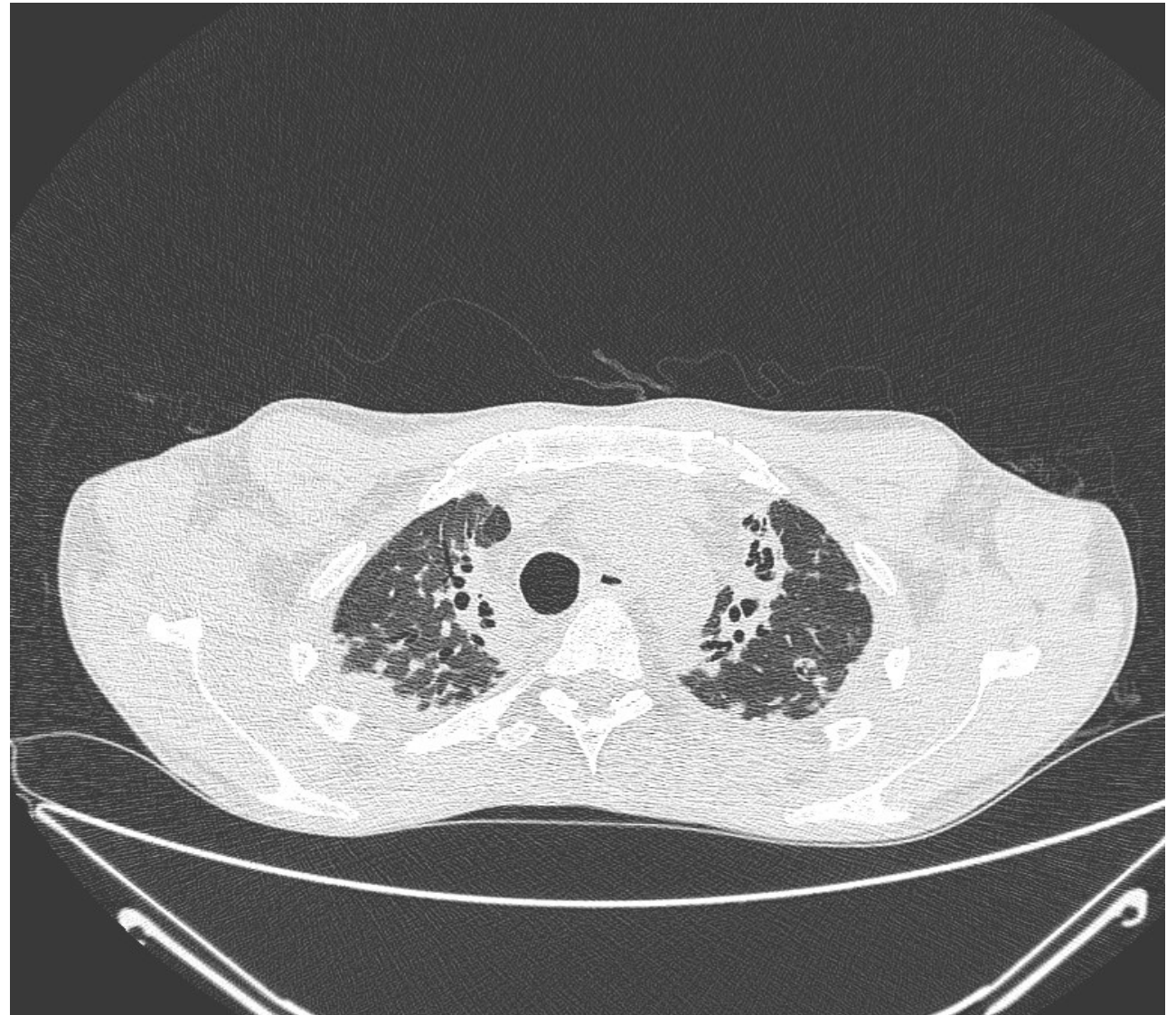


# TC TÓRAX DE ALTA DEFINICIÓN

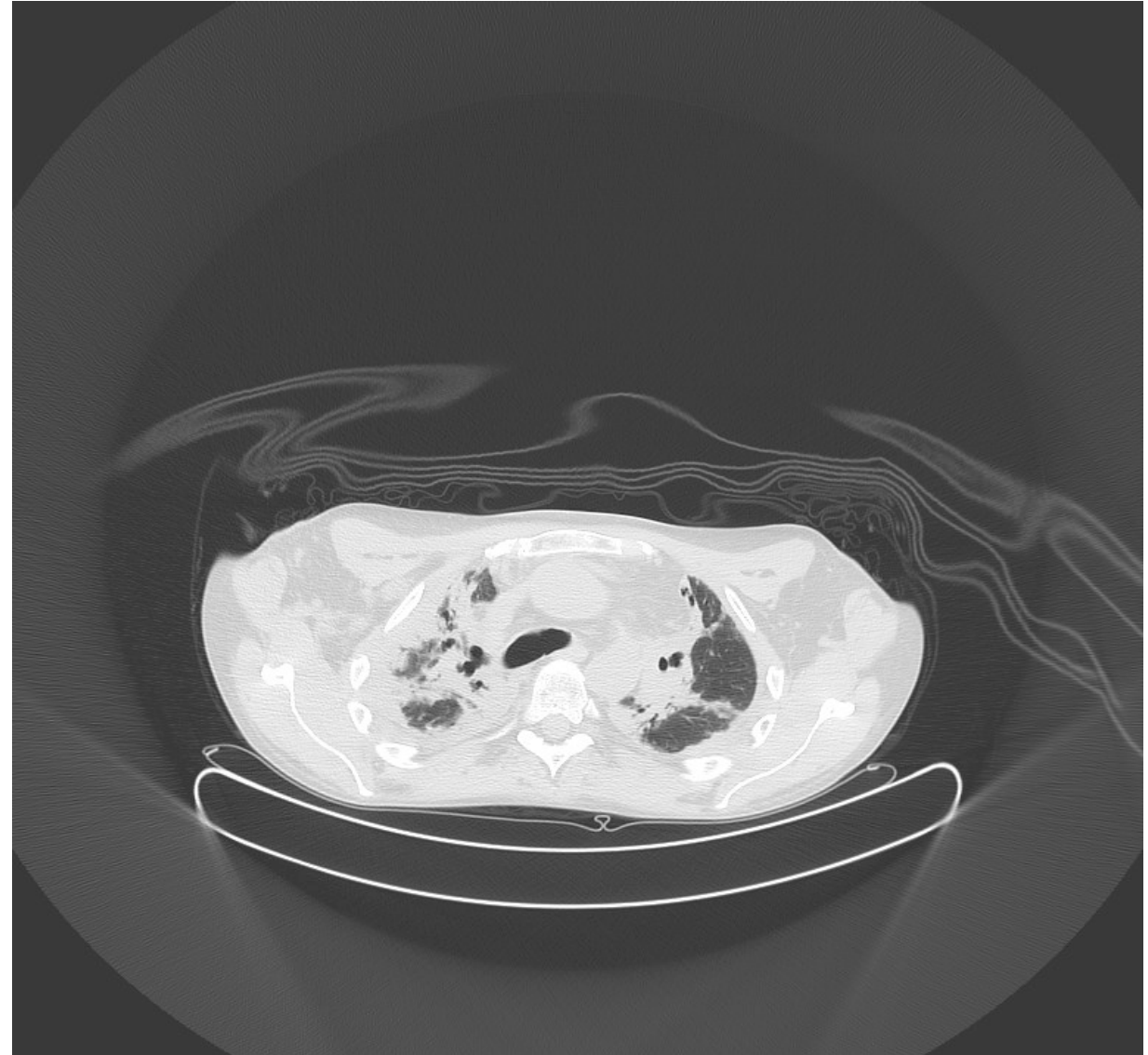
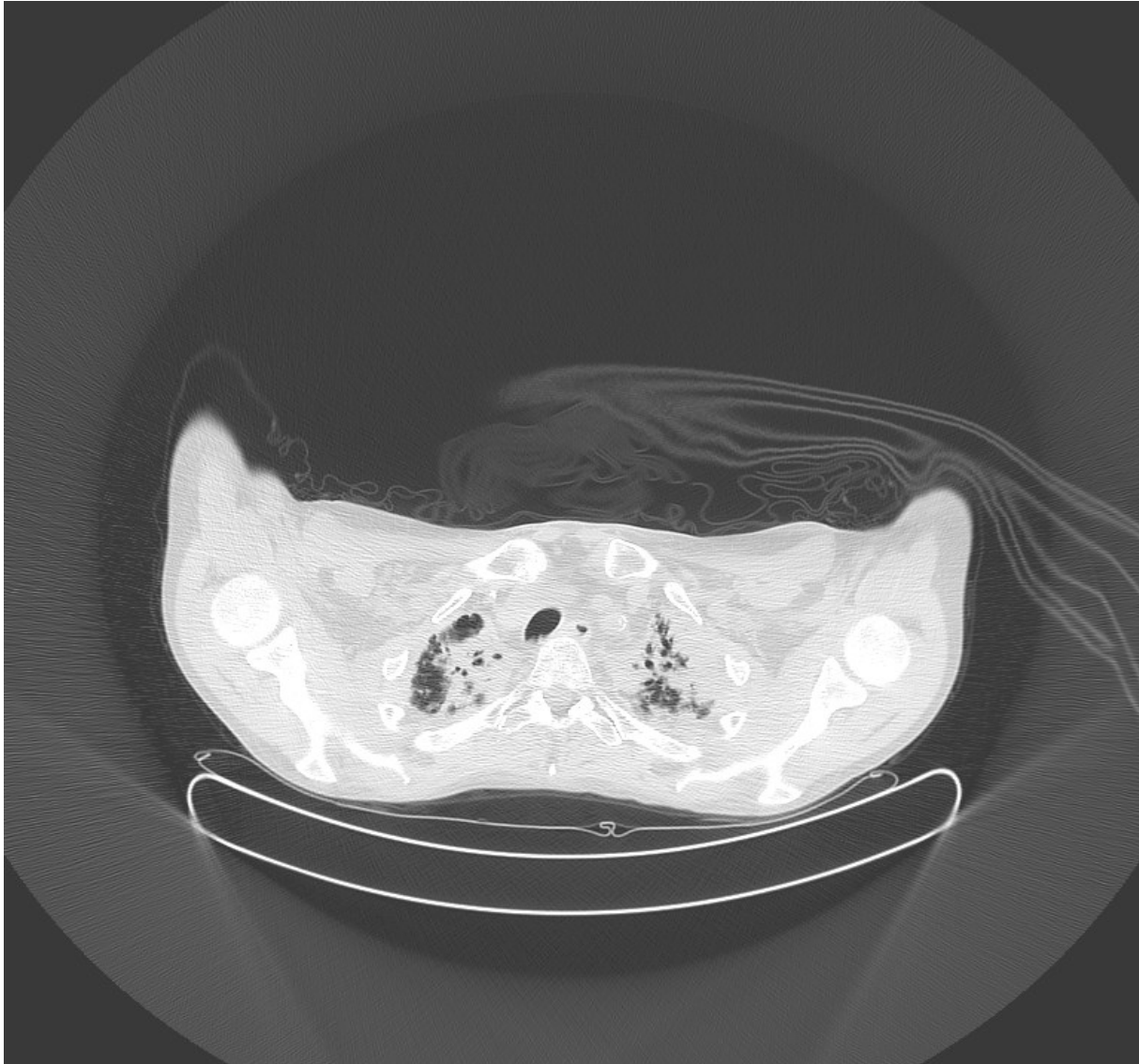
2013



2015

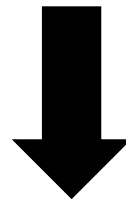


# TC TÓRAX DE ALTA DEFINICIÓN 2016



# ÚLTIMO INGRESO 2016

- ESTUPOROSO
- DETERIORO CLÍNICO CON TENDENCIA A LA SOMNOLENCIA
- INSUFICIENCIA RESPIRATORIA HIPERCÁPNICA

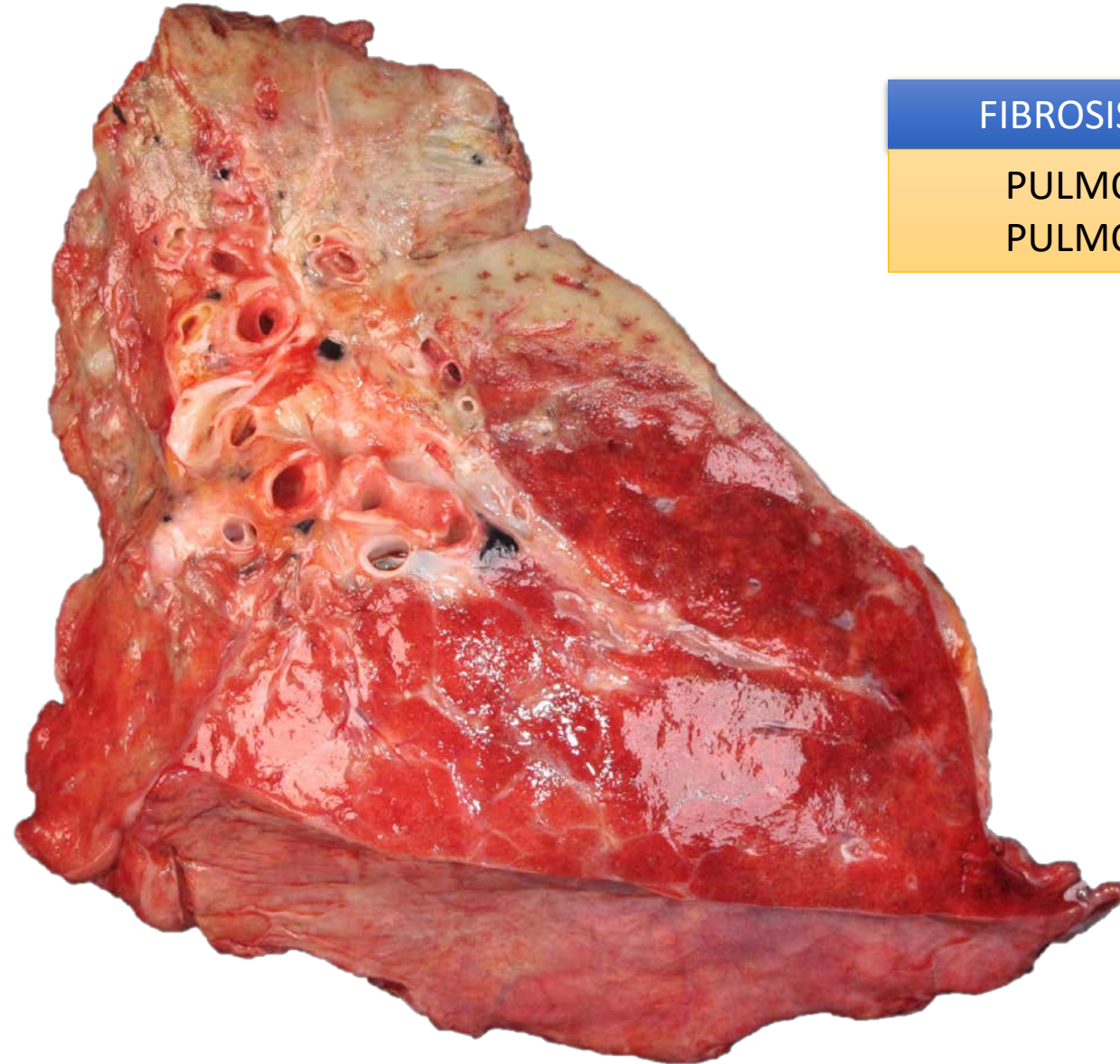


MEDIDAS DE CONFORT  
FALLECIMIENTO A LOS 3 DÍAS



# AUTOPSIA

## ESTUDIO MACROSCÓPICO PULMONAR

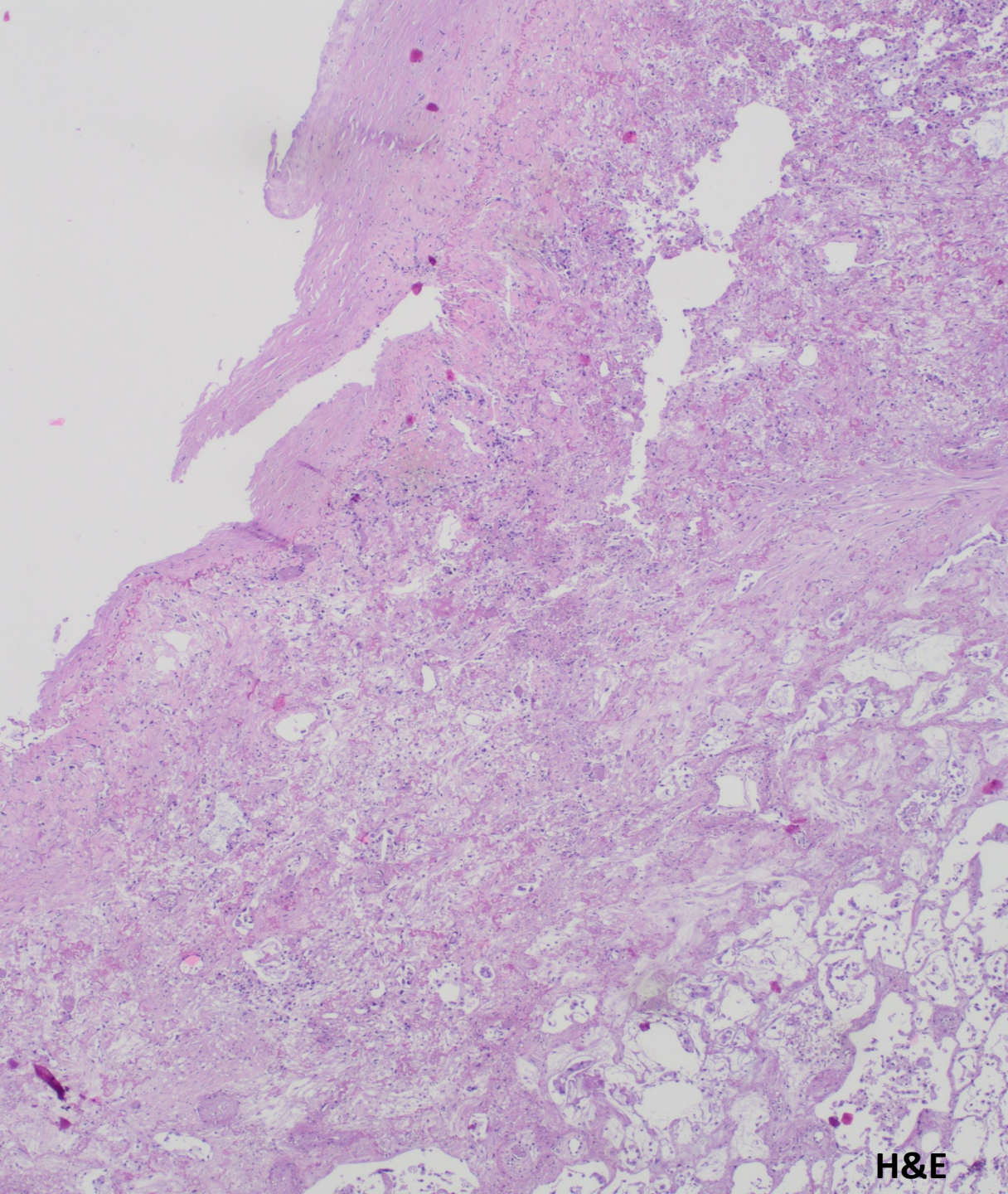


FIBROSIS APICAL BILATERAL

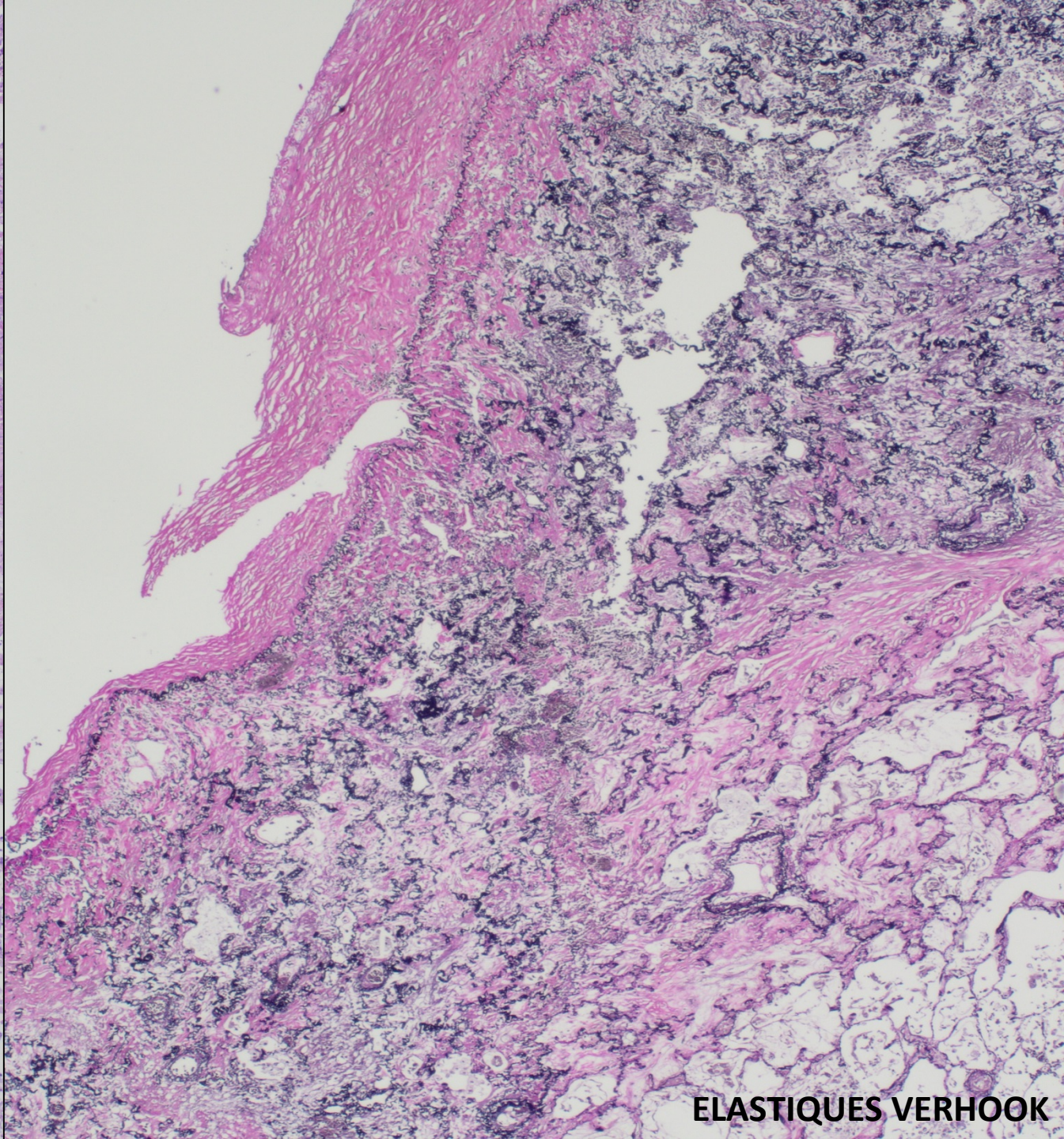
PULMÓN DERECHO 635 gr.

PULMÓN IZQUIERDO 565 gr.



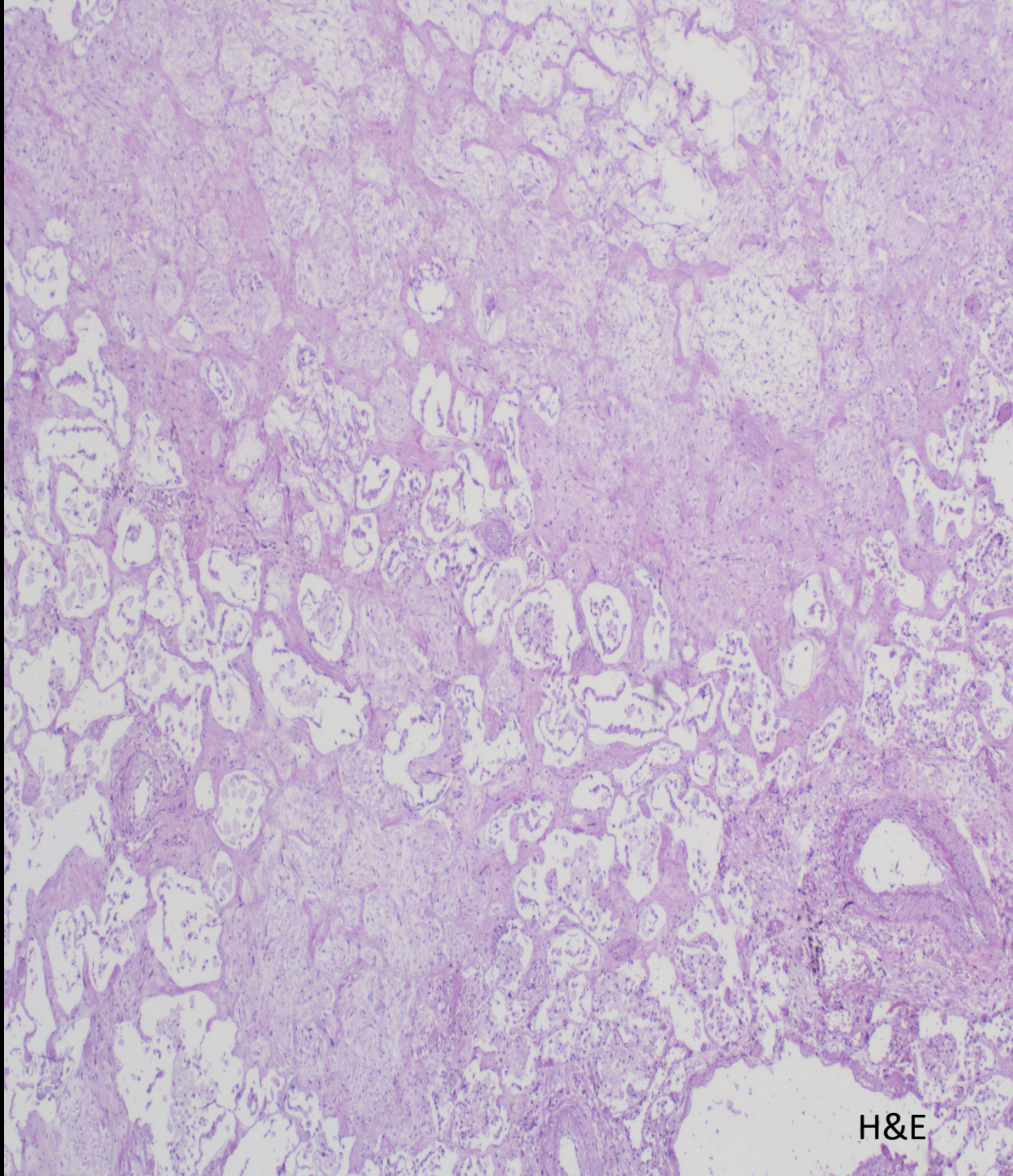


H&E

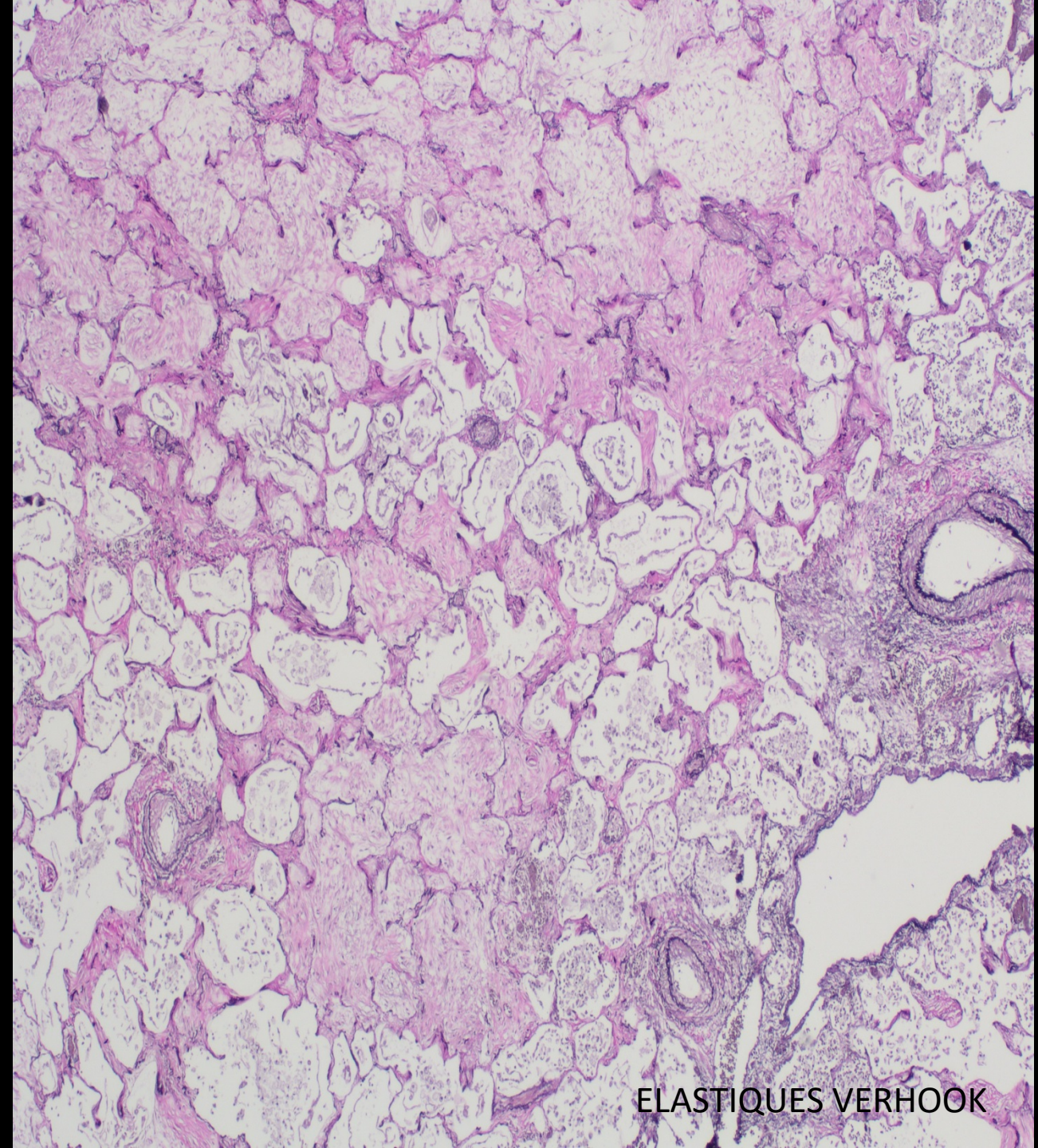


ELASTIQUES VERHOOK



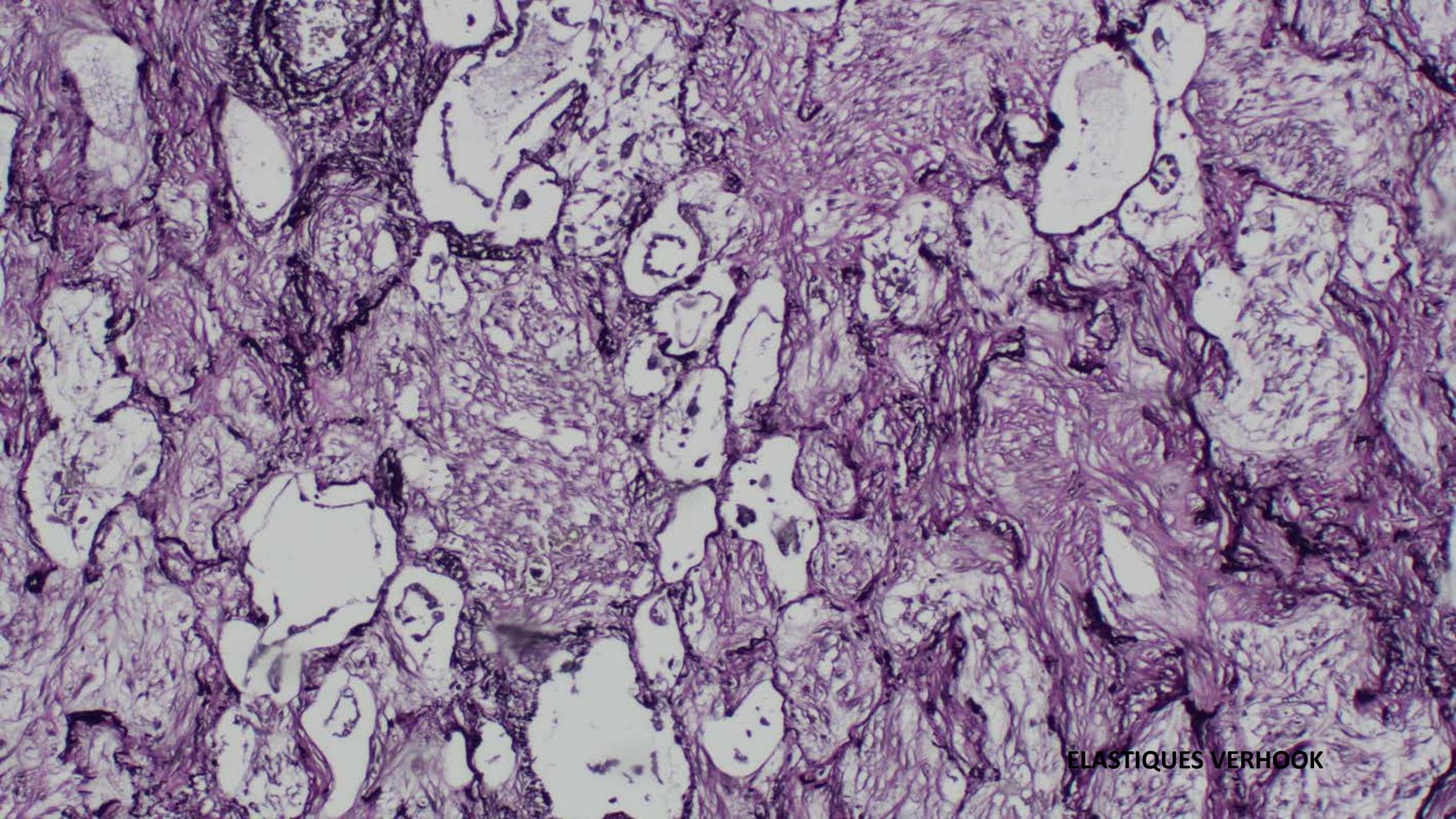


H&E



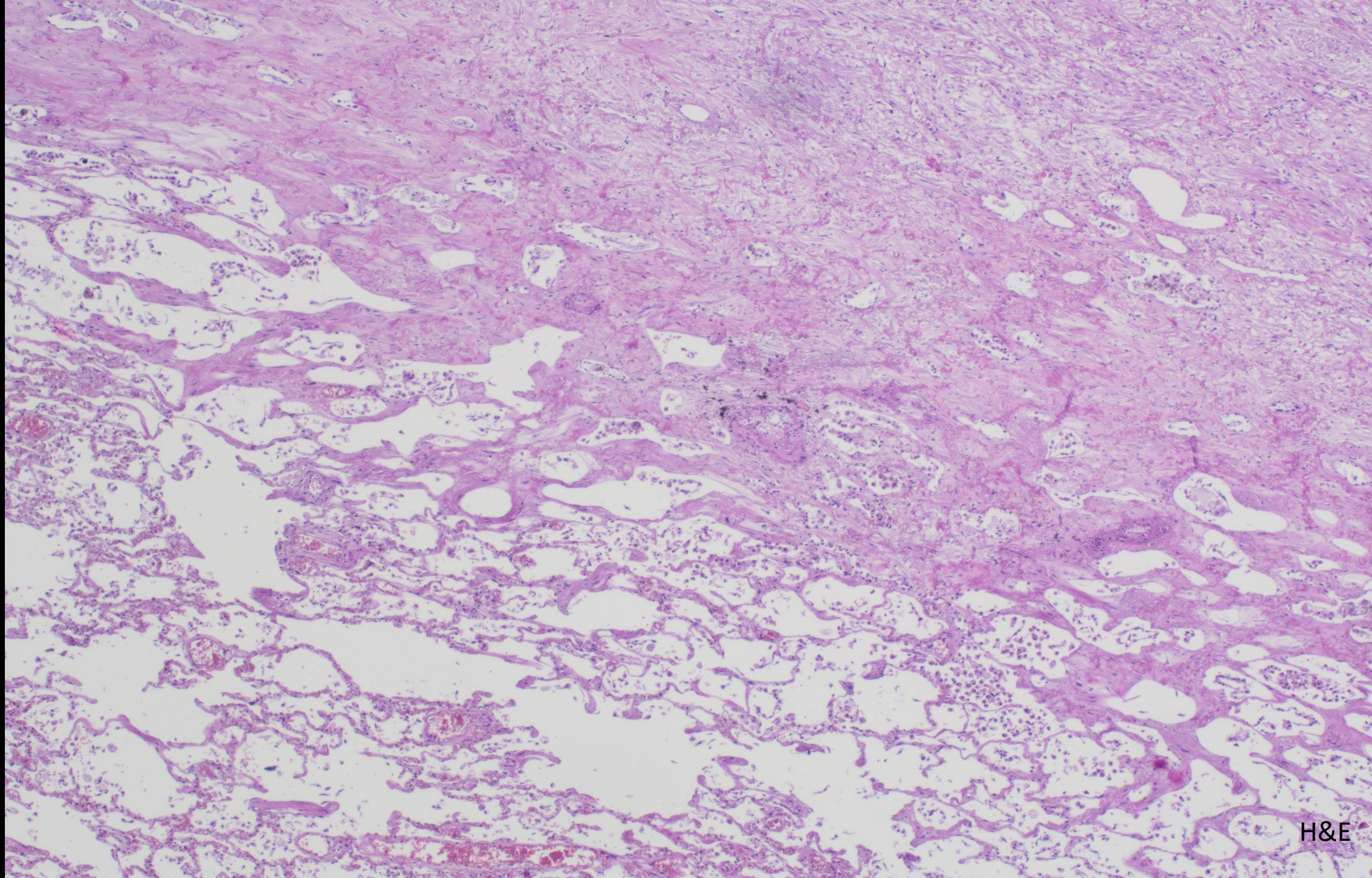
ELASTIQUES VERHOOIK





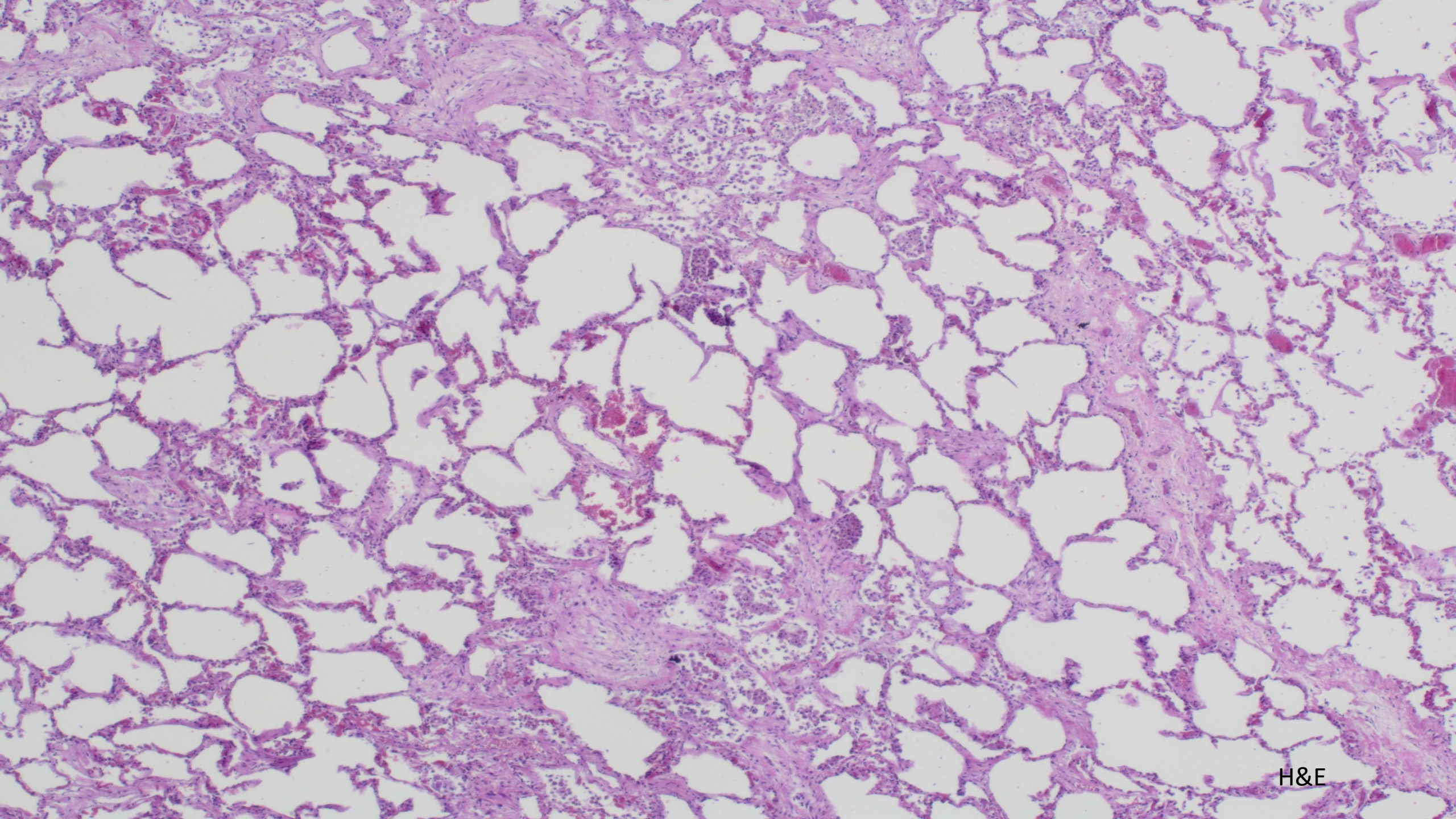
ELASTIQUES VERHOOK





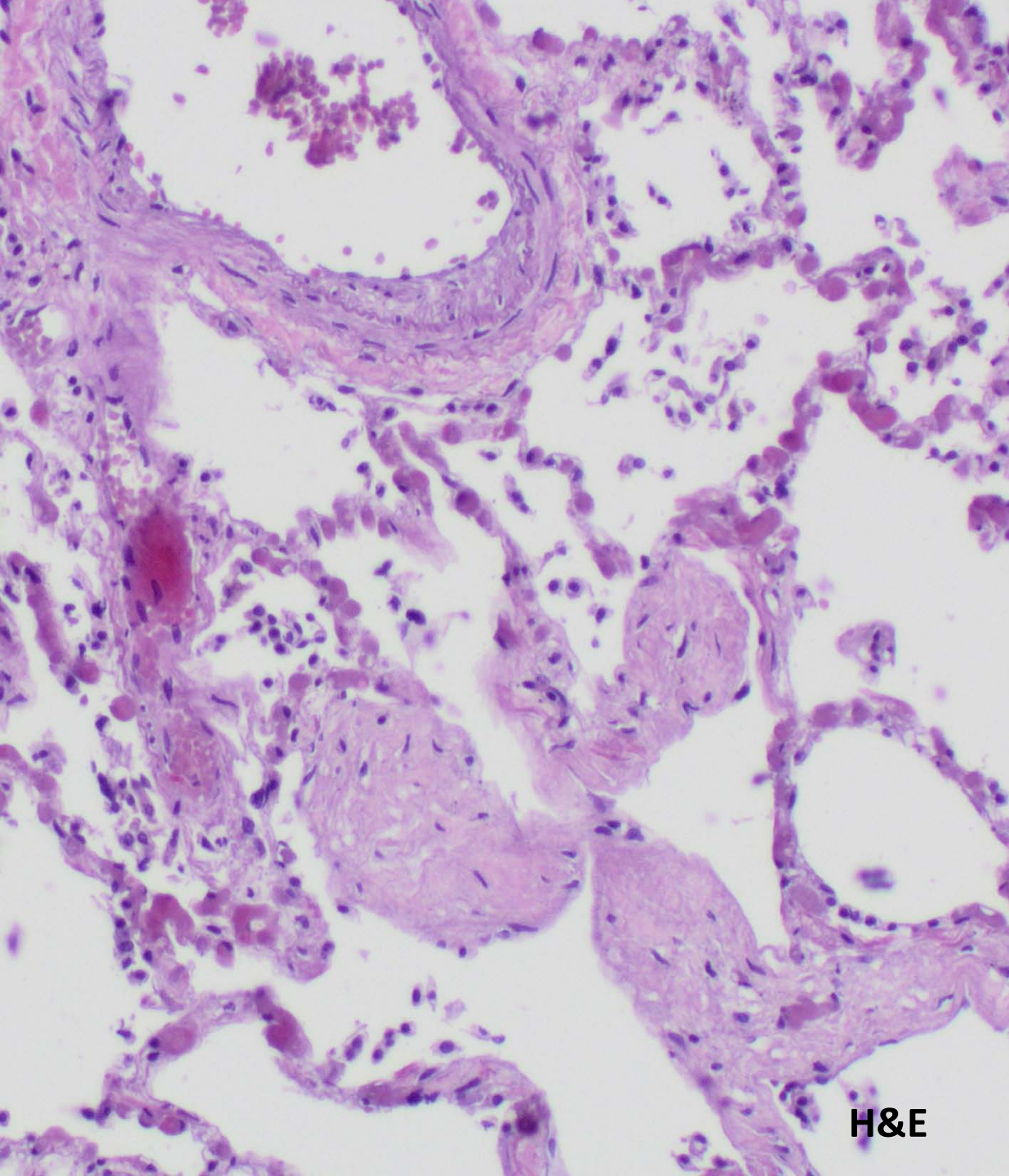
H&E



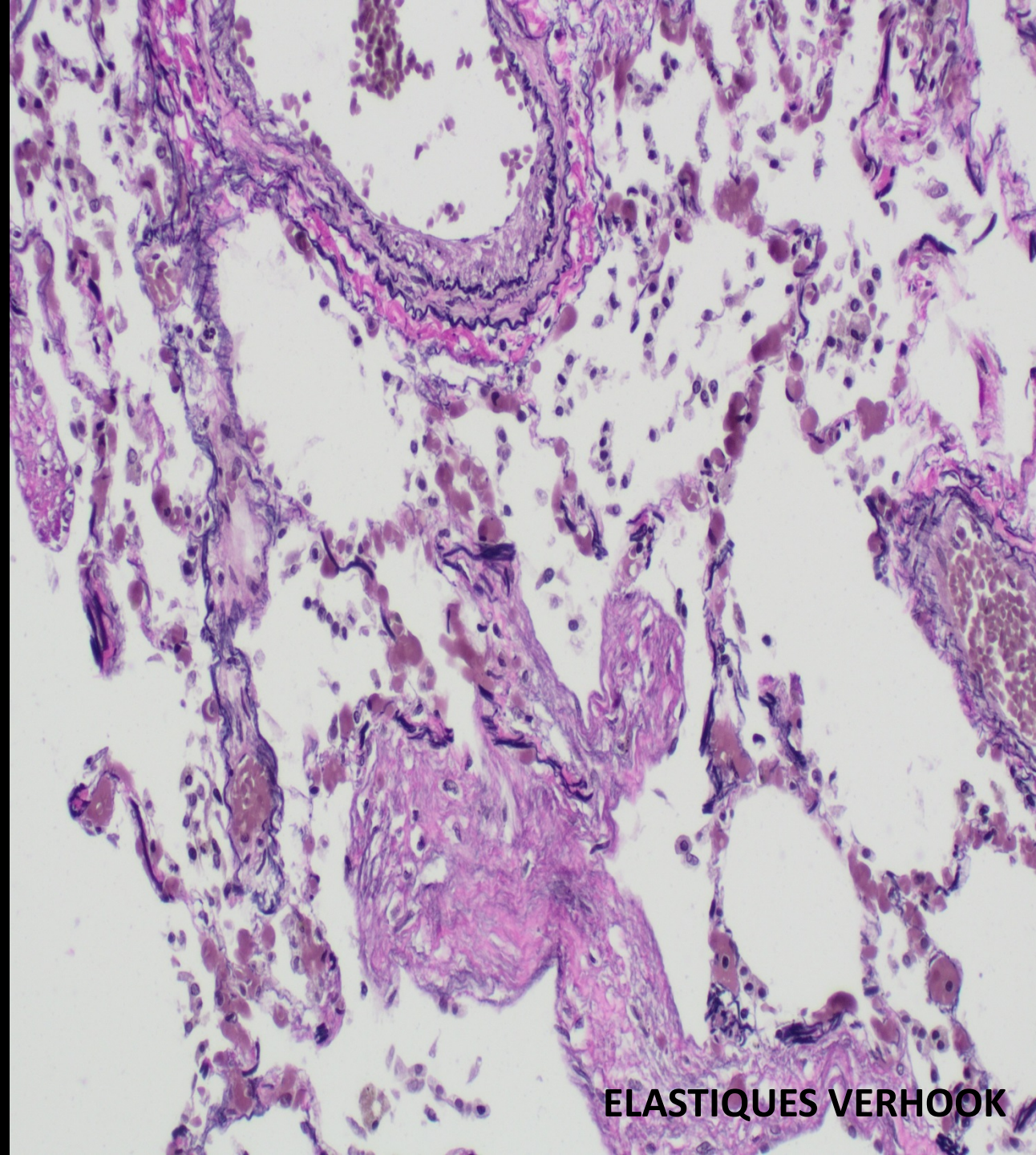


H&E



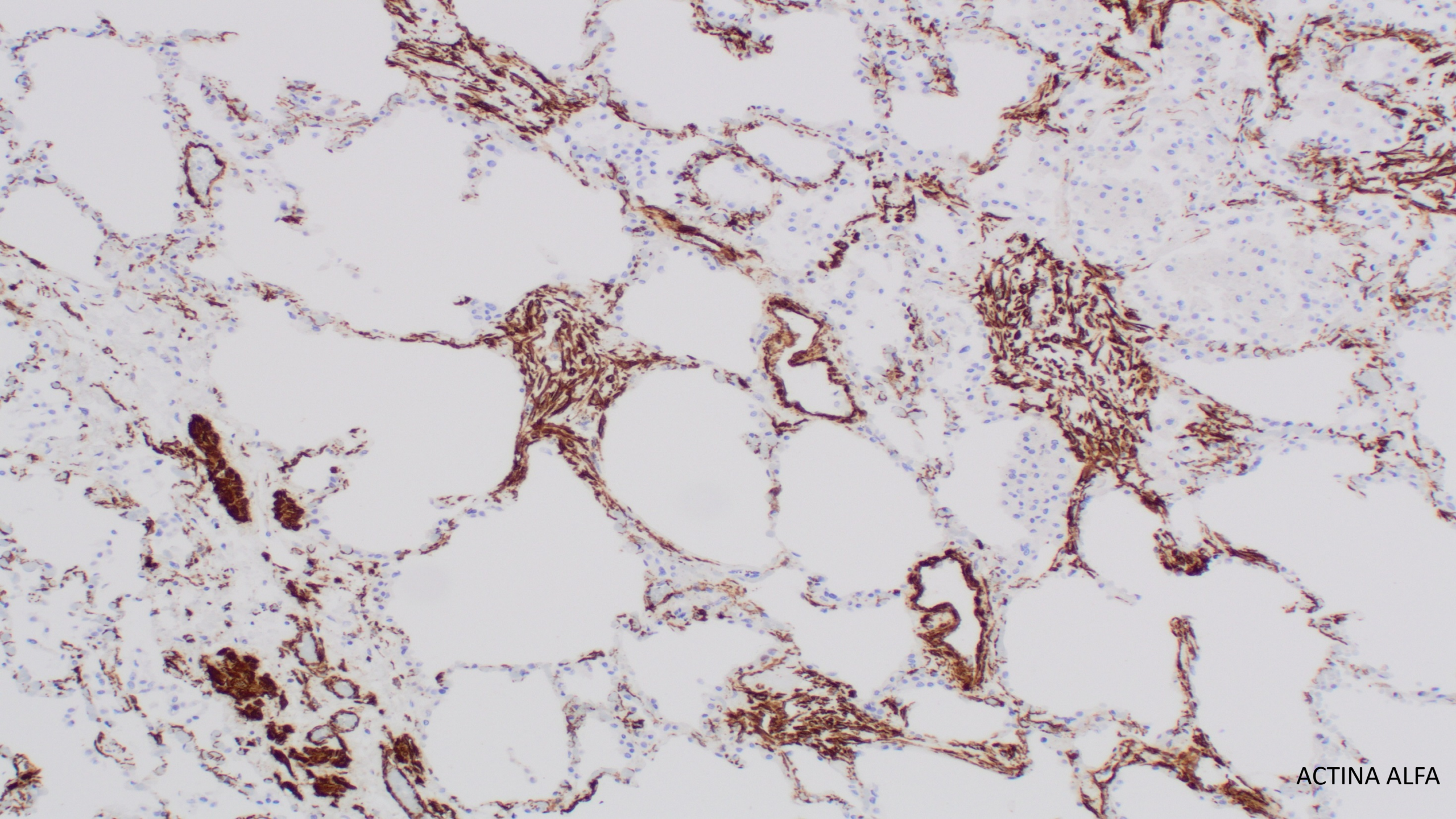


**H&E**



**ELASTIQUES VERHOOEK**





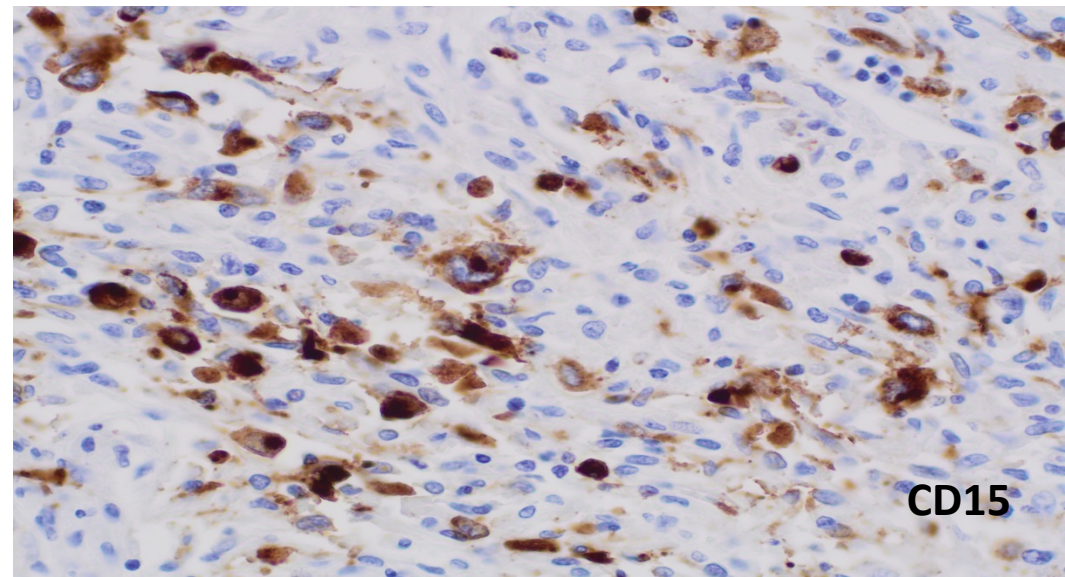
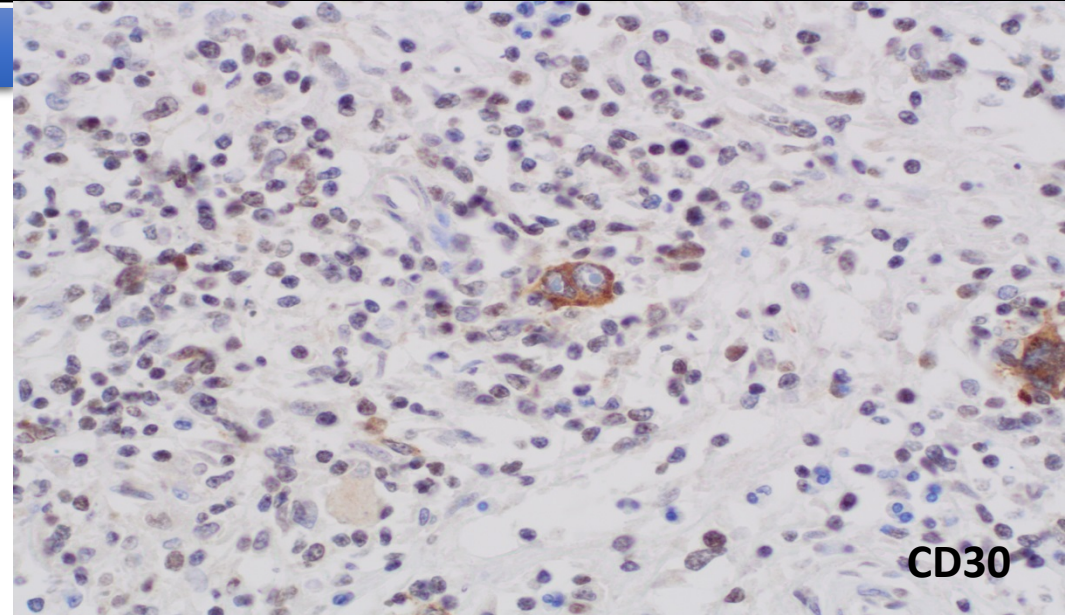
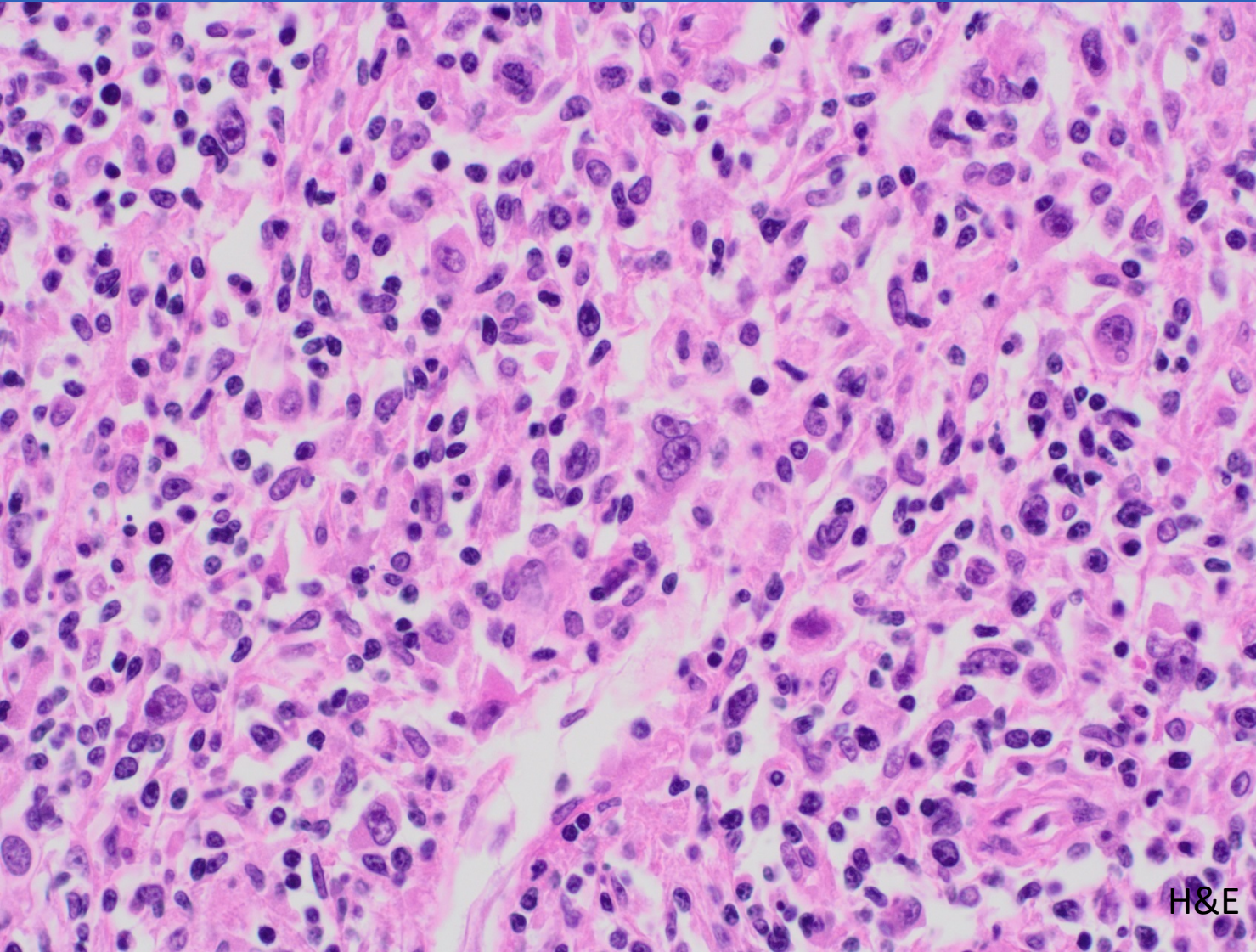
ACTINA ALFA



# LINFOMA HODGKIN CLÁSICO

## tipo Esclerosis Nodular

GANGLIO PERIAÓRTICO





# AUTOPSIA DIAGNÓSTICO DEFINITIVO

## Proceso fundamental:

- **FIBROELASTOSIS PLEUROPARENQUIMATOSA** secundaria a alotrasplante de médula ósea



**Causa de muerte:** Insuficiencia respiratoria hipercápnica

**Otros Hallazgos:** Recidiva de Linfoma Hodgkin Clásico en riñones, ganglios linfáticos peri-aorticós.

# HISTORIA

## FIBROELASTOSIS PLEUROPARENQUIMATOSA

**1992** JAPAN

« AMITANI'S DISEASE »

O PULF= PULMONARY UPPER-LOBE FIBROSIS

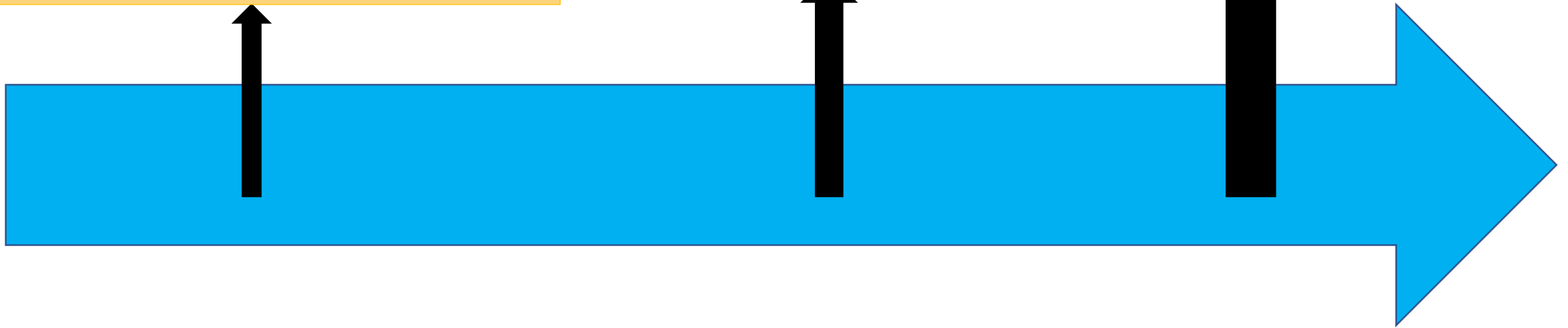
**2004**

« IDIOPATHIC PLEUROPARENCHYMAL  
FIBROELASTOSIS » O PPFE

FRANKEL

**2013**

INCLUIDA EN NEUMONÍAS  
INTERSTICIALES IDIOPÁTICAS  
ATS/ERS





# PREVALENCIA FIBROELASTOSIS PLEUROPARENQUIMATOSA



- 5,9% DE 205 BIOPSIAS EN 1622 PACIENTES CON ENFERMEDAD PULMONAR INTERSTICIAL
- **0,28% DE 700 TRASPLANTES DE MÉDULA ÓSEA**
- 7,5% DE 57 TRASPLANTES PULMONARES

Mariani F, Gatti B, Rocca A, Bonifazi F, Cavazza A, Fanti S, et al. Pleuroparenchymal fibroelastosis: the prevalence of secondary forms in hematopoietic stem cell and lung transplantation recipients. *Diagn Interv Radiol* 2016;22;400-406.

# CLÍNICA

- Edad media: 53 años
- **Bimodal:** pico en década de los 30 y 60 años

**SÍNTOMAS:** - Disnea, Tos, Pérdida de peso, Dolor Pleurítico  
- Episodios de **NEUMOTÓRAX**

**SIGNOS:** - Platitórax

# PRUEBAS RESPIRATORIAS



- PATRON VENTILATORIO RESTRICTIVO PARENQUIMATOSO
- DLCO DISMINUIDO
- HIPOXEMIA CON HIPERCAPNIA

# TAC CRITERIOS RADIOLÓGICOS

- **ENGROSAMIENTO PLEURAL Y FIBROSIS SUBPLEURAL**
- **PREDOMINIO APICAL BILATERAL**

➤ **ENGROSAMIENTO IRREGULAR DE PLEURA VISCERAL**

➤ **BULLAS SUBPLEURALES**

➤ **MOSAICISMO**



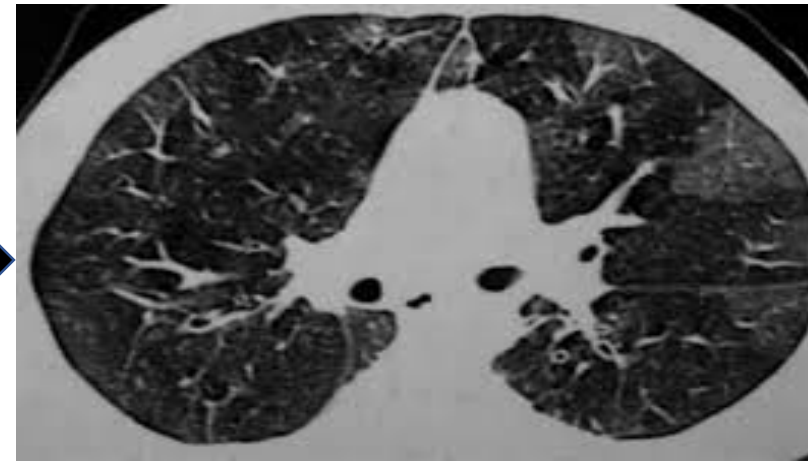
Bronquiolitis  
obliterans



➤ **FIBROSIS IRREGULAR PLEUROPARENQUIMAL**

➤ **FIBROSIS INTERSTICIAL FINA**

➤ **VIDRIO DESLUSTRADO**

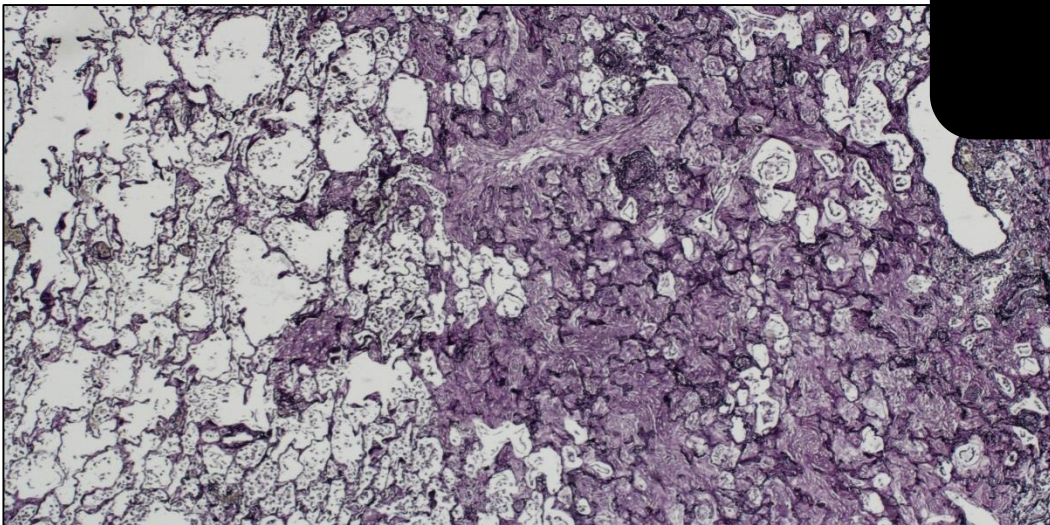


**CAMBIOS  
ABRUPTOS**

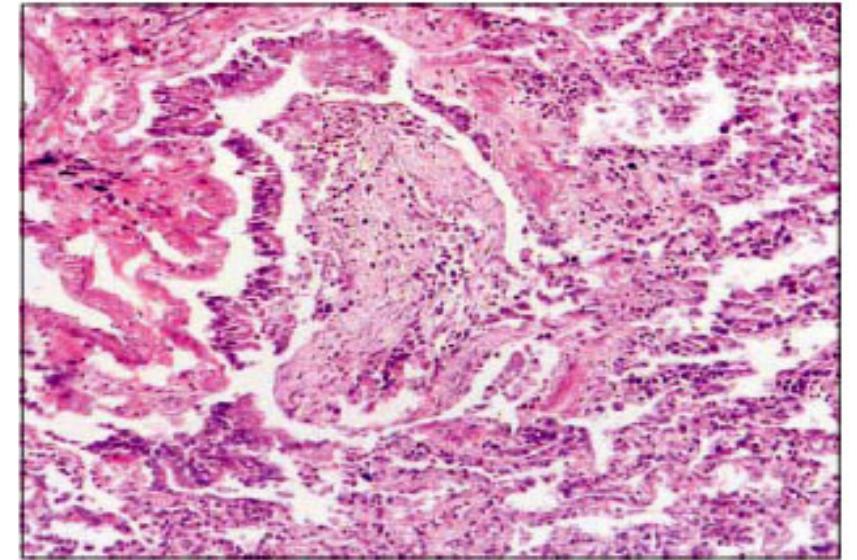
# CRITERIOS HISTOPATOLÓGICOS FIBROELASTOSIS PLEUROPARENQUIMATOSA

- FIBROSIS PLEURAL BIAPICAL
- FIBROSIS INTRAALVEOLAR SUBPLEURAL
- ELASTOSIS ALVEOLAR SEPTAL

CAMBIOS  
ABRUPTOS



➤ BRONQUIOLITIS OBLITERANTE



**Figura 2.** Bronquiolitis obliterante. Tinción de hematoxilina y eosina de 10 X.



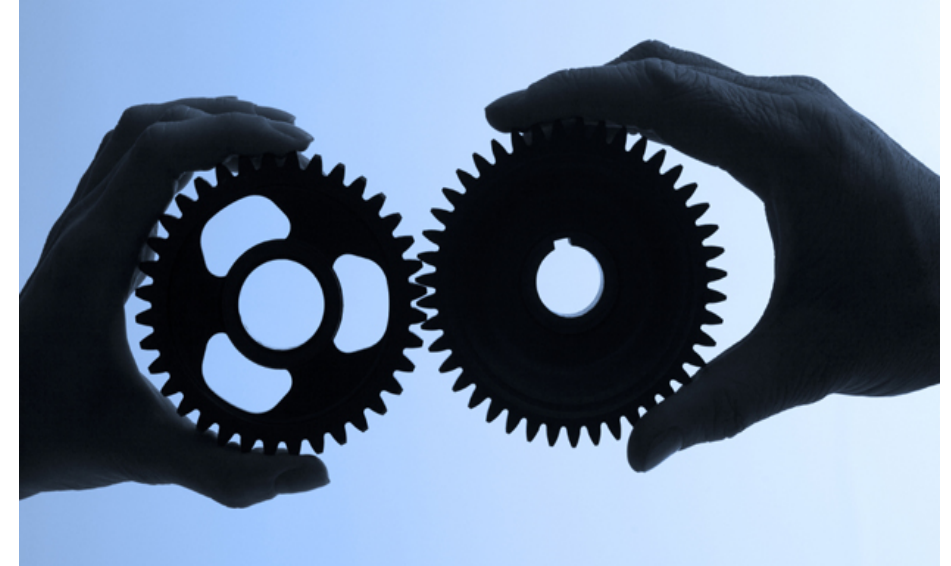
# PATOGENIA

## EICH CON BRONQUIOLITIS OBLITERANS

**INCIDENCIA BO**  
EN FIBROELASTOSIS  
PLEUROPARENQUIMATOSA **71%**



FIBROELASTOSIS  
PLEUROPARENQUIMATOSA



**TRASPLANTES MÉDULA ÓSEA/PULMÓN**  
QUIMIOTERAPIA  
RADIOTERAPIA

Cause of pleuroparenchymal fibroelastosis following allogeneic hematopoietic stem cell transplantation. Respiratory investigation 57 (2019) 321-324

# DIAGNÓSTICO DIFERENCIAL ENFERMEDAD PULMONAR POST TRASPLANTE MEDULAR

## ➤ SÍNDROME DE BRONQUIOLITIS OBLITERANS = EICH PULMONAR

➤ NEUMONÍA ORGANIZADA

➤ FIBROELASTOSIS PLEUROPARENQUIMATOSA

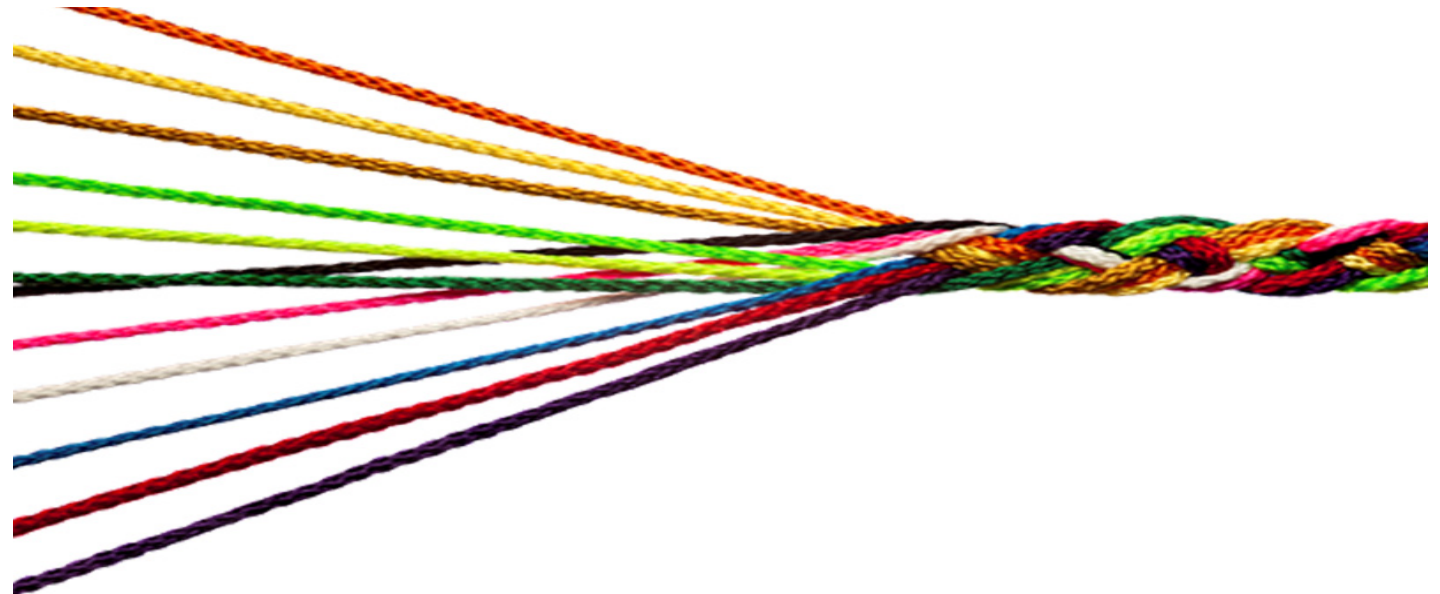
➤ OTRAS: Daño Aveolar Difuso, Neumonía Organizada Fibrinosa Aguda

ENFERMEDAD INTERSTICIAL

# CONCLUSIONES

## FIBROELASTOSIS PLEUROPARENQUIMATOSA

- ES UN PATRÓN EVOLUTIVO HACIA UN PROCESO FIBRÓTICO TERMINAL
- SE ASOCIA A TRASPLANTE DE MÉDULA ÓSEA/PULMÓN
- PRONÓSTICO INFAUSTO





# BIBLIOGRAFÍA

- Hisao Higo, Nobuaki Miyahara, Akihiko Taniguchi, Yoshinobu Maeda, Katsuyuki Kiura. Cause of pleuroparenchymal fibroelastosis following allogeneic hematopoietic stem cell transplantation. *Respiratory investigation* 57 (2019) 321-324
- Felix Chua, Sujal R. Desai, Andrew G. Nicholson, Anand Devaraj, Elisabetta Renzoni, Alexandra Rice, Athol U. Wells. Pleuroparenchymal fibroelastosis. A review of clinical, radiological, and pathological characteristics. *Ann Am Thorac Soc* Vol 16, No 11, pp 1351-1359, Nov 2019.
- Jesus Duque-Alfonso, Gabriele Ihorst, Miguel Waterhouse, Robert Zeiser, Ralph Wasch, Hartmut Bertz, Joachim Muller-Quernheim, Jurgen Finke, Reinhard Marsk, Antje Prasse. Impact of lung Function on bronchiolitis obliterans syndrome and outcome after allogeneic hematopoietic cell transplantation with reduced-intensity conditioning. *Biol Blood Marrow Trasplant* 24 (2018) 2277-2284. doi: 10.1016/j.bbmt.2018.06.024.
- Mariani F, Gatti B, Rocca A, Bonifazi F, Cavazza A, Fanti S, et al. Pleuroparenchymal fibroelastosis: the prevalence of secondary forms in hematopoietic stem cell and lung transplantation recipients. *Diagn Interv Radiol* 2016;22;400-406.
- Anne Bergeron, MD, PhD, Guang-Shing Cheng, MD. Bronchiolitis obliterans syndrome and other late pulmonary complications after allogeneic hematopoietic stem cell transplation. *Clin Chest Med* 38 (2017) 607-621.
- Lee Gazourian, Laura Spring, Emily Meserve, David Hwang, Alejandro A. Diaz, Samuel Y. Ash, Vincent T.Ho, Lynette M. Sholl, George R. Washko. Pulmonary Clinicopathological correlation after allogeneic hematyopoietic stem cell transplantation: an autopsy series. *Biol Blood Marrow Trasplant* (2017) 1767-1772. doi: 10.1016/j.bbmt.2018.06.024.
- Jan H von der Thusen, David M Hansell, Masaki Tominaga, Paul A Veys, Michael T Ashworth, Catherine M Owens and Andrew G Nicholson. Pleuroparenchymal fibroelastosis in patient with pulmonary disease secondary to bone marrow transplantation. *Modern Pathology* (2011) 24, 1633-1639.
- Martina Bonifazi, M. Angeles Montero, Elisabetta A. Renzoni. Idiopathic pleuroparenchymal fibroelastosis. *Nutrition and clinical care. Curr Pulmonol Rep* (2017) 6:9-15.
- Shaun Kian Hong Cheng, MB ChB, Khoon Leong Chuah, MBBS, FRCPA. Pleuroparenchymal fibroelastosis of the lung. A review. *Arch Pathol Lab Med- Vol* 140, August 2016.
- Ho Namkoong, Makoto Ishii, Takehiko Mori, Hiroaki Sugiura; Sadatomo Tasaka, Masatoshi Sakurai, Yuya Koda, Jun Kato, Naoki Hasegawa, Shinichiro Okamoto and Tomoko Betsuyaku. Clinical and radiological characteristics of patients with late-onset severe restrictive lung defect after hematopoietic stem cell transplantation. *BMC Pulmonary Medicine* (2017) 17:123.