

Pulmonary veno-occlusive disease

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Introduction

- A clinicopathologic syndrome
- A subgroup of pulmonary arterial hypertension

Eur Respir J 2009; 33: 189–200.



Epidermiology

- True incidence is unknown
- Estimate 0.1-0.2 case per million person in general population

Am J Respir Crit Care Med 2000; 162: 1964–1973.

- Age range from 9 days old neonate to 67 years old man
- In pediatric population both sexes are equally affected

Chest 2000; 118: 1671–1679.



Etiology

- Genetic risk factor has been suggested by several reports of PVOD in siblings
- BMPR2 gene mutation have been report in some PVOD case

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- Infection : *Toxoplasma gondii* and measles has been documented around the time when PVOD was diagnosed
- Feature suggestive of EBV & CMV around the time of diagnosis

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Etiology

- Toxic exposure
 - a case report of 2 years history of ingesting and sniffing powdered cleaning product
 - Association between some cases of PVOD and treatment of tumor (Bleomycin, mitomycin, carmustine)
- Thrombotic Diathesis
 - Several reports increase platelet adhesiveness in PVOD

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