

ANCA-associated vasculitides

- In the proper clinical setting, a positive c-ANCA has sufficient PPV that biopsy may be deferred
- A positive p-ANCA lacks sensitivity and provides no more than suggestive evidence of CSS, MPA, or idiopathic pauci-immune RPGN because it can be found in a wide variety of settings (i.e. rheumatoid arthritis and Goodpasture's syndrome)
- There is insufficient sensitivity and specificity of an isolated rise in ANCA titer to accurately predict disease relapse in WG or other vasculitis

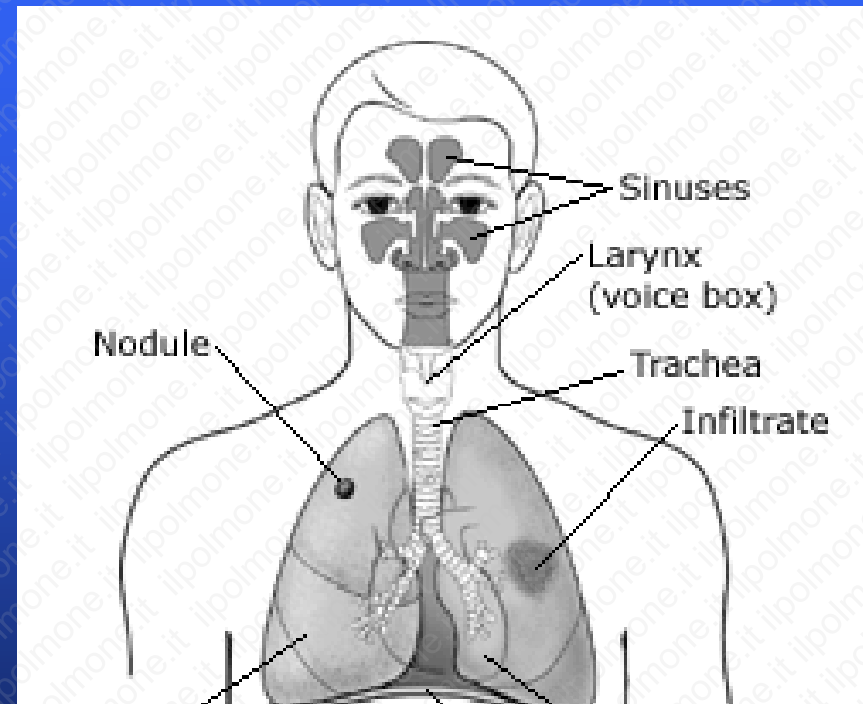
Other Laboratories

- Culture of blood and other affected organs (when applicable) must be obtained to exclude infection
- Routine laboratories
- An \uparrow ERS and C-reactive protein are expected
- Urinalysis with the microscopic examination on a fresh sample

Wegener's Granulomatosis

It is clinically characterized by the triad of:

- ◆ Upper airway disease
- ◆ Lower respiratory tract disease
- ◆ Glomerulonephritis



Key Message

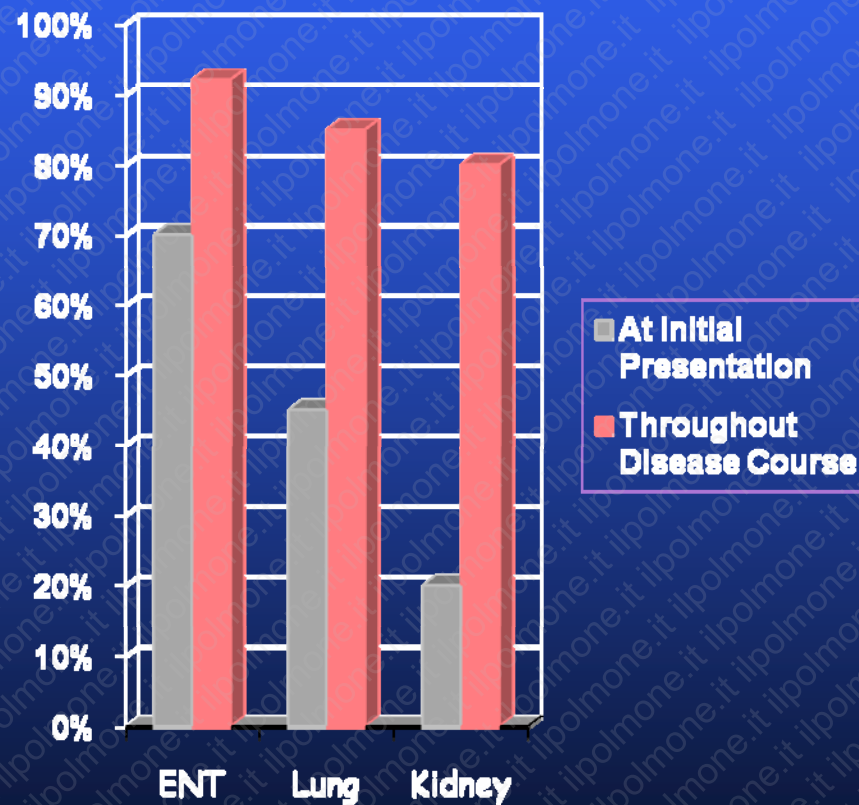
The complete triad is frequently not present at initial presentation

Symptom Prevalence in Wegener's Granulomatosis

| Feature of disease | Prevalence at onset | Prevalence during course of disease |
|---------------------|---------------------|-------------------------------------|
| Ear Nose and Throat | 70% | 90% |
| Lung | 45% | 85% |
| Eye | 15% | 50% |
| Glomerulonephritis | 20% | 70% |
| Musculoskeletal | 30% | 70% |

Hoffman et al, 1998

Wegener's Granulomatosis



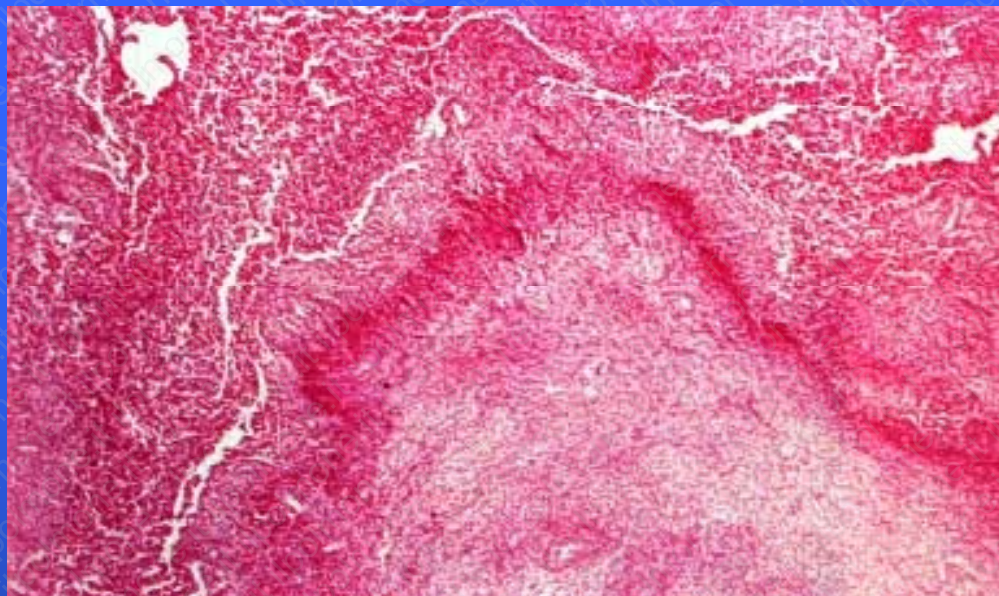
Klippel, 1998

- About 50% have no lung involvement at presentation
- Lung involvement:
 - ◆ Infiltrates
 - ◆ Nodules
 - ◆ Hemoptysis
 - ◆ Pleuritis
- 33% with lung involvement are asymptomatic
- About 80% have no renal involvement at presentation

Wegener's Granulomatosis

clinical manifestations

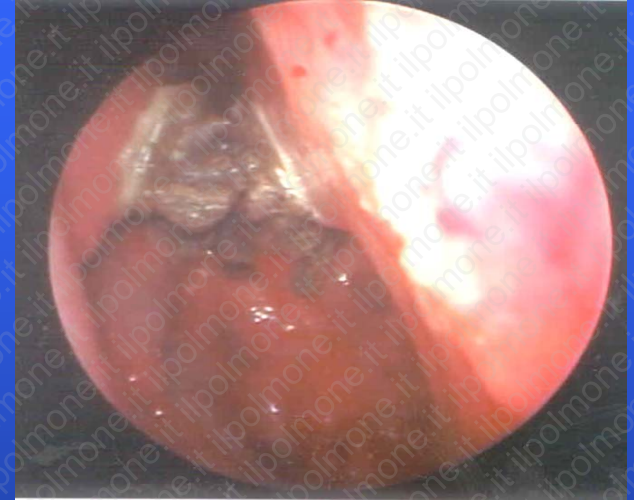
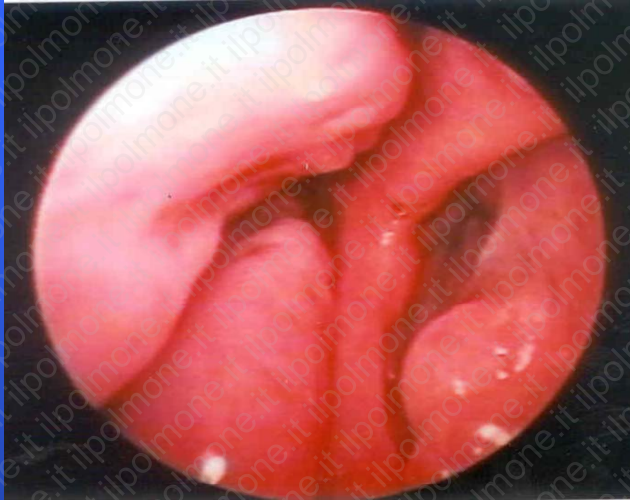
| <i>Clinical manifestations</i> | <i>Frequency (%)</i> |
|---|----------------------|
| Pulmonary (cough, hemoptysis, dyspnea, chest pain) | 70-95 |
| Upper airway (epistaxis, sinusitis, rhinorrhea, otitis, hearing impairment, ear pain, destructive lesions/bony deformities ulcerations) | 70-95 |
| Tracheobronchial (subglottic stenosis, bronchial Stenosis, endobronchial lesion) | 10-55 |
| Renal/glomerulonephritis | 50-85 |
| Cutaneous (purpura, ulcers, vesicles or nodules) | 45-60 |
| Musculoskeletal (arthralgias, myalgias, arthritis) | 30-70 |
| Ocular (conjunctivitis, uveitis, episcleritis, scleritis, proptosis) | 25-55 |



Wegener's Granulomatosis



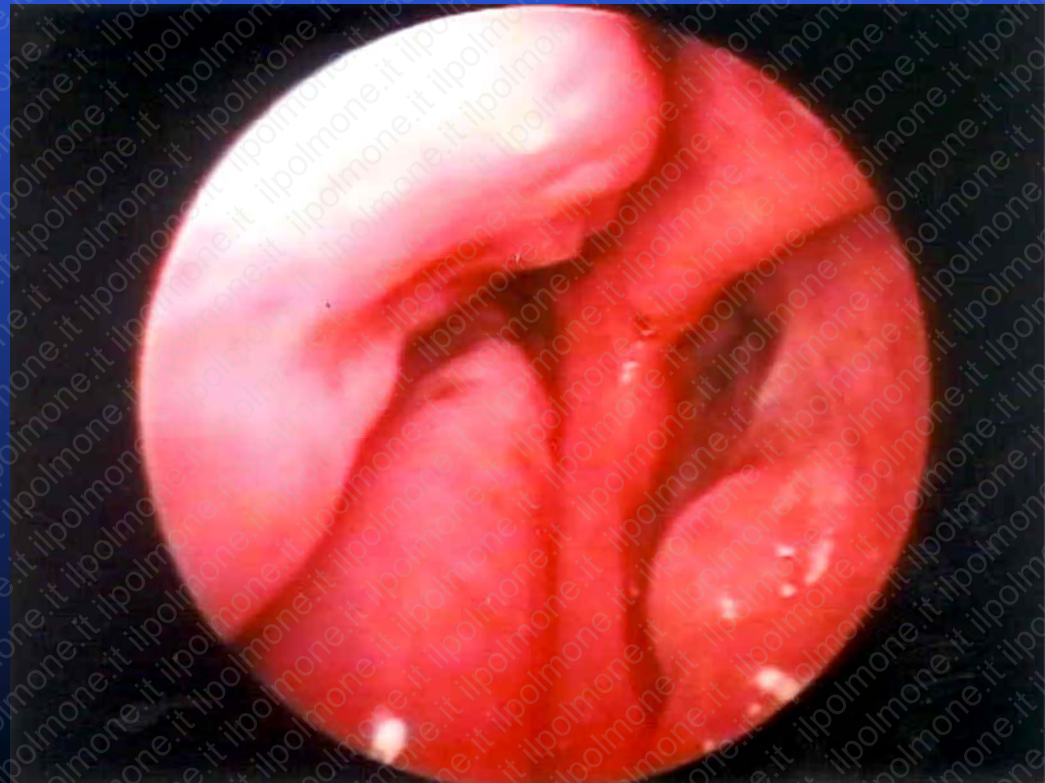
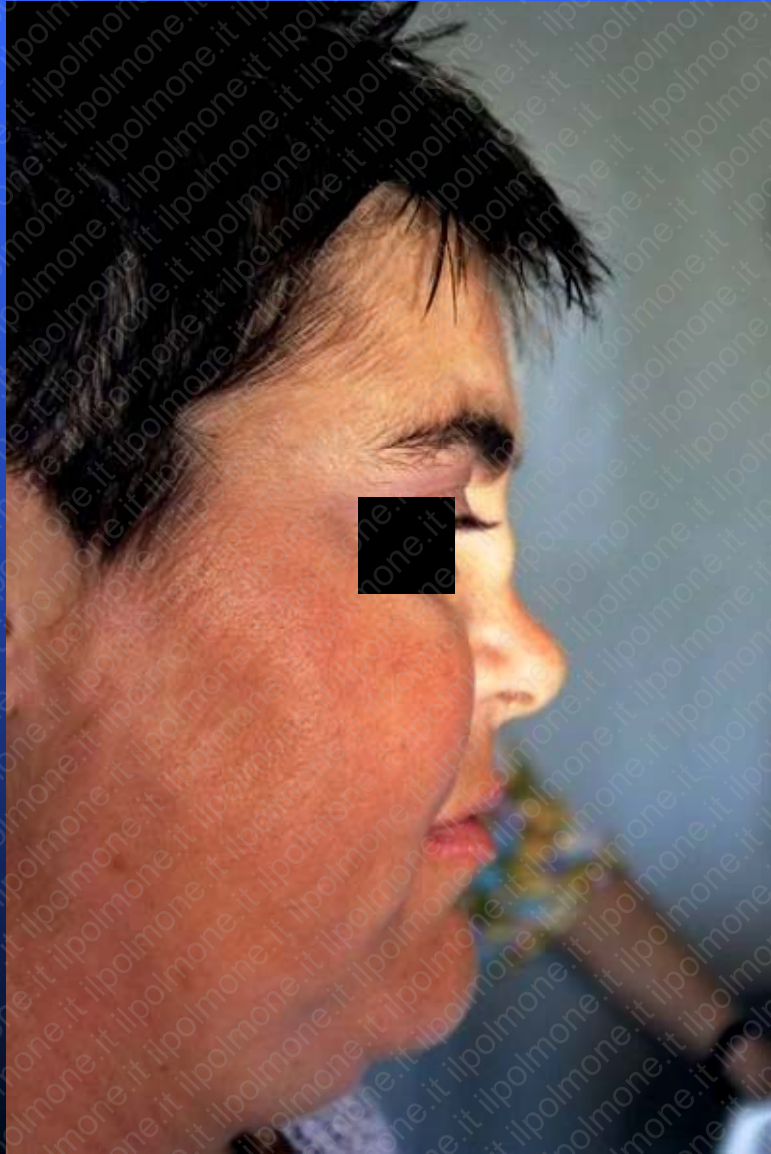
Upper Airway Involvement in Wegener's Granulomatosis



Saddle nose

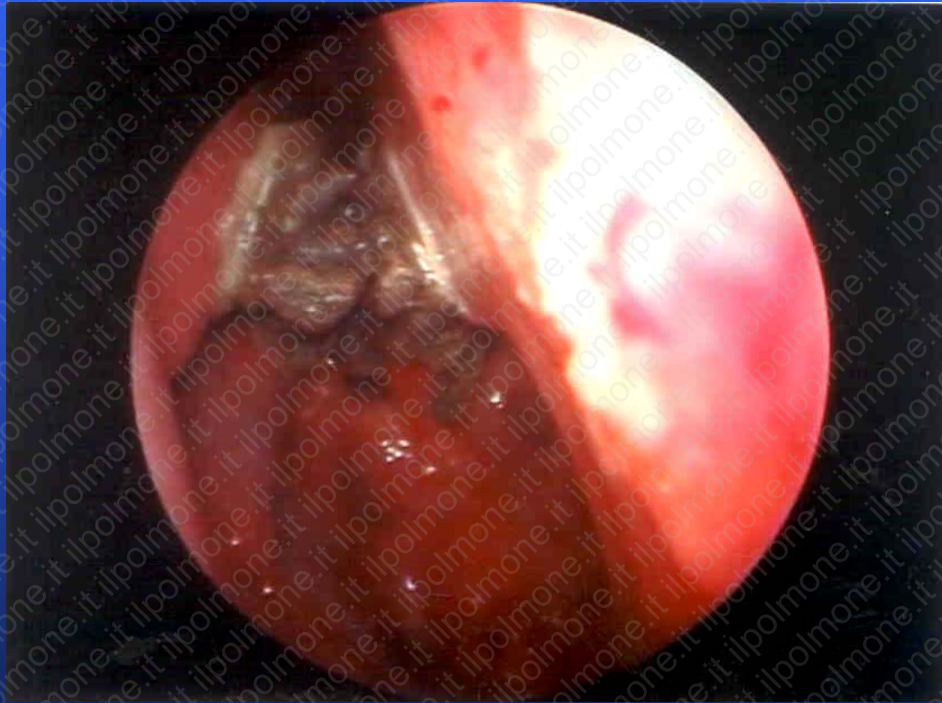
13%

Perforated nasal septum

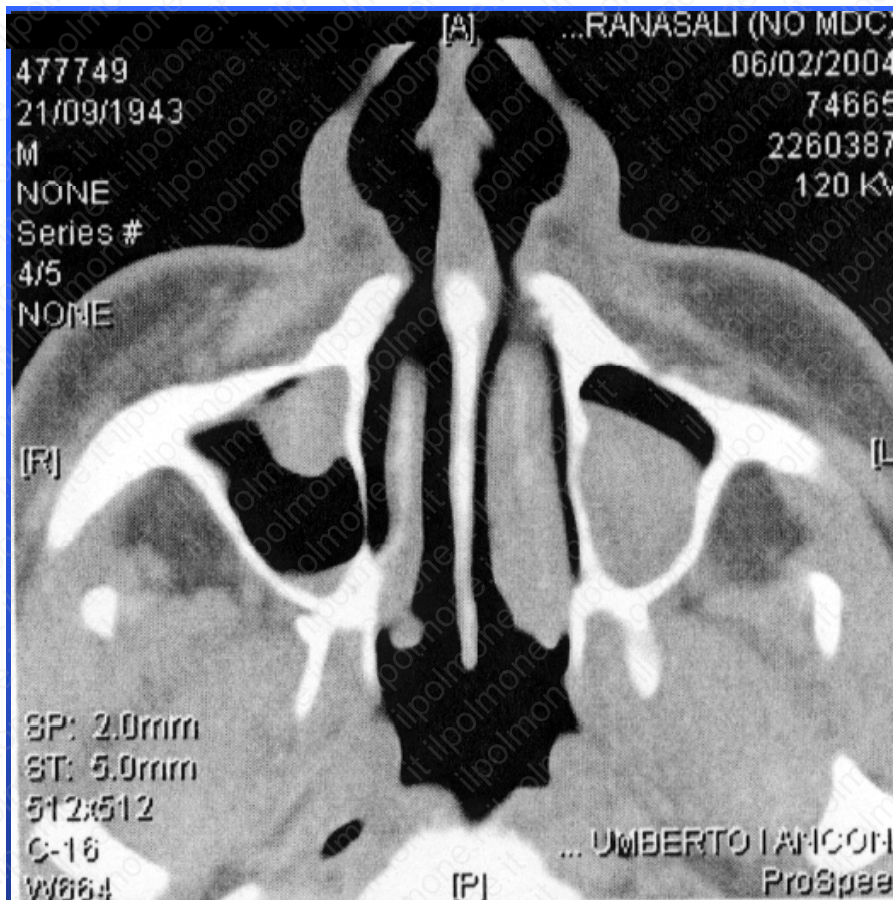


Crust between nasal septum and turbinate

65%



4x3cm



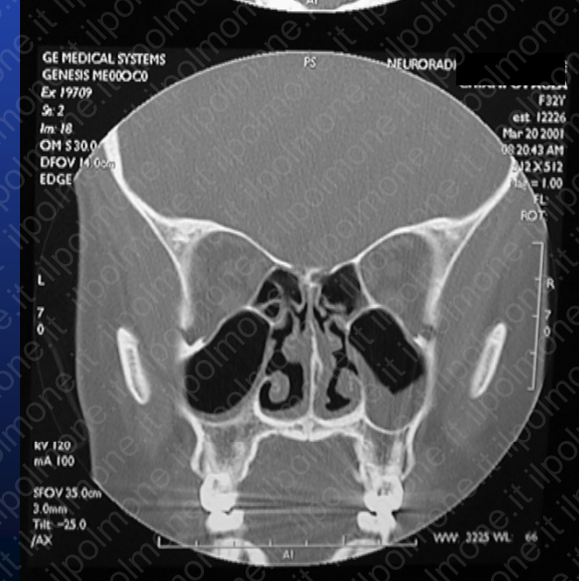
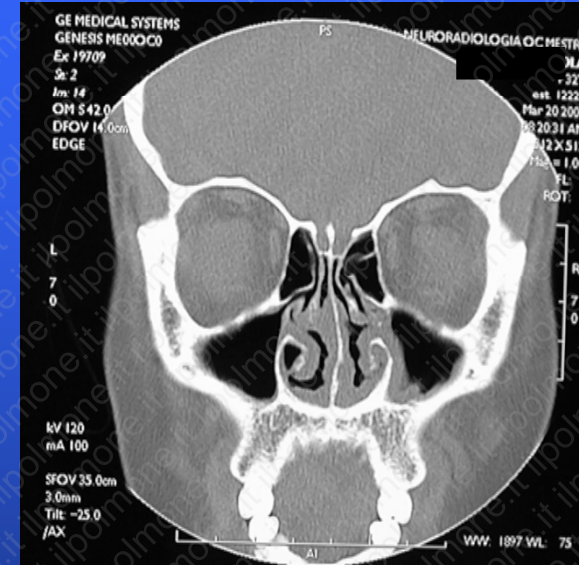
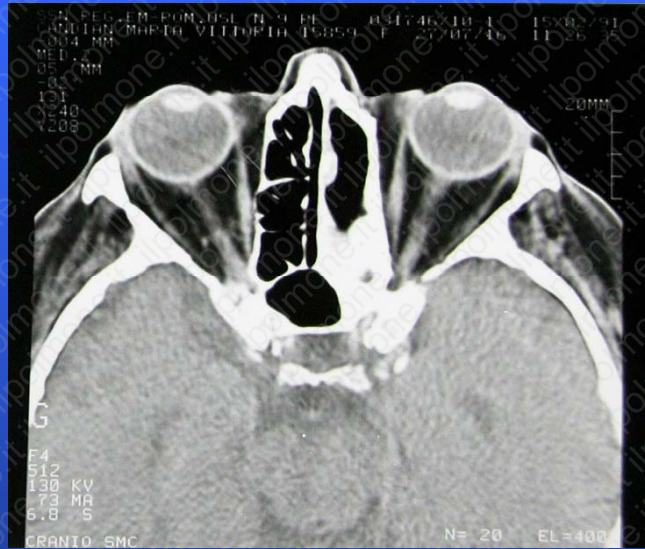
Erosion of the medial wall of
the right maxillary sinus
Osteosclerosis of
right maxillary sinus
Disappearance of right
turbinate
Left-sided sinusitis

Sinusitis

20%

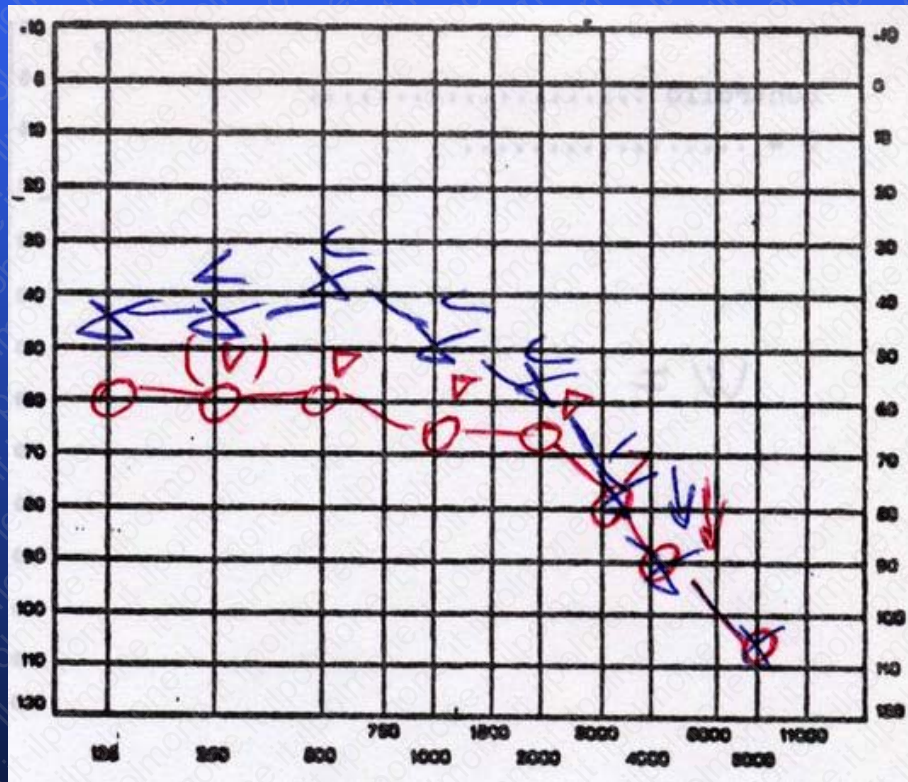


Upper Airway Involvement in Wegener's Granulomatosis

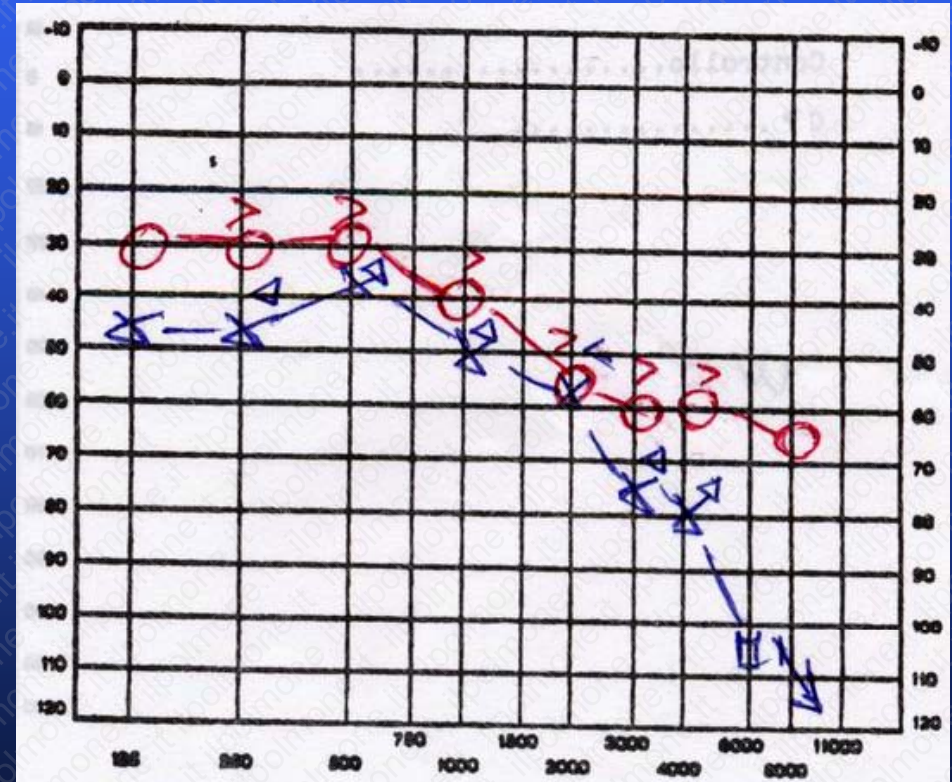


Hearing loss

20%



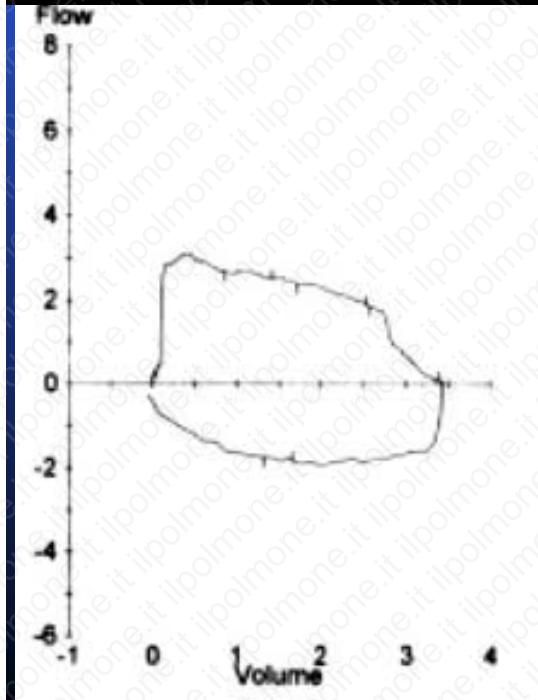
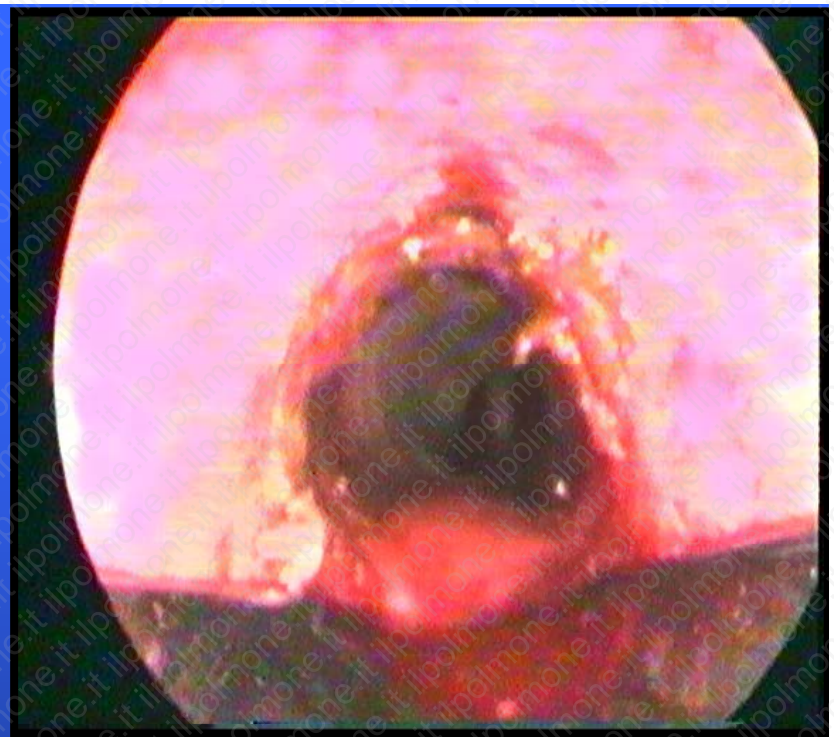
Before treatment



After treatment



13%



Subglottic stenosis
Therapeutic management
 Flow-volume loops

