Idiopathic (Immune) Thrombocytopenic Purpura

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Idiopathic (Immune) Thrombocytopenic Purpura

- Thrombocytopenia in the absence of other blood cell abnormalities (normal RBC & WBC, normal peripheral smear)

- No clinically apparent conditions or medications that can account for thrombocytopenia

Statistics of ITP

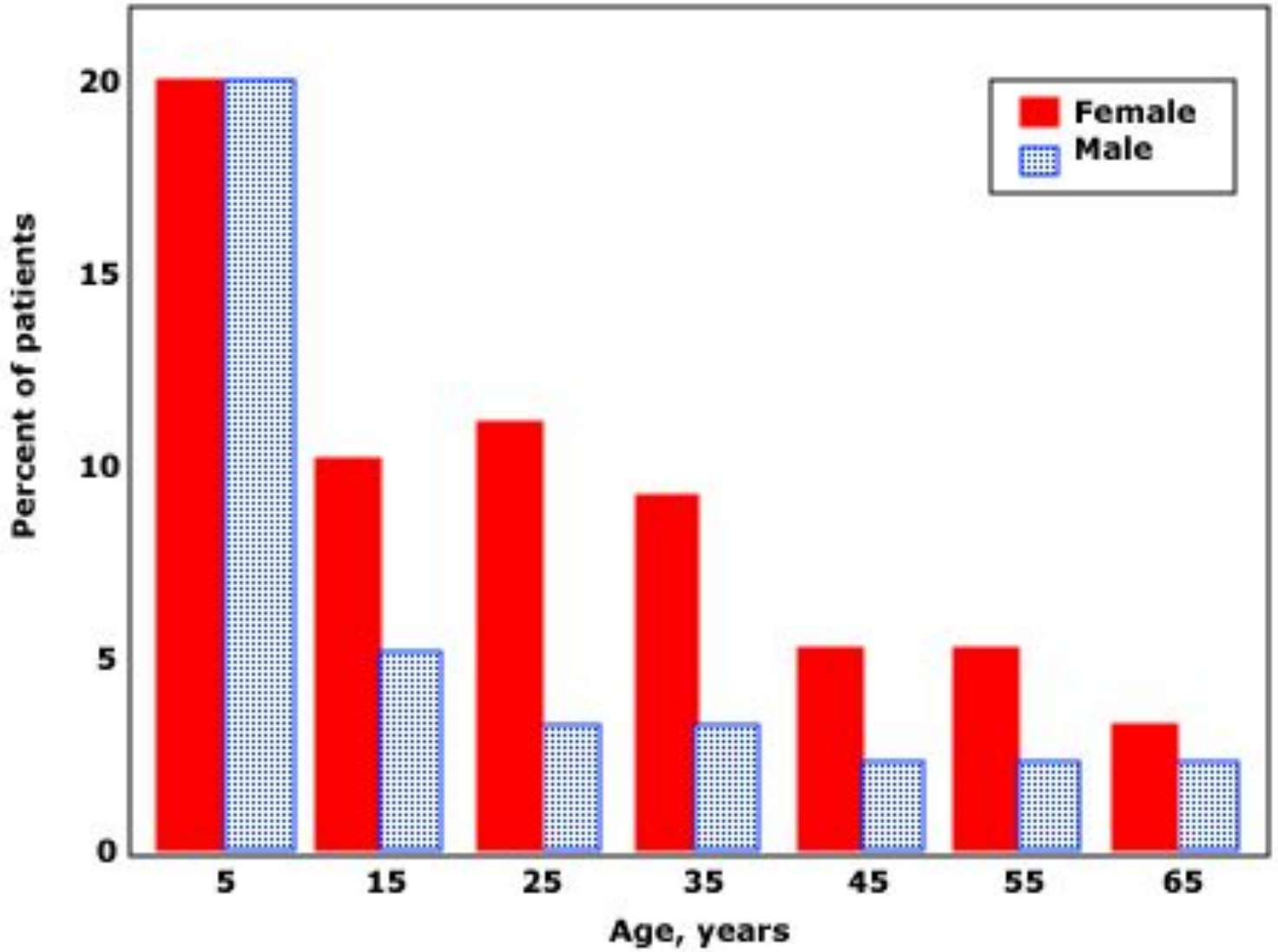
- Incidence of 22 million/year in one study

- Prevalence greater as often **chronic**

  *Segal et al 100 million/year

  *age-adjusted prevalence 9.5/100,000

  *1.9 :1 females / males


Female
Male
Percent of patients
20
15
10
5
0
5
15
25
35
45
55
65
Age, years

Clinical Manifestations

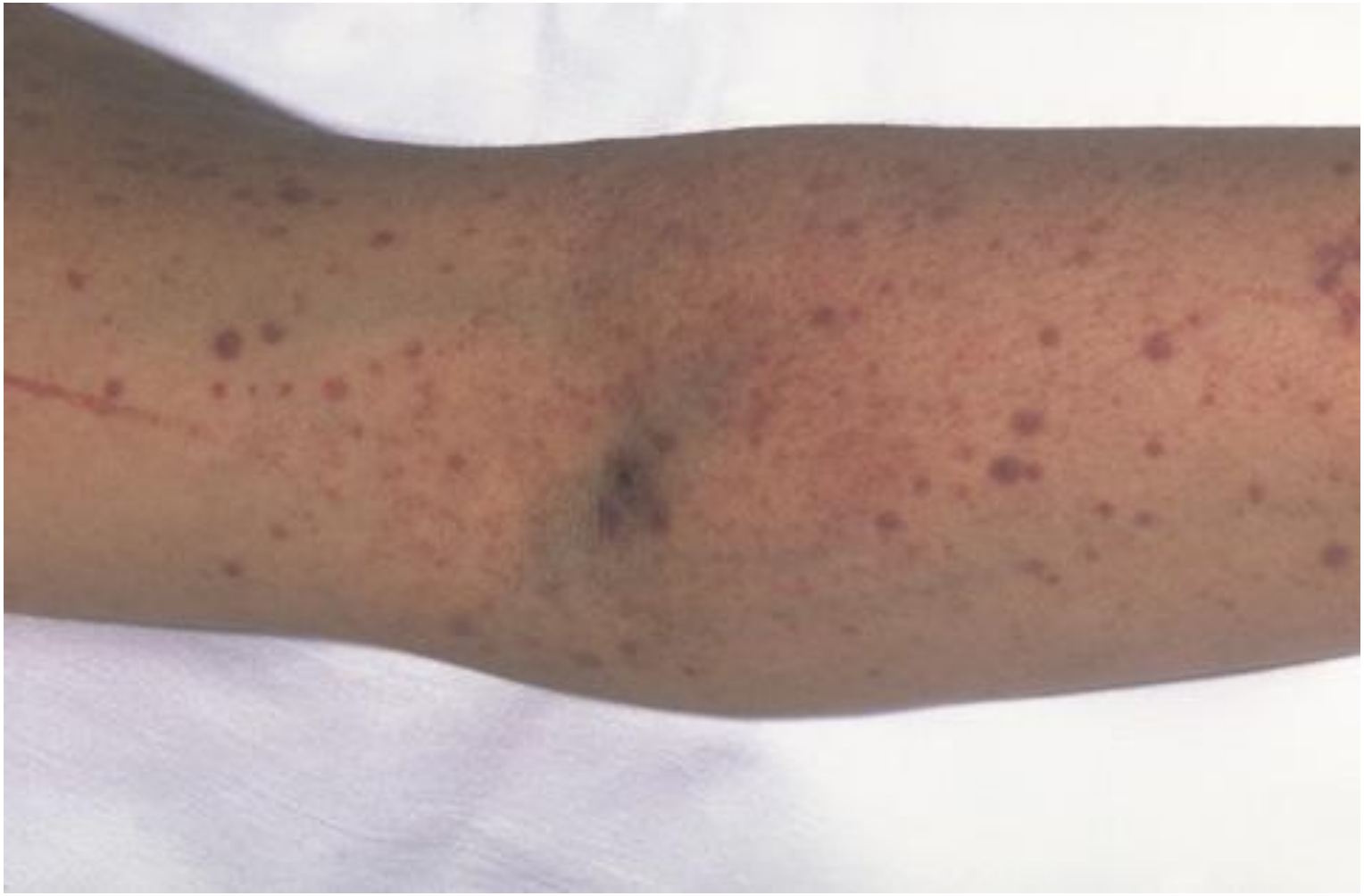
- May be acute or insidious onset
- **Mucocutaneous Bleeding**

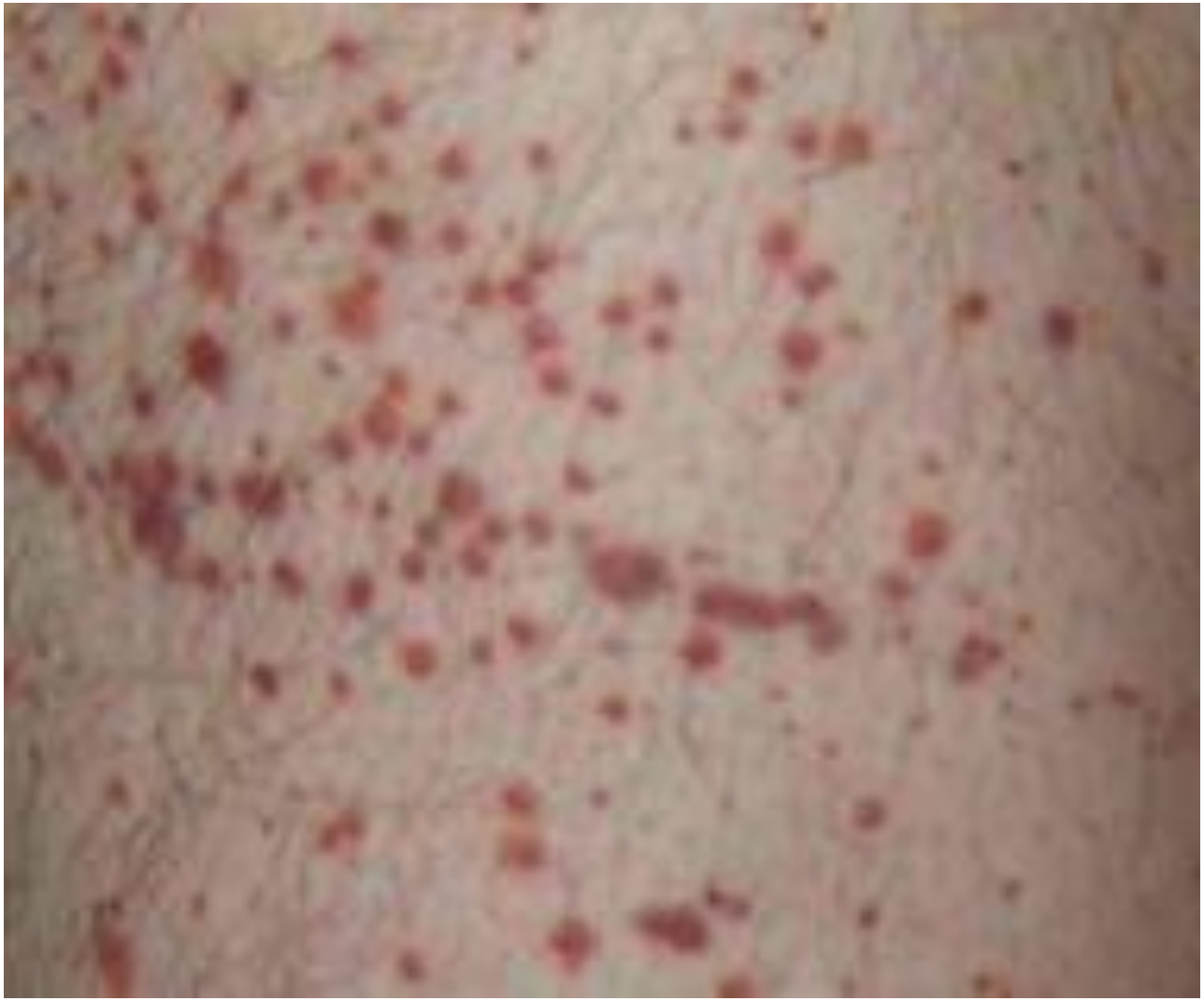
  *petechiae, purpura, ecchymosis

  *epistaxis, gum bleeding

  *menorrhagia

  ***GI bleed, CNS bleed = RARE**





Etiology of ITP : Children

- Often after **infection** (viral or bacterial)

- Theories:

  *antibody cross-reactivity

  **H. pylori*

  *bacterial lipopolysaccharides

Diagnosis *(of Exclusion)*

- Rule out other causes:

*lab error / PLT clumping

*drug / medication interaction

*infections (HIV, Hepatitis C)

*thyroid / autoimmune disease

*destructive / consumptive processes (TTP/HUS)

*bone marrow disease (leukemias, MDS)

Diagnosis *(of Exclusion)*

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To Marrow or Not to Marrow?

- Bone marrow aspiration & biopsy if…

- Patient 60 yrs. or older
- Poorly responsive to tx
- Unclear clinical picture

Anti-Platelet Antibody Testing

- **NOT recommended by ASH Practice Guidelines**
- Poor positive/negative predictive values, poor sensitivity with all current testing methods…
- …and doesn't change the management!

Management of ITP

- Goal = prevention of bleeding, NOT cure!

General Principles of Therapy

- Major bleeding **rare** if PLT > 10,000
- Goal = get PLT count to safe level to prevent bleeding…
- …**not** to specifically cure the ITP!

“Safe” Platelet Counts

- “moderately” t-penic = 30-50,000
- Probably safe if asymptomatic
- Caution with elderly (CNS bleeds)

When Planning Therapy…

- Tailor therapy and decision to treat to the individual patient
- Weigh bleeding vs. therapy risks

Initial Therapy

- **Prednisone 1 mg/kg/day**
  *usually response within 2 weeks
- Taper off after PLT response
- **Duration** of use = controversial

Second-Line Therapy

- IV Immune Globulin (IVIg)
  1 gram/kg/day x 2 days

- WinRho (anti-D) – if pt is Rh+
  50-75 mcg/kg/day

Treatment Side-Effects

- Steroids

  *bone density loss *GI effects

  *muscle weakness *weight gain

- IVIG/anti-D

  *hypersensitivity *headache

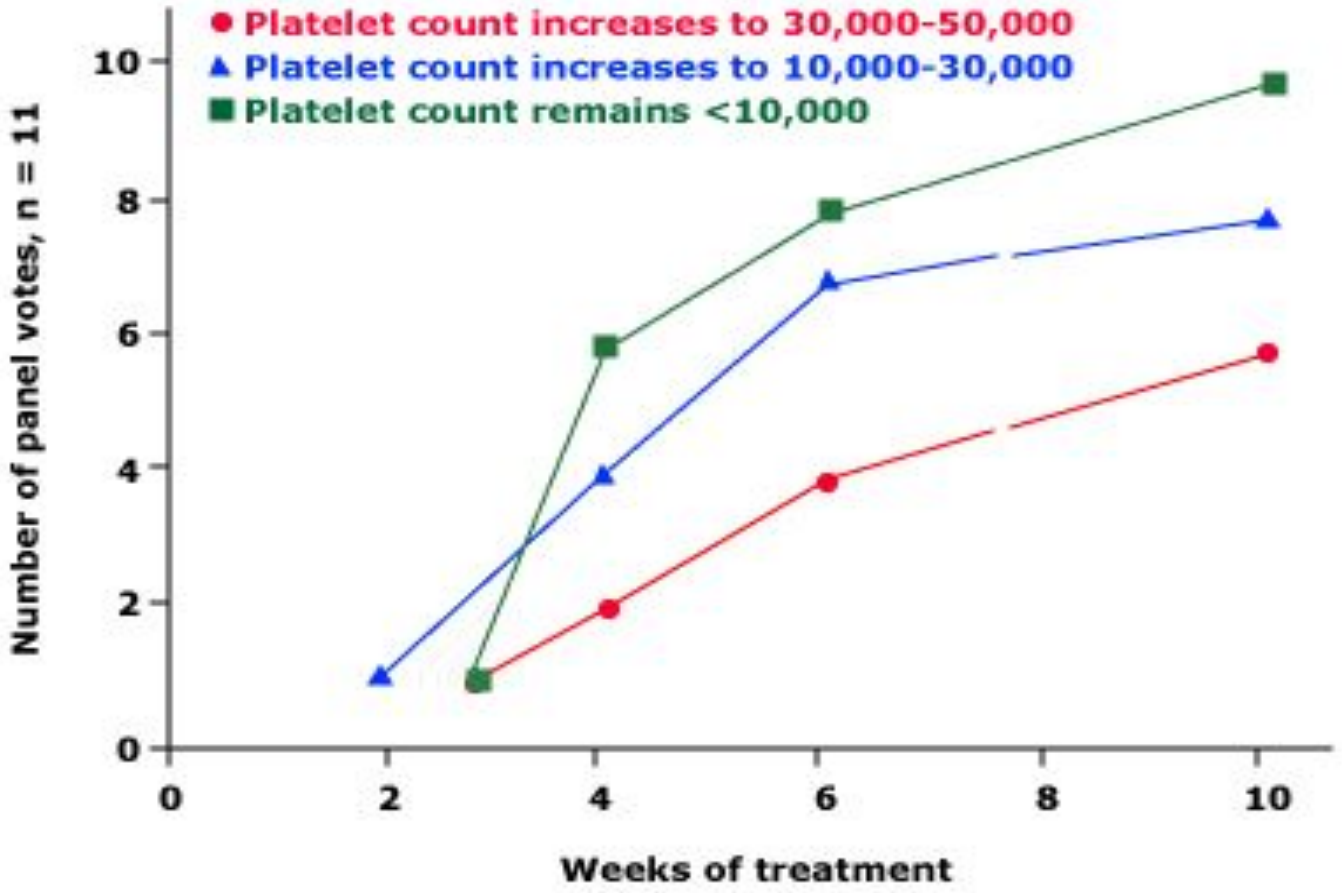
  *renal failure *nausea/vomiting

  *alloimmune hemolysis

Splenectomy

- Usually reserved for treatment failure
- Consider risk of bleeding, pt lifestyle
- **RISKS**
  *surgical procedure
  *loss of immune function vaccinations

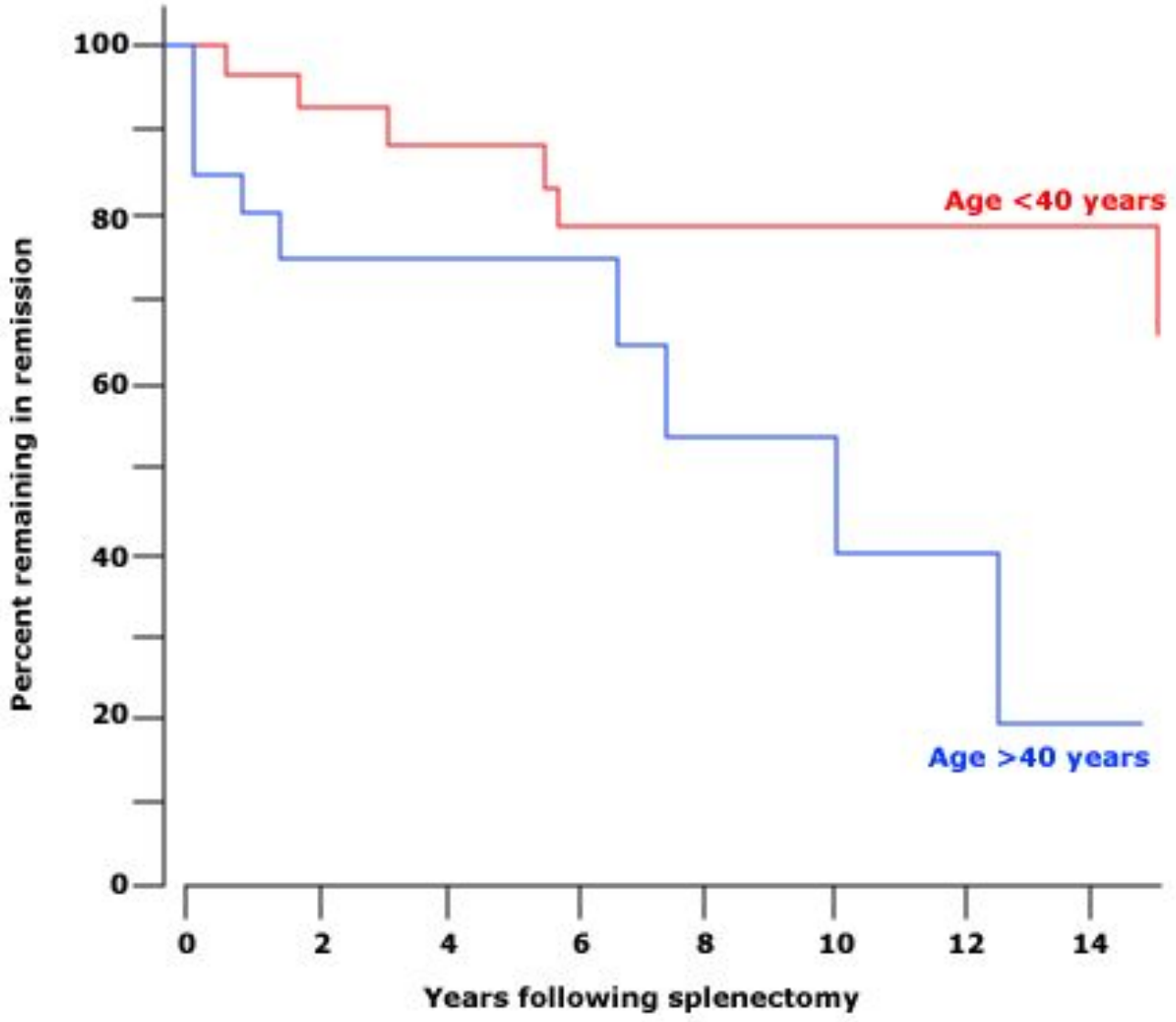
When to do Splenectomy?



Data from George, JN, Woolf, SH, Raskob, GE, et al. Blood 1996; 88:3.

Response Post-Splenectomy

- Usually normalized PLTs within 2 weeks (often immediately)
- Younger pts do better
- Kojouri et al (*Blood* 2004) □ 65% CR



Data from Fabris, F, et al. Br J Haematol 2001; 112:637.

Chronic Refractory ITP

- Persistent > 3 months
- PLT < 50,000
- Failure to respond to splenectomy

When all else fails…

- Steroids
- IVIg / anti-D
- Rituximab (anti-CD20)
- Cyclophosphamide
- Danazol
- Accessory splenectomy
- *H. pylori* eradication

Wrapping it up…

- ITP is often a chronic disease in adults
- Multiple therapies may be needed over time
- Goal = prevention of complications
- Therapy needs to be tailored to the individual patient


THANK
YOU
THANK

- Return to Main