



# WEGENER 'S GRANULOMATOSIS



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# WEGENER'S GRANULOMATOSIS

- ❖ A rare primary systemic vasculitis ( PSV ) of childhood.
- ❖ Necrotizing granulomatous inflammation of small to medium vessels.
- ❖ Typically affecting the upper and lower respiratory tract and the kidneys.
- ❖ Incidence 0.03-3.2 per 100,000 children per year.



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## ❖ Definition and classification criteria of WG

ACR 1990 criteria (2/4 )	EULAR/PRESS criteria (3/6)
<ul style="list-style-type: none"><li>- Nasal or oral inflammation</li><li>- Abnormal chest radiograph (nodules, fixed infiltrates or cavities )</li><li>- Abnormal urinary sediment ( microhematuria or red cell cast )</li><li>- Granulomatous inflammation on biopsy</li></ul>	<ul style="list-style-type: none"><li>- Nasal or sinus inflammation</li><li>- Abnormal chest radiograph or chest CT scan</li><li>- Abnormal urinalysis ( hematuria and/or significant proteinuria )</li><li>- Granulomatous inflammation on biopsy/necrotizing pauci-immune GN</li><li>- Subglottic, tracheal, or endobronchial stenosis</li><li>- Anti-PR3 ANCA or c-ANCA staining</li></ul>



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## ❖ Clinical subgroup of WG according to the definitions of the EUVAS

Subgroup	Organ involvement	Constitutional Symptoms	Presence of ANCA
Localized	Upper and/or lower respiratory tract	No	+/-
Early systemic	Any except renal or imminent organ failure	Yes	Usually +
Generalized	Renal with serum creatinine $\leq 500$ $\mu\text{mol/l}$ and/or other imminent organ failure	Yes	+
Severe renal	Renal with serum creatinine $> 500$ $\mu\text{mol/l}$	Yes	+
Refractory	Progressive disease despite therapy with corticosteroids and cyclophosphamide	Yes	+/-

EUVAS = The European Vasculitis Study Group



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## ❖ Pathophysiology : granulomatous lesion

- WG starts as granulomatous disease in the respiratory tract and systemic vasculitis develops subsequently.
  - early foci of fibrinoid necrosis could be a consequence of PR3-ANCA-induced necrotizing capillitis.
  - the granulomatous lesions are built up by CD4<sup>+</sup> T-cells, CD8<sup>+</sup> T-cells, histiocytes, CD20<sup>+</sup> B-lymphocytes, neutrophil granulocytes, CD68<sup>+</sup> macrophages and CD68<sup>+</sup> multinucleated giant cells surrounding a central necrosis
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