GATA2-related MDS

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Wlodarski MW, Hirabayashi S, Pastor V, Starý J, Hasle H, Masetti R, Dworzak M, Schmugge M, van den Heuvel-Eibrink M, Ussowicz M, De Moerloose B, Catala A, Smith OP, Sedlacek P, Lankester AC, Zecca M, Bordon V, Matthes-Martin S, Abrahamsson J, Kühl JS, Sykora K-W, Albert MH, Przychodzien B, Maciejewski JP, Schwarz S, Göhring G, Schlegelberger B, Cseh A, Noellke P, Yoshimi A, Locatelli F, Baumann I, Strahm B, Niemeyer CM for the EWOG-MDS. Prevalence, clinical characteristics, and prognosis of GATA2-related myelodysplastic syndromes in children and adolescents. *Blood*. 2016;127(11):1387-1397.

1. Your patient is a 12-year-old boy diagnosed with myelodysplastic syndrome (MDS). According to the genetic testing done on samples from 2 consecutive prospective studies on pediatric MDS by Wlodarski and colleagues, which of the following statements about the prevalence of germline *GATA2* mutations among children and adolescents with MDS is correct?

- ☐ Germline *GATA2* mutations account for 32% of advanced pediatric MDS
- ☐ Germline *GATA2* mutations account for 15% of primary pediatric MDS
- ☐ Germline *GATA2* mutations occur in 5% of children with MDS secondary to therapy or acquired aplastic anemia
- ☐ Germline *GATA2* mutations occur in 37% of all pediatric patients with MDS and monosomy 7, and in 72% of adolescents with MDS and monosomy 7

2. On the basis of the samples from 2 consecutive prospective studies and stratified analysis according to karyotype by Wlodarski and colleagues, which of the following statements about other genetic features and clinical characteristics of germline *GATA2* mutation carriers vs wild-type cases of MDS is correct?

- ☐ Monocytosis was more frequent in patients with *GATA2* mutations, independent of monosomy 7
- ☐ Germline *GATA2* mutations were associated with significantly worse overall survival than in wild-type MDS
- ☐ *GATA2* mutation carriers with MDS were older at diagnosis and were more likely to present with advanced disease than patients with no *GATA2* mutations
- ☐ Outcome of hematopoietic stem cell transplantation (HSCT) was worse in mutation carriers than in patients with wild-type MDS

3. On the basis of the studies and stratified analysis according to karyotype by Wlodarski and colleagues, which of the following statements about the clinical implications of the findings regarding pediatric MDS is correct?

- ☐ GATA2 analysis should be included in the workup of pediatric MDS only if family history is positive
- ☐ Early diagnosis of GATA2 deficiency can avoid unnecessary diagnostic procedures, enable tailored surveillance, and limit the use of noncurative therapies specifically avoiding treatment with immunosuppression
- ☐ HSCT should be delayed, given the low risk for progression to advanced disease
- ☐ Known risk factors do not affect decisions on timing and preparative regimen for HSCT in GATA2-related MDS

Activity Evaluation (where 1 is strongly disagree and 5 is strongly agree)

1. The activity supported the learning objectives.
   1 2 3 4 5
2. The material was organized clearly for learning to occur.
   1 2 3 4 5
3. The content learned from this activity will impact my practice.
   1 2 3 4 5
4. The activity was presented objectively and free of commercial bias.
   1 2 3 4 5

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