New MDS Nomenclature and changes

An update of the 1997 International Prognostic Scoring System (IPSS-R) was published in 2012 to include a refinement of cytogenetic risk grouping, a more detailed use of cytopenias and MDS with <5% blasts were separated into <= 2% and >2%- <5% blasts. There was a change in MDS nomenclature to better reflect how they are diagnosed as shown in Table 1. However, the WHO classification of MDS will still be based on morphology. MDS with multilineage dysplasia and ring sideroblasts will be reinstated. Molecular mutations will increasingly impact diagnosis and prognosis. MDS-SLD with pancytopenia or 1% circulating blasts should be considered MDS-U only if 1% blasts are detected on at least 2 separate occasions. MDS-SLD RS and MDS-MLD with ring sideroblasts include either >= 15% ring-sideroblasts or any ring sideroblasts and SF3B1 mutation. MDS with isolated del(5q) allows one additional but non-chromosome 7 abnormality and recommends testing for TP53 mutation since it confers poor prognosis to del (5q) patients treated with lenalidomide. In addition to that, cases with significant granulocytic dysplasia are now excluded from a MDS del (5q) classification.Refractory anemia with ring sideroblasts associated with marked thrombocytosis (RARS-T) has now moved from a provisional to a full entity. However, it must be a de novo diagnosis, not evolved from MDS with RS. RARS-T also involves a common co-mutation of JAK2 and SF3B1.

Table 1: New MDS Nomenclature and changes

WHO MDS Nomenclature 2008	New WHO MDS Nomenclature 2016	Blood	Marrow	Changes
No excess blasts				
Refractory Anemia with rings sideroblasts **(RARS)**	MDS with single lineage dysplasia and ring sideroblasts **(MDS- RS)**	Anemia No blasts No Auer rods. <1 G/L monocytes	Dyserythropoiesis only < 5% Blasts >15% Ring sideroblasts	Includes either >=15% ring-sideroblasts **or** >5% ring-sideroblasts and SF3B1 mutation
Refractory cytopenia with unilineage dysplasia **(RCUD-A,N,T)**	MDS with single lineage dysplasia **(MDS-SLD)**	Anemia or bicytopenia No Blasts No Auer rods. <1 G/L monocytes	only one cell lineage with dysplasia in >10% <5%Blasts <15% Ring sideroblasts	

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GWT-TUD GmbH
Medical Consulting

Managing Director: Claus-Peter Held

Address:

GWT-TUD GmbH
Blasewitzer Straße 43,
01307 Dresden
Germany

Phone:
+49.351.25933-100

Email:
contact@gwtonline.de

https://www.gwtonline.de

registered at Dresden District Court (Amtsgericht)
HRB 13 840
VAT-ID DE 182 302 853

WHO MDS Nomenclature 2008	New WHO MDS Nomenclature 2016	Blood	Marrow	Changes
No excess blasts				
Refractory cytopenia with multi-lineage dysplasia with or without ring sideroblasts **(RCMD)**	MDS with multilineage dysplasia **(MDS-MLD)**	Cytopenia (s) = 1% blasts No Auer rods. <1 G/L monocytes	Dysplasia in >10% of the cells of at least 2 cell lines < 5% Blasts, no Auer rods. < 15% ring sideroblasts (without RS)	
	MDS with multilineage dysplasia with ringsideroblasts **(MDS-MLD RS)**	Cytopenia (s) = 1% blasts No Auer rods. <1 G/L monocytes	Dysplasia in >10% of the cells of at least 2 cell lines < 5% Blasts, no Auer rods. >= 15% ring sideroblasts (with RS)	**MDS-MLD** with RS include either >=15% ring-sideroblasts **or** 5% ring-sideroblasts and SF3B1 mutation
MDS with isolated del(5q)	MDS with isolated del(5q)	Anemia, or bicytopenia Normal or elevated platelets <1% blasts	<5% blasts, no Auer rods Hypolobulated megakar-yocytes	- One additional non-chromosomal 7 abnormality al-lowed - Recommend testing for TP53 mutation - Cases with signif-icant myeloid dys-plasia excluded.
MDS, unclassifiable (MDS-U)	MDS, unclassifiable (MDS-U) 3 types a) MDS SLD with pancytopenia b) MDS SLD or MDS with 1% peripheral blasts c) MDS without clear dysplasia but but cytogenetic abnormal-ity considered as presumptive for MDS	Cytopenia	< 5% blasts	

WHO MDS Nomenclature 2008	New WHO MDS Nomenclature 2016	Blood	Marrow	Changes
Excess blasts				
Refractory anemia with excess blasts **(RAEB1)**	MDS with excess blasts **(MDS-EB1)**	Cytopenia (s) <5% Blasts No auer rods <1 G/L Monocytes	Unilineage or multilineage dysplasia No Auer rods 5-9% Blasts	- Now include most cases previ-ously classified as acute erythroid leukemia - Categorized as MDS-EB1 or MDS-EB2 based on blast % of total marrow cells
Refractory anemia with excess blasts **(RAEB2)**	MDS with excess blasts **(MDS-EB2)**	Cytopenia(s) 5-19% blasts Auer rods possible <1 G/L Monocytes	Unilineage or multilineage dysplasia Auer rods possible 10-19% Blasts	
	Refractory anemia with ring sideroblasts associated with marked thrombocytosis **(RARS-T)**	Anemia No blasts ≥450 x 109/L platelets	Erythroid dysplasia only <5% blasts ≥15% ring sideroblasts proliferation of large megakaryocytes	- Moved from a provisional to a full entity - Must be a de novo diagnosis, not evolved from MDS with RS - Common co-mutation of JAK2 and SF3B1