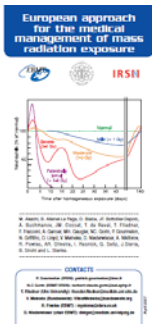
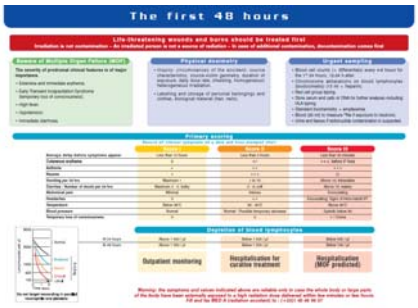
EBMT 2017
European Society for Blood and Marrow Transplantation

EBMT Pocket Guide





最初の48時間の留意点

- MOF所見がないか注意する
- 被ばく量推定
- 迅速なサンプリング

The first 48 hours

Life-threatening wounds and burns should be treated first
Irradiation is not contamination – An irradiated person is not a source of radiation – In case of additional contamination, decontamination comes first

Beware of Multiple Organ Failure (MOF)

The severity of prodromal clinical features is of major importance.

- Extensive and immediate erythema.
- Early Transient Incapacitation Syndrome (temporary loss of consciousness).
- High fever.
- Hypotension.
- Immediate diarrhoea.

Physical dosimetry

- Inquiry: circumstances of the accident, source characteristics, source-victim geometry, duration of exposure, daily dose rate, shielding, homogeneous/ heterogeneous irradiation.
- Labelling and storage of personal belongings and clothes, biological material (hair, nails).

Urgent sampling

- Blood cell counts (+ differentials) every 4-8 hours for the 1st 24 hours, 12-24 h after.
- Chromosome abberations on blood lymphocytes (biodosimetry) (15 ml + heparin).
- Red cell group typing.
- Store serum and cells or DNA for further analyses including HLA typing.
- Standard biochemistry + amylasemia.
- Blood (20 ml) to measure ^{24}Na if exposure to neutrons.
- Urine and faeces if radionuclide contamination is suspected.

MOF所見に注意する

- 急性発症の顕著な紅斑
- 意識障害
- 高熱
- 血圧低下
- 急性発症の下痢

Beware of Multiple Organ Failure (MOF)

The severity of prodromal clinical features is of major importance.

- Extensive and immediate erythema.
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被ばく量推定

- 問診
  - 被ばく状況、被ばく源
  - 被ばく源の性質
  - 被害者の位置関係
  - 被ばく期間
  - 一日被ばく線量
  - 遮蔽の有無
- 患者の所有物、爪、毛髪の保存

Physical dosimetry

- Inquiry: circumstances of the accident, source characteristics, source-victim geometry, duration of exposure, daily dose rate, shielding, homogeneous/ heterogeneous irradiation.
- Labelling and storage of personal belongings and clothes, biological material (hair, nails).

迅速なサンプリング（EBMT)

Urgent sampling

- Blood cell counts (+ differentials) every 4-8 hours for the 1st 24 hours, 12-24 h after.
- Chromosome abberations on blood lymphocytes (biodosimetry) (15 ml + heparin).
- Red cell group typing.
- Store serum and cells or DNA for further analyses including HLA typing.
- Standard biochemistry + amylasemia.
- Blood (20 ml) to measure ^{24}Na if exposure to neutrons.
- Urine and faeces if radionuclide contamination is suspected.

- 血算（分画）
  - 最初24時間は4-8時間毎、以降12-24時間毎
- 血液リンパ球の染色体異常（生物学的線量測定）
- 血液型
- 血清、細胞、DNA保存,HLA typing.
- アミラーゼを含む生化学検査
- 中性子被ばくの時は^{24}Na
- 内部被ばくが疑われる場合は、尿・便を保存する

初期重症度決定資料

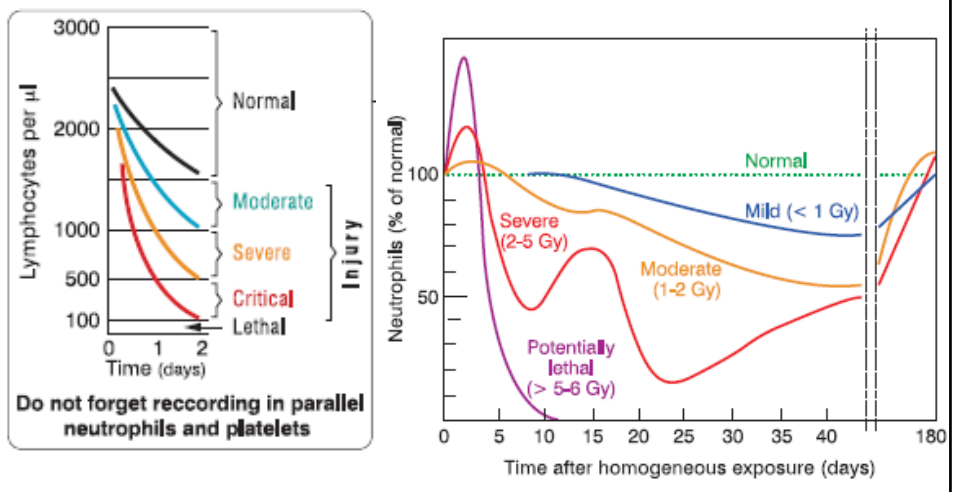
リンパ球数と重症度(EBMT)

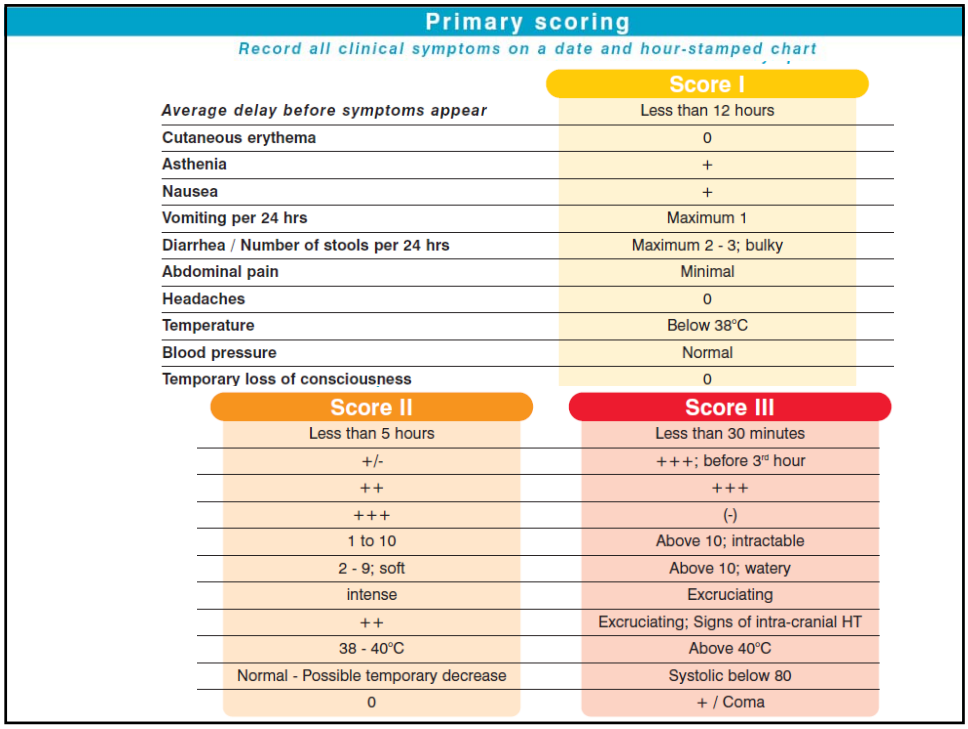
Primary scoring

Record all clinical symptoms on a date and hour-stamped chart

	Score I	Score II	Score III
	Depletion of blood lymphocytes		
At 24 hours	Above 1 500 / µl	Below 1 500 / µl	Below 500 / µl
At 48 hours	Above 1 500 / µl	Below 1 500 / µl	Below 100 / µl
	Outpatient monitoring	**Hospitalisation for curative treatment**	**Hospitalisation (MOF predicted)**

Warning: the symptoms and values indicated above are reliable only in case the whole body or large parts of the body have been externally exposed to a high radiation dose delivered within few minutes or few hours. Fill and fax MED A (radiation accident) to : (+33)1 40 46 96 07





Primary scoring

Record all clinical symptoms on a date and hour-stamped chart

	Score I	Score II	Score III
Average delay before symptoms appear	Less than 12 hours	Less than 5 hours	Less than 30 minutes
Cutaneous erythema	0	+/-	+++; before 3rd hour
Asthenia	+	++	+++
Nausea	+	+++	(-)
Vomiting per 24 hrs	Maximum 1	1 to 10	Above 10; intractable
Diarrhea / Number of stools per 24 hrs	Maximum 2 - 3; bulky	2 - 9; soft	Above 10; watery
Abdominal pain	Minimal	intense	Excruciating
Headaches	0	++	Excruciating; Signs of intra-cranial HT
Temperature	Below 38°C	38 - 40°C	Above 40°C
Blood pressure	Normal	Normal - Possible temporary decrease	Systolic below 80
Temporary loss of consciousness	0	0	+ / Coma

重症度決定資料（EBMT）

		Primary scoring		
		Score1	Score2	Score3
症状が出るまでの平均的時間		12時間以内	5時間以内	30分以内
紅斑		0	+/-	+++
衰弱		+	++	3時間以内+++
嘔気		+	+++	(-)
一日の嘔吐回数		最大で1回	1 to 10	10回以上頑固
一日の下痢回数		最大2 - 3; 形あり	2 - 9; 軟便	10回以上; 水様
腹痛		軽度	強度	激痛
頭痛		0	++	激痛; 脳圧亢進
体温		38°C未満	38 - 40°C	40°C以上
血圧		正常	正常または一過性低下	80未満
一過性意識消失		0		0+ / Coma
Depletion of blood lymphocytes	24h	Above 1 500 / µl	Below 1 500 / µl	Below 500 / µl
	48h	Above 1 500 / µl	Below 1 500 / µl	Below 100 / µl
注意： 本表は全身が被ばくした場合に使用		Outpatient	monitoring Hospitalisation for curative treatment	Hospitalisation (MOF predicted)

48時間以降

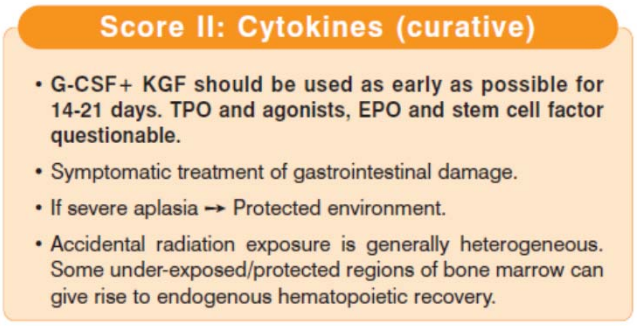
Beyond the first 48 hrs, a second patient scoring is done by organs (Neurovascular, Hemopoesis, Cutaneous, Gut) according to the METREPOL document for therapeutical management and Multiple Organ Failure (MOF) prediction.**

- 48時間以降は、2回目のScoringをMETREPOLの神経、血液、皮膚、消化管の臓器評価に従って行い、治療とMOF予測を行う。

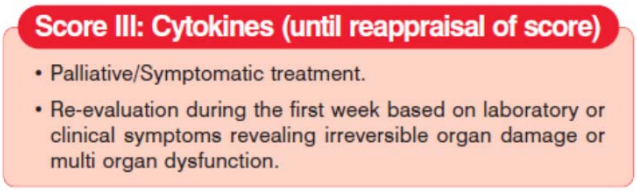
Score I: Monitoring. No cytokine

- Outpatient clinical monitoring.
- Blood count: - every day for 6 days,
  - then once a week for 2 months.

- 外来フォロー
- 最初の6日は毎日血算を行う
- その後は毎月1回で6週まで



- G-CSF+KGFを14-21日可及的速やかに使用
- TPO,EPO,SCFについてのエヴィデンスは不明
- 胃腸障害の治療
- 骨髄不全であれば防護環境（無菌治療室）
- 放射線事故による被ばくは一般にheterogeneous
- 被ばくされていない骨髄からの自力回復が可能



- 緩和的治療、対症療法
- 最初の１週で検査結果、臨床症状で再評価を行い、回復不可能な臓器障害、多臓器不全かどうかを明らかにする

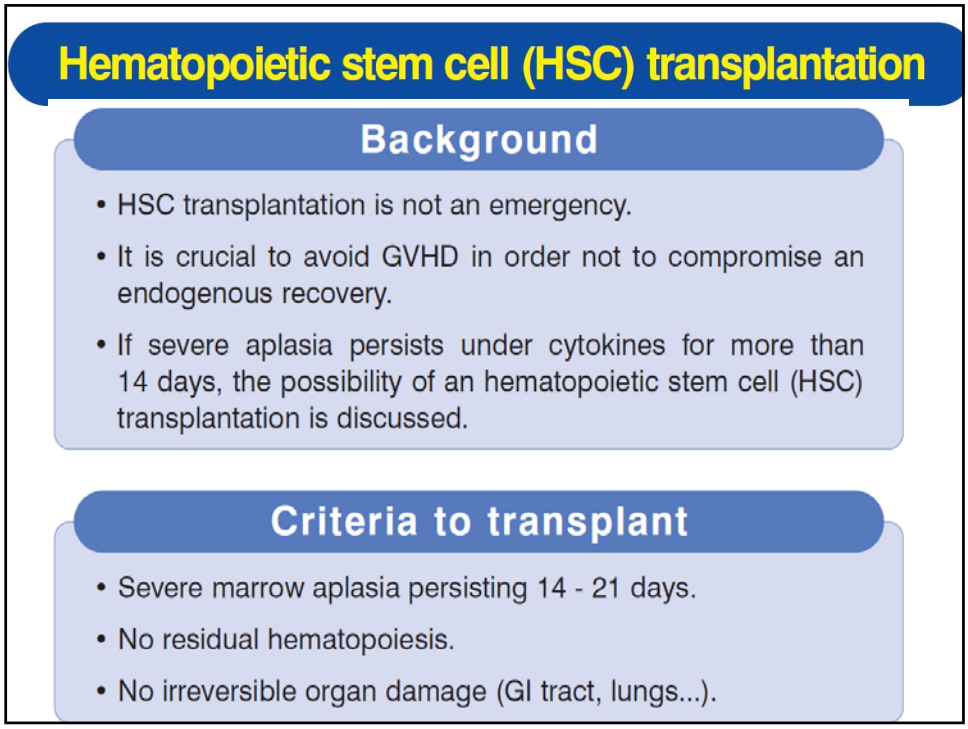
All blood products should be irradiated

Severe radiological skin lesions have a peculiar torpid evolution and require specialist treatment.

- すべての血液製剤は照射すること
- 重症の皮膚障害は進行が遅く、専門医による治療を必要とする

Hematopoietic stem cell (HSC) transplantation

造血幹細胞移植



背景・適応

- 造血幹細胞移植は緊急に行うものでない
- 自己造血の回復を複雑にするGVHDを避けることが重要である
- 14日間以上のG-CSF投与しても重症の造血不全が続く場合に、造血幹細胞移植の適応について考える
- 自己造血の回復の見込みがないこと
- 不可逆的な臓器不全がないこと

Graft

- **Type of graft:**
  - Bone marrow.
  - Peripheral blood HSC (depleted or not).
  - Cord blood.
- **Donor in the following order of priority:**
  - HLA-identical sibling or 7/8 matched.
  - HLA-identical unrelated donor or 9/10 matched.
  - Cord blood > 4/6 matched.
- **Doses of cells to be grafted:**

  At least: - $2x10^6$ CD34 cells.kg^{-1} (peripheral blood);
  - $2x10^8$ nucleated cells.kg^{-1} (bone marrow);
  - $3x10^7$ nucleated cells (cord blood).

Conditioning and GVHD prevention

- **Non myelotoxic conditioning:**
  - Fludarabine (30 mg.m^{-2}.d^{-1} for 3 days) ± anti-lymphocyte globulins.
- **GVHD prevention:**
  - No Methotrexate.

- ドナー選択、ＨＬＡ適合度、輸注細胞数については通常の移植と同じ
- 減量レジメンを使用する
- GVHD予防にMTXを使用しない