

# MDS 2020

## Paient Discussion Forum

A young patient with progressive LR-MDS

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# Patient SM: History (I)

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- **A 41 yo**, male, engineer
- **3/2019:**
  - Fatigue
  - Macrocytic anemia (Hb 9.3)
- **2017:**
  - Asymptomatic
  - Anemia (Hb 11.0)
- **PMH:**
  - Pericarditis
  - Fatty liver
  - Smoking (20 pack yr)

# Patient SM: FamilyHistory

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- **Parents:**

- Cousins

- **Father**

- Ischemic heart disease
- COPD

- **Siblings:**

- Brother died in childhood (neuroblastoma)
- Two healthy: brother and siser

# Patient SM: Workup (2019) - I

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- **PE:** Unremarkable
- **CBC:**
  - Hb 8.8, MCV 106,
  - WBC 4.4 , ANC 3.0, PLT 343
- **Chemistry:**
  - Normal (except: GGT 203u)
- **Anemia workup** - negative
- **Diseases excluded:**
  - Collagen vascular; Infections;
  - Systemic inflammatory
- **Imaging:** Fatty liver; Spleen (15 cm)

# Patient SM: Workup (2019) - II

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- **Bone Marrow:**
  - Hypercellular (80%)
  - **Erythroid:**
    - Mild hyperplasia
    - Dyserythropoiesis
  - Myeloid: Blasts 3%
  - **Megakaryocytes:**
    - Increased numbers
    - Slightly atypical
  - Ringed **sideroblasts**
- **Cytogenetics:** 46XY
- **Genetics:** SF3B1 mutation

# Patient SM: Course (2019-20)

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- **Dg:**
  - MDS – RARS - Lower Risk
- **Hb decline**
  - 9.3 – 8.8 – 8.3 – 7.8
- **RBC transfusions**
  - 1-2/4-6 wk
- **Erythropoietin**
  - Eprex 40ku – 80ku/wk
- **No response**

# Patient SM :Summary

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- A 41 yo male
- RARS - Lower risk MDS
- 46XY
- SF3B mutation
- Hb decline
- RBC transfusion dependent
- ESA resistant
- Sister – matched donor

# Patient SM : Questions

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- **Allo- SCT ?**
  - Now ? Later ?
  - Pre-Allo-SCT treatment ? (Aza ?)
- **Experimental treatment?**
  - Ruxodustat ?
  - Luspatercept ?
  - Other ?
- Azacitidine ?
- Azacitidine + Venetoclax ?