



Pulmonary vasculitis



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Systemic diseases and the lung

- relatively rare in pediatric practice
- CNT diseases : SLE, dermatomyositis
- Pulmonary vasculitis
- Inherited CNT disorders : Ehlers-Danlos
- Mucopolysaccharidoses, familial dysautonomia, sarcoidosis
- LCH, pulmonary lymphangiomatosis

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Systemic diseases and the lung

- may cause significant respiratory complications
- treatment : often requires corticosteroids & immunosuppressants
- outcome : variable
- **when they affect the lung, carry a small but significant mortality**

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Systemic vasculitis

- true incidence in children is not known
- around 2-5 cases/million/year
- pathology of vasculitis : cellular inflammation, vessel destruction, tissue necrosis
- clinical features : site, size and type of vessel involved

Classification ของโรคในกลุ่ม **Vasculitis**

Primary idiopathic vasculitis

- **Small vessel**
 - **Wegener's granulomatosis**
 - **Microscopic polyangitis**
 - **Churg-Strauss syndrome**
 - **Idiopathic pauci-immune rapidly progressive glomerulonephritis**
 - **Isolated pauci-immune pulmonary capillaritis**