How to document complicated patient histories?

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How to document complicated patient histories?

- Patient case
- Diagnosis indication for HSCT
- Subclassification of the disease at diagnosis at HSCT
- First line therapy or pre-HSCT treatment
- Disease status at HSCT
- Complications after HSCT treatment
- Relapse
- Additional disease treatment

Patient case (part 1)

- Male patient, first diagnosed at the age of 2 ½ years
- Diagnosed with Langerhans Cell Histiocytosis (LCH), in1992
- Received chemotherapy treatment with excellent response
- Received radiotherapy treatment
- Multiple reactivations of the disease in the following years
- Disease maintenance with a single dose chemotherapy

Patient case (part 2)

- Developed Myelodysplastic Syndrome (MDS) in 2011
- Received allogeneic HSCT with identical sibling donor in 2012
- Complete Remission (CR) after transplant
- No complications within the first 100 days.
- Developed series of complications after 100 days

Main indication diagnosis for HSCT

- How to determine which diagnosis is indication for transplant?
  - Langerhans Cell Histiocytosis (LCH)?
  - Myelodysplastic syndrom (MDS)?
  - Both diagnoses?

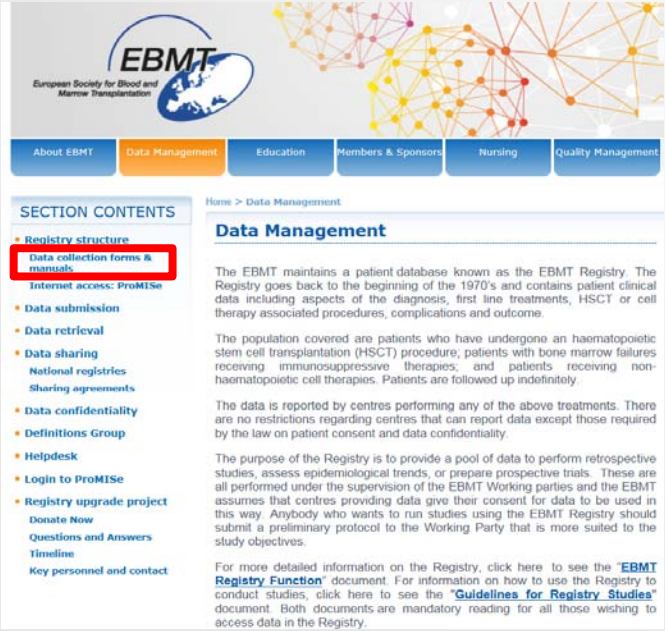
Diagnosis indication for HSCT (cont.)

- MDS was reported – why?
- Reported as therapy-related MDS or secondary disease
- Exposure to therapeutic agents or radiation
- Primary disease LCH, reported as diagnosis non indication for transplant

Disease subclassification at diagnosis

- How to determine the MDS subclassification at diagnosis

  - Subclassification according to WHO (World health organization)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European Society for Blood and Marrow Transplantation
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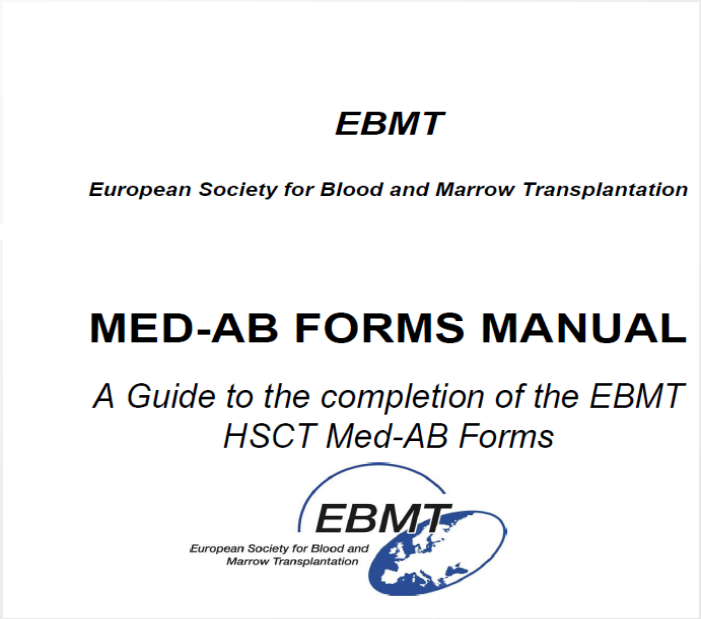
Data Management
The EBMT maintains a patient database known as the EBMT Registry. The Registry goes back to the beginning of the 1970's and contains patient clinical data including aspects of the diagnosis, first line treatments, HSCT or cell therapy associated procedures, complications and outcome.
The population covered are patients who have undergone an haematopoietic stem cell transplantation (HSCT) procedure; patients with bone marrow failures receiving immunosuppressive therapies; and patients receiving non-haematopoietic cell therapies. Patients are followed up indefinitely.
The data is reported by centres performing any of the above treatments. There are no restrictions regarding centres that can report data except those required by the law on patient consent and data confidentiality.
The purpose of the Registry is to provide a pool of data to perform retrospective studies, assess epidemiological trends, or prepare prospective trials. These are all performed under the supervision of the EBMT Working parties and the EBMT assumes that centres providing data give their consent for data to be used in this way. Anybody who wants to run studies using the EBMT Registry should submit a preliminary protocol to the Working Party that is more suited to the study objectives.
For more detailed information on the Registry, click here to see the "EBMT Registry Function" document. For information on how to use the Registry to conduct studies, click here to see the "Guidelines for Registry Studies" document. Both documents are mandatory reading for all those wishing to access data in the Registry.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MED-AB FORMS MANUAL
A Guide to the completion of the EBMT HSCT Med-AB Forms
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☐ MYELODYSPLASTIC SYNDROME VMDS

(WHO)Subclassification

Peripheral blood and bone marrow findings in myelodysplastic syndromes (MDS)

Disease	Blood findings	Bone marrow findings
Refractory cytopenias with unilineage dysplasia (RCUD); Refractory anaemia (RA); Refractory neutropenia (RN); Refractory thrombocytopenia (RT)	Unicytopenia or bicytopenia[1] No or rare blasts (<1%)	Unilineage dysplasia: ≥10% of cells in one lineage <5 blasts <15% of erythroid precursors are ring sideroblasts
Refractory anaemia with ring sideroblasts (RARS)	Anaemia No blasts	≥15% erythroid precursors are ring sideroblasts Erythroid dysplasia only <5% blasts
MDS associated with isolated del(5q)	Anaemia Usually normal or increased platelet count No or rare blasts (<1%)	Normal to increased megakaryocytes with hypolobated nuclei <5% blasts Isolated del(5q) cytogenetic abnormality No Auer rods
Refractory cytopenia with multilineage dysplasia (RCMD)	Cytopenia(s) No or rare blasts (<1%)[2] No Auer rods <1x10^9/L monocytes	Dysplasia in ≥10% of the cells in ≥ two myeloid lineages (neutrophil and/or erythroid precursors and/or megakaryocytes) <5% blasts in marrow No Auer rods ±15% ring sideroblasts
Refractory anaemia with excess blasts-1 (RAEB-1)	Cytopenia(s) <5% blasts[2] No Auer rods <1x10^9/L monocytes	Unilineage or multilineage dysplasia 5-9% blasts No Auer rods
Refractory anaemia with excess blasts-2 (RAEB-2)	Cytopenia(s) 5-19% blasts Auer rods ±[3] <1x10^9/L monocytes	
Myelodysplastic syndrome – unclassified (MDS-U)	Cytopenias ≤1% blasts	Unequivocal dyslasia in less than 10% of cells in one or more myeloid cell lines when accompanied by a cytogenetic abnormality considered as presumptive evidence for a diagnosis of MDS <5% blasts

Bicytopenia may occasionally be observed. Cases with pancytopenia should be classified as MDS-U.

[2] If the marrow myeloblast percentage is <5% but there are 2-4% myeloblasts in the blood, the diagnostic classification is RAEB 1.

Cases of RCUD and RCMD with 1% myeloblasts in the blood should be classified as MDS, U.

[3] Cases with Auer rods and <5% myeloblasts in the blood and <10% in the marrow should be classified as RAEB 2.

Subclassification and status of disease at HSCT

- Subclassifications at HSCT

  - WHO subclassification

- Disease status at HSCT

-**Primary refractory phase of disease**
Treatment with intent to achieve remission was given, but no remission was achieved
-**Complete remission (CR)**
Complete remission was achieved: marrow blast count below 5% and a normalisation of peripheral blood counts for at least 4 weeks
Indicate the number of this CR
-**Improvement but no CR**
Bone marrow blasts decreased by >=50% over pretreatment but still > 5%
All CR criteria if abnormal before treatment
-**Relapse**
At least one complete remission was achieved with a previous treatment but the patient has relapsed since then.
Indicate the number of this relapse
-**Progression/worse**
More blasts in BM than before treatment
-**Never treated** (Supportive care or treatment without chemotherapy)
No treatment was given (blood transfusions are not considered treatment in this context)

First line therapy or pre-HSCT treatment

- The first line therapy or pre-HSCT treatment for MDS
  - none
- The chemotherapy/radiotherapy for LCH
  - Not reported

Complications after HSCT treatment

- Complications at 100 days
  - Acute GvHD? - No
  - Chronic GvHD? - No
  - Infection related complications? - No
  - Non-infection related complications? -NO

Complications after HSCT (cont.)

- Yearly follow-up complications
  - Acute GvHD
  - Chronic GvHD
  - Infections
  - Idiopatic pneumonia syndrome (IPS)
  - Acute Respiratory Distress Syndrom (ARDS)
  - Renal failure with dialysis

Complications after HSCT (cont.)

- Yearly follow-up complications (cont.)
  - Femoral caput necrosis
  - Crohn’s disease
  - Decubitus
  - Epstein Barr virus/post transplant lymphoproliferative disease (EBV-PLTD)
  - Hemophagocytic lymphohistiocytosis (HLH)

Relapse

- Relapse of the disease after HSCT treatment
  - Relapse of MDS?
  - Relapse of LCH?

Additional disease treatment

- Chemotherapy treatment after HSCT:
  - Puri-nethol/Mercaptapur6
  - Methotrexate
  - MabThera/Rituximab
  - Methylprednisolone

Recommendations

- Read the patient’s journal in details
- Clarify with your principal investigator or physician
- Always refer to the «Data management page & sections»
- Be updated for changes in «data management page & sections»
- Attend data entry training courses and educational sessions offered by the EBMT
- Ask for help or assistance from the registry helpdesk