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)
- Nasal or oral inflammation - Abnormal chest radiograph (nodules,fixed infiltrates or cavities) - Abnormal urinary sediment (microhematuria or red cell cast) - Granulomatous inflammation on biopsy	- Nasal or sinus inflammation - Abnormal chest radiograph or chest CT scan - Abnormal urinalysis (hematuria and/or significant proteinuria) - Granulomatous inflammation on biopsy/necrotizing pauci-immune GN - Subglottic, tracheal,or endobronchial stenosis -Anti-PR3 ANCA or c-ANCA staining

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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 ≤500 µmol/l and/or other imminent organ failure	Yes	+
Severe renal	Renal with serum creatinine > 500 µmol/l	Yes	+
Refactory	Progressive disease despite therapy with corticosteroids and cyclophosphamide	Yes	+/-

EUVAS = The European Vasculitis Study Group

WEGENER'S GRANULOMATOSIS

- 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 capillalitis.

  - the granulomatous lesion are built up by $CD4^+$ T-cells, $CD8^+$ T-cells, histiocytes, $CD20^+$ B-lymphocytes, neutrophil granulocytes, $CD68^+$ macrophages and $CD68^+$ multinucreated giant cells surrounding a central necrosis