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IgG4-related lung disease

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IgG4-related disease:

- The etiology of IgG4-related disease remains unclear.
- Involvement of complex **genetic susceptibility factors** and resultant **abnormality in immune responses** have been suggested to underlie development of the disease.
- Aberrant innate and adaptive immunity are considered as the main pathogenesis of IgG4-RD.

IgG4-related disease:

• REFERENCES:

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An update on IgG4-related lung disease

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BRIEF COMMUNICATION

IgG4-related respiratory disease

IgG4-related disease: a new challenging diagnosis mimicking lung cancer

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Case report

 OPEN ACCESS

IgG4-related lung disease as a differential diagnosis of a lymphoproliferative disease: atypical presentation of an atypical disease

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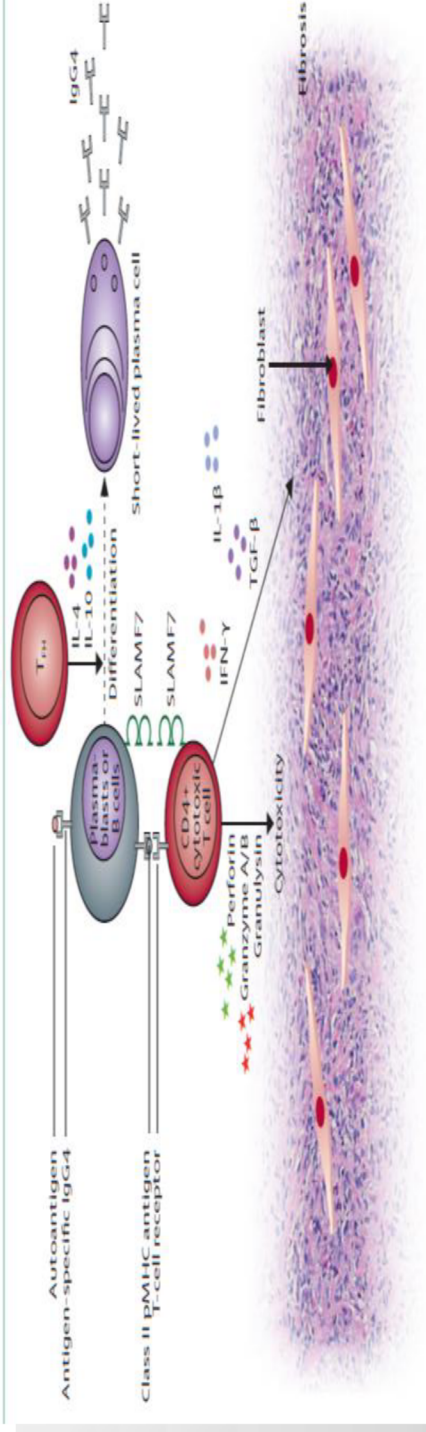
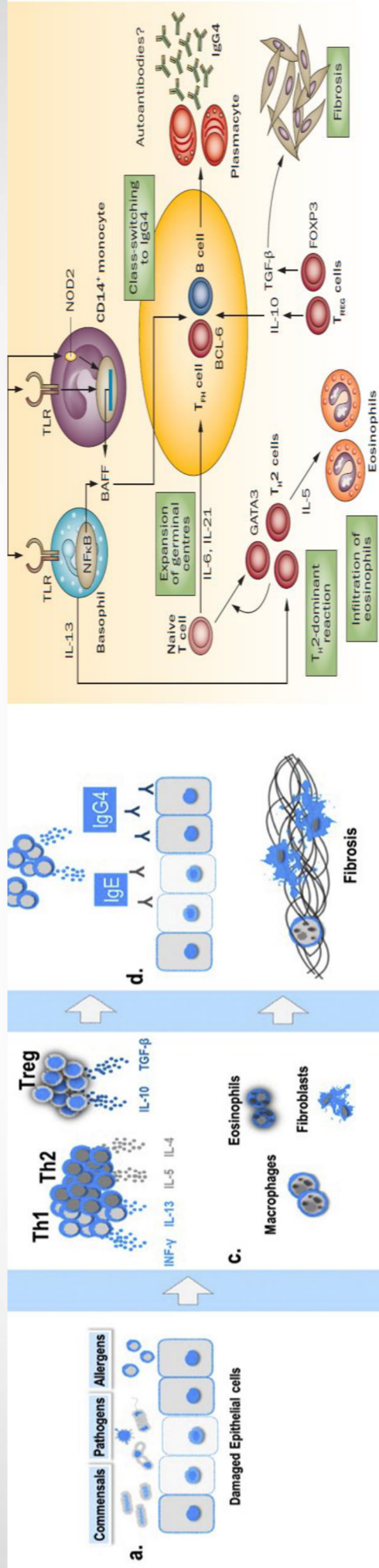
IgG4-related disease:

- IgG4-RD was reported first in the late 1990's.
- IgG4-RD is a systemic fibroinflammatory process characterized by substantial infiltration of plasma cells with IgG4 in target organs, regardless of plasma IgG4 level.
- only about 50% of patients have elevated IgG4 levels in serum.
- IgG4-related lung disease (IgG4-RLD) was first reported in 2004.
- IgG4-RLD is primarily composed of four clinical pulmonary syndromes :inflammatory lung pseudotumor, central airway disease, interstitial pneumonia, and pleuritis.

IgG4-related disease:

- **Characteristic radiologic manifestations** in the lung include nodules, ground glass opacities, alveolar and interstitial disease, and bronchovascular bundle thickening.

• Pathophysiology



IgG4-related disease:

Epidemiology: In general, IgG4-RD is predominantly seen in adults, mostly older males (70–80%).

- AIP type I is the only disease subtype that has been studied in detail .
- Lung specific involvement in AIP is reported in 9.5% to 51.2% of patients.
- There is a subgroup of patients that experienced IgG4-RD in lungs without involvement of other organs.

IgG4-related disease:

Clinical presentation:

- Pulmonary lesions have been subclassified based on their intrathoracic compartments.
- **Airway manifestations** :Airway manifestations may include rhino-sinusitis, asthmatic symptoms, bronchial tumors, and airway stenosis.
- **Mediastinal manifestations**: Hilar and mediastinal lymphadenopathy and Mediastinal fibrosis are the most common findings in IgG4-RRD.