Myelodysplastische Syndrome

Mikroskopierkurs am Hofgarten

12.11.2024

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Unter der Schirmherrschaft der

DGHO
DEUTSCHE GESELLSCHAFT FÜR
HÄMATOLOGIE UND MEDIZINISCHE ONKOLOGIE

Verbund Katholischer Kliniken Düsseldorf VKKD

UKE
HAMBURG

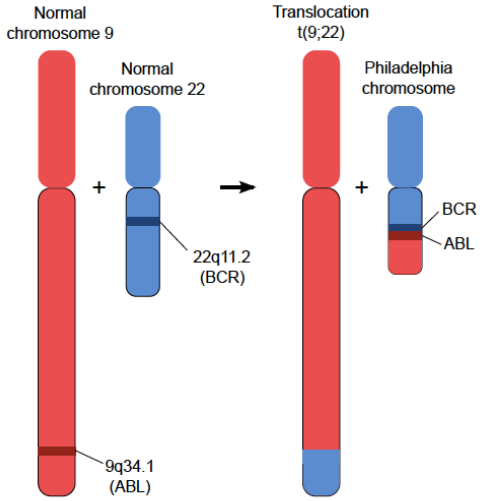
Malteser
…weil Nähe zählt.

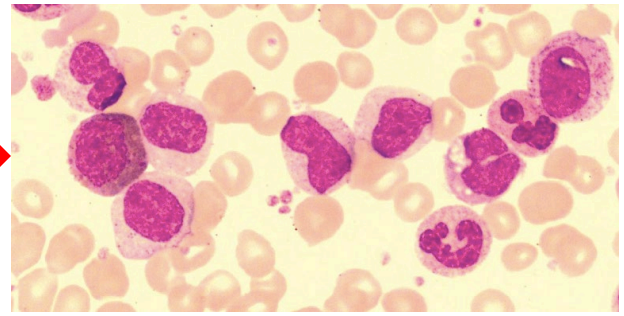
eLCH Curriculum Hämatologie e-Learning

Digitale Mikroskopierplattform der DGHO
Nächster Virtueller Mikroskopierkurs: 18.12.2024

meistens

Was ein MDS NICHT ist… Phänotyp-Genotyp-Korrelation





- Monoallelische Erkrankung: CML
- Bis auf wenige Ausnahmen (5q-, SF3B1-Mutation) keine eindeutige Genotyp-Phänotyp-Korrelation oder mutationsgerichtete Therapie beim MDS

MDS-Diagnosekriterien

- **Ohne periphere Zytopenie kein MDS!**
  (Hb < 10g/dl,
  Thromobzyten < 100/nl,
  Neutrophile < 1,0/nl)

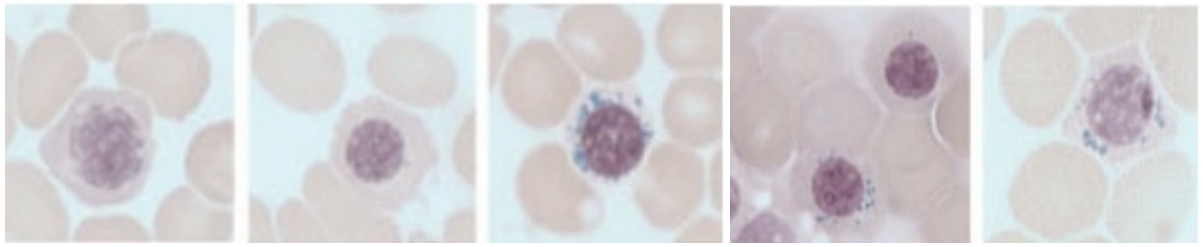
- **Dysplasien?**
  (wenn ja, in wie vielen Linien?)
- **Blastenanteil in % ?**
  (exakt!)

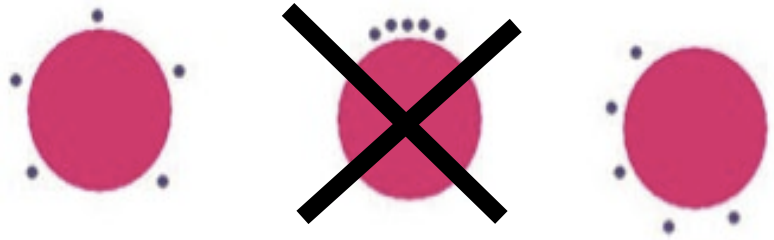
MDS-Klassifikation

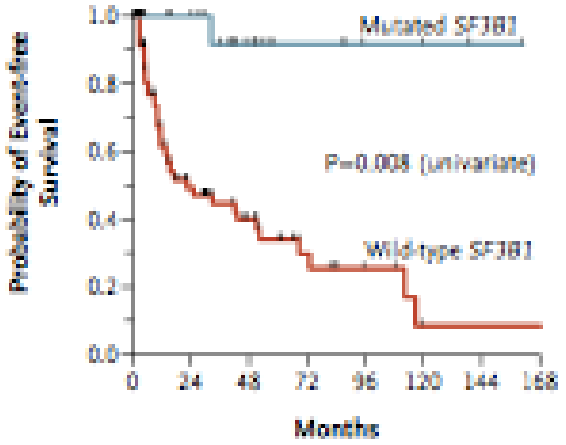
MDS with defining genetic abnormalities	Blasts
MDS with low blasts and isolated 5q deletion (MDS-5q)	<5% BM and <2% PB
MDS with low blasts and *SF3B1* mutation (MDS-*SF3B1*)	<5% BM and <2% PB
MDS with biallelic *TP53* inactivation (MDS-biTP53)	<20% BM and PB
MDS, morphologically defined	
MDS with low blasts (MDS-LB)	
MDS, hypoplastic (MDS-h)	
MDS with increased blasts (MDS-IB)	
MDS-IB1	5%–9% BM or 2%–4% PB
MDS-IB2	10%–19% BM; or 2%–19% PB
MDS with fibrosis	5%–19% BM; or 2%–19% PB

Khoury/Solary et al. Leukemia 2022

Ringsideroblasten







Fast immer assoziiert mit SF3B1-Mutationen

Riskoklassifikation nach IPSS-R

	IPSS-R risk	IPSS-R score	Median OS, years	Median time to 25% AML transformation, years
Lower-risk	Very low	≤1.5	8.8	NR
	Low	>1.5–3	5.3	10.8
	Intermediate	>3–4.5	3.0	3.2
Higher-risk	High	>4.5–6	1.6	1.4
	Very high	>6	0.8	0.7

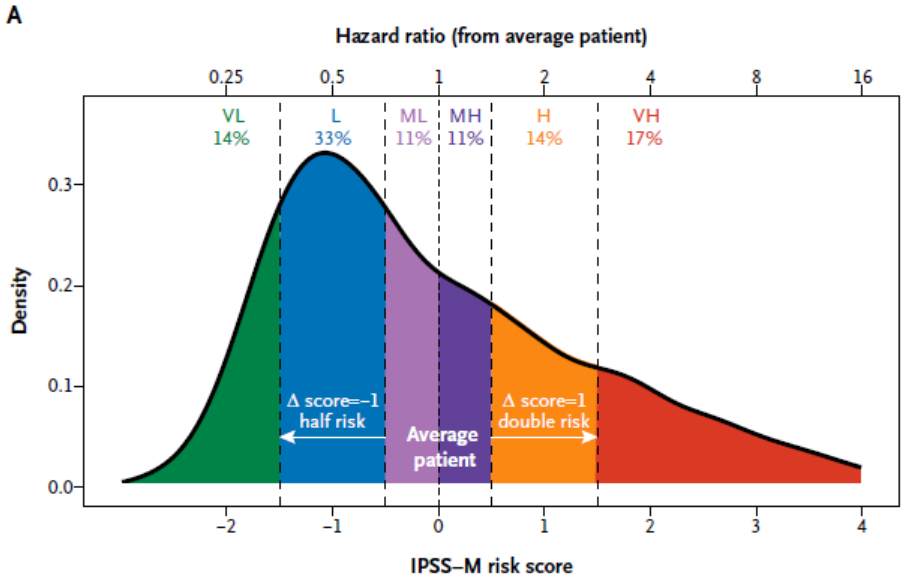
AML, acute myeloid leukaemia; IPSS-R, revised International Prognostic Scoring System; MDS, myelodysplastic syndromes; NR, not reached; OS, overall survival

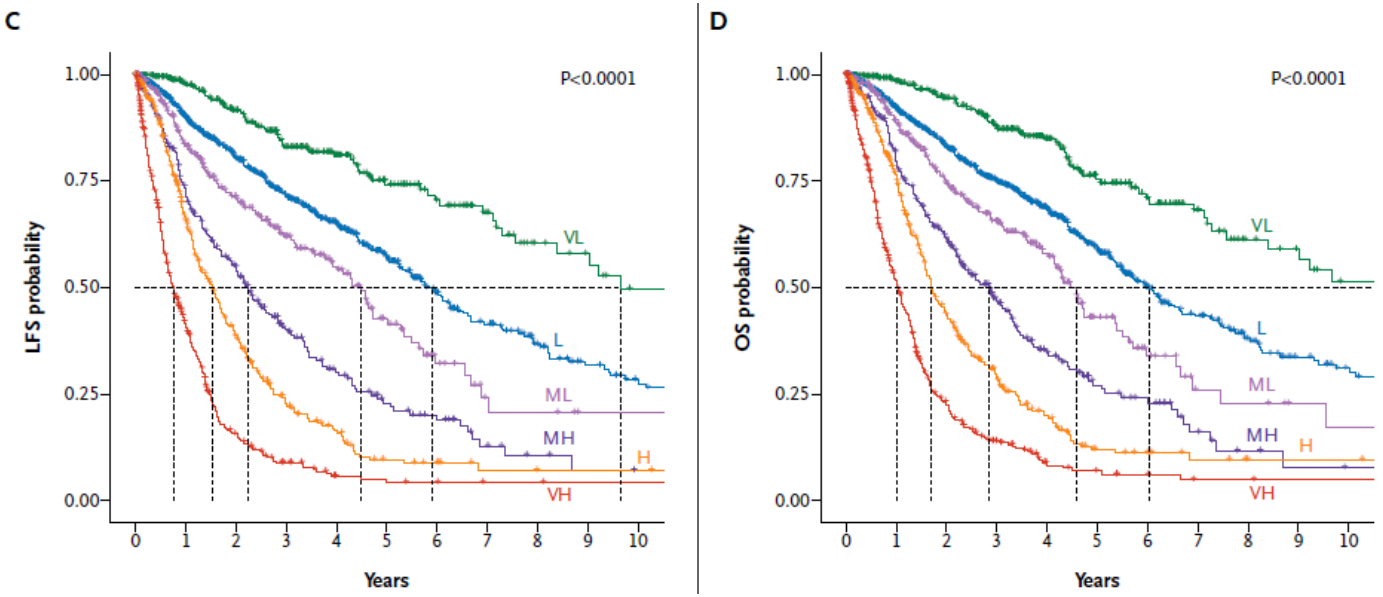
Greenberg et al. Blood 2012

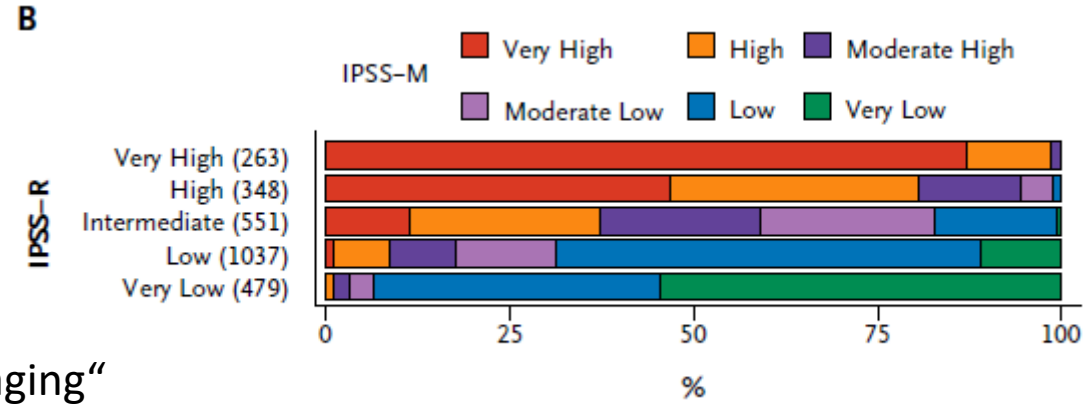
Risikoklassifikation nach IPSS-M

1. Blastenanteil pB und KM

2. Molekulares Risikoprofil







- Reklassifikation von 46% der Pt, davon 75% „Upstaging“
- Ggf. Therapieindizierend (alloSZT, HMA…)

Bernard et al, NEJM Evid 2022

MDS-Pathophysiologische Konzepte