What’s Routine?
General Medical Care in Patients with MPNs

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Joyce Niblack Memorial
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Communication

- Information about Patient
- Hematologist
- Primary Care Doctor
- Information about disease
Today’s Goals

• Review unique risks faced by MPN patients
• Review targets for optimizing
  – Optimizing cardiac health
  – Minimizing infection risks
  – Limiting other cancers
• Discuss strategies for increasing provider-to-provider communication
Overall Risks

Risk of Infections: Bacterial, Viral, Fungal

Risk of Bleeding

Risk of Heart Attack

\(^1\)Hultcrantz M et al., JCO Vol 33; No 20; July 2015
Overall Risks

Risk of Blood clots in the legs, lung, liver and skull veins

Risk for arterial clots that cause stroke
Control what you can

“You’re fifty-seven years old. I’d like to get that down a bit.”
Infections

• Large studies show increased rate of fatal infections

• Why might that be?
  – Disease → irregularities in immune system
  – Low blood counts: Neutropenia
  – Splenic dysfunction
  – Treatment with JAK inhibitors
Neutropenia

• Determination
  – Blood smear examined
  – Percentage neutrophils among total white blood cells calculated

• Treatment
  – If asymptomatic, can watch
  – If repeated infections, then prophylaxis recommended
Preventing Infection

• Prophylaxis only in selected patients

  NCCN: “antibiotic prophylaxis for recurrent infections is recommended”

  – Bacterial: Fluoroquinolone antibiotic
  – Viral: Acyclovir or other
  – Fungal: Fluconazole, Posaconazole, Voriconazole

• Vaccination

  – Influenza vaccine to patient and family
  – If immunocompromised (i.e. no spleen, on JAK inhibitor)
    • Avoid live-attenuated virus vaccination, i.e. no Zoster vaccination
    • 7-days contact avoidance from family members
Infections related to treatment

• Most common:
  – Bacterial infections of respiratory and urogenital tract
  – Frequency decreases with length on therapy
  – Severe infections no more common than patients on placebo

• Likely increased risk for rare infections
  – For example, tuberculosis reactivation
  – Shingles (Herpes zoster)
Figure 2. Cumulative incidence of infectious complications according to IPSS and splenomegaly.
Care while on therapy

- Avoid starting therapy while actively infected
- Ensure no tuberculosis risk prior to starting treatment
- Watch for signs/symptoms of zoster
- Rule out Hep B prior to starting therapy
- No routine recommendations for prophylaxis against infections
Arterial Clotting Events
Younger Individuals

Cause-Specific Mortality Rates when Aged 18-49 at diagnosis

- Cardiovascular Disease: MPN 8.8, Control 1.0
- Strokes: MPN 8.7, Control 1.0
- Solid Tumor: MPN 2.5, Control 1.0

Data from Hultcrantz M et al., JCO Vol 33; No 20; July 2015
Arterial or venous blood clotting complications

### At diagnosis
- ET: ~13%
- PV: 34-39%
- MF: 10-29%

### Follow-up
- ET: 8 to 31%
- PV: 8 to 19%
- MF: ~10%

Elliot, MA Seminars in Thrombosis and Hemostasis 2007; Barbui, T Blood 2010; Elliot Haematologica 2010: Slide from B Stein
Why more common?

Abnormal interaction between the blood and blood vessels

- Elevated RBCs
- Elevated WBCs
- Genetic mutations: JAK 2
Cardiac Risk factors

- Age
- Smoking
- Total Cholesterol
- "Good cholesterol"
- Diabetes
- Blood pressure
- Activity level
- Weight

60 yo male; no diabetes, smoker, total cholesterol of 210; good cholesterol of 30; blood pressure of 140/70 mm/hg

Risk of heart attack in next 10 years: 23%

If he is not a smoker, takes medication or diets to reduce cholesterol by 30 points and lowers his BP to 130/70: Risk drops to 13%
# Blood Pressure Targets

<table>
<thead>
<tr>
<th>Blood Pressure Category</th>
<th>Systolic mm Hg (upper #)</th>
<th>Diastolic mm Hg (lower #)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>less than 120</td>
<td>less than 80</td>
</tr>
<tr>
<td>Prehypertension</td>
<td>120 – 139</td>
<td>or</td>
</tr>
<tr>
<td>High Blood Pressure (Hypertension) Stage 1</td>
<td>140 – 159</td>
<td>or</td>
</tr>
<tr>
<td>High Blood Pressure (Hypertension) Stage 2</td>
<td>160 or higher</td>
<td>or</td>
</tr>
<tr>
<td>Hypertensive Crisis (Emergency care needed)</td>
<td>Higher than 180</td>
<td>Higher than 110</td>
</tr>
</tbody>
</table>

Guidelines: American Heart Association
Targets

• Complete abstinence from smoking
• Heart-healthy eating patterns
  – American Heart Association
    • Use up the calories you take in
    • Choose high-nutrient foods: Fruits and vegetables, whole grains, low-fat dairy, poultry and fish without the skin, nuts and legumes, non-tropical vegetable oils
• Keep your body-mass index between 18-24.9
  – AHA has a calculator
Cholesterol?

**Metrics**

- Total cholesterol = HDL + LDL + 20 percent of your triglyceride level.
- HDL (good) cholesterol: High is better
- LDL (bad) cholesterol: Low is better
- Targets are complicated – depend on your age, prior history, presence or absence of diabetes
- Frequency of testing: discuss with PCP

**Interventions**

- Dietary interventions
  - Fish, Omega-3 fatty acids
  - Oatmeal, oat bran, high soluble fiber
- Exercise
  - Boost HDL, decreases danger of LDL
- “Statins”
  - Lower LDL
- Nicotinic Acid “Niacin”
  - Raise HDL
- Fibrates
  - Lower triglycerides
Exercise

• Aerobic Exercise
  – Lowers cholesterol and BP
  – Increases endurance
  – Lower resting heart rate
  – Weight loss and maintenance
  – Stress relief
  – Improved sleep

• 30 min; 5-7 days a week
What about Venous Clots?

• Most common locations
  – Legs (deep venous thrombosis “DVT”)  
  – Lungs (pulmonary embolism “PE”)
• Can also occur in abdominal vasculature and brain drainage
## Treatment of PE/VTE or Other

<table>
<thead>
<tr>
<th>Protection from Blood Clots: “Prophylaxis”</th>
<th>Initial Treatment</th>
<th>Extended Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>MPN Disease Control</td>
<td></td>
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</tr>
<tr>
<td>Around the time of surgery</td>
<td>Blood Thinning medications for at least 3-6 months</td>
<td>Consider extended treatment for abdominal clots, SVT, life threatening clots, second clot</td>
</tr>
<tr>
<td></td>
<td>Usually pause aspirin while on blood thinner unless risk &gt;benefit</td>
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</tbody>
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What about bleeding?

- Can occur in patients with very high platelet count, i.e. over 1.5 million/uL
- Called acquired Von Willebrand’s disease
- Treatment
  - Avoid aspirin
  - Lower platelet count
Acquired VWD

Normal Blood Vessel

Increase in platelets
## Modifying Surgical Risk

<table>
<thead>
<tr>
<th>Planning</th>
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<tbody>
<tr>
<td></td>
<td>--Assessment by hematologist</td>
</tr>
<tr>
<td></td>
<td>--Optimize blood counts</td>
</tr>
<tr>
<td></td>
<td>--Especially platelets if splenectomy planned</td>
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<table>
<thead>
<tr>
<th>Preoperative</th>
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<tbody>
<tr>
<td></td>
<td>--Discontinue ASA</td>
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<th>Postoperative</th>
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<td></td>
<td>--Anticoagulation – LMWH</td>
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<tr>
<td></td>
<td>--Clinical vigilance re hemorrhage</td>
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<td></td>
<td>--US of abdominal veins</td>
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Other cancers?

• Slightly increased risk for solid tumors
  – Italian Study
    • Roughly 4 times as likely to acquire lymphoma-type diseases
  – Danish Study
    • Incidence of solid tumors slightly higher in patients with ET, PV and CML
  – Swedish Study
    • Increased incidence of thyroid and parathyroid cancers and skin cancers
  – MD Anderson Study
    • Statistically significant increase in solid tumors
Preventative Health

• Everybody
  – No tobacco
  – Good sun protection;
  – Regular skin evaluations
  – Healthy diet; Weight control
  – Limit alcohol

• Age-specific (start date depends on family history)
  – Mammogram, colonoscopies, PAP smears, prostate exams
  – Low-dose CT scans if smoker
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Provider Collaboration

• Communication between providers critical
• Lots of ways you can facilitate
  – Keep your own records to bring back and forth
    • i.e. information about labs, clinical trials
  – Ask about sharing EMR
  – Encourage communication
  – Work with ancillary staff
  – Engage in your care
Thank You for all you’ve done to advance this field

Happy to take any questions