Appendix Z: Primary Disease and Disease Inserts Due

E-mail comments regarding the content of the CIBMTR Forms Instruction Manual to: CIBMTRFormsManualComments@nmdp.org. Comments will be considered for future manual updates and revisions. For questions that require an immediate response, please contact your transplant center's CIBMTR CRC.

Primary Disease and Disease Inserts Due

This appendix is useful for determining the disease inserts that should be completed for the disease selected on a recipient's Pre-TED form. The disease inserts should appear in the recipient's forms list in FormsNet for those on the comprehensive reporting track after the Pre-TED is submitted. No disease inserts are due for those on the TED reporting track.

Acute Myelogenous Leukemia (AML or ANLL)

Primary Disease	Disease Subtype(s)	Disease Inserts
Acute myelogenous leukemia (AML or ANLL)	☐ AML with t(9;11) (p22;q23); MLLT 3-MLL (5) ☐ AML with t(6;9) (p23;q24); DEK-NUP214 (6) ☐ AML with inv(3) (q21;q26.2) or t(3;3) (q21;q26.2); RPN1-EVI1 (7) ☐ AML (megakaryoblastic) with t(1;22) (p13;q13); RBM15-MKL1 (8) ☐ AML with t(8;21); (q22; q22); RUNX1/RUNX1T1 (281) ☐ AML with inv(16); (p13;1q22) or t(16;16) (p13.1; q22); CBFB/MYH11 (282) ☐ APL with t(15;17); (q22;q12); RARA;PML (283) ☐ AML with 11q23 (MLL) abnormalities (i.e., t(4;11), t(6;11), t(9;11), t(11;19)) (284) ☐ AML with myelodysplasia – related changes (285) ☐ Therapy related AML (t-AML) (9) ☐ Myeloid sarcoma (295) ☐ Blastic plasmacytoid dendritic cell neoplasm (296) ☐ AML or ANLL, not otherwise specified (280) ☐ AML, minimally differentiated (M0) (286) ☐ AML without maturation (M1) (287) ☐ AML with maturation (M2) (288) ☐ Acute myelomonocytic leukemia (M4) (289) ☐ Acute monoblastic / acute monocytic leukemia (M5) (290) ☐ Acute erythroid leukemia (erythroid / myeloid and pure erythroleukemia) (M6) (291) ☐ Acute megakaryoblastic leukemia (M7) (292) ☐ Acute basophilic leukemia (293) ☐ Acute panmyelosis with myelofibrosis (294)	Form 2010 & 2110

Acute Lymphoblastic Leukemia (ALL)

Primary Disease	Disease Subtype(s)	Disease Inserts
Acute lymphoblastic leukemia (ALL)	☐ t(9;22)(q34;q11); BCR/ABL1 (192) ☐ t(v;11q23); MLL rearranged (193) ☐ t(1;19)(q23;p13) TCF3-PBX1 (194) ☐ t(12;21) (p12;q22); TEL-AML1 (195) ☐ t(5;14) (q31;q32); IL3-IGH (81) ☐ Hyperdiploidy (51-65 chromosomes) (82) ☐ Hypodiploidy (<45 chromosomes) (83) ☐ B-cell ALL, NOS {L1/L2} (191) ☐ T-cell lymphoblastic leukemia / lymphoma (Precursor T-cell ALL) (196) ☐ ALL, NOS (190)	Form 2011 & 2111

Other Acute Leukemia

Primary Disease	Disease Subtype(s)	Disease Inserts
Other Acute Leukemia	☐ Acute undifferentiated leukemia (31) ☐ Biphenotypic, bilineage or hybrid leukemia (32) ☐ Acute mast cell leukemia (33) ☐ Other acute leukemia (89)	Form 2010 & 2110

Chronic Myelogenous Leukemia (CML)

Primary Disease	Disease Subtype(s)	Disease Inserts
Chronic myelogenous leukemia (CML)	☐ Ph+ / bcr+ (41) ☐ Ph+ / bcr- (42) ☐ Ph+ / bcr unknown (43) ☐ Ph- / bcr+ (44) ☐ Ph unknown / bcr+ (47)	Form 2012 & 2112

Myelodysplastic (MDS) / Myeloproliferative (MPN) Diseases

Primary Disease	Disease Subtype(s)	Disease Inserts
Myelodysplastic (MDS) / myeloproliferative (MPN) diseases	**MDS** ☐ Refractory cytopenia with unilineage dysplasia (RCUD) (includes refractory anemia (RA)) (51) ☐ Refractory anemia with ringed sideroblasts (RARS) (55) ☐ Refractory anemia with excess blasts-1 (RAEB-1) (61) ☐ Refractory anemia with excess blasts-2 (RAEB-2) (62) ☐ Refractory cytopenia with multilineage dysplasia (RCMD) (64) ☐ Childhood myelodysplastic syndrome (Refractory cytopenia of childhood (RCC)) (68) ☐ Myelodysplastic syndrome with isolated del(5q) (5q– syndrome) (66) ☐ Myelodysplastic syndrome (MDS), unclassifiable (50) **MPN** ☐ Chronic neutrophilic leukemia (165) ☐ Chronic eosinophilic leukemia, NOS (166) ☐ Essential thrombocythemia (includes primary thrombocytosis, idiopathic thrombocytosis, hemorrhagic thrombocythemia) (58) ☐ Polycythemia vera (PCV) (57) ☐ Primary myelofibrosis (includes chronic idiopathic myelofibrosis (CIMF), agnogenic myeloid metaplasia (AMM), myelofibrosis/sclerosis with myeloid metaplasia (MMM), idiopathic myelofibrosis) (167) ☐ Myeloproliferative neoplasm (MPN), unclassifiable (60) **MDS / MPN** ☐ Chronic myelomonocytic leukemia (CMMoL) (54) ☐ Myelodysplastic / myeloproliferative neoplasm, unclassifiable (69)	Form 2014 & 2114
	☐ Juvenile myelomonocytic leukemia (JMML/JCML) (no evidence of Ph[1] or BCR/ABL) (36)	Form 2015 & 2115
	☐ Atypical chronic myeloid leukemia, Ph-/bcr/abl- {CML, NOS} (45) ☐ Atypical chronic myeloid leukemia, Ph-/bcr unknown {CML, NOS} (46) ☐ Atypical chronic myeloid leukemia, Ph unknown/bcr- {CML, NOS} (48) ☐ Atypical chronic myeloid leukemia, Ph unknown/bcr unknown {CML, NOS} (49)	Form 2010 & 2110

Other Leukemia (Includes CLL)

Primary Disease	Disease Subtype(s)	Disease Inserts
Other Leukemia	☐ Chronic lymphocytic leukemia (CLL), NOS (34) ☐ Chronic lymphocytic leukemia (CLL), B-cell / small lymphocytic lymphoma (SLL) (71) ☐ Hairy cell leukemia (35) ☐ Prolymphocytic leukemia (PLL), NOS (37) ☐ PLL, B-cell (73) ☐ PLL, T-cell (74)	Form 2013 & 2113
	☐ Other leukemia, NOS (30) ☐ Other leukemia (39)	Form 2010 & 2110

Hodgkin Lymphoma

Primary Disease	Disease Subtype(s)	Disease Inserts
Hodgkin Lymphoma	☐ Nodular lymphocyte predominant Hodgkin lymphoma (155) ☐ Lymphocyte-rich (151) ☐ Nodular sclerosis (152) ☐ Mixed cellularity (153) ☐ Lymphocyte depleted (154) ☐ Hodgkin lymphoma, NOS (150)	Form 2018 & 2118

Non-Hodgkin Lymphoma

Primary Disease	Disease Subtype(s)	Disease Inserts
Non-Hodgkin Lymphoma	**B-cell neoplasms:** ☐ Splenic marginal zone B-cell lymphoma (124) ☐ Extranodal marginal zone B-cell lymphoma of mucosal associated lymphoid tissue type (MALT) (122) ☐ Nodal marginal zone B-cell lymphoma (± monocytoid B-cells) (123) ☐ Follicular, predominantly small cleaved cell (Grade I follicle center lymphoma) (102) ☐ Follicular, mixed, small cleaved and large cell (Grade II follicle center lymphoma) (103) ☐ Follicular, predominantly large cell (Grade IIIA follicle center lymphoma) (162)	Form 2018 & 2118

Primary Disease	Disease Subtype(s)	Disease Inserts
	☐ Follicular, predominantly large cell (Grade IIIB follicle center lymphoma) (163) ☐ Follicular (grade unknown) (164) ☐ Mantle cell lymphoma (115) ☐ Intravascular large B-cell lymphoma (136) ☐ Primary mediastinal (thymic) large B-cell lymphoma (125) ☐ Primary effusion lymphoma (138) ☐ Diffuse, large B-cell lymphoma — NOS (107) ☐ Burkitt lymphoma (111) ☐ B-cell lymphoma, unclassifiable, with features intermediate between DLBCL and Burkitt lymphoma (140) ☐ B-cell lymphoma, unclassifiable, with features intermediate between DLBCL and classical Hodgkin Lymphoma (149) ☐ T-cell / histiocytic rich large B-cell lymphoma (120) ☐ Primary diffuse large B-cell lymphoma of the CNS (118) ☐ Other B-cell lymphoma (129) **T-cell / NK cell neoplasms:** ☐ Extranodal NK / T-cell lymphoma, nasal type (137) ☐ Enteropathy-type T-cell lymphoma (133) ☐ Hepatosplenic T-cell lymphoma (145) ☐ Subcutaneous panniculitis-like T-cell lymphoma (146) ☐ Mycosis fungoides (141) ☐ Sezary syndrome (142) ☐ Primary cutaneous CD30+ T-cell lymphoproliferative disorders [Primary cutaneous anaplastic large-cell lymphoma (C-ALCL), lymphoid papulosis] (147) ☐ Peripheral T-cell lymphoma (PTCL), NOS (130) ☐ Angioimmunoblastic T-cell lymphoma (131) ☐ Anaplastic large-cell lymphoma (ALCL), ALK positive (143) ☐ Anaplastic large-cell lymphoma (ALCL), ALK negative (144) ☐ T-cell large granular lymphocytic leukemia (126) ☐ Aggressive NK-cell leukemia (27) ☐ Adult T-cell lymphoma / leukemia (HTLV1 associated) (134) ☐ Other T-cell / NK-cell lymphoma (139)	
	☐ Waldenstrom macroglobulinemia / Lymphoplasmacytic lymphoma (173)	Form 2019 & 2119

Multiple Myeloma/ Plasma Cell Disorder (PCD)

Primary Disease	Disease Subtype(s)	Disease Inserts
Multiple myeloma / plasma cell disorder (PCD)	☐ Multiple myeloma-IgG (181) ☐ Multiple myeloma-IgA (182) ☐ Multiple myeloma-IgD (183) ☐ Multiple myeloma-IgE (184) ☐ Multiple myeloma-IgM (not Waldenstrom macroglobulinemia) (185) ☐ Multiple myeloma-light chain only (186) ☐ Multiple myeloma-non-secretory (187) ☐ Plasma cell leukemia (172) ☐ Solitary plasmacytoma (no evidence of myeloma) (175) ☐ Amyloidosis (174) ☐ Osteosclerotic myeloma / POEMS syndrome (176) ☐ Light chain deposition disease (177) ☐ Other plasma cell disorder (179)	Form 2016 & 2116

Solid Tumors

Primary Disease	Disease Subtype(s)	Disease Inserts
Solid Tumors	☐ Breast cancer (250)	Form 2020 & 2120
	☐ Lung, small cell (202) ☐ Lung, non-small cell (203) ☐ Lung, not otherwise specified (230)	Form 2021 & 2121
	☐ Germ cell tumor, extragonadal (225) ☐ Testicular (210)	Form 2022 & 2122
	☐ Ovarian (epithelial) (214)	Form 2023 & 2123
	☐ Bone sarcoma (excluding Ewing family tumors) (273) ☐ Ewing family tumors of bone (including PNET) (275) ☐ Ewing family tumors, extraosseous (including PNET) (276) ☐ Fibrosarcoma (244) ☐ Hemangiosarcoma (246) ☐ Leiomyosarcoma (242)	Form 2024 & 2124

Primary Disease	Disease Subtype(s)	Disease Inserts
	☐ Liposarcoma (243) ☐ Lymphangio sarcoma (247) ☐ Neurogenic sarcoma (248) ☐ Rhabdomyosarcoma (232) ☐ Synovial sarcoma (245) ☐ Soft tissue sarcoma (excluding Ewing family tumors) (274)	
	☐ Central nervous system tumor, including CNS PNET (220) ☐ Medulloblastoma (226)	Form 2025 & 2125
	☐ Neuroblastoma (222)	Form 2026 & 2126
	☐ Head / neck (201) ☐ Mediastinal neoplasm (204) ☐ Colorectal (228) ☐ Gastric (229) ☐ Pancreatic (206) ☐ Hepatobiliary (207) ☐ Prostate (209) ☐ External genitalia (211) ☐ Cervical (212) ☐ Uterine (213) ☐ Vaginal (215) ☐ Melanoma (219) ☐ Wilm tumor (221) ☐ Retinoblastoma (223) ☐ Thymoma (231) ☐ Other solid tumor (269) ☐ Solid tumor, not otherwise specified (200)	No disease insert required
	☐ Renal cell (208)	Form 2027 & 2127

Severe Aplastic Anemia

Primary Disease	Disease Subtype(s)	Disease Inserts
Severe Aplastic Anemia	☐ Acquired severe aplastic anemia, not otherwise specified (301) ☐ Acquired SAA secondary to hepatitis (302) ☐ Acquired SAA secondary to toxin / other drug (303) ☐ Acquired amegakaryocytosis (not congenital) (304) ☐ Acquired pure red cell aplasia (not congenital) (306) ☐ Other acquired cytopenic syndrome (309)	Form 2028 & 2128
	☐ Dyskeratosis congenital (307)	No disease insert required

Inherited Abnormalities of Erythrocyte Differentiation or Function

Primary Disease	Disease Subtype(s)	Disease Inserts
Inherited abnormalities of erythrocyte differentiation or function	☐ Paroxysmal nocturnal hemoglobinuria (PNH) (56) ☐ Shwachman-Diamond (305) ☐ Diamond-Blackfan anemia (pure red cell aplasia) (312) ☐ Other constitutional anemia (319)	Form 2028 & 2128
	☐ Sickle thalassemia (355) ☐ Sickle cell disease (356)	Form 2030 & 2130
	☐ Beta thalassemia major (357) ☐ Other hemoglobinopathy (359)	No disease insert required
	☐ Fanconi anemia (311)	Form 2029 & 2129

Disorders of the Immune System

Primary Disease	Disease Subtype(s)	Disease Inserts
Disorders of the immune system	☐ Adenosine deaminase (ADA) deficiency / severe combined immunodefiency (SCID) (401) ☐ Absence of T and B cells SCID (402) ☐ Absence of T, normal B cell SCID (403)	Form 2031 & 2131

Primary Disease	Disease Subtype(s)	Disease Inserts
	☐ Omenn syndrome (404) ☐ Reticular dysgenesis (405) ☐ Bare lymphocyte syndrome (406) ☐ Other SCID (419) ☐ SCID, not otherwise specified (410) ☐ Ataxia telangiectasia (451) ☐ HIV infection (452) ☐ DiGeorge anomaly (454) ☐ Common variable immunodeficiency (457) ☐ Leukocyte adhesion deficiencies, including GP180, CD-18, LFA and WBC adhesion deficiencies (459) ☐ Kostmann agranulocytosis (congenital neutropenia) (460) ☐ Neutrophil actin deficiency (461) ☐ Cartilage-hair hypoplasia (462) ☐ CD40 ligand deficiency (464) ☐ Other immunodeficiencies (479) ☐ Immune deficiency, not otherwise specified (400)	
	☐ Chediak-Higashi syndrome (456) ☐ Griscelli syndrome type 2 (465) ☐ Hermansky-Pudlak syndrome type 2 (466)	Form 2056 & 2156
	☐ Chronic granulomatous disease (455)	Form 2055 & 2155
	☐ Wiskott-Aldrich syndrome (453)	Form 2033 & 2133
	☐ X-linked lymphoproliferative syndrome (458)	Form 2034 & 2134

Inherited Abnormalities of Platelets

Primary Disease	Disease Subtype(s)	Disease Inserts
Inherited abnormalities of platelets	☐ Congenital amegakaryocytosis / congenital thrombocytopenia (501)	Form 2035 & 2135
	☐ Glanzmann thrombasthenia (502) ☐ Other inherited platelet abnormality (509)	No disease insert required

Inherited Disorders of Metabolism

Primary Disease	Disease Subtype(s)	Disease Inserts
Inherited disorders of metabolism	☐ Osteopetrosis (malignant infantile osteopetrosis) (521)	Form 2036 & 2136
	☐ Metachromatic leukodystrophy (MLD) (542) ☐ Adrenoleukodystrophy (ALD) (543) ☐ Krabbe disease (globoid leukodystrophy) (544)	Form 2037 & 2137
	☐ Lesch-Nyhan (HGPRT deficiency) (522) ☐ Neuronal ceroid lipofuscinosis (Batten disease) (523) **Mucopolysaccharidosis:** ☐ Hurler syndrome (IH) (531) ☐ Scheie syndrome (IS) (532) ☐ Hunter syndrome (II) (533) ☐ Sanfilippo (III) (534) ☐ Morquio (IV) (535) ☐ Maroteaux-Lamy (VI) (536) ☐ β-glucuronidase deficiency (VII) (537) ☐ Mucopolysaccharidosis (V) (538) ☐ Mucopolysaccharidosis, not otherwise specified (530) ☐ Gaucher disease (541) ☐ Niemann-Pick disease (545) ☐ I-cell disease (546) ☐ Wolman disease (547) ☐ Glucose storage disease (548) ☐ Mucolipidoses, not otherwise specified (540) **Polysaccharide hydrolase abnormalities:** ☐ Aspartyl glucosaminidase (561) ☐ Fucosidosis (562) ☐ Mannosidosis (563) ☐ Polysaccharide hydrolase abnormality, not otherwise specified (560) ☐ Other inherited metabolic disorder (529) ☐ Inherited metabolic disorder, not otherwise specified (520)	Form 2038 & 2138

Histiocytic Disorders

Primary Disease	Disease Subtype(s)	Disease Inserts
Histiocytic disorders	☐ Hemophagocytic lymphohistiocytosis (HLH) (571)	Form 2039 & 2139
	☐ Langerhans cell histiocytosis (histiocytosis-X) (572)	Form 2040 & 2140
	☐ Hemophagocytosis (reactive or viral associated) (573) ☐ Malignant histiocytosis (574) ☐ Other histiocytic disorder (579) ☐ Histiocytic disorder, not otherwise specified (570)	No disease insert required

Autoimmune Diseases

Primary Disease	Disease Subtype(s)	Disease Inserts
Autoimmune diseases	☐ Rheumatoid arthritis (603)	Form 2041 & 2141
	☐ Psoriatic arthritis / psoriasis (604) ☐ Juvenile idiopathic arthritis (JIA): systemic (Stills disease) (640) ☐ JIA: oligoarticular (641) ☐ JIA: polyarticular (642) ☐ JIA: other (643) ☐ Other arthritis (633)	Form 2042 & 2142
	☐ Multiple sclerosis (602)	Form 2043 & 2143
	☐ Systemic sclerosis (scleroderma) (607)	Form 2044 & 2144
	☐ Systemic lupus erythematosis (SLE) (605)	Form 2045 & 2145
	Connective tissue diseases: ☐ Sjögren syndrome (608) ☐ Polymyositis / dermatomyositis (606) ☐ Antiphospholipid syndrome (614) ☐ Other connective tissue disease (634) **Vasculitis:** ☐ Wegener granulomatosis (610) ☐ Classical polyarteritis nodosa (631) ☐ Microscopic polyarteritis nodosa (632) ☐ Churg-Strauss (635) ☐ Giant cell arteritis (636)	No disease insert required

Primary Disease	Disease Subtype(s)	Disease Inserts
	☐ Takayasu (637) ☐ Behcet syndrome (638) ☐ Overlap necrotizing arteritis (639) ☐ Other vasculitis (611) **Other neurologic autoimmune disease:** ☐ Myasthenia gravis (601) ☐ Other autoimmune neurological disorder (644) **Hematologic autoimmune disease:** ☐ Idiopathic thrombocytopenic purpura (ITP) (645) ☐ Hemolytic anemia (646) ☐ Evan syndrome (647) ☐ Other autoimmune cytopenia (648) **Bowel disease:** ☐ Crohn's disease (649) ☐ Ulcerative colitis (650) ☐ Other autoimmune bowel disorder (651)	

Other Disease

Primary Disease	Disease Subtype(s)	Disease Inserts
Other disease	Specify other disease:_____	No disease insert required