

In the following situations, order **G6PDZ / Glucose-6-Phosphate Dehydrogenase (G6PD) Full Gene Sequencing**:

- To determine carrier status (assigned female at birth) or affected status (assigned male at birth)
- Previous G6PD enzyme test result is inconclusive
- High white blood cell count (may elevate G6PD results)
- Reticulocytosis (may elevate G6PD results)
- Family history of unknown G6PD variant
- Provider preference

Assigned male at birth (hemizygous)

WHO Class A Variant*

- <20% enzyme activity expected
- Deficient with chronic nonspherocytic hemolytic anemia (CNSHA) phenotype (chronic hemolysis)

WHO Class B Variant*

- <45% enzyme activity expected
- Deficient with possible episodic/triggered hemolysis

WHO Class U Variant*

- Uncertain clinical significance
- Indeterminate result
- Order **G6PD1 / Glucose 6- Phosphate Dehydrogenase Enzyme Activity, Blood** unless previously performed.

WHO Class C Variant*

- 60%-150% enzyme activity expected
- Normal

Therapeutic Recommendation:
No G6PD-related contraindication of drugs/ compounds.

WHO Class A + A Variants*

- Each allele predicted with <20% enzyme activity
- Deficient with CNSHA phenotype (chronic hemolysis)

WHO Class A + B Variants*

- One allele predicted with <20% enzyme activity
- One allele predicted with <45% enzyme activity
- Deficient, at-risk for CNSHA phenotype

WHO Class B + B Variants*

- Each allele predicted with <45% enzyme activity
- Deficient

WHO Class (A or B) + U Variants*

- One allele predicted with either <20% enzyme activity or <45% enzyme activity
- One allele with uncertain clinical significance
- Deficient with CNSHA phenotype (chronic hemolysis)

WHO Class C Variant*

- Predicted 60%-150% enzyme activity
- Normal

■ Indeterminate result
■ Order **G6PD1** unless previously performed.

Therapeutic Recommendation:
No G6PD-related contraindication of drugs/compounds.

Therapeutic Recommendation:
Contraindication for drugs associated with hemolytic anemia in G6PD-deficient patients. See [Pharmacogenomic Association Tables](#)

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Contraindication for drugs associated with hemolytic anemia in G6PD-deficient patients. See [Pharmacogenomic Association Tables](#)

Assigned female at birth (heterozygous)

WHO Class A, B, or U Variant*

- Uncertain clinical significance
- Indeterminate result
- Order **G6PD1** unless previously performed.

WHO Class C Variant*

- 60%-150% enzyme activity expected
- Normal

Therapeutic Recommendation:
No G6PD-related contraindication of drugs compounds.

* World Health Organization (WHO) classification of individual alleles
 WHO Class A Variant: Severe G6PD enzyme deficiency (<20% enzyme activity) and CNSHA
 WHO Class B Variant: Deficient G6PD enzyme activity (<45% enzyme activity) and episodic/triggered hemolytic anemia
 WHO Class C Variant: 60%-150% G6PD activity, considered normal/adequate enzyme activity
 WHO Class U Variant: Uncertain clinical significance

Legacy WHO classification of individual alleles (Updated in 2022 to the current classification above)
 WHO Class I Variant: Severe enzyme deficiency (<10% enzyme activity) with CNSHA
 WHO Class II Variant: Severe enzyme deficiency (<10% enzyme activity)
 WHO Class III Variant: Moderate to mild enzyme deficiency (10%-60% enzyme activity)
 WHO Class IV Variant: Very mild or no enzyme deficiency (60%-150% enzyme activity)
 WHO Class V Variant: Increased enzyme activity (more than twice normal enzyme activity)