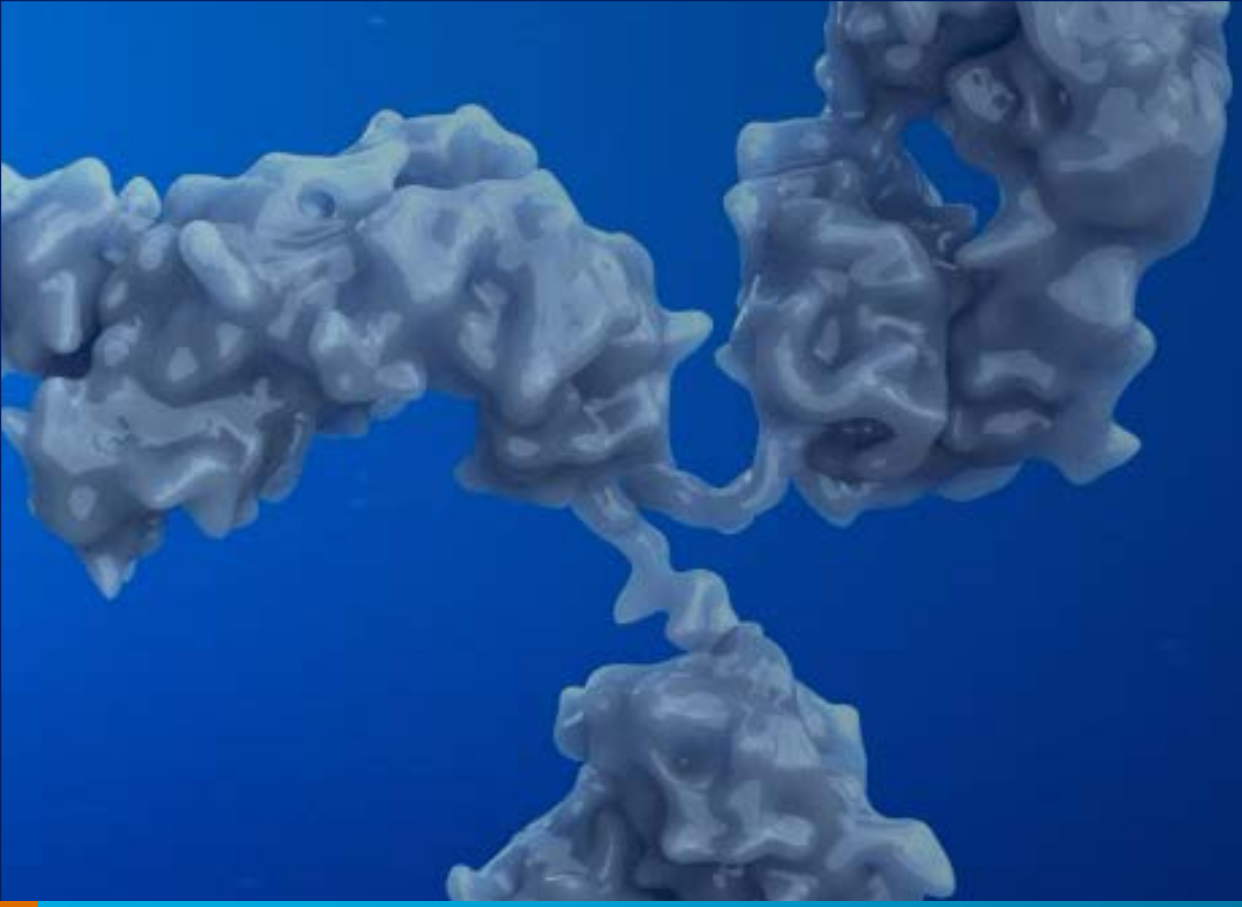

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Hematopoietic Stem Cell Transplant-Associated Thrombotic Microangiopathy (HSCT-TMA/TA-TMA)

Center for Medicare & Medicaid Services
ICD-10 Coordination and Maintenance Committee Meeting
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Request for an ICD-10-CM code for HSCT-TMA

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- HSCT-TMA (also called TA-TMA) is a life-threatening condition that occurs post-HSCT
- HSCT treatment protocols and associated complications, survival rates, and quality of care are rigorously monitored by transplant centers, clinical societies, and governmental agencies
  - Stem Cell Therapeutic and Research Act of 2005
- Absence of a specific ICD-10-CM code for HSCT-TMA has led to underreporting of the diagnosis, inaccurate coding, and challenges to monitoring the quality of care and outcomes associated with HSCT-TMA
- A specific ICD-10-CM code will assist centers with identifying and treating HSCT-TMA, enhancing patient care and potentially saving patient's lives

Incidence: Severe HSCT-TMA

HSCT-TMA is a serious multisystem and life-threatening complication of HSCT presenting with signs of microangiopathic hemolytic anemia, consumptive thrombocytopenia in the absence of coagulopathy, and microvascular thrombosis with end-organ damage that is associated with highly unfavorable post-HSCT survival.

of patients with HSCT-TMA display at least one **high-risk** feature

50%

of HSCT-TMA cases may be **severe**

of severe cases of HSCT-TMA can be **fatal**

HSCT-TMA is a Distinct Diagnosis

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- HSCT expert clinicians and researchers, and the FDA, identify HSCT-TMA as a specific clinical diagnosis
- A national clinical working group, with international representation, is conducting a retrospective study to create consensus diagnostic criteria for HSCT-TMA
- Other complications post-HSCT have specific ICD-10-CM codes
  - Veno-occlusive Disease: K76.5
  - Acute Graft-versus-Host Disease: D89.810
  - Chronic Graft-versus–Host Disease: D89.811
- HSCT-TMA deserves an ICD-10 code to allow for the accurate tracking of this life-threatening complication and associated treatment, survival and long-term outcomes

Current Status of HSCT-TMA

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- The current standard of care involves removal of immunosuppressive therapies, but recent evidence suggests this does not meaningfully improve outcomes for these patients
- In those who survive, HSCT-TMA may be associated with long-term morbidity and chronic organ injury
- The FDA is currently monitoring several therapies under study for the specific indication of HSCT-TMA

Example of HSCT Complications

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	HSCT-TMA	VOD/SOS	aGVHD
	No **ICD-10-CM code**	**K76.5**	**D89.810**
	Transaminitis > indirect Bili elevated (hemolysis); **no coagulopathy**	Direct Bili elevated (cholestasis) > **coagulopathy** > transaminitis	Transaminitis > Direct Bili elevated (cholestasis) without coagulopathy
	Seizures with hypertension/PRES	Encephalopathy due to liver failure, may have elevated ammonia	No primary CNS injury
	AKI, **nephrotic proteinuria**, renal failure (RRT), severe **hypertension**	Secondary AKI, **hypotension** due to capillary leak, RRT for fluid removal	No primary renal injury
	Right heart failure due to pulmonary hypertension or pericardial effusion	Serositis due to third spacing due to liver failure	No primary cardiac injury
	Pulmonary hypertension, interstitial pulmonary bleeding leading to ARDS (often clear lungs at presentation)	Respiratory failure is secondary to **fluid overload** and third spacing ("wet" lungs at presentation)	Acute presentation is not common. Evolving hypoxemia, abnormal PFTs, bronchiolitis obliterans in chronic phase
	Intestinal bleeding due to ischemic colitis, severe pain. Histology shows vascular endothelial injury	No primary bowel injury. Possible abdominal compartment syndrome due to ascites	Intestinal bleeding in advanced cases. Histology shows epithelial injury. Need to eval for TMA
	Intravascular hemolysis (free plasma Hb), thrombocytopenia due to vascular microthrombi formation	Anemia (extravascular hemolysis if any), thrombocytopenia due to organomegaly/consumption	Not common with acute GVHD

Used with permission from Sonata Jodele, MD; ICD-10-CM codes.

Need for specific HSCT-TMA ICD-10-CM code

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- A specific ICD-10-CM code would:
  - Provide accurate coding as to the specific disease entity
  - Allow accurate tracking of disease incidence and treatment
    - The reported incidence of HSCT-TMA in the medical literature ranges from 0.5% to 76%; recent studies show an incidence of approximately 40%
  - Support observational research
  - Provide accurate data on diagnosis and associated drug utilization
  - Help provide access to treatment for patients and support tracking and understanding of the needs for such treatment

Summary

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- Identification of a specific ICD-10-CM code for HSCT-TMA will assist healthcare clinicians to code accurately for this disease separate from other conditions that occur during and after HSCT that require different treatments
- Improved coding granularity will increase healthcare provider awareness of the disease and allow nationwide monitoring based on real-world evidence to enable earlier and more accurate diagnosis as well as the potential to improve the treatment of HSCT-TMA
- Addition of a specific ICD-10-CM code for HSCT-TMA is also consistent with the goals set out by the CDC for the development of the ICD-10 coding system
- Without a unique ICD-10-CM code, there is no ability to accurately characterize and report on the true incidence, morbidity and mortality associated with HSCT-TMA