ICD 10 Diagnosis Codes for Bronchiolitis Obliterans Syndrome (BOS)

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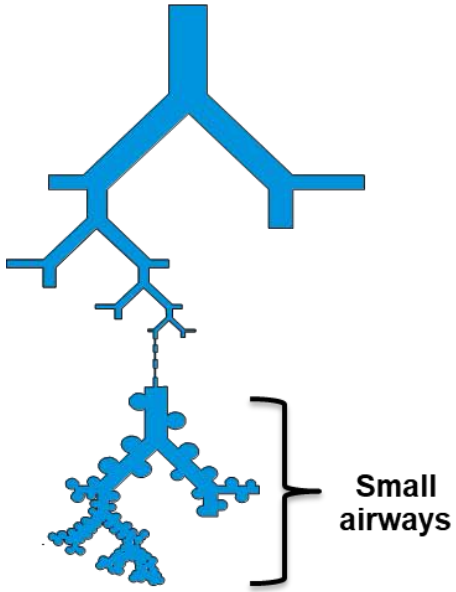
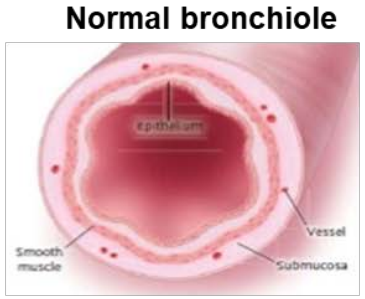
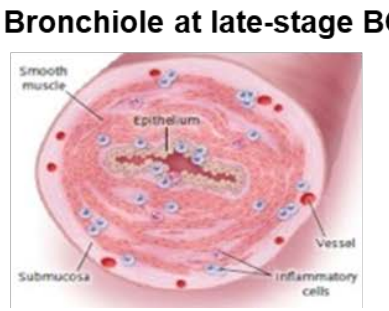
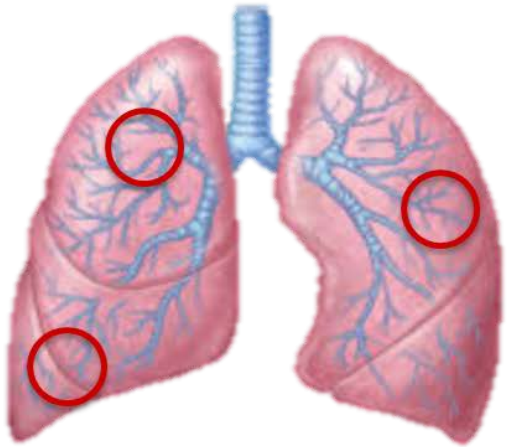
Bronchiolitis Obliterans Syndrome

- Bronchiolitis obliterans syndrome (BOS) is a clinical syndrome characterized by airflow limitation not reversible with inhaled bronchodilators which may be associated with progressive dyspnea.
- It was first clearly described in early 1980s in the context of lung transplant as a rare fibrotic disorder involving terminal and respiratory bronchioles.[1,2]
- The histologic hallmark of BOS is obliterative bronchiolitis (OB), which consists of a fibrotic luminal obliteration of the respiratory and terminal bronchioles.
- BOS is classified as a rare disease.[3]





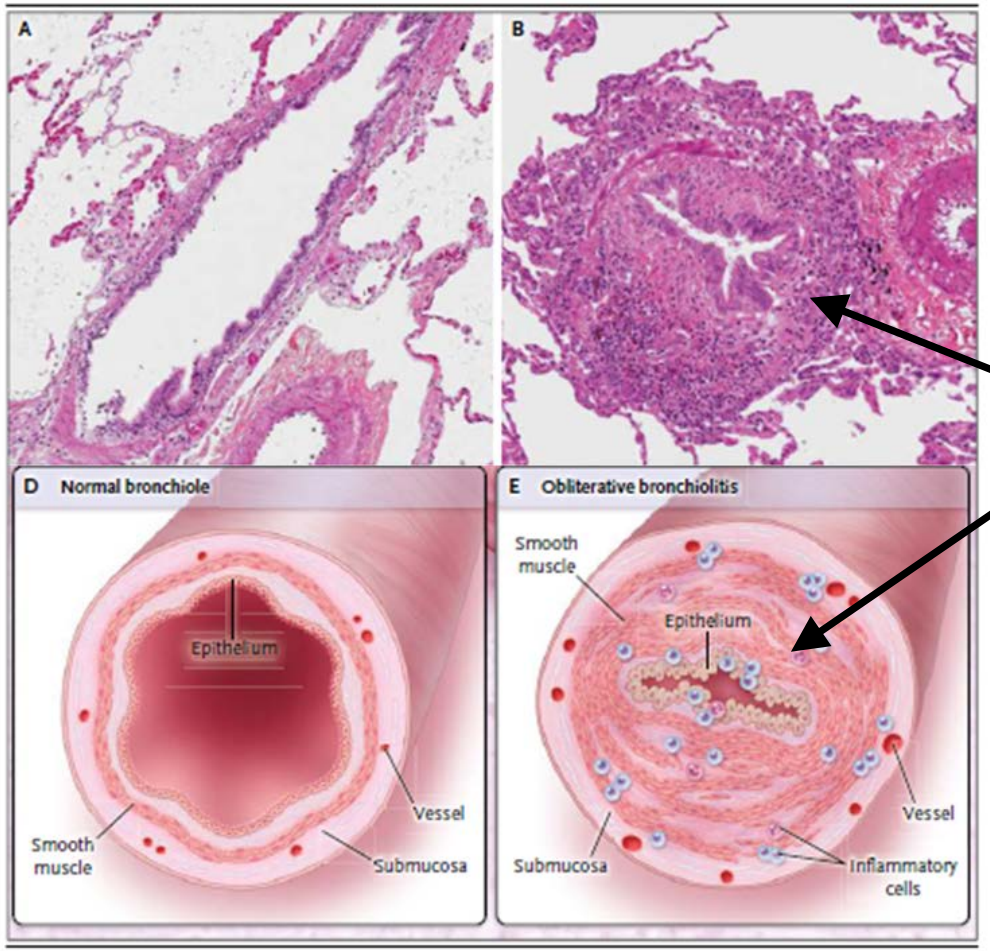




Reference(s):
[1]. Seminars in Respiratory and Critical Care Medicine. 33 (5): 509–32; [2]. Pathology of Solid Organ Transplantation. doi.org/10.1007/978-3-540-79343-4_7; [3]. Orphanet ORPHA: 1303

Pathology of Bronchiolitis Obliterans

Compared to normal lung bronchiole, the airway lumen is narrowed in BOS



Lung biopsy specimens of a normal bronchiole (left) and a bronchiole from a patient with obliterative bronchiolitis (right)

Thickened bronchiolar wall by inflammatory fibrosis located between epithelium and smooth muscle

Cross section of a normal bronchiole (left) and a bronchiole affected by obliterative bronchiolitis (right)

BOS as a manifestation of CLAD after lung transplant

- BOS is associated with injury of small airways due to infections, systemic and autoimmune diseases, and certain inhaled agents, but appears most frequently after:
  - Lung transplantation as a manifestation of chronic lung allograft dysfunction (CLAD), or
  - Allogenic hematopoietic stem cell transplantation (alloHSCT) as the pulmonary manifestation of chronic graft-versus-host-disease (cGVHD).
- BOS is the most common manifestation (or phenotype) of CLAD, accounting for nearly 50% to 70% of the cases. Up to 30% of patients with CLAD develop a restrictive defect called restrictive allograft syndrome (RAS).[4]
- The pulmonary council of the International Society for Heart and Lung Transplantation (ISHLT) classifies CLAD into four clinical sub-types—BOS, RAS, mixed and unknown.[4]

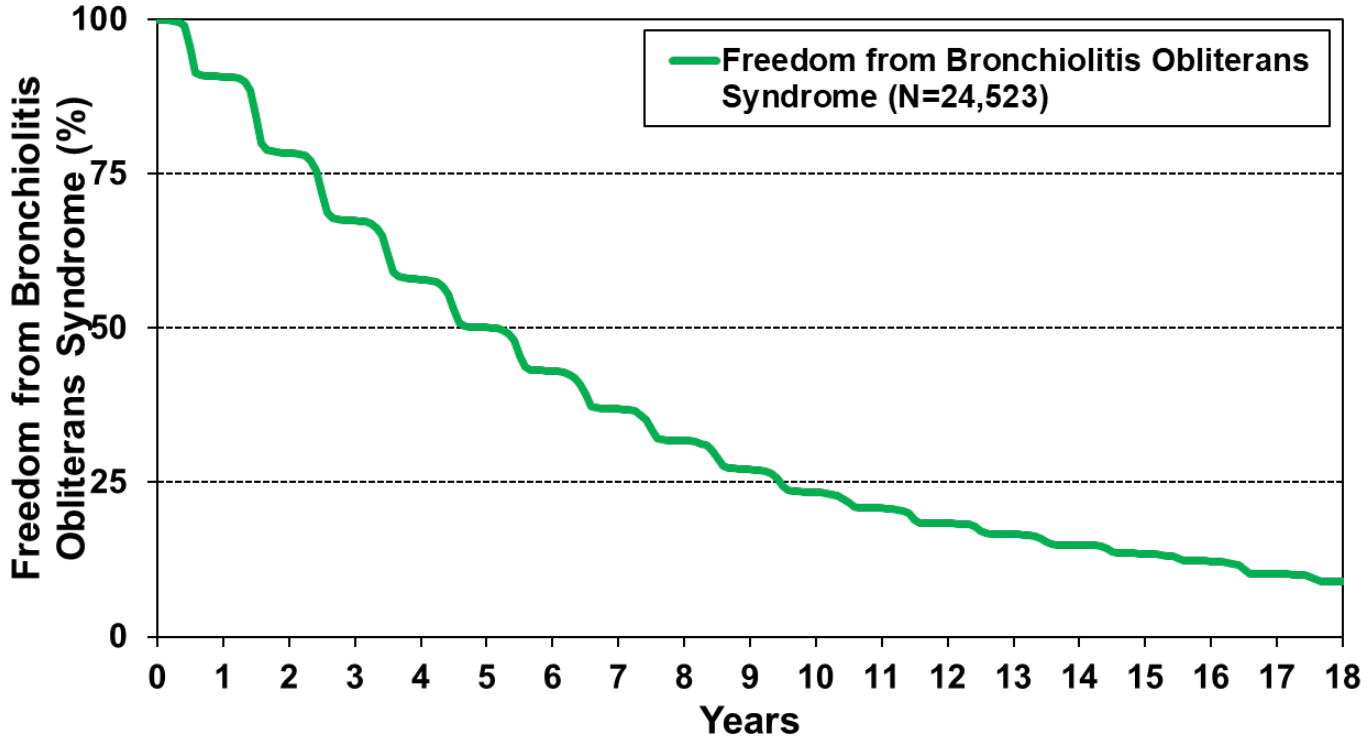
Classification of CLAD after lung transplant	
Bronchiolitis Obliterans Syndrome (BOS)	Most common manifestation (or phenotype) of CLAD, accounting for nearly 50% to 70% of the cases
Restrictive Allograft Syndrome (RAS)	~30% of patients with CLAD develop a restrictive defect
Mixed	BOS with other measures of chronic lung allograft dysfunction
Unknown	BOS due to environmental, chemical exposure and other reasons

Reference(s):
[4]. J Heart Lung Transplant. 2019 May;38(5):493-503

Incidence of BOS after lung transplant

Within first 4.5 years after lung transplant, half of the patients develop BOS

(Transplants: January 1994 – June 2016)[5]



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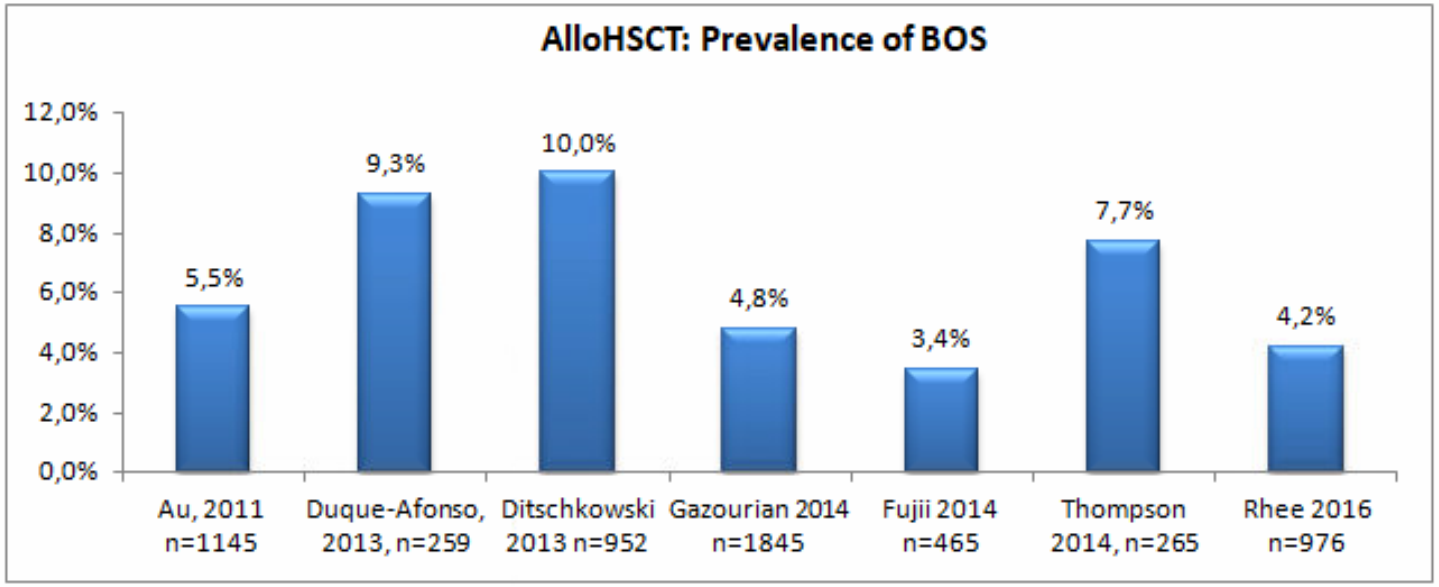
Reference(s):
[5]. JHLT. 2018; 37:1155-1206

BOS as a manifestation of cGVHD after allogenic stem cell transplant

- In alloHSCT patients, BOS occurs as a pulmonary manifestation of cGVHD.
- In alloHSCT patients afflicted by cGVHD, BOS is an important contributor to morbidity and mortality. It is associated with poor prognosis with a 5-year survival rate of 60%.[7]
- It is the most common non-infectious pulmonary complication of alloHSCT, typically presenting after the first 100 days following transplantation, with most cases presenting between 12 to 18 months after transplantation.[8]
- The exact incidence and prevalence of BOS after alloHSCT are not known but individual studies have reported that the prevalence of BOS in alloHSCT patients ranges from 3.4% to 10%.[9]



Reference(s):
[7]. Bone Marrow Transplant. 2019 Mar;54(3):383-392; [8]. Bone Marrow Transplant. 2013 Sep;48(9):1224-9 [9]. Blood 2019; 134 (Supplement_1): 5678.

Challenges due to absence of an ICD code specific to BOS

- Currently, there is no specific code in the ICD-10 system for CLAD and its phenotypes (BOS, RAS, mixed and undefined) following lung transplant.

- Similarly, there are no codes available for BOS and other pulmonary manifestations of cGVHD after alloHSCT.

- Because BOS after these procedures requires frequent healthcare encounters, therapeutic interventions and regular monitoring, a coding strategy that helps to accurately identify BOS after lung transplant and alloHSCT could help in
  - accurate measurement of the quality, safety and efficacy of care in these two separate patient populations
  - conducting more effective research, epidemiological studies, and clinical trials
  - effective monitoring of resource utilization in these patients

Support for ICD 10 code application from an international panel of clinical experts

PRECISIONheor

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Thank you!