

# ICD 10 Diagnosis Codes for Bronchiolitis Obliterans Syndrome (BOS)

---

**Dr. Ajay Sheshadri, MD, MSCI**

Assistant Professor, Department of Pulmonary Medicine, Division of Internal Medicine,  
The University of Texas MD Anderson Cancer Center

September, 2022



# Agenda: Proposal for ICD-10-CM code expansion

- Clinical background: Bronchiolitis Obliterans and Bronchiolitis Obliterans Syndrome

## Why are we looking for a ICD-10-CM code expansion for BOS after lung transplant and alloHSCT?

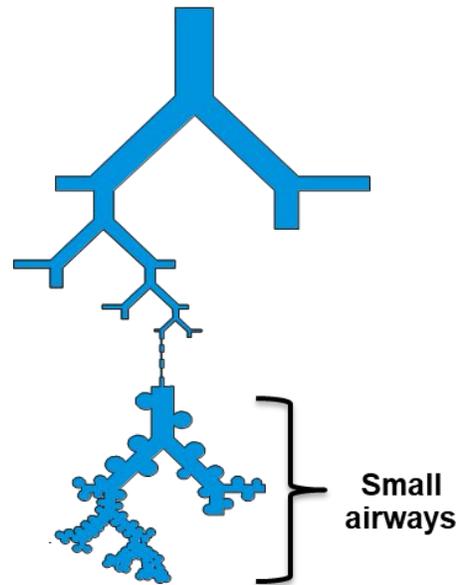
- accurate measurement of the quality, safety and efficacy of care in these two separate patient populations
- conducting more effective research, epidemiological studies, and clinical trials
- effective monitoring of resource utilization in these patients



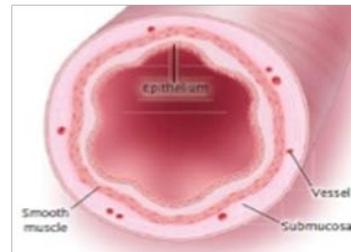
Clinical background:  
Bronchiolitis Obliterans and  
Bronchiolitis Obliterans Syndrome

# Bronchiolitis Obliterans Syndrome (BOS)

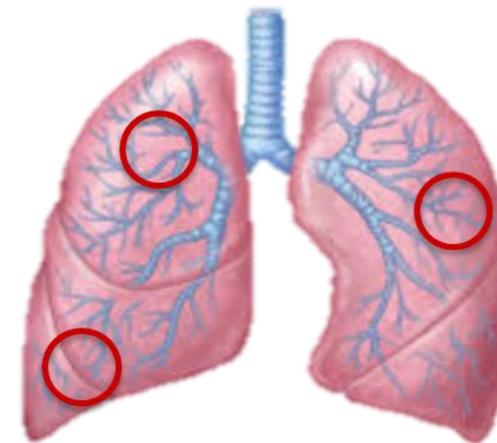
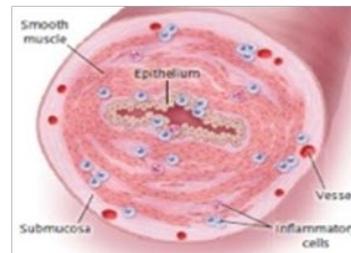
- Bronchiolitis obliterans syndrome (BOS) is a clinical syndrome characterized by airflow limitation not reversible with inhaled bronchodilators which may be associated with progressive dyspnea.
- It was first clearly described in early 1980s in the context of lung transplant as a rare fibrotic disorder involving terminal and respiratory bronchioles.<sup>1,2</sup>
- 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.<sup>3</sup>



Normal bronchiole



Bronchiole at late-stage BO



Bronchioles with BO, a patchy infestation

**BOS occurs after lung transplant or allogeneic stem cell transplant**

After lung transplant as a manifestation of CLAD

After allogeneic stem cell transplant as a manifestation of cGVHD

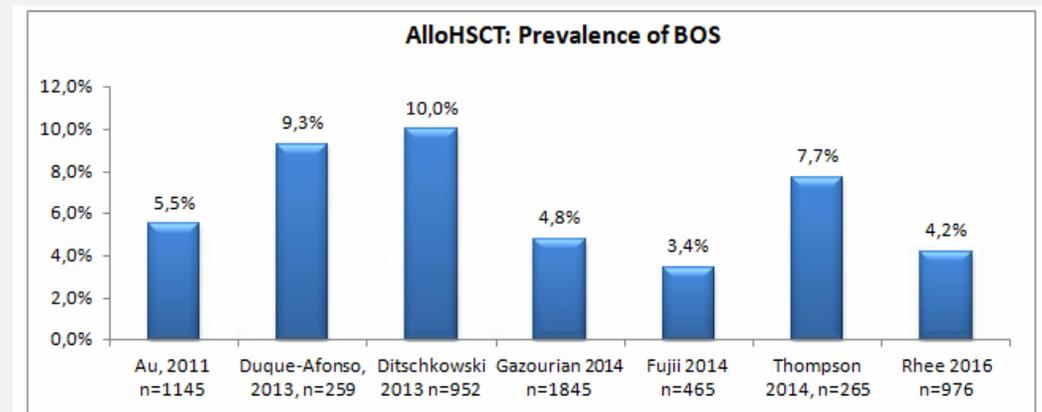
# BOS as a manifestation of CLAD after lung transplant

- 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
  - Allogeneic 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).<sup>4</sup>
- 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.<sup>4</sup>

Classification of CLAD after lung transplant	Description
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

# BOS as a manifestation of cGVHD after allogeneic 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%.<sup>7</sup>
- 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.<sup>8</sup>
- 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%.<sup>9</sup>



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.

Reference(s):

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

# Challenges due to absence of an ICD-10-CM code specific to BOS

Currently, there is no specific code in the ICD-10-CM system for CLAD and its phenotypes (BOS, RAS, mixed and undefined) following lung transplant or for BOS and other pulmonary manifestations of cGVHD after alloHSCT.

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 would 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-CM code application from an international panel of clinical experts

KOL	Specialty	Affiliation (location)	Country
Adriana Balduzzi	Allo-HSCT (pediatric hematology)	University of Milan-Bicocca, San Gerardo Hospital	Italy
Ajay Sheshadri	Pulmonary medicine	MD Anderson Cancer Center	USA
Alan Glanville	Lung transplant	University of New South Wales	Australia
Aldo Iacono	Pulmonary medicine, Lung Tx	University of Maryland	USA
Anne Bergeron	Pulmonary medicine	Hôpital Saint-Louis, Service de Pneumologie	France
Claus Neurohr	Pulmonary medicine, Lung Tx	Department for Pneumology and Respiratory Medicine, Robert Bosch Hospital	Germany
Daniel Wolff	Hematology, HSCT	University of Regensburg Faculty of Medicine	Germany
Fabio Ciceri	Hematology, HSCT	Bone Marrow Transplantation, IRCCS Ospedale San Raffaele	Italy

KOL	Specialty	Affiliation (location)	Country
Federica Melloni	Pulmonary medicine, Lung Tx	Department of Hematological, Pneumological and Cardiovascular Sciences, University of Pavia	Italy
Geert Verleden	Pulmonary medicine, Lung Tx	Dept. of Respiratory disease, University Hospital Gasthuisberg KU Leuven	Belgium
Howard Huang	Pulmonary medicine, Lung Tx	Houston Methodist J.C. Walter Jr. Transplant Center	USA
Jens Gottlieb	Pulmonary medicine, Lung Tx	Hannover Medical School, Center for Internal Medicine	Germany
Kirsten M Williams	Pediatric Hematology/ Oncology	Aflac Cancer and Blood Disorders Center, Children's Healthcare of Atlanta	USA
Michael Perch	Pulmonary medicine, Lung Tx	Dept. of Cardiology, section for Lung transplantation, Copenhagen Uni Hospital	Denmark
Nikolaus Kneidinger	Pulmonary medicine, Lung Tx	Department of Internal Medicine V, Comprehensive Pneumology Center; University of Munich	Germany

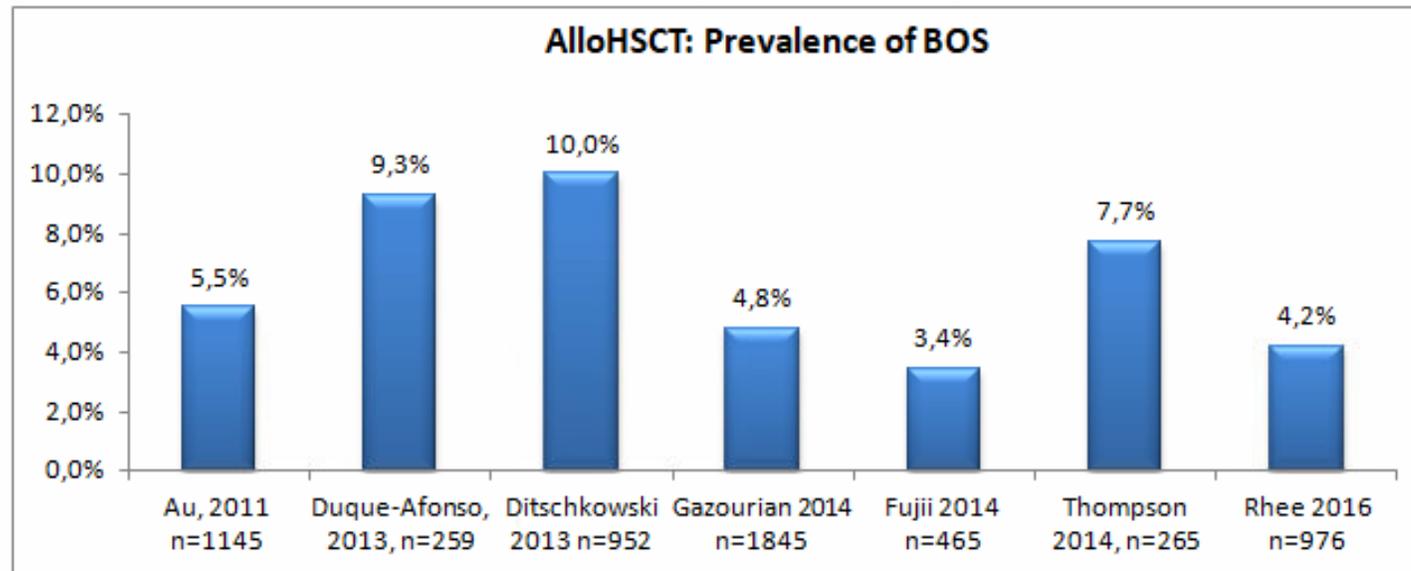
Thank you!



Appendix

# BOS as a manifestation of cGVHD after allogeneic 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%.<sup>7</sup>
- 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.<sup>8</sup>
- 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%.<sup>9</sup>



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.