

MPN Symptome und Diagnostik

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MPN gemäss WHO Definition

Table 1. Myeloproliferative neoplasms.

Chronic myeloid leukaemia

Polycythaemia vera

Essential thrombocythaemia

Primary myelofibrosis

Chronic neutrophilic leukaemia

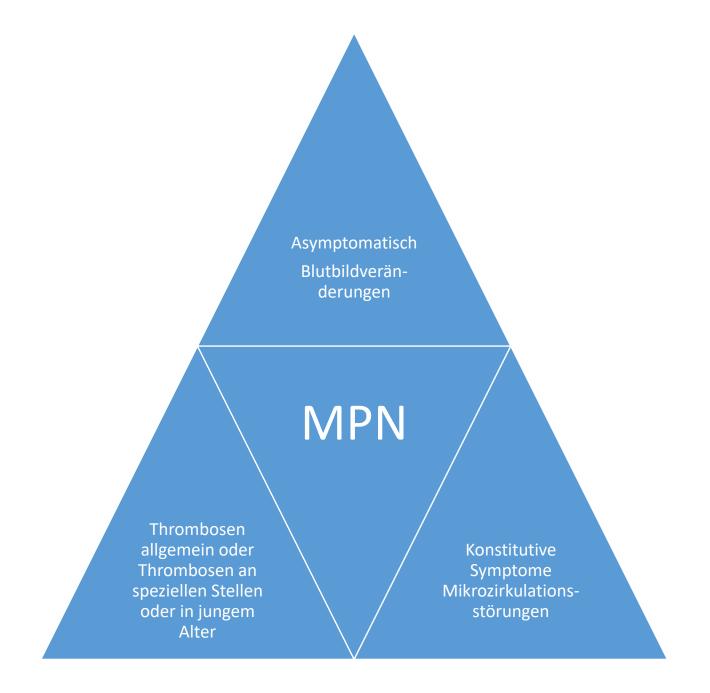
Chronic eosinophilic leukaemia

Juvenile myelomonocytic leukaemia

Myeloproliferative neoplasm, not otherwise specified

Leukemia (2022) 36:1703 – 1719







Asymptomatisch

Was sind die Laborwerte die auf MPN hinweisen?

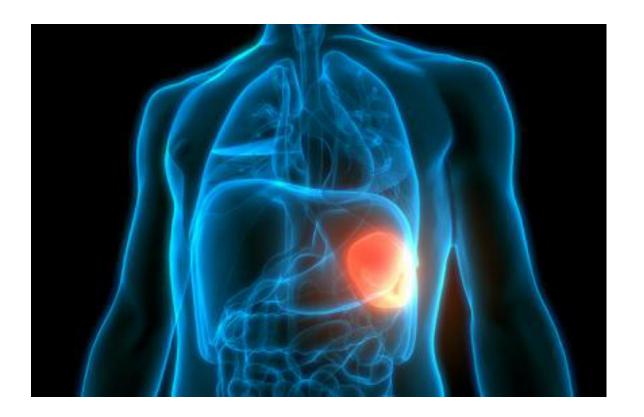
- Polyzythämia vera:
 - Hb: ♂>165 g/l/Hkt >49%; ♀>160 g/l/Hkt >48%
- Essentielle Thrombozythämie
 - Thrombozyten >450 G/I



Asymptomatisch/Symptomatisch

• Weitere Befunde, die nebenbei gefunden werden können

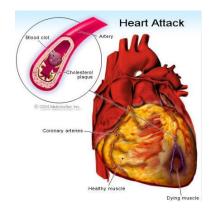
Grosse Milz

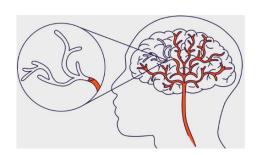




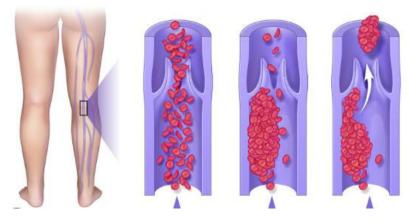
Symptomatisch

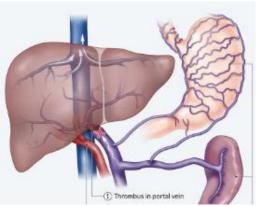
Thrombose in Arterie





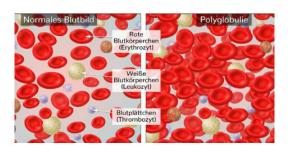
Thrombose in Vene







Symptomatisch: Mikrozirkulation



Kopfschmerzen



• Juckreiz/Brennen: verstärkt durch Wärme (Duschen)



Sehstörungen





Symptomatisch: Entzündung/Botenstoffe

Gewichtsverlust



Nachtschweiss



Fatigue/Müdigkeit/Konzentration





Diagnose

Diagnosis

unlikely

Diagnostic algorithm for myeloproliferative neoplasms Polycythemia vera Essential thrombocythemia Primary myelofibrosis suspected suspected suspected Hb >16.5 g/dL Men Platelets ≥450 x 10(9)/L Anemia Hb >16 g/dL Women Splenomegaly Leukoerythroblastosis Blood JAK2V617F/CALR/MPL Blood JAK2 mutation mutation screening (RT-PCR) Bone marrow biopsy Screening (RT-PCR) with mutation screening (V617F and exon 12) and cytogenetics JAK2 60% CALR Negative Positive 22% Negative Positive JAK2 TN ET likely but ET still 10% a possibility not certain MPL 8% Diagnosis likely "Triple-negative" JAK2V617F 57% CALR 20%

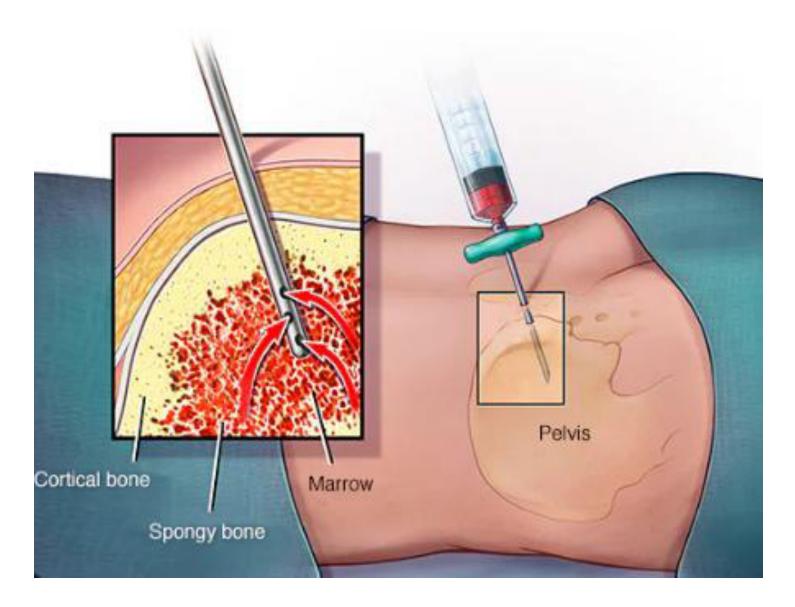
MPL 3%

Diagnosis considered If bone marrow

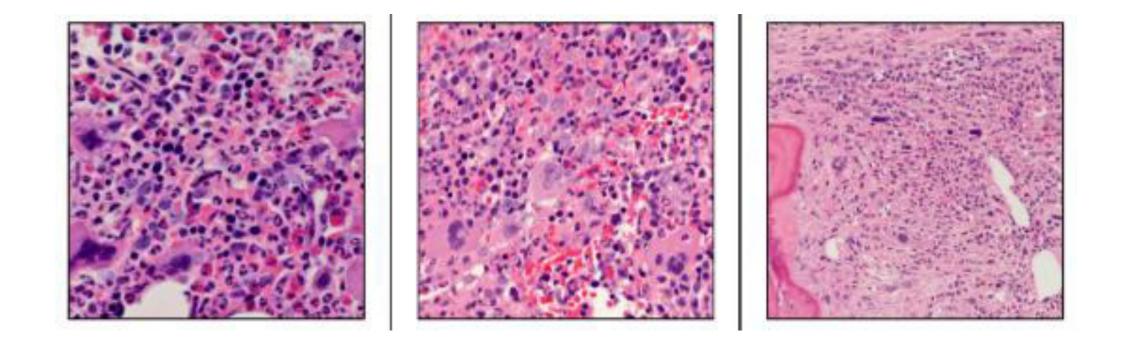
trisomy 9 or del(13q) present or
Other myeloid malignancies are excluded

morphology is consistent with PMF and 1. JAK2 CALR or MPL mutated or

Spital Thurgau









Bildgebung: Sonographie oder CT/MRI





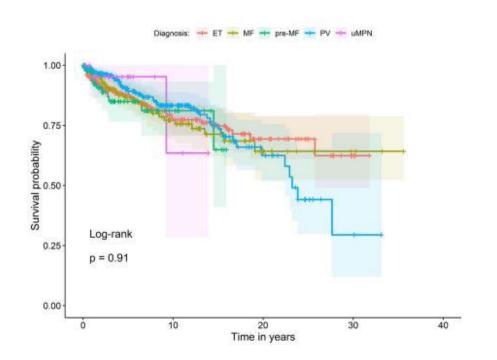
Komplikationen

- Verhinderbare Komplikationen:
 - Thrombosen
 - Blutungen

- Zielwerte, die erreicht werden müssen:
 - Polyzythämia vera: Hkt<45%
- Therapien
 - Polyzythämie vera: Aspirin
 - Weiteres: sh. Referat Dr. Lehmann

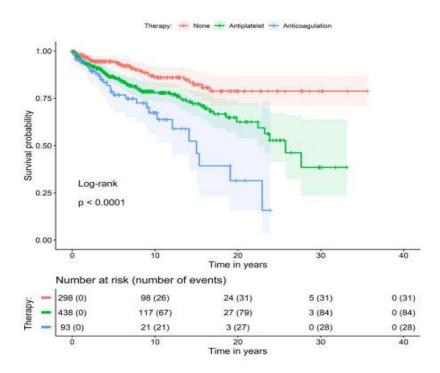


Blutungen?



	Bleeding events (n = 143)	Minor bleeding events (n = 96)	Major bleeding events (n = 47)
Localization n (%)			
Mucocutaneous bleedings	56 (39.2)	52 (54.2)	4 (8.5)
Postoperative bleeding	22 (15.4)	21 (21.9)	1 (2.1)
Epistaxis	21 (14.7)	8 (8.3)	13 (27.7)
Gastrointestinal bleeding excluding esophageal variceal bleeding	17 (11.9)	-	17 (36.2)
Other bleedings ^a	13 (9.1)	7 (7.3)	6 (12.8)
Menorrhagia	8 (5.6)	8 (8.3)	_
CNS bleeding	3 (2.1)	-	3 (6.4)
Esophageal variceal bleeding	3 (2.1)	-	3 (6.4)

aMajor bleedings: intraoperative rupture of spleen (n = 1), macrohematuria (n = 1), spontaneous bleeding into musculus ileopsoas (n = 1), subcapsular intrasplenic hemorrhage (n = 1), bleeding of a corpus luteum cyst (n = 1), gastrointestinal bleeding of unknown localisation (n = 1) minor bleedings: hemorrhagic cystitis (n = 1), hemorrhoidal bleeding (n = 1), hemorrhoidal bleeding (n = 1), witreous bleeding (n = 1), gingival bleeding (n = 2), intraabdominal bleeding after TIPSS (n = 1)



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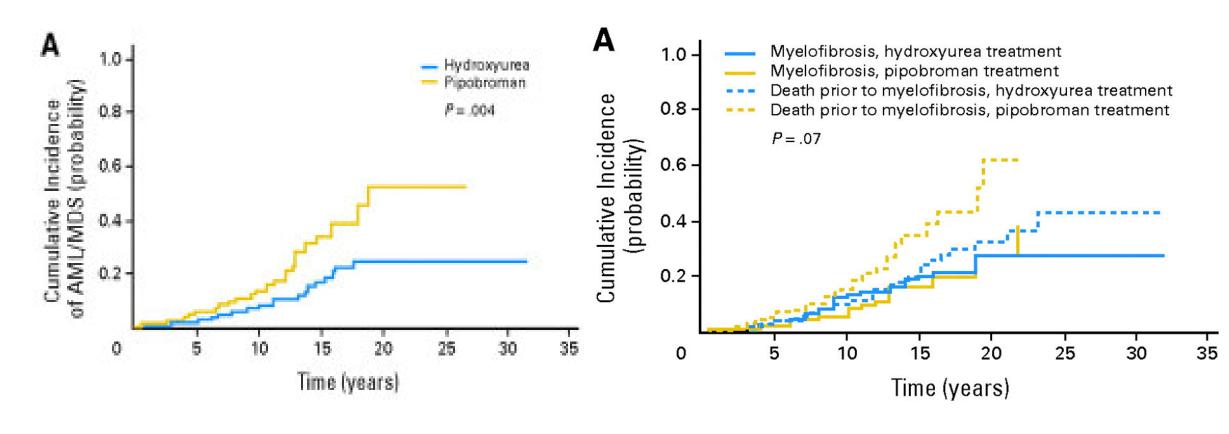


Blutungen

- Risiko hoch bei hohen Thrombozyten > 1000 G/I
- Wachsam sein va auch vor Operationen
 - Thrombozyten situativ senken
 - Stop der Blutverdünnung in Abhängigkeit der Vorgeschichte und der Grösse des Eingriffes
 - Lokal kann auch Tranexamsäure angewendet werden



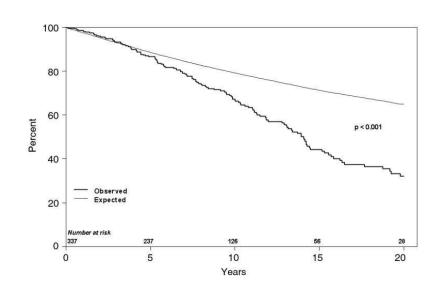
Langzeitrisiko für AML oder sek MF





Konsequenzen der Diagnose P. vera

- Überleben
- Beeinflussbare Folgen:
 - Arterielle Thrombosen
 - Venöse Thrombosen
 - Blutungen
- Nicht beeinflussbare Folgen: (?)
 - Sekundäre Myelofibrose (post PV Myelofibrose)
 - Akute myeloische Leukämie





Diskrepante Ziele Patient-Arzt

- Arzt
 - Blutwerte (Hkt)
 - Jak2 VAF
 - Usw.

- Patient
 - Symptome
 - Lebensqualität
 - Unabhängigkeit
 - USW.

