Ms PB is a 51 yo woman with no PMH presenting to OSH with nausea, vomiting and abdominal pain, found to have a large cecal mass causing obstruction.

**Extreme Thrombocytosis: Reactive or Proliferative?**

<table>
<thead>
<tr>
<th></th>
<th>Reactive</th>
<th>MPN</th>
<th>Uncertain</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean peak count</td>
<td>1,195 x10^9</td>
<td>1,808 x10^9</td>
<td>1,437 x10^9</td>
</tr>
<tr>
<td>Maximum count</td>
<td>2,092 x10^9</td>
<td>2,900 x10^9</td>
<td>2,194 x10^9</td>
</tr>
<tr>
<td>Peak &gt;2,000 (%)</td>
<td>6.9%</td>
<td>34%</td>
<td>18%</td>
</tr>
<tr>
<td>Bleeding (%)</td>
<td>5%</td>
<td>24%</td>
<td>46%</td>
</tr>
<tr>
<td>Vaso-occlusion (%)</td>
<td>1%</td>
<td>24%</td>
<td>46%</td>
</tr>
</tbody>
</table>


To the OR...

- A successful right hemicolectomy with re-anastomosis was performed.
- Pathology revealed a “polypoid inflammatory mass... associated with ischemic change involving the colonic mucosa and submucosa.” All 24 lymph nodes sampled were negative for malignancy.


Success?

- The patient was discharged home on POD4 feeling well.
- Discharge labs:

  - WBC: 9.8
  - Hgb: 43.1
  - Platelets: 777
The patient returned to the ER with abdominal pain, nausea and vomiting.

ER labs:

Two Days Later…
CT was performed…

CT was performed…

Back to the OR…
New anatomy

Exploratory laparotomy revealed ischemia of the small bowel from the ligament of Treitz to the anastomosis necessitating surgical resection.
OSH hematology was consulted and recommended therapeutic anticoagulation and a thrombophilia workup

The next morning…
Another CT was performed…
IR and hematology consults were requested...

Aspirin was started rectally
Successful thromboaspiration was performed
Additional workup:

- Factor II: 25
- Factor VII: 35
- Factor VIII: 376
- Factor X: 25
- Factor XI: 49
- Fibrinogen: 358
- HIT ELISA: negative
- PNH flow: negative
- Lupus anticoagulant: negative
- Anticardiolipin antibody: negative
- Anti Beta 2 Glycoprotein: negative

A diagnostic procedure was performed...

Peripheral blood

Core biopsy

Core biopsy

Reticulin Stain
**JAK2V617F**: positive

Morphology: hypercellular marrow with trilineage hematopoiesis and panmyelosis including megakaryocytic hyperplasia.

**Final diagnosis**: essential thrombocytosis vs pre-fibrotic myelofibrosis

**Sort of...**
- **Risk of Recurrent Thrombosis**
  - 494 Italians with PV or ET and prior venous or arterial event
  - Overall recurrence: 7.6% patient-year
  - Antiplatelet rx: HR 0.72
  - Cytoreductive rx: HR 0.53
  - Antiplatelet + cytoreduction: HR 0.67
  - Anticoagulation + cytoreduction: HR 0.33


**Now what?**
- Pt already on aspirin and anticoagulation
- What about cytoreduction?


**What’s the Goal?**
- Normal
- <600
- <1 million

No Data

- Thrombosis rates actually **decrease** at higher platelet counts (1-1.5 million)
- Rates of hemorrhage **increase** at higher platelet counts.

**Which agent?**
- Hydroxyurea
- Anagrelide
- Interferon
- Busulfan
- Chemotherapy?
**Peg-interferon in MPN**

- Weekly SQ injection
- Titrate to effect (80 or 90mcg/wk starting)
- Hematologic response rate in ET 60-80%


**In the meantime...**

**Platelet Depletion**

**Not just platelets...**

**Bleeding in Thrombocytosis**

- Acquired Von Willebrand's (?)
- Mechanism unknown
- Decreased ristocetin cofactor activity
- Treatment: decrease the platelet count

**More Platelet Depletion...**

Line removed
Extreme thrombocytosis in a young person is most likely reactive... except when it isn’t.
Cytoreductive therapy is key in patients with MPN and thrombosis.
Peg interferon is a reasonable option for cytoreduction in patients who cannot absorb po.
Patients with extreme thrombocytosis are at greater risk of bleeding than clotting.
When all else fails... pheresis.

Take-Away Points

Thank you!

- Dr. Dan Martin
- Dr. Sindhu Cherian
- Dr. David Garcia
- Dr. Shweta Jain
- Dr. Siobhan Keel