Imatinib (Gleevec) for treatment of severe mast cell activation disease (a subset of mast cell activation syndrome - MCAS)

There is clinical experience and published case reports showing that Imatinib (Gleevec) can be effective in drug-resist, aggressive mast cell activation disease/syndrome/indolent mastocytosis. This medication is currently approved for use in aggressive systemic mastocytosis and various leukemias and gastrointestinal stromal tumors (GIST). The dose of Imatinib is lower for MCAS than for malignancy – 100-200 mg vs. 400 mg by mouth each day

Potential side effects from taking Gleevec include:

- Edema (swelling) and severe fluid retention have occurred. You should weigh yourself regularly. Report any rapid weight gain so you can be treated by drug interruption and diuretics.
- Abnormal blood counts, particularly anemia, neutropenia, and thrombocytopenia, have occurred. You need to have complete blood counts weekly for the first month, biweekly for the second month, and monthly thereafter. If there is a problem we will treat with dose reduction, dose interruption, or discontinuation of the medication.
- Severe congestive heart failure and left ventricular dysfunction have been reported, particularly in patients with comorbidities and risk factors. Report any shortness of breath so we can monitor and treat patients with cardiac disease or risk factors for cardiac failure.
- Severe hepatotoxicity including fatalities may occur. Assess liver function before initiation of treatment and monthly thereafter or as clinically indicated.
- Note that in other patients with lymphoma and GI cancer, GI bleeding has been reported.
- Gastrointestinal perforations, some fatal, have been reported.
- Cardiogenic shock/left ventricular dysfunction has been associated with the initiation of Gleevec in patients with specific conditions associated with high eosinophil levels
- Bullous dermatologic reactions (e.g., erythema multiforme and Stevens-Johnson syndrome) have been reported with the use of Gleevec.
- Hypothyroidism has been reported in thyroidectomy patients undergoing levothyroxine replacement. We will monitor the TSH levels in such patients.
- Fetal harm can occur when administered to a pregnant woman. Avoid pregnancy completely when taking Gleevec.
- Growth retardation occurring in children and pre-adolescents receiving Gleevec has been reported. Close monitoring of growth in children under Gleevec treatment is recommended.
- Reports of motor vehicle accidents have been received in patients receiving Gleevec. Caution driving a car or operating machinery – make sure there is no fatigue during these events
- Renal toxicity. A decline in renal function may occur in patients receiving Gleevec. We will evaluate renal function at baseline and during therapy.

The most frequently reported adverse reactions (greater than or equal to 30%) were edema, nausea, vomiting, muscle cramps, musculoskeletal pain, diarrhea, rash, fatigue and abdominal pain.

Consent:

I am read the above information and wish to take Imatinib/Gleevec therapy. I will comply with lab monitoring.** I understand and accept the risks and understand that this therapy is used in cases of mast cell diseases where the symptoms have not responding to numerous treatments and the quality of life is so impaired that innovative therapy is required. I have been given this form in advance of taking the medication and was allowed to read it and ask my physician questions regarding this medication.

Signature/date: _____ _____

**Labs: Baseline CBC, CMP and if taking thyroid medicine a TSH. During therapy have CBC (complete blood count) weekly for the first month, biweekly for the second month, and then monthly; CMP (complete metabolic panel) monthly

References

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