[7] Adapted from Durie BG, Salmon SE: A clinical staging system for multiple myeloma: Correlation of measured myeloma cell mass with presenting clinical features, response to treatment, and survival. *Cancer*. 1975;36:842-54.

Questions 594-596: Stage at Diagnosis: I.S.S.

> ! Currently there is an issue on Form 2400 regarding the ISS Staging. Stage I requires albumin greater or equal to 3.5 g/dL.

Report the recipient's lab values from diagnosis and the ISS stage of myeloma.

I.S.S. Staging System for Multiple Myeloma[8]

Stage	Description
Stage I	Serum β2-microglobulin < 3.5 mg/L and serum albumin ≥ 3.5 g/dL
Stage II	Serum β2-microglobulin < 3.5 mg/L and serum albumin < 3.5 g/dL OR Serum β2-microglobulin 3.5 to <5.5 mg/dL irrespective of serum albumin level
Stage III	Serum β2-microglobulin ≥ 5.5 mg/L irrespective of serum albumin level

[8] Greipp, P. R., San Miguel, J., Durie, B. G., Crowley, J. J., Barlogie, B., Bladé, J., … & Westin, J. (2005). International staging system for multiple myeloma. *Journal of Clinical Oncology*, 23(15), 3412-3420.

Question 597: Were cytogenetics tested (conventional or FISH)?

Cytogenetics is the study of chromosomes. Cytogenetic assessment involves testing blood or bone marrow for the presence of a known chromosomal abnormality that reflects the recipient's disease. Testing methods you may see include conventional chromosome analysis (karyotyping) or fluorescence *in situ* hybridization (FISH). For more information about cytogenetic testing and terminology, see Appendix R, Cytogenetic Abbreviations and Terminology.

Indicate if cytogenetic studies were obtained at any time prior to the start of the preparative regimen. If cytogenetic studies were obtained, select "yes" and continue with question 598.

If no cytogenetic studies were obtained or if it is unknown if chromosome studies were performed, select "no" or "unknown" and continue with question 619.

Question 598: Results of test:

If cytogenetic studies identified abnormalities, indicate “abnormalities identified” and continue with question 599.

If cytogenetic studies yielded “no evaluable metaphases” or there were “no abnormalities” identified, continue with question 619.

Questions 599-618: Specify abnormalities identified at any time prior to the start of the preparative regimen:

Report all abnormalities identified by all methods of cytogenetic assessment at any time prior to the start of the preparative regimen by selecting “yes” or “no” for each question. Do not leave any response blank. If one or more abnormalities are best classified as “other abnormality” select “yes” for question 617 and specify the abnormality in question 618.

Question 619: What was the disease status?

Indicate the disease status of the PCD at the last evaluation prior to the start of the preparative regimen. **See Multiple Myeloma Response Criteria for disease status definitions.**

> ! Currently there is an issue on Form 2400 regarding the number of plasma cells required for CR. CR requires less than (but not equal to) 5 % plasma cells in the bone marrow.

At any response level, if some but not all criteria are met, the disease status should be downgraded to next lower level of response.

The percentage of plasma cells in the bone marrow aspirate and/or biopsy may also be identified on a flow cytometry report. A flow cytometry report may **NOT** be used to confirm CR (e.g., < 5% plasma cells in the bone marrow).

For more information on determining how to report disease status prior to the preparative regimen, see Appendix V.

If the disease response prior to transplant is unknown, select “unknown” and continue with the signature line.

If the recipient had amyloidosis or POEMS syndrome, but no evidence of myeloma, select “not applicable” and continue with the signature line.

Example 1: A 62-year-old man is diagnosed with IgG Kappa multiple myeloma. He receives initial therapy with 6 cycles of bortezomib and lenalidomide/dexamethasone; and achieves a near complete remission (nCR). The values used to determine disease status at transplant are the values obtained at diagnosis.

Time Point	BMBX	SPEP	SIFE	UPEP	UIFE	Skeletal Survey	Treatment	Disease Status
10/31/08	27% plasma cells	3.3 g/dL	+	336 mg/24 hours	+	Negative	Bortezomib/ Lenalidomide/ Dexamethasone	Diagnosis: IgG Kappa
4/3/09	3% plasma cells							
4/17/09		Negative	+	Negative	Negative			nCR
5/13/09		Negative	+	Negative	Negative			nCR (confirmatory)
5/17/09							Autologous HCT	

Example 2: A 59-year-old woman is diagnosed with IgA Lambda multiple myeloma. She receives bortezomib and thalidomide/dexamethasone as initial treatment and achieves a CR. A few months later she has evidence of relapse. She is then treated with lenalidomide/dexamethasone and achieves a PR. The patient receives high-dose cyclophosphamide as part of an autologous stem cell harvest. The values used to determine disease status at transplant would be the values obtained at the time of relapse.

Time Point	BMBX	SPEP	SIFE	UPEP	UIFE	Skeletal Survey	Treatment	Disease Status
1/27/10		4.5 g/dL	+	Negative	Negative			
2/01/10	Aspirate=18% plasma cells; biopsy= sheets of plasma cells							Diagnosis: IgA lambda
2/05/10						Negative	Bortezomib/ Thalidomide/ Dexamethasone	
3/05/10		2.6 g/dL	+					
4/5/10		1.7 g/dL	+					

5/5/10		0.5 g/dL	+					
6/4/10		0.03 g/dL	+	Negative	Negative			
8/18/10	1% plasma cells	0.01 g/dl	+					
9/15/10		Not detected	+					
10/15/10		Not detected	Negative					CR
11/15/10		Not detected	Negative				(no treatment given)	CR (confirmatory)
12/15/11		Not detected	Negative					
1/15/11		1.9 g/dL	+	Negative	Negative			Relapse
2/15/11	7% plasma cells	2.2 g/dL	+			Negative	Lenalidomide/ Dexamethasone	Relapse (confirmatory)
3/15/11		1.4 g/dL	+					
4/15/11		0.9 g/dL	+					PR
5/15/11		0.7 g/dL	+					PR (confirmatory)
6/15/11	3% plasma cells	0.5 g/dL	+					
7/31/11							Autologous HCT	

Question 620: Date Assessed:

Enter the date of the most recent assessment of disease status prior to the start of the preparative regimen. Report the date the blood/urine was collected for the laboratory evaluations (e.g., SPEP/UPEP, serum/urine immunofixation) or report the date the bone marrow was collected for pathological evaluation. A PET scan may be used if a previous PET scan had been obtained and only in limited circumstances (e.g., plasmacytomas, lytic lesions).

If the exact date is not known, use the process for reporting partial or unknown dates as described in General Instructions, Guidelines for Completing Forms.

Q621-622: Solid Tumors

Questions 621-622: Specify the solid tumor classification:

Indicate the solid tumor disease classification at the time of diagnosis. Germ cell tumors that originate in the ovary or testes should be reported as *ovarian* or *testicular*, respectively. If the subtype is not listed, report as "Other solid tumor" and specify the reported malignancy in question 622. If a certain disease becomes a common indication for HCT, the CIBMTR will add the disease as a separate category.

Q623-624: Severe Aplastic Anemia

Questions 623-624: Specify the severe aplastic anemia classification:

Indicate the severe aplastic anemia disease classification at diagnosis. If the subtype is not listed, report as “other acquired cytopenic syndrome” and specify the reported disease. If a certain disease becomes a common indication for HCT, the CIBMTR will add the disease as a separate category.

Q625-627: Inherited Abnormalities of Erythrocyte Differentiation or Function

Questions 625-627: Specify the inherited abnormalities of erythrocyte differentiation or function classification

Indicate the inherited abnormalities of erythrocyte differentiation or function disease classification at diagnosis. If the subtype is not listed, report as “other constitutional anemia” or “other hemoglobinopathy” and specify the reported disease. If a certain disease becomes a common indication for HCT, the CIBMTR will add the disease as a separate category.

Q628-630: Disorders of the Immune System

Questions 628-630: Specify disorder of immune system classification:

Indicate the disorder of the immune system's disease classification at diagnosis. If the subtype is not listed, report as "other SCID" or "other immunodeficiency" and specify the reported disease. If a certain disease becomes a common indication for HCT, the CIBMTR will add the disease as a separate category.

Q631-632: Inherited Abnormalities of Platelets

Questions 631-632: Specify inherited abnormalities of platelets classification:

Indicate the inherited abnormalities of platelets disease classification at diagnosis. If the subtype is not listed, report as “other inherited platelet abnormality” and specify the reported disease. If a certain disease becomes a common indication for HCT, the CIBMTR will add the disease as a separate category.

Q633-634: Inherited Abnormalities of Metabolism

Questions 633-634: Specify inherited abnormalities of metabolism classification:

Indicate the inherited abnormalities of metabolism disease classification at diagnosis. If the subtype is not listed, report as “inherited metabolic disorder, not otherwise specified” and specify the reported disease. If a certain disease becomes a common indication for HCT, the CIBMTR will add the disease as a separate category.

Q635-636: Histiocytic Disorders

Questions 635-636: Specify the histiocytic disorder classification:

Indicate the histiocytic disorder disease classification at diagnosis. If the subtype is not listed, report as “other histiocytic disorder” and specify the reported disease in question 636. If a certain disease becomes a common indication for HCT, the CIBMTR will add the disease as a separate category.

Q637-644: Autoimmune Diseases

Questions 637-644: Specify autoimmune disease classification:

Indicate the autoimmune disease classification at diagnosis. If the subtype is not listed, report as “other arthritis,” “other connective tissue disease,” “other vasculitis,” “other autoimmune neurological disorder,” “other autoimmune cytopenia,” or “other autoimmune bowl disorder,” and specify the reported disease. If a certain disease becomes a common indication for HCT, the CIBMTR will add the disease as a separate category.

Q645: Other Disease

Question 645: Specify other disease:

Before using this category, check with a transplant physician to determine whether the disease can be classified as one of the listed options in the Disease Classification questions. Examples include: erythropoietic protoporphyria (EPP), and dystrophic epidermolysis bullosa (DEB).