ABMTRR
Australasian
Bone Marrow Transplant
Recipient Registry

Guidelines for completing Registration Form (Form A)

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Introduction

A Registration Form (Form A) should be completed for each transplant. These should be submitted within a month of the transplant. A transplant is defined as the following:

The major criterion for a haematopoietic stem cell transplant is that it involves an infusion of haematopoietic stem cells with the intention of repopulating the bone marrow and hence the recovery of haematopoiesis in all lineages. This is usually done subsequent to either full or reduced intensity conditioning chemotherapy.

Registration is NOT required:

- If patients who were scheduled for transplant (including administration of the pre-transplant conditioning chemotherapy) did not proceed for medical or other reasons.

- If infusion of cells is a Donor Cellular Infusion (DCI)
  A DCI is commonly used to treat infections (e.g. viral) or recurrent disease. In the setting of recurrent disease, the DCI is used to create a graft-versus-leukaemia/tumour effect. A DCI may also be given to treat GVHD or promote engraftment when chimerism studies reveal less than 100% donor cells. Conditioning treatment is not given prior to receiving the additional donor cells since replacement of the marrow is not the goal.

In some cases there may be difficulty applying these definitions eg with donor lymphocyte infusions, haematopoietic cells are also present. In this case the intention of the treatment should be taken into account i.e. if the intention was to repopulate the patient marrow with haematopoietic cells, then the procedure is considered a transplant. This includes additional donor cells given for failed or poor ANC recovery, loss of graft, or late graft failure.

Privacy and consent

All transplant recipients should be asked for their consent for the transfer of information describing themselves and their transplant procedures to central data registries.

If consent is not obtained, limited information on the transplant is still required to be sent to the Registry. This involves no details that may identify the patient. Please contact the Registry for details.

Date format: All dates to be formatted as day/month/year

Patient Details

1. Patient UPN

This is the Unique Patient Number assigned to each patient or transplant at your centre to identify transplant recipients (rather than the Hospital Patient Record Number)

3. First four letters of surname

Enter first four letters, ignoring little ‘c’, apostrophes eg McKN enter as MCKN, O’ROU enter as OROU

4. Place of usual residence

Enter State and postcode if patient resides in Australia, otherwise specify the country if resides overseas e.g. New Zealand

Transplant Details

8a. Transplant date

Only enter date of first day if the infusion is given over more than one day. It will be regarded as one transplant.

Autologous staged - subsequent dates can be entered on the same form if the infusion has already taken place, otherwise complete a new registration form including Questions 1 to 15 (Disease classification is not required as this information should be unchanged).

8b. Transplant number

Chronological number of transplant

8c. If more than one transplant

Completing this section will allow the Registry to link this transplant to an existing patient in the database if the prior transplant(s) have been registered.

If the transplant was performed at a different centre, an approximate date may only be known.

9. Type of transplant

Select Allogeneic, Autologous or Autologous staged**.**
Autologous staged – involves two or more planned infusions at set intervals.

10. Mobilisation

Select all agents given (to an autologous recipient or an allogeneic donor) for mobilisation of cells.

11. Transplant source

Select all cell sources that apply
If there are more than one donor e.g. double cord transplant, then compete Question 25.
If there are more than two donors, then complete additional copies of Question 25

12. Allogeneic donor - complete for allogeneic transplants only

Specify relationship of donor to recipient - if "**HLA matched other relative**" or "**HLA mismatched other relative**" is selected, e.g. parent, aunt etc. This would include siblings who are not HLA identical and all other blood related relatives. Adoptive or step-parents/children should be reported as unrelated

HLA-mismatched other relative:

- indicate degree of mismatch
  - 1 HLA antigen mismatch
  - ≥ 2 HLA antigen mismatch
- if donor is known to be haploidentical, please indicate in the checkbox

Unrelated donor:

- record donor registry country
- complete table for HLA matching, includes antigenic and allelic
- include adoptive or step-parents/children here

13. Were any of following components of this transplant performed substantially as outpatient procedures?

If more than half the time is spent as an outpatient during the following procedures:

- conditioning,
- infusion of the cells,
- acute post transplant period, defined as up to 30 days post transplant

please indicate here.

14a. Conditioning agents

Select all agents given.

- Indicate dose options for melphalan and TBI if given
- Indicate source of ATG (ALG, ALS, ATS) if given.

Indicate if conditioning was intended to be myeloablative for allogeneic transplants only.

15. Latest date patient is seen

It is important to enter a date here as this will contribute to the survival analysis.

This date may be obtained from correspondence or pathology results etc. If the patient has died, then this will be the death date.

16. Date diagnosed of primary disease for this transplant

This is the date of diagnosis of the indication for which the patient is being transplanted.

Note. If the diagnosis is MDS/MDP transformed to AML, then record the MDS/MDP diagnosis date here. The date of transformation to AML will be recorded in the MDS/MPD disease specific section.

DISEASE CLASSIFICATION

The classification methods reflect standard practice in stem cell transplant research as far as possible. ABMTRR classifications are similar to those used by the Centre for International Blood and Marrow Transplant Research (CIBMTR) and the European Group for Blood and Marrow Transplantation (EBMT)

Please indicate the **most specific** disease classification from the Disease Classification in this section or alternatively enter a WHO classification code. For more information, go to: http://www.who.int/classifications/icd/en/

17. ACUTE LEUKAEMIA

Please indicate the **most specific** disease classification in this section or alternatively enter the WHO classification code

17.1. Acute myeloid leukaemia

AML with recurrent genetic abnormalities

- AML with t(8;21)(q22;q22) (AML1/ETO)
- AML with abnormal BM eosinophils and inv(16)(p13q22) or t(16;16)(p13;q22) CBFβ/MYH11)
- APL with t(15;17)(q22;q12),(PML/RARα) and variants (M3)
- AML with 11q23(MLL) abnormalities
- AML with multi-lineage dysplasia (myelodysplasia-related changes)

AML, not otherwise categorised

- AML, minimally differentiated (MO)
- AML without maturation (M1)
- AML with maturation (M2)
- AML, Acute myelomonocytic leukaemia (M4)
- Acute monoblastic/ acute monocytic leukaemia (M5)
- Acute erythroid leukaemia (erythroid/myeloid and pure erythroleukemia) (M6)
- Acute megakaryoblastic leukaemia (M7)
- Acute basophilic leukaemia
- Acute panmyelosis with myelofibrosis
- Myeloid sarcoma
- AML, not otherwise specified

Did AML transform from MDS/MPS?

If yes, then complete both Questions 17 and 20 (Myelodysplasia/Myeloproliferative Diseases)

Was AML treatment related?

If the AML is related to previous treatment with chemotherapy or radiotherapy then select "Yes"

17.2. Acute Lymphoblastic Leukaemia

- Precursor B-cell ALL
  Indicate subtype if known:
  - t(9;22)(q34;q11);bcr/abl+
  - t(v;11q23);MLL rearranged
  - t(1;19)(q23;p13)E2A/PBX1
  - t(12;21)(p12;q22)ETV/CBFα
- Precursor T-cell ALL
- ALL, not otherwise specified

17.3. Other Acute Leukaemias

- Acute undifferentiated leukaemia
- Biphenotypic, bilineage or hybrid leukaemia
- Acute mast cell leukaemia
- other, specify

17.4. Disease Status At Transplant – Acute Leukaemia

Disease Status	Definition
Never Treated	Never received treatment for Acute Leukaemia This does not include treatment for prior MDS/MPD, eg. MDS was initially diagnosed and treated, then transformed into AML and proceeded immediately to transplant instead of first treating the AML.
Primary Induction Failure (PIF)	Never achieved complete remission. Not limited to the number of treatments used unsuccessfully.
Complete Remission *	**All** of the following are met for at least four weeks:* • < 5% blasts in the bone marrow • Normal maturation of all cellular components in the bone marrow (myeloid, erythroid, and megakaryocytic lineages) • No blasts with Auer rods (AML only) • No extramedullary disease (e.g. CNS or soft tissue involvement) • ANC > 1 x10^9/L • Platelets ≥ 100 x 10^9/L • Transfusion independent *Note: There may not be a four-week interval between completion of treatment and assessment immediately prior to transplant. CR should still be reported as the status at transplant if it represents the "best assessment" prior to transplant. The pre-transplant disease status ***should not be changed*** based on early relapse or disease assessment ***post-HSCT.*** Recipients who meet all the criteria of CR, but have persistent cytogenetic and/or molecular abnormalities, should be reported as being in CR. Report cytogenetic and/or molecular abnormalities in the appropriate section Recipients with extramedullary disease are considered to have persistent or relapsed disease. **NOTE: Recipients with MDS that transformed to AML** If the recipient has residual MDS following treatment for AML, report the AML disease status as either PIF or relapse (i.e. cannot be in an AML CR if there is evidence of MDS at time of assessment.)
Relapse *	Recurrence of disease after CR. Defined as: • >5% blasts in the marrow • Extramedullary disease • Reappearance of cytogenetic abnormalities and/or molecular markers associated with the diagnosis at levels that, as determined by a physician, represent relapse.

*** Indicate Number of CR or Relapse**

18. CHRONIC MYELOGENOUS LEUKAEMIA

Please indicate the **most specific** disease classification in this section or alternatively enter the WHO classification code

- Ph+; bcr/abl+
- Ph+; bcr/abl-
- Ph+; bcr/abl unknown
- Ph- ; bcr/abl+
- Ph unknown; bcr/abl+

The diagnosis of CML must include the following (WHO Classification requirements):

- Philadelphia chromosome, complex variation and/or variant form, see table below
- or bcr/abl gene rearrangement

Philadelphia chromosome t(9;22)(q34;q11)	An exchange of genetic material between region q34 of chromosome 9 and region q11 of chromosome 22.
Complex variation	Translocation of three or more chromosomes, one of which must be chromosome 22 [eg. t(3; 9; 22)].
Variant form	Any translocation involving 22q11, or 22.q11.2 in which CML is the suspected diagnosis [eg. t(13; 22)(p3;q11)].

If none of these are identified, but CML is suspected, report under "Other Leukaemias" as "Atypical chronic myeloid leukaemia"

18.1. Disease Status At Transplant - CML

Disease Response	Definition
Chronic Phase *	None of the characteristics of accelerated phase or blast crisis Also indicate if the response qualify as a haematological, cytogenetic and molecular remission Haematological CR is when all of following are met: • WBC < 10 x 10^9/L, without immature granulocytes and with < 5% basophils. • Platelet count < 450 x 10^9/L. • Non-palpable spleen.
Accelerated Phase *	**One or more** of the following: • 10%-19% blasts in blood or marrow • ≥ 20% basophils in peripheral blood • Clonal marrow cytogenetic abnormalities in addition to the single Philadelphia chromosome (clonal evolution) • Increasing spleen size, unresponsive to therapy • Increasing WBC, unresponsive to therapy • Thrombocytopenia (platelets < 100 x 10^9/L), unrelated to therapy • Thrombocytosis (platelets > 1,000 x 10^9/L), unresponsive to therapy
Blast Crisis *	**One or more** of the following: • ≥ 20% blasts in the peripheral blood or bone marrow. • extramedullary blast proliferation • large foci or clusters of blasts on BM biopsy

*** Indicate Number of Chronic Phase, Accelerated Phase or Blast Crisis.**

19. OTHER LEUKAEMIAS

- Chronic lymphocytic leukaemia (CLL), not otherwise specified
- CLL, B-cell/small lymphocytic lymphoma
- Hairy cell leukaemia
- Prolymphocytic leukaemia (PLL), not otherwise specified
    - PLL, B-cell
    - PLL, T-cell
- other leukaemia, specify

19.1. Disease Status At Transplant – CLL

Disease Status at transplant	**Definition**
Never Treated	Never received treatment for CLL.
Complete Remission (CR)	Requires **all** the following: • No radiographic evidence of lymphadenopathy • No organomegaly • Neutrophils > 1.5 x 10^9/L • Platelets > 100 x 10^9/L • Haemoglobin >110g/L • Lymphocytes < 4 x 10^9/L • Bone marrow < 30% lymphocytes • Absence of constitutional symptoms (e.g., fatigue, fevers, night sweats)
Nodular Partial Remission (nPR)	Complete response with persistent lymphoid nodules in bone marrow.
Partial Remission (PR)	Requires **all** of the following: • ≥ 50% decrease in peripheral blood lymphocyte count from pretreatment value • ≥ 50% reduction in lymphadenopathy if present pretreatment • ≥ 50% reduction in liver and spleen size if enlarged pretreatment **AND one or more** of the following: • Neutrophils ≥ 1.5x10^9/L or 50% above baseline • Platelets > 100x10^9/L or 50% improvement over baseline • Haemoglobin > 110g/L or 50% improvement over baseline
No Response/Stable Disease (NR/SD)	No change. Not achieve complete or partial response, or have progressive disease.
Progression	Requires **one or more** of the following: • ≥ 50% increase in the sum of the products of ≥ 2 lymph nodes (≥ 1 node must be ≥ 2 cm) or new nodes • ≥ 50% increase in liver or spleen size, or new hepatomegaly or splenomegaly • ≥ 50% increase in absolute lymphocyte count to ≥ 5 x 109/L • transformation to a more aggressive histology (eg Richter's syndrome)
Relapse (untreated)	The re-appearance of disease after complete recovery. Relapse should be determined by one or more diagnostic tests.

19.2. Disease Status At Transplant – Hairy Cell Leukaemia

Disease Status at transplant	Definition
Never Treated	Never received treatment for HCL.
Complete Remission (CR)	Requires **all** of the following: • Neutrophils ≥ 1.5 x 10^9 /L • Haemoglobin ≥ 120g/L • Platelets ≥ 100 x 10^9/L • Absence of hairy cells on peripheral blood smear • No palpable lymphadenopathy or hepatosplenomegaly
Nodular Partial Remission (nPR)	Not applicable
Partial Remission (PR)	Requires **all** of the following: • ≥ 50% reduction in the absolute hairy cell count in the peripheral blood and the bone marrow • ≥ 50% improvement of all cytopenias • ≥ 50% reduction in abnormal lymphadenopathy or hepatosplenomegaly
No Response/Stable Disease (NR/SD)	Not applicable
Progression	Not applicable
Relapse (untreated)	**Relapse after CR:** • Reappearance of hairy cells in the peripheral blood smear and/or bone marrow (regardless of the degree of infiltration) • Development of peripheral blood cytopenias • Splenomegaly **Relapse after PR:** • ≥ 50% increase of residual hairy cells in the marrow • Development of cytopenias • Splenomegaly insufficient to qualify as PR **Or** • Reappearance of hairy cells in the bone marrow of those patients classified as partial responders based on residual splenomegaly only

19.3. Disease Status At Transplant - Prolymphocytic Leukaemia

Disease Status at transplant	Definition
Never Treated	Never received treatment for PLL.
Complete Remission (CR)	Requires **all** the following: • No radiographic evidence of lymphadenopathy • No organomegaly • Neutrophils > 1.5 x 10^9/L • Platelets > 100 x 10^9/L • Haemoglobin > 110g/L • Lymphocytes < 4 x 10^9/L • Bone marrow < 30% lymphocytes • Absence of constitutional symptoms (e.g., fatigue, fevers, night sweats)
Nodular Partial Remission (nPR)	Complete response with persistent lymphoid nodules in bone marrow.
Partial Remission (PR)	Requires **all** of the following: • ≥50% decrease in peripheral blood lymphocyte count from pretreatment value • ≥50% reduction in lymphadenopathy if present pretreatment • ≥50% reduction in liver and spleen size if enlarged pretreatment **AND one or more** of the following: • Neutrophils ≥ 1.5x10^9/L or 50% above baseline • Platelets > 100x10^9/L or 50% improvement over baseline • Haemoglobin > 110g/L or 50% improvement over baseline
No Response/ Stable Disease (NR/SD)	No change. Not complete response, partial response, or progressive disease.
Progression	Requires **one or more** of the following: • ≥ 50% increase in the sum of the products of ≥ 2 lymph nodes (≥ 1 node must be ≥ 2 cm) or new nodes • ≥ 50% increase in liver or spleen size, or new hepatomegaly or splenomegaly • ≥ 50% increase in absolute lymphocyte count to ≥ 5 x 10^9/L • Transformation to a more aggressive histology
Relapse (untreated)	The re-appearance of disease after complete recovery. Relapse should be determined by one or more diagnostic tests.

20. MYELODYSPLASTIC/MYELOPROLIFERATIVE DISORDERS (MDS/MPD)

Please indicate the **most specific** disease classification in this section or alternatively enter the WHO classification code

20.1. Myelodysplastic Syndromes (MDS)

- RA
- RARS
- RAEB-1
- RAEB-2
- RCMD
- RCMD/RS
- Isolated del(5q)
- transformed to AML – require date of transformation
- MDS unclassifiable/not otherwise specified

20.2. Myeloproliferative Disease (MPD)

- Chronic Neutrophilic Leukaemia
- Chronic Eosinophilic Leukaemia (hypereosinophilic syndrome)
- Chronic Idiopathic myelofibrosis (primary/acute myelofibrosis, myelosclerosis, myelofibrosis with myeloid metaplasia)
- Chronic Myeloproliferative Disease, not otherwise specified
- Essential thrombocythemia (ET)
- Polycythemia vera (PCV)

20.3. Combined Myelodysplastic/Myeloproliferative Disease

- Atypical CML, Ph-; bcr/abl-
- Atypical CML, Ph-; bcr/abl unknown
- Atypical CML, Ph unknown; bcr/abl-
- Atypical CML, Ph unknown; bcr/abl unknown
- Chronic myelomonocytic leukaemia (CMMol, CMML)
- Juvenile myelomonocytic leukaemia (JMML, JCML, JCMML)

Therapy related

Please indicate if the MDS/MPD was related to prior exposure to therapeutic drugs or radiation, ie. therapy related

NOTE:
" transformed to AML "
If the recipient is being transplanted for AML that has transformed from MDS/MPS/CMML, complete **both sections for MDS/MPD and AML.**
Enter the diagnosis date of the MDS/MPS as the diagnosis date (Question 16), and the date of AML diagnosis as the date of transformation in the MDS/MPD section (Question 20)

20.4. Disease Status At Transplant – MDS/MPS/CMML

Disease Status	Definition
Supportive care or treatment without chemotherapy (Never treated)	Examples include but are not limited to: • Observation with periodic blood count tests ("watch and wait") • Blood transfusions and iron chelation therapy • Erythropoietin (EPO) and other blood cell growth factors • Antithymocyte globulin (ATG) • Immune modulation agents including thalidomide and lenalidomide
Relapsed after CR *	**one or more** of the following: • Return to pre-treatment bone marrow blast percentage • Decrease of ≥ 50% from maximum response levels in granulocytes or platelets • Transfusion dependence or Hb level ≥ 15g/L lower than prior to therapy Do not include PRs when calculating the number of relapses

Complete Remission (CR) *	**all** of the following and maintained for ≥ 4 weeks*:
	• Bone marrow evaluation:
	  • < 5% myeloblasts with normal maturation of all cell lines
	• Peripheral blood evaluation:
	  • Hb ≥ 110g/L untransfused, without erythropoietin support
	  • ANC ≥ 1 x 10^9/L without myeloid growth factor support
	  • platelets ≥ 100 x 10^9/L without thrombopoietic support
	  • 0% blasts
	*If the period between achieving CR and Day 0 is less than four weeks, and the recipient is documented to be in CR, report the status at transplant as CR.

Improvement, but no CR	**one or more** of the following, maintained for ≥ 8 weeks, no ongoing cytotoxic therapy:
	• Haematologic Improvement (HI)-E
	  – Hb increase of ≥ 15 g/L untransfused
	  – For RBC transfusions given for Hb ≤ 90 g/L, reduction in RBC units transfused in 8 weeks by ≥ 4 units compared to the pre-treatment transfusion number in the previous 8 weeks
	• HI-P
	  – For pre-treatment platelet count of > 20 x 10^9/L, platelet absolute increase of ≥ 30 x 10^9/L
	  – For pre-treatment platelet count of < 20 x 10^9/L, platelet absolute increase of ≥ 20 x 10^9/L and ≥ 100% from pre-treatment level
	• HI-N
	  – Neutrophil count increase of ≥ 100% from pre-treatment level and an absolute increase of ≥ 0.5 x 10^9/L

No response (NR)	Does not meet the criteria for "Improvement, but no CR" category, and no evidence of disease progression.
Progressive/Worse	**one or more** of the following, in the absence of another explanation (e.g., infection, bleeding, ongoing chemotherapy, etc.): • ≥ 50% reduction from maximum response levels in granulocytes or platelets • Reduction in haemoglobin by ≥ 15 g/L • Transfusion dependence

*** Indicate Number of CR, or Relapse**

20.5. Disease Status At Transplant – Atypical CML

Disease Status at transplant	Definition
Never Treated	Never received treatment for Atypical CML.
Complete Remission (CR)	**All** of the following criteria are met and maintained for ≥ 4 weeks: • Marrow with normal maturation of all cellular components • ≤ 5% blasts in the marrow • No signs or symptoms of the disease If the time between achieving CR and Day 0 of the transplant is less than four weeks, and the recipient is believed to be in CR, report as CR. Include recipients with persistent cytogenetic abnormalities who otherwise meet all the criteria of CR. Recipients with extramedullary disease should be considered to have persistent disease, or to be in relapse.
No Response (NR)	**Never achieved CR** with any therapy. PIF is not limited to the number of treatments used unsuccessfully. This status only applies to recipients who have **never** been in CR.
Progression	Not applicable for atypical CML.
Relapse after CR	Defined as: • >5% blasts in the marrow • extramedullary disease • reappearance of cytogenetic abnormalities and/or molecular markers associated with the diagnosis at levels that, as determined by a physician, represent relapse.

20.6. Disease Status At Transplant – JMML

Disease Status	Definition
Continued Complete Response	Achieved and remained in CR. Continued absence of all known disease after achieving CR following a previous line of therapy.
Complete Response	Normalization of WBC and organomegaly.
Partial Response	≥ 50% reduction in WBC and/or organomegaly.
Minimal Response	**one or more** of the following: • 25%-50% reduction in WBC and organomegaly • partial response in WBC but no change in organomegaly • partial response in organomegaly but no change in WBC
Stable Disease	≤ 25% reduction in WBC and/or organomegaly.
Progressive Disease	Increase in WBC and/or organomegaly.
Not Assessed	No assessment of the recipient's disease has been done.

21. LYMPHOMA

Please indicate the **most specific** disease classification in this section or alternatively enter the WHO classification code

21.1. Hodgkin Lymphoma

- Nodular, lymphocyte predominant
- Lymphocyte-rich
- Nodular sclerosis
- Mixed celluarity
- Lymphocyte depleted
- Hodgkin lymphoma, not otherwise specified

21.2. Non Hodgkins Lymphoma

B-cell neoplasms

- Burkitt's lymphoma/Burkitt cell leukaemia (ALL L3)
- High grade B-cell lymphoma, Burkitt-like (provisional entity)
- Diffuse large B-cell lymphoma
  Indicate subtype if known:
  - Intravascular large B-cell
  - Mediastinal large B-cell
  - Primary effusion
- Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue (MALT)
- Follicular lymphoma
  Indicate grade if known:
  - Grade I (predominantly small cleaved cell)
  - Grade II (Follicular, mixed, small cleaved and large cell)
  - Grade III (Follicular, predominantly large cell)
  - grade unknown
- Lymphoplasmacytic lymphoma
- Mantle cell lymphoma
- Nodal marginal zone B-cell lymphoma (± monocytoid B-cells)
- Primary CNS lymphoma
- Splenic marginal zone B-cell lymphoma
- Waldenstrom's Macroglobulinaemia
- Unclassifiable, features intermediate between Diffuse large B cell and Burkitt lymphoma
- Unclassifiable, features intermediate between Diffuse large B cell and Classical Hodgkin lymphoma
- other B-cell lymphoma, specify

T-Cell & NK-cell neoplasms

- Adult T-cell lymphoma/leukaemia (HTLV1+)
- Aggressive NK-cell leukaemia
- Anaplastic large-cell lymphoma, T/null cell, primary cutaneous
- Anaplastic large-cell lymphoma, T/null cell, primary systemic
- Angioimmunoblastic T-cell lymphoma (AILD)

- Enteropathy-type T-cell lymphoma
- Extranodal NK/T-cell lymphoma, nasal type
- Hepatosplenic gamma-delta T-cell lymphoma
- Mycosis fungoides
- Peripheral T-cell lymphoma, not otherwise specified
- Subcutaneous panniculitis-like T-cell lymphoma
- Sezary syndrome
- Large T-cell granular lymphocytic leukaemia
- Other T-cell/NK cell lymphoma, specify

If Non-Hodgkin's Lymphoma transforms from one subtype to another prior to transplant, report the most current subtype. Report the initial subtype in "Prior histology if transformed"

21.3. Disease Status At Transplant - Lymphoma

Disease Status	Definition
Never Treated	Never received treatment for lymphoma.
Primary Refractory (less than PR to initial therapy)/ PIF res	The response to treatment is less than in a partial response. Includes recipients who achieved a prior PR (but never CR) but are not in either PR or relapse immediately prior to transplant.
Partial Response *	Reductions of ≥ 50% in greatest diameter of all sites of known disease and no new sites. Distinguish the type of PR with: "**without** prior CR" and "**with** prior CR," Note: "with prior CR" is analogous to relapse-sensitive (Rel sen). Indicate which relapse number.
Complete Remission Confirmed *	Complete disappearance of all known disease for ≥ 4 weeks. "Confirmed" is defined as a laboratory and/ pathological radiographic evidence.
Complete Remission Unconfirmed *	Complete disappearance of all known disease for ≥ 4 weeks with the exception of persistent scan abnormalities of unknown significance.
Relapse *	Recurrence of disease after CR. This may involve an increase in size of known disease or new sites of disease. Guideline for calculating number of relapses: – 1st relapse indicates one prior CR – 2nd relapse indicates 2 prior CRs – 3rd or higher indicates 3 or more prior CRs Do not include PRs when calculating the number of relapses
Progression	Worsening of disease, previously assessed as not in CR or where CR lasted less than 3 months

*** Indicate Number of PR, CR, or Relapse**

For relapse only - indicate if disease is sensitive to chemotherapy
Sensitivity is measured based on the **last chemotherapy given within the six months prior to transplant**.

Sensitive: ≥ 50% reduction in the bi-dimensional diameter of all disease sites with no new sites of disease
Resistant: < 50% reduction in the diameter of all disease sites or development of new disease sites
Untreated: no chemotherapy was given within the six months prior to the pre-transplant conditioning.
Unknown: not assessed or not available

22. PLASMA CELL DISORDERS

Please indicate the **most specific** disease classification in this section or alternatively enter the WHO classification code

22.1. Multiple Myeloma

- IgG
- IgA
- IgD
- IgE
- IgM (not Waldenstrom macroglobulinemia)
- Light chain only – indicate which light chain type i.e. kappa or lambda
- Non-secretory - neither kappa nor lambda light chains will be present

Light chain type

- kappa
- lambda

22.2. Stage at Diagnosis: Indicate which staging system is used

Durie-Salmon System

Stage I	All of the following: • Haemoglobin >99 g/L • Serum calcium <2.65mmol/L • No lytic lesions or one single minor lesion • Monoclonal IgG < 50g/L or IgA <30 g/L (for `common' type myeloma), Or light chains in urine <4 g/24 hrs (for light chain myeloma)
Stage II	Neither stage I or III
Stage III	One or more of the following: • Haemoglobin < 85 g/L • Serum calcium > 2.65mmol/L • Monoclonal IgG >70 g/L, IgA >50 g/L ('common' type) or light chains in urine > 12 g/24 hrs • Multiple skeletal lesions and/or pathologic fractures
Sub-classification	
A	Relatively normal renal function (serum creatinine < 180µmol/L)
B	Abnormal renal function (serum creatinine ≥ 180µmol/L)

I.S.S. System

Stage I	β2 microglobulin < 3.5 mg/L, albumin ≥ 35 g/L
Stage II	β2 microglobulin < 3.5 mg/L and albumin <35 g/L or β2 microglobulin 3.5 – ≤ 5.4 mg/L irrespective of albumin
Stage III	β2 microglobulin > 5.5 mg/L

22.2. Other Plasma Cell Disorders

- Plasma cell Leukaemia
- Solitary plasmacytoma
- Primary amyloidosis
- Other, specify

Other Plasma Cell Disorder, specify
On occasion, a recipient could have two heavy-chain types; this is known as biclonal myeloma (e.g., IgA Kappa and IgG Kappa). Other examples of "other plasma cell disorders" include: Light Chain Deposition Disease (LCDD), light chain only myeloma with associate LCDD, and osteosclerotic myeloma (POEMS). In this instance, report "other plasma cell disorder" and specify the type.

22.3. Disease Status At Transplant – Plasma Cell Disorders

Disease Status	**Definition**
Never Treated	No treatment given in the six months prior to transplant
Stringent Complete Remission * (sCR)	Follow criteria for CR as defined below, **plus all of the following**: • Normal free light chain ratio, • Absence of clonal cells in the bone marrow by immunohistochemistry or immunofluorescence (confirmation with repeat BM biopsy not needed). (Presence and/or absence of clonal cells is based upon the κ/λ ratio. An abnormal κ/λ ratio by immunohistochemistry and/or immunofluorescence requires a minimum of 100 plasma cells for analysis. An abnormal ratio reflecting the presence of an abnormal clone is κ/λ of > 4:1 or < 1:2.) sCR requires two consecutive assessments (of the same method) made at any time before the start of any new therapy. If radiographic studies were performed, there must be no known evidence of progressive or new bone lesions. Radiographic studies are not mandatory.
Complete Remission *	**All of the following**: • Negative immunofixation on serum and urine samples • Disappearance of any soft tissue plasmacytomas • < 5% plasma cells in the bone marrow Recipients with light chain only myeloma, **all** of the following must be met: • Normal serum free light chain ratio • Negative immunofixation on urine samples • Disappearance of any soft tissue plasmacytomas • < 5% plasma cells in the BM Recipients with non-secretory myeloma, **all** of the following must be met: • Disappearance of all soft tissue plasmacytomas • < 5% plasma cells in the bone marrow (confirmation with repeat bone marrow biopsy not needed) CR requires two consecutive assessments (of the same method) made at any time before the start of any new therapy.

	If radiographic studies were performed, there must be no known evidence of progressive or new bone lesions. Radiographic studies are not mandatory.
Very Good Partial Response (VGPR) *	**One or more** of the following must be present: • Serum and urine M-protein detectable by immunofixation but not on electrophoresis • ≥ 90% reduction in serum M-protein and urine M-protein level < 100 mg/24 hours. VGPR requires two consecutive assessments (of the same method) made at any time before the institution of any new therapy. If radiographic studies were performed, there must be no known evidence of progressive or new bone lesions. Radiographic studies are not mandatory.
Partial Response (PR) *	**Both** of the following must be present: • ≥ 50% reduction in serum M-protein • Reduction in 24-hour urinary M-protein by ≥ 90% or to < 200 mg/24 hours. If the serum and urine M-protein are not measurable (i.e. do not meet the following criteria): • serum M-protein ≥ 1 g/dL, • urine M-protein ≥ 200 mg/24 hours; then a ≥ 50% decrease in the difference between involved and uninvolved free light chain levels is required in place of the M-protein criteria (provided the serum free light chain assay shows involved level > 10 mg/dL and the serum free light chain is abnormal). If serum and urine M-protein *and* serum-free light assay are not measurable, a ≥ 50% reduction in bone marrow plasma cells is required in place of M-protein, provided the baseline bone marrow plasma cell percentage was ≥ 30%. In addition to the above listed criteria, a ≥ 50% reduction in the size of soft tissue plasmacytomas is also required, if present at baseline. PR requires two consecutive assessments (of the same method) made at any time before the institution of any new therapy. If radiographic studies were performed, there must be no known evidence of progressive or new bone lesions. Radiographic studies are not mandatory.
Stable Disease (SD)	Does not meet the criteria for CR, VGPR, PR, or PD. SD requires two consecutive assessments (of the same method) made at any time before the start of any new therapy. If radiographic studies were performed, there must be no known evidence of progressive or new bone lesions. Radiographic studies are not mandatory.
Progressive Disease (PD) *	**One or more** of the following: Increase of ≥ 25% from the lowest response value achieved in: • Serum M-component with an absolute increase ≥ 0.5 g/dL (for progressive disease, serum M-component increases of ≥ 1 g/dL are sufficient if the starting M-component is ≥ 5 g/dL) • Urine M-component with an absolute increase ≥ 200 mg/24 hours • For recipients without measurable serum and urine M-protein levels, the

	difference between involved and uninvolved free light chain levels with an absolute increase > 10 mg/dL • Bone marrow plasma cell percentage with absolute percentage ≥ 10% • Definite development of new bone lesions or soft tissue plasmacytomas, or definite increase in the size of any existing bone lesions or soft tissue plasmacytomas • Development of hypercalcemia (corrected serum calcium > 11.5 mg/dL or 2.65 mmol) , attributed solely to the plasma cell proliferative disorder. PD requires two consecutive assessments (of the same method) made at any time before classification as disease progression, and/or the institution of any new therapy.
Relapse from CR (untreated) *	**One or more** of the following: • Reappearance of serum or urine M-protein by immunofixation or electrophoresis • Development of ≥ 5% plasma cells in the bone marrow (relapse from CR has a 5% cutoff vs. 10% for other categories) • Appearance of any other sign of progression (e.g., new plasmacytoma, lytic bone lesion, hypercalcemia). Relapse requires two consecutive assessments (of the same method) made at any time before classification as relapse, and/or the start of any new therapy.

*** Indicate the number of the CR, sCR, VGPR, PR, Progression or Relapse**

23. SOLID TUMOURS

Please indicate the **most specific** disease classification in this section

- Bone sarcoma (excluding Ewing family tumours)
- Central nervous system tumours (include CNS PNET)
- Colorectal
- Ewing sarcoma/PNET, extra skeletal
- Ewing sarcoma/PNET, skeletal
- Germ cell tumour, extragonadal only
- Hepatobiliary
- Lung cancer, non-small cell
- Lung cancer, small cell
- Medulloblastoma
- Melanoma
- Neuroblastoma
- Ovary
- Pancreas
- Prostate
- Renal cell
- Retinoblastoma
- Rhabdomyosarcoma
- Soft tissue sarcoma
- Testicular
- Thymoma
- Wilm tumour
- Other solid tumour, specify

23.1. Disease Status At Transplant – Solid Tumours

Disease Status	WHO Definition	RECIST Definition
Adjuvant	Treatment given after primary cancer treatment to increase chances of cure. May include chemotherapy, radiation therapy, hormone therapy, or biological therapy.	Not applicable
Never Treated	Recipient was not treated for the malignancy prior to the preparative regimen.	Not applicable
Complete Response (CR) *	Complete disappearance of all known disease for ≥ 1 month. Includes disappearance of all signs and symptoms of disease with normalization of all biochemical and radiologic parameters, as well as a negative repeat biopsy.	Disappearance of all target lesions for a period of at least one month.
Complete Response Unconfirmed (CRU) *	Disappearance of all signs and symptoms of disease with normalization of all biochemical and radiologic parameters, but with persistent, unchanging imaging abnormalities of unknown significance.	Complete response with persistent imaging abnormalities of unknown significance.
Partial Response (PR)	Decrease of ≥ 50% in total tumour load of the lesions that have been measured for at least 4 weeks. Distinguish the type of PR as either: "**without** prior CR" or "**with** prior CR."	At least a 30% decrease in the sum of the longest diameter of measured lesions (target lesions), taking as reference the baseline sum of longest diameter.
Stable Disease/ No response (NR/SD)	After treatment, the size of one or more lesions has neither increased 25% or more, nor has total tumour size	Neither sufficient shrinkage to qualify for PR nor sufficient increase to qualify for PD, taking as reference the smallest sum of the

	decreased 50% or more.	longest diameter of measured lesions (target lesions), since the treatment started.
Progressive Disease (PD)	Increase of ≥ 20% in the size of one or more measurable lesions, or the appearance of new lesions.	At least a 20% increase in the sum of the longest diameter of measured lesions (target lesions), taking as reference the smallest sum of the longest diameter recorded since the treatment started or the appearance of one or more new lesions.
Relapse (untreated) *	Reappearance of disease after a complete remission. Should be determined by one or more diagnostic tests.	Not applicable.

*** Indicate Number of CR, CRU, or Relapse**

Sensitivity to chemotherapy (complete only for relapse)
Indicate if the disease is sensitive to chemotherapy. Sensitivity is measured based on the last chemotherapy given prior to HSCT; chemotherapy must include ≥ 2 cycles of treatment given ≤ 6 months prior to HSCT. Indicate the sensitivity to chemotherapy using the following guidelines:

Sensitive: ≥ 50% reduction in the bi-dimensional diameter of all disease sites with no new sites of disease
Resistant: < 50% reduction in the diameter of all disease sites or development of new disease sites
Untreated: if last treatment was not chemotherapy
Unknown: only if there no documentation of the recipient's response following treatment

NON MALIGNANT DISEASES

24.1. ANAEMIA (BONE MARROW FAILURE SYNDROMES)

- Acquired Severe Aplastic Anaemia (SAA), not otherwise specified
- Acquired SAA, secondary to hepatitis
- Acquired SAA, secondary to toxin/other drug
- Acquired Amegakaryocytosis (for congenital see under inherited disorders)
- Acquired Pure Red Cell Aplasia (PRCA) (not congenital)
- Other acquired cytopenic syndrome, specify
- Paroxysmal nocturnal haemoglobinuria (PNH)
- Fanconi anaemia
- Diamond-Blackfan anaemia (congenital PRCA)
- Shwachman-Diamond
- Other constitutional anaemia, specify

24.2. HAEMOGLOBINOPATHY

- Sickle cell disease
- Sickle thalassemia
- Thalassemia, not otherwise specified
- Other haemoglobinopathy, specify

24.3. PLATELET DISORDERS

- Congenital amegakaryocytosis/congenital thrombocytopenia
- Glanzmann thrombasthenia
- Other inherited platelet abnormalities, specify

24.4. HISTIOCYTIC DISORDERS

- Histiocytic disorders, not otherwise specified
- Familial erythro/haemophagocytic lymphohistiocytosis (FELH)
- Langerhans Cell Histiocytosis (Histiocytosis-X)
- Haemophagocytosis (reactive or viral associated)
- Malignant histiocytosis
- Other histiocytic disorder, specify

24.5. INHERITED DISORDERS OF METABOLISM/OSTEOPETROSIS

- Adrenoleukodystrophy (ALD)
- Aspartyl glucosaminuria
- B-glucuronidase deficiency (VII)
- Fucosidosis
- Gaucher disease
- Glucose storage disease
- Hunter syndrome (II)
- Hurler syndrome (IH)
- I-cell disease
- Krabbe disease (globoid leukodystrophy)
- Lesch-Nyhan (HGPRT deficiency)
- Mannosidosis
- Maroteaux-Lamy (VI)
- Metachromatic leukodystrophy (MLD)
- Morquio (IV)
- Mucolipidoses, not otherwise specified
- Mucopolysaccharidosis (V)
- Mucopolysaccharidosis, not otherwise specified
- Neimann-Pick disease
- Neuronal ceriod – lipofuscinosis (Batten disease)
- Osteopetrosis (malignant infantile osteopetrosis)
- Sanfilippo (III)
- Scheie syndrome (IS)
- Wolman disease
- Other inherited disorder of metabolism, specify
- Inherited Disorders of Metabolism, not otherwise specified

24.6. IMMUNE DEFICIENCIES

- Ataxia telangiectasia
- Bare lymphocyte syndrome
- DiGeorge anomaly
- CD 40 Ligand deficiency
- Cartilage hair hypoplasia
- Chediak-Higashi syndrome
- Chronic granulomatous disease
- Common variable immunodeficiency
- HIV infection
- Immune Deficiencies, not otherwise specified
- Leukocyte adhesion deficiencies
- Kostmann syndrome-congenital neutropenia
- Neutrophil actin deficiency
- Omenn syndrome
- Reticular dysgenesis
- SCID, ADA deficiency severe combined immune deficiency
- SCID, Absence of T and B cells
- SCID, Absence of T, normal B cell
- SCID, not otherwise specified
- SCID other, specify:
- Wiskott Aldrich syndrome
- X-linked lymphoproliferative syndrome
- Other immune deficiency, specify

24.7. AUTOIMMUNE DISORDERS

Additional data is required where indicated for some diseases, see pages 26 to 28.

- Indicate the involved organs and/or clinical problems at the time of original diagnosis, and whether that involvement was the primary reason for the transplant.
- For each antibody listed, indicate whether the result was “normal,” “elevated,” or “not done.”

Print the relevant page and submit with Form A.

If no additional information is required, specify the diagnosis in Question 24 on the registration form.

24.8. OTHER DISEASE, specify

Use this option only if the disease does not belong to any of the previous categories.

Hospital:	Patient UPN:	Name ID: __ __ __ __ __ __

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Connective Tissue Disease

Systemic sclerosis (scleroderma)

Involved organs/clinical problem/s (select all that apply)	Primary reason for transplant	Antibodies	normal	elevated	Not done
☐ diffuse cutaneous	☐ yes ☐ No	Scl 70 pos	☐	☐	☐
☐ limited cutaneous	☐ yes ☐ No	ACA pos	☐	☐	☐
☐ lung parenchyma	☐ yes ☐ No	ANA	☐	☐	☐
☐ pulmonary hypertension	☐ yes ☐ No				
☐ systemic hypertension	☐ yes ☐ No				
☐ renal, biopsy type:_____	☐ yes ☐ No				
☐ oesophagus	☐ yes ☐ No				
☐ other GI tract	☐ yes ☐ No				
☐ Raynaud	☐ yes ☐ No				
☐ CREST	☐ yes ☐ No				
☐ other, specify:_____	☐ yes ☐ No				

Systemic lupus erythematosus SLE

☐ renal, biopsy type:_____	☐ yes ☐ No	ANA	☐	☐	☐
☐ CNS, type:_____	☐ yes ☐ No	ds DNA	☐	☐	☐
☐ PNS, type:_____	☐ yes ☐ No	C3	☐	☐	☐
☐ lung	☐ yes ☐ No	C4	☐	☐	☐
☐ serositis	☐ yes ☐ No	Total			
☐ arthritis	☐ yes ☐ No	complement	☐	☐	☐
☐ skin, type:_____	☐ yes ☐ No	Other,specify:	☐	☐	☐
☐ haematological, type:_____	☐ yes ☐ No	_____			
☐ vasculitis, type:_____	☐ yes ☐ No				
☐ other, specify:_____	☐ yes ☐ No				

Polymyositis-dermatomyositis

☐ proximal weakness	☐ yes ☐ No	CPK	☐	☐	☐
☐ generalised weakness (including bulbar)	☐ yes ☐ No	Typical biopsy	☐	☐	☐
☐ pulmonary fibrosis	☐ yes ☐ No	Typical EMG	☐	☐	☐
☐ vasculitis, type:_____	☐ yes ☐ No	Typical rash	☐	☐	☐
☐ malignancy, type:_____	☐ yes ☐ No	(DM)			
☐ other, specify:_____	☐ yes ☐ No				

Antiphospholipid syndrome

☐ thrombosis, type:_____	☐ yes ☐ No	Anticardiolipin IgG	☐	☐	☐
☐ CNS, type:_____	☐ yes ☐ No	Anticardiolipin IgM	☐	☐	☐
☐ abortion	☐ yes ☐ No	Lupus anticoagulant	☐	☐	☐
☐ skin (livedo, vasculitis)	☐ yes ☐ No				
☐ haematological, type:_____	☐ yes ☐ No				
☐ other, specify:_____	☐ yes ☐ No				

Hospital:	Patient UPN:	Name ID: __ __ __ __ __ __

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(Connective Tissue Disease continued)

Sjögren syndrome

	Primary reason for transplant
☐SICCA	☐yes ☐No
☐exocrine gland swelling	☐yes ☐No
☐other organ lymphocytic infiltration	☐yes ☐No
☐lymphoma, paraproteinemia	☐yes ☐No
☐vasculitis	☐yes ☐No
☐other, specify:_____	☐yes ☐No

Other connective tissue disease, specify

Vasculitis

Wegener granulomatosis

Involved organs/clinical problem/s (select all that apply)	Primary reason for transplant	Antibodies	normal	elevated	Not done
☐upper respiratory tract	☐yes ☐No	c-ANCA pos	☐	☐	☐
☐pulmonary	☐yes ☐No	Anti Pr3	☐	☐	☐
☐renal, biopsy type:_____	☐yes ☐No	Anti MPO	☐	☐	☐
☐skin	☐yes ☐No	c-ANCA IFA	☐	☐	☐
☐other, specify:_____	☐yes ☐No	p-ANCA IFA	☐	☐	☐

Polyarteritis nodosa

☐classical ☐microscopic

☐renal, type:_____	☐yes ☐No	p-ANCA pos	☐	☐	☐
☐mononeuritis multiplex	☐yes ☐No	c-ANCA pos	☐	☐	☐
☐pulmonary haemorrhage	☐yes ☐No	Hepatitis			
☐skin	☐yes ☐No	serology	☐	☐	☐
☐GI tract	☐yes ☐No				
☐other, specify:_____	☐yes ☐No				

Other vasculitis

☐Churg-Strauss
☐Giant cell arteritis
☐Takayasu
☐Behçet's Syndrome
☐overlap necrotizing arteritis
☐other, specify:_____

Hospital:	Patient UPN:	Name ID: __ __ __ __ __ __

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Arthritis

Rheumatoid arthritis

Involved organs/clinical problem/s (select all that apply)	Primary reason for transplant
☐ destructive arthritis	☐ yes ☐ No
☐ necrotizing vasculitis	☐ yes ☐ No
☐ eye, type:_____	☐ yes ☐ No
☐ pulmonary	☐ yes ☐ No
☐ extra-articular, specify:_____	☐ yes ☐ No
☐ other, specify:_____	☐ yes ☐ No

Psoriatic arthritis/psoriasis

☐ destructive arthritis	☐ yes ☐ No
☐ psoriasis	☐ yes ☐ No
☐ other, specify:_____	☐ yes ☐ No

Other

☐ Juvenile idiopathic arthritis: systemic (Stills disease)
☐ Juvenile idiopathic arthritis: Oligoarticular
☐ Juvenile idiopathic arthritis: Polyarticular
☐ Juvenile idiopathic arthritis: Other, specify: _____ _____
☐ other, arthritis, specify:_____

Multiple sclerosis

clinical problem/s (select all that apply)	Primary reason for transplant
☐ primary progressive	☐ yes ☐ No
☐ secondary progressive	☐ yes ☐ No
☐ relapsing/ remitting	☐ yes ☐ No
☐ other, specify:_____	☐ yes ☐ No

Other Neurological Autoimmune Disease

- Myasthenia gravis
- other autoimmune neurological disorder, specify

Haematological Autoimmune Disease

- Idiopathic thrombocytopenic purpura (ITP)
- Haemolytic anaemia
- Evan syndrome
- other autoimmune cytopenia, specify

Bowel Disease

- Crohn's disease
- Ulcerative colitis
- other autoimmune bowel disorder, specify

References

1. Instructions for Pre-Transplant Essential Data (Pre-TED) Form. Center for International Blood and Marrow Transplant Research. A00413 Version 2.3 (8/28/2012)

2. MED-AB Forms Manual. A guide to the completion of the EBMT HSCT Med-AB Forms 2010 . European Group for Blood and Marrow Transplantation. (Last modified 17/07/2012)