Vascular Ehlers-Danlos syndrome

Pneumothorax

Pneumothorax is frequently associated with vascular Ehlers-Danlos syndrome and is the most frequent respiratory manifestation of the disease. It can occur from adolescence onwards and sometimes precedes genetic diagnosis. Treatment usually involves the insertion of a chest tube (complete pneumothorax).



WHAT IS RECOMMENDED

• In the event of a failure to remove the chest tube, a mechanical pleurodesis should be discussed at the time of the first occurrence.



WHAT YOU SHOULD NOT DO

- Pleurodesis in the event of first episode without a failure to remove the chest tube.
- Pleurodesis in cases of partial or prophylactic pneumothorax with asymptomatic bullous lesions in a patient without pneumothorax.